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Clinical Studies on the Occurrence and the Pathogenesis of Optociliary Veins

Yoshimasa Masuyama, M.D., Yoshihisa Kodama, M.D., Yoshifumi Matsuura, M.D., Atsushi Sawada, M.D., Kazumichi Harada, M.D., and Toshinori Tsuchiya, M.D.

We reviewed retrospectively 20 cases of optociliary vein over the past 5 years. Fifteen of the 20 cases (75%) were over 40 years of age. There was no sex differences (9 males and 11 females). Causative ocular diseases were: central retinal vein occlusion (14 cases, 70%); optic disc drusen (2 cases, 10%); and optic nerve sheath meningioma, high myopia, glaucoma, congenital anomaly (1 case each, total 20%). The number of patients with an optociliary vein was high in central retinal vein occlusion (14/190 cases, 7.4%). We concluded that occurrence of optociliary vein is not restricted to cases with optic nerve sheath meningioma and this shunt vessel may occur under other conditions in which central retinal venous return is seriously disturbed. The most common cause of optociliary vein is central retinal vein occlusion.

Key Words: Central retinal occlusion—Fluorescein fundus angiography—Optic nerve sheath meningioma—Optociliary vein.

An optociliary vein rarely occurs between the central retinal vein and the peripapillary choroidal vein in the pre laminar region of the optic nerve head. This anomalous shunt vessel is thought to represent enlargement of a pre-existing capillary vessel between the central retinal vein and the choroid when the retinal venous return is obstructed (1,2). Salzmann (3) reported an evidence of optociliary vein for the first time in a case of myxosarcoma of the optic nerve in 1893. An optociliary vein is a well-known characteristic finding of the optic nerve sheath meningioma, and therefore, it has been mentioned as one of the triad symptoms in sphenoorbital meningiomas, including optic nerve sheath meningioma (the Hoyt-Spencer's sign) (4-9). However, these shunt vessels are encountered in other ocular disorders such as optic disc drusen (10), arachnoid cyst of optic nerve (11), chronic papilledema (12-15), optic nerve glioma (16), central retinal vein occlusion (1,17-19), phakomatosis (20), and chronic glaucoma (21). In addition to the above acquired optociliary veins, a congenital optociliary vein has been reported even in the normal eye (17,21,22). We have experienced 20 cases with optociliary vein during the past 5 years. The purpose of this study is to clarify the occurrence and the pathogenesis of the optociliary vein.

PATIENTS AND METHODS

A retrospective review was made of those cases diagnosed as optociliary vein at the general clinics of the Department of Ophthalmology, Miyazaki Medical College Hospital and Miyazaki Central Eye Hospital for 5 years from 1978 to 1982. The appearance of an optociliary vein was examined with fundus photography and fluorescein fundus...
RESULTS

In our study, we found 20 total cases of optociliary vein. The occurrence was 11 of 6,851 patients at Miyazaki Medical College Hospital and 9 of 296,948 patients at the Miyazaki Central Eye Hospital, respectively. As to the age distribution, 15 patients (75%) were above 40 years of age, and of these eight (40%) were in their 60s, indicating a high incidence after middle age. No sex difference was noted in this study, which consisted of 9 men and 11 women. The right eye and the left eye were involved in 10 cases, each without any preference for the affected side. As to the causative diseases, central retinal vein occlusion was found to be the most frequent cause, in 14 of 20 cases (70%), followed by optic disc drusen in two cases (10%), and optic nerve sheath meningioma, high myopia, glaucoma, and congenital anomaly in one case each (5%) (Fig. 1).

We examined the number of the patients with optociliary vein in this patient population. The number of the patients with central retinal vein occlusion was 190 cases; 65 cases at Miyazaki Medical College Hospital and 125 cases at Miyazaki Central Eye Hospital. Of these cases with central retinal vein occlusion, 14 cases (7.4%), including two cases of hemi-central retinal vein occlusion, had optociliary vein; seven cases (10.8%) and seven cases (5.6%) in respective hospitals. When evaluated by the $\chi^2$ test the frequencies for these two hospitals were not statistically significant ($p < 0.05$). Patients with branch retinal vein occlusion examined in both hospitals totaled 575 cases, but we could not find any cases involving an optociliary vein. Of 1,465 glaucoma patients and 1,920 high myopia patients, optociliary vein was seen in one case each (0.07% and 0.05%). The incidence of the optociliary vein in the population with the same disease was significantly higher in central retinal vein occlusion patients than in glaucoma and high myopia patients ($p < 0.01$). Of seven patients with optic disc drusen, optociliary vein was seen in two cases. Only one optic nerve sheath meningioma patient was found, and they had an optociliary vein. These conditions showed no statistical significance when compared with that of central retinal vein occlusion. Congenital optociliary vein was found in one case in this study making its incidence extremely low.

In these 20 cases, 25 optociliary veins were found in all. Out of the four quadrants of the disc, nine (36%) were found on the temporal quadrant, seven (28%) on the nasal, five (20%) on the lower, and 4 (16%) on the upper quadrant (Fig. 2). Representative cases of optociliary vein are discussed as follows.

Case 1: Central Retinal Vein Occlusion

A 31-year-old woman had a 1-month history of blurred vision in her left eye. She had suffered from renal hypertension in the past. On examination, visual acuity was 20/20 in the right eye and 20/40 in the left eye. Intraocular pressure was 16 mmHg in the right eye and 14 mmHg in the left eye. The right fundus showed no abnormalities. The left fundus revealed venous dilation and loop

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**FIG. 1.** The incidence of the causative ocular diseases on the optociliary vein.

**FIG. 2.** The site of the optociliary vein on four quadrants of the optic disc.
formation near the disc margin at the 12 o'clock position and a tortuous optociliary vein was present at the temporal margin. Fluorescein angiography indicated that this vein filled in the arteriovenous phase from the central retinal vein to the choroid and fluoresced along with the retinal vein until the late stage. The optociliary vein did not leak fluorescein dye (Fig. 3).

Case 2: Hemi-Central Retinal Vein Occlusion

A 39-year-old man presented with a 3-month history of blurred vision in his right eye. He had no other ocular or neurological symptoms, and his past medical history and family history were unremarkable. Visual acuity was 20/60 in the right eye and 20/20 in the left eye. Ophthalmoscopy of the left eye revealed no abnormalities. In the right eye, retinal hemorrhages spread in the inferior half of the fundus due to occlusion of the inferior retinal venous trunk. There was a venous loop connection between the superior and inferior major venous trunk at the nasal disc margin. An optociliary vein was seen between the venous loop and the choroid at the 9 o'clock position of the disc margin. Fluorescein angiography demonstrated the optociliary vein filled in the arteriovenous phase from the central retinal vein to the choroid and remained fluorescent until the late stage (Fig. 4).

Case 15: Optic Disc Drusen

A 46-year-old woman was seen with a chief complaint of blurred vision in the right eye. Visual acuity was 20/25 in the right eye and 20/20 in the left eye. In the right fundus, the disc margins were poorly demarcated and slightly elevated with drusen. The retinal veins were mildly dilated but no retinal hemorrhage was present. An optociliary vein was found on the temporal aspect of the disc, partially hidden by the overlying nerve fibers (Fig. 5). On fluorescein angiography, this optociliary vein filled in the arteriovenous phase from the central retinal vein to the choroid. The drusen remained hyperfluorescent after the late stage even after the veins had emptied (Fig. 6). The optic nerve head showed high acoustic reflectivity on echography. The optociliary vein remains unchanged on follow-up 5 years later.

Case 17: Optic Nerve Sheath Meningioma

A 58-year-old woman had a recurrent optic nerve sheath meningioma in the left eye. Her initial surgery was 6 years previous to this examination. Visual acuity was 20/20 in the right eye and

FIG. 3. Fluorescein angiogram of case 1 with central retinal vein occlusion. Loop formation of the dilated veins is seen at the upper disc margin. The tortuous optociliary shunt vessel was present at 3 o'clock (arrowhead) and filled from the central retinal vein to the choroid during the arteriovenous phase.

FIG. 4. Fluorescein angiogram of case 2 with hemi-central retinal vein occlusion. The optociliary vein is present near the margin of the disc at the 9 o'clock position (arrowhead). This vessel was filled in the arteriovenous phase from the central retinal vein and remained fluorescent until the late stage.

FIG. 5. Fluorescein angiogram of case 15 with optic disc drusen. Loop formation of the dilated veins is seen at the upper disc margin. The tortuous optociliary shunt vessel was present at 3 o'clock (arrowhead) and filled from the central retinal vein to the choroid during the arteriovenous phase.
no light perception in the left eye. Exophthalmometry was 12 mm in the right eye and 21 mm in the left eye. Ophthalmoscopy before her second operation revealed two optociliary veins in the upper quadrant and one in the nasal quadrant. On fluorescein angiography, these three shunt veins filled in the early arteriovenous phase from the central retinal vein and followed the venous pattern, remaining eufluorescent into the late stage. These veins did not leak fluorescein dye (Fig. 7).

Case 18: High Myopia

A 57-year-old woman had a history of high myopia. Visual acuity was 20/100 (better with -7.00) in the right eye and 20/200 (better with -7.00) in the left eye. There was a mild cortical opacity to the lens in each eye. Ophthalmoscopy of the left eye revealed a temporally distorted disc with conus and posterior staphyloma. There was neither retinal hemorrhage or vascular sheathing. Only mild dilation of the retinal veins was present. An optociliary vein was seen at the temporal conus margin (Fig. 9). Fluorescein angiography indicated that the optociliary vein communicating with the
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FIG. 9. Fundus photograph of case 18 with high myopia. The fundus revealed temporally distorted disc and posterior staphyloma. There are coiling of retinal vein on the disc and venous dilation. An optociliary vein is seen at the temporal aspect of the disc (arrowhead).

choroid filled from the central retinal vein during the early venous phase and fluoresced along with the retinal vein until the late stage (Fig. 10).

Case 19: Congenital Glaucoma

A 16-year-old girl had a history of congenital glaucoma in both eyes. Her highest intraocular pressure readings were 35 mm Hg in the right eye and 33 mm Hg in the left eye, with a corneal radius of 13 mm in both eyes. The right eye was more seriously affected due to the prolonged high pressure. She had had a trabeculectomy in both eyes 4 years before and her intraocular pressure has been well controlled. On examination after surgery, visual acuity with best correction was 20/400 in the right eye and 20/40 in the left eye. Intraocular pressure was 16 mm Hg in the right eye and 14 mm Hg in the left eye. Ophthalmoscopy of the right eye revealed the disc to be pale with a large cup. At the 7 o'clock position of the disc margin an optociliary vein was seen communicating with the choroid (Fig. 11). Fluorescein angiography indicated this shunt vein to be filling during the arteriovenous phase from the inferotemporal retinal vein (Fig. 12). The optociliary vein was present before the operation and remains afterwards. The left fundus showed marked optic atrophy with a large cup but had no optociliary vein.

Case 20: Congenital Anomaly

A 32-year-old woman was seen who complained of itching in the left eye. Her past medical history was unremarkable and her present health was excellent. Visual acuity in each eye was 20/20 with full visual fields. The eye examination was normal with the exception of the right fundus, which showed a prominent optociliary vein at the inferior disc margin. This vessel displayed a light red color and straight course (Fig. 13). On fluorescein angiography this vein filled during the early arteriovenous phase and remained as equally fluorescent as the retinal veins during the late venous phase, and was identified as a veno-venous anastomosis. The direction of flow was from the choroid to the retinal vein. There were no other abnormalities in the right fundus (Fig. 14). She had no neurological abnormalities and computerized tomographic scans showed a normal optic nerve and no retro-
The disc has a large glaucomatous cup. The optociliary shunt vessel (arrowhead) is seen communicating between the inferotemporal retinal vein and the choroid. This vein filled from the retinal vein during the arteriovenous phase.

bulbar lesions. The optociliary vein has remained unchanged over a 5-year follow-up.

DISCUSSION

We reviewed retrospectively 20 cases with optociliary vein during the past 5 years. The important fact to emphasize from this study is that central retinal vein occlusion is the most common cause of optociliary vein among the various causative diseases, accounting for 14 cases (70%). Central retinal vein occlusion has a high prevalence in middle and advanced age. For this reason optociliary vein is seen more frequently in this same age group.

The incidence of the optociliary vein was high in central retinal vein occlusion patients among various causative diseases (7.4%). This explanation is supported by the evidence that the central retinal vein is seriously blocked in the prelaminar region of the optic disc.

Branch retinal vein occlusion, however, was not a causative factor of optociliary vein because the venous return is blocked at the arteriovenous crossing, which is away from the disc. Collateral vessels occur among the adjoining retinal veins. We could not find the optociliary vein in branch retinal vein occlusion patients in this study. Anderson et al. (21) reported four cases of branch retinal vein occlusion with optociliary vein. From the fundus and fluorescein photographs of case 8 published in their article, the patient was thought to have a hemi-central retinal vein occlusion because narrowing of the inferior trunk of central retinal vein was apparently found in the prelaminar region. Hemi-central retinal vein occlusion should be sharply distinguished from branch retinal vein occlusion. We emphasize that optociliary vein occurs only in a condition where venous return is blocked or impaired in the prelaminar region of the optic disc.

An optociliary vein has appeared in monographs as one of the characteristic clinical features of optic nerve sheath meningioma. In the single optic nerve sheath meningioma patient in this series, optociliary veins developed and grew slowly for 6 years. These veins disappeared after tumor excision that has been previously reported (7,14,15). Boschetti et al. (23) reported differences
in fluorescein angiograms between optociliary veins developing secondary to retinal vein occlusion and those developing with optic nerve sheath meningioma. In the optic nerve sheath meningioma patients, the shunt vessels filled in the arteriovenous phase and demonstrated hyperfluorescence in the late venous phase when compared with the retinal veins. During fluorescein angiography of our patient (case 17), the optociliary veins initially filled in the arteriovenous phase as seen in cases of Boschetti et al. However, we could not find any hyperfluorescence in these veins during all of the studies phases. Hyperfluorescence seen in Boschetti et al.’s cases may suggest the presence of an arteriovenous shunt or drainage from the tumor behind the optic nerve head to the retinal vein. Further clinical studies will be needed to clarify the fluorescein angiographic differences between these two conditions.

Two cases with optic disc drusen in this series showed large and well-developed optociliary veins. The deep-seated drusen or their conglomerates are the most probable obstacle for blood flow in the prelaminar section of the central retinal vein, which the dilated optociliary communications try to bypass. Reports (10,24) that optic disc drusen disturb the retinal venous return are supported by our observations. In the case of high myopia, the extension of the axis of the globe seems to have affected the optic disc somehow, probably interfering with venous return in the central retinal vein. Retinal venous dilation seen in this case may indicate the presence of impaired blood return. In glaucoma the optociliary vein is most likely the result of sustained high intraocular pressure. The sustained high intraocular pressure is probably the cause for the distortion of the lamina cribrosa, as well as causing the corneal enlargement. As a result the optociliary vein remains even after the pressure returns to normal.

Usually the optociliary vein is an acquired condition as described above. The occurrence of optociliary veins as a congenital vascular malformation is extremely rare. A few authors (21,22) indicated that the direction of blood flow is from the choroid into the central retinal vein. The optociliary vein in our case (case 20) revealed the same direction of flow. Acquired optociliary veins were tortuous and dark red, and the blood flow was from the retinal to the choroidal circulation. Our congenital optociliary vein patient showed a light red color, indicating a higher oxygen content, and the vein’s blood flow was from the choroidal to the retinal circulation. This vein is straight in course and stable without other intraocular lesions. These findings indicate that a congenital optociliary vein is not concerned with disturbance of retinal venous return.

Fluorescein angiography helps in identification of the optociliary vein and may ascertain the direction of blood flow. Optociliary veins are veins of larger caliber as compared with the new vessels on the disc, and do not leak fluorescein. They fill during the arteriovenous phase or the venous phase of the angiogram and are seen connecting the central retinal vein with the choroidal vessel. New vessels, on the contrary, are much finer structures that originate from a vein and end in the same vessels, and are a network of fine vessels that leak fluorescein profusely. This differentiation is important because of the frequent interpretation of collaterals on the disc as new vessels. Other congenital vascular abnormalities on the optic disc, for example, arteriovenous anastomosis associated with Wyburn-Mason syndrome (25) and prepapillary vascular loops (26) are easily differentiated with fluorescein angiography. Little attention has been paid to the site of optociliary veins at the optic disc. From the results of this study, an optociliary vein tends to occur more frequently on the temporal quadrant on the optic disc. This quadrant of the disc is thought to have more vascularity than others and as a result cilioretinal vessels or optociliary veins sometimes arise in this region.

Finally, optociliary vein is well known as one of Hoyt-Spencer’s signs in patients with optic nerve sheath meningioma. However, we emphasize that the occurrence of the optociliary vein is not restricted to cases of meningioma and this shunt vessel may be seen in other conditions in which central retinal venous return is seriously disturbed in the prelaminar region of the optic disc. The most common cause of optociliary vein is central retinal vein occlusion.

Acknowledgment: We wish to thank Dr. Prem S. Virdi for his valuable comments on the manuscript.

REFERENCES


Primary Chiasmal Germinoma
A Case Report and Review of the Literature

C. Bradley Bowman, M.D. and Bradley K. Farris, M.D.

A case is presented in which an adult man with painless progressive loss of vision subsequently was found to have a primary suprasellar/perichiasmal germinoma (ectopic pinealoma). A review of the literature revealed 93 similar cases of germinoma occurring in the perichiasmal region and these are tabulated. The diagnosis and management of this lesion are discussed, including the recognition of the characteristic neuroendocrinologic triad of diabetes insipidus, visual changes, and hypopituitarism. Of 64 cases from the literature wherein presenting symptoms were reported, 56 (87.5%) had diabetes insipidus, 53 (82.8%) visual changes, and 36 (56%) hypopituitarism. A review of the literature suggests that diabetes insipidus is usually the initial symptom in suprasellar germinoma. However, we wish to emphasize the ophthalmologic presentation of this entity, because patients in the age group most affected (adolescents) will often not recognize symptoms of diabetes insipidus, but will first seek medical attention for painless progressive loss of vision suggestive of chiasmal compression. The radiosensitivity of this lesion is also discussed. Of 61 patients receiving irradiation therapy, 42 (68%) were surviving at the time of their individual case report. No patient in the review survived without irradiation therapy.

Key Words: Atypical teratoma—Chiasm—Chiasmal compression syndrome—Ectopic pinealoma—Junctional scotoma.

Suprasellar germinomas, also referred to as ectopic pinealomas or atypical teratomas, are relatively rare tumors that have attracted considerable attention during the past few years. Reports of the remarkable sensitivity of these germ cell tumors to irradiation therapy, and improved imaging of midline central nervous system (CNS) lesions with magnetic resonance imaging (MRI) have contributed to the importance of making a rapid and correct tissue diagnosis (1-4). In 1961, Kageyama and Belsky described a neuroendocrinologic triad associated with suprasellar germinoma consisting of diabetes insipidus, visual disturbances, and hypopituitarism (5). This same triad has been well documented by several authors in the neurosurgical and radiological literature (1,2,6-8). However, the more common ophthalmologic presentation of this entity, with visual loss suggestive of chiasmal compression, has not yet been appropriately emphasized.

CASE REPORT

A 44-year-old white male oil field worker presented to a local ophthalmologist in September 1988, complaining of painless, progressive loss of vision in both eyes over the previous 5 months. One week before our examination his visual acuity had been noted to be 20/40 right eye, and light perception left eye. A routine computerized axial tomographic scan (CT) of the brain was obtained and was stated to be normal. He was subsequently referred to the Dean A. McGee Eye Institute for further evaluation.

Neuro-ophthalmologic examination on September 19, 1988, revealed a best corrected visual acuity of 20/20 + 2 RE, and 20/400 LE. Pupils were 6.0 mm RE and 5.5 mm LE, with 3+ and equal reaction to light in both eyes. There was no afferent
pupillary defect noted. Color vision by Ishihara plates was 11.0/15 correct right eye, and 1.5/15 correct left eye. Tangent screen testing demonstrated a complete temporal hemianopsia RE, and a cecocentral scotoma LE, suggestive of a left junctional scotoma.

Funduscopic examination demonstrated temporal atrophy of the right optic nerve, and diffuse atrophy of the left. The macula, vessels, and periphery were normal bilaterally. The remainder of the neuro-ophthalmological examination was unremarkable.

Suspecting a left anterior chiasmal compression syndrome, an MRI of the brain was obtained that demonstrated a diffuse high intensity signal emanating from a thickened optic chiasm. The thickening was more prominent on the left with marked gadolinium enhancement (Fig. 1). A lumbar puncture was not performed. Additional history subsequently obtained from the patient re-
revealed a 30-pound weight gain over the past year. Also, when questioned specifically, the patient admitted to, but did not complain of, a 3-year history of polydipsia, and polyuria. In fact, he recalled a daily water consumption of 2-3 gallons. He denied any changes in hair distribution, hat or shoe size, or libido. The patient also denied headaches. Endocrinologic evaluation confirmed diabetes insipidus, hypotestosteronism, and hypothyroidism.

On October 21, 1988, the patient underwent a left frontal craniotomy. On gross inspection in situ, the chiasm was noted to be markedly thickened just posterior to the left optic nerve, and the left carotid artery was somewhat displaced. Several small biopsy specimens taken from the outer portion of the mass subsequently revealed "gliosis and chronic inflammation suggestive of germinoma" (Fig. 2). The mass was felt to be nonresectable and the patient recovered well from the procedure. Subsequently, the patient received 5,040 rad of total ventricular irradiation therapy over a period of 6 weeks, after which the chiasmal mass was felt to have almost completely resolved by MRI (Fig. 3).

On March 22, 1989, follow-up neuro-ophthalmologic examination revealed a best corrected visual acuity of 20/20 right eye and 20/400 left eye. Pupils were equal and reactive to light in both eyes, with a subtle afferent pupillary defect on the left eye. Tangent screen and Goldmann visual field testing revealed a right homonymous hemianopsia with a coexisting ceco-central scotoma LE. Funduscopic examination was unchanged. The remainder of the neuro-ophthalmological examination was normal. Symptoms of diabetes insipidus had cleared.

REVIEW OF THE LITERATURE

A review of the literature revealed 93 cases of primary suprasellar (ectopic) germinoma (Table 1). Suprasellar germinoma secondary to metastasis from a primary in the pineal body will not be discussed. Primary tumors of the pineal region comprise a separate and distinct entity and do not usually present with the neuroendocrinologic triad previously described. The patients ranged in age from 6 to 41 years with the average age being 14.9 years. The male/female ratio for the group as a whole was near 1:1, with 49 women and 44 men. Fig. 4 illustrates the frequency of this tumor at various ages. Of the 64 cases in which the presenting symptoms were specified by the authors, 56 (87.5%) had diabetes insipidus, 53 (82.8%) had visual changes (decreased acuity, diplopia, or visual field defects), and 36 (56%) had hypopituitarism (Fig. 5). Of the 61 patients who received irradiation therapy, 42 (69%) were reported to be surviving to date of the individual case report, and of all 93 case reports, no patient survived without irradiation therapy. The average amount of irradiation given was 5,255 rad over 5-6 weeks, with eight patients receiving spinal irradiation.

DISCUSSION

Pierce (9) and Ramsey (10) showed that the fine structure of the pineal and ectopic germinoma is identical to that of the classic testicular seminoma. Many theories have been proposed as to how these germ cell tumors find their way into the cranial vault (11-15). However, it seems that most authors now accept the proposal of Witschi, which
FIG. 3. T1-weighted MRI scans of the brain. Top: sagittal postinfusion (gadolinium), postirradiation, demonstrating near complete resolution of tumor with only residual gadolinium uptake. Bottom: coronal postinfusion, postirradiation, also demonstrating tumor resolution.

suggests that germ cells, which originate in the yolk sac endoderm and migrate widely throughout the embryo before localizing in the gonadal ridges, sometimes find their way into the head of the embryo (16). Simson et al. go further to state that the pineal, 3rd ventricle, and suprasellar regions seem to have in common the properties necessary to sequester and maintain aberrant germ cells (2). These cells may then produce neoplasms that are identical to germ cell tumors found elsewhere in the body.

The classification of intracranial germ cell tumors has not been formally unified. However, Takakura has recently categorized them into three types: germinoma (two-cell pattern pinealoma), mature teratoma (benign), and immature teratoma (malignant, including choriocarcinoma, endodermal sinus tumor, embryonal carcinoma, etc.) (4). Kageyama and Belsky have devised a classification that divides these tumors of the infundibul-suprasellar region into three types. One reaches the pituitary stalk and optic pathways after arising in the pineal region and metastasizing anteriorly. A second originates within the anterior part of the third ventricle and spreads to involve the optic nerves, chiasm, pituitary, and hypothalamus. The last type, located primarily extra-axially is the ectopic pinealoma of the chiasmal region (5). This separation is important, as patients with primary chiasmal and perichiasmal germinomas will often notice visual field defects and/or decreased visual acuity, and thus present initially to the ophthalmologist.

In 1961, Kageyama (8) was one of the first authors to describe the characteristic triad of diabetes insipidus, hypopituitarism, and visual disturbances that has since defined ectopic pinealoma as a distinct diagnostic entity (5,8). This same triad has subsequently been re-emphasized and documented by several other authors (1-4,6,7). Most authors report diabetes insipidus as the initial symptom, sometimes preceding the onset of visual disturbances and hypopituitarism by 2-3 years (1,2,7,17). Others, however, emphasize that symptoms of diabetes insipidus often go unrecognized, especially in children, and it is usually the visual disturbance that brings the patient to seek medical attention (1,2,8). Most authors agree with Kageyama when he states that this syndrome rarely presents with signs or symptoms indicating increased intracranial pressure (5,8).

Age distribution was studied in 70 cases of suprasellar germinomas by Camins and Mount in their literature review (17). They found that female subjects were more affected under the age of 10 years, while male subjects were more affected over the age of 20 years. Between the ages of 10 and 20 years, both sexes were affected equally. These findings are also supported by our literature review (Fig. 4). Takeuchi noted that this may suggest that the growth of suprasellar germinomas might be influenced by a sex factor (7). Interestingly, intracranial germ cell tumors are much more com-
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<td>Akamatsu (31)</td>
<td>Case 1</td>
<td>22/m</td>
<td>Dx. visual changes, hypopituitarism, headache</td>
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<td>Death, time not reported</td>
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<tr>
<td>Russell et al. (32)</td>
<td>Case 1</td>
<td>10/f</td>
<td>Dx. visual changes, hypopituitarism, headache</td>
<td>Incomplete resection</td>
<td>Death 13 mo later with metastases to spinal cord</td>
</tr>
<tr>
<td>Russell (33)</td>
<td>Case 1</td>
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<td>Dx. visual changes, hypopituitarism, headache</td>
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<td>Post-op death 1 mo later</td>
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<td>Horrax and Wyatt (34)</td>
<td>Case 1</td>
<td>12/m</td>
<td>Dx. visual changes, hypopituitarism, headache</td>
<td>Incomplete resection, 4,000 rads</td>
<td>Surviving 4 yr after diagnosis</td>
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<td>Troland and Brown (35)</td>
<td>Case 1</td>
<td>14/m</td>
<td>Headache, precocious puberty</td>
<td>Incomplete resection, 2,000 rad?</td>
<td>Surviving 2 yr after diagnosis</td>
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<td>Baker and Rucker (14)</td>
<td>Case 1</td>
<td>17/m</td>
<td>Dx. visual changes, hypopituitarism, headache</td>
<td>Biopsy, irradiation, amount unspecified</td>
<td>Death 1.5 yr after diagnosis with spinal cord metastases</td>
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<td>Rand and Lemmen (36)</td>
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<td>Kahn et al. (38)</td>
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<td>Case 2</td>
<td>41/m</td>
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<td>Dx. visual changes, hypopituitarism, headache</td>
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<td>Busch (41)</td>
<td>Case 1</td>
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<td>Biopsy and incomplete resection</td>
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<td>Héon and Scoville (42)</td>
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<td>Death 11 d post-op</td>
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<td>Death 5 yr later with renal insufficiency</td>
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<td>Biopsy and 5,000 rad</td>
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<td>Dx. visual changes, hypopituitarism, headaches</td>
<td>Biopsy and 5,000 rad</td>
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<td>Dx. headache</td>
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<td>Death 2 yr after initial presentation</td>
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<td>Dayan et al. (45)</td>
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<td>Hypopituitarism, headache</td>
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<td>Biopsy and irradiation, amount unspecified</td>
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<td>Ref.</td>
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<td>Age/Sex</td>
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<td>Treatment</td>
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<td>Biopsy and partial excision, 4,500 rad</td>
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<td>Biopsy and partial excision, total of 7,600 rad with recurrence</td>
<td>Death 5 yr later with widely infiltrative tumor</td>
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<td>13/f</td>
<td>Di. visual changes, hypopituitarism</td>
<td>Biopsy and partial excision, 6,000 rad</td>
<td>Death 1 yr later from residual suprasellar tumor</td>
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<td>12/m</td>
<td>Di. visual changes</td>
<td>Biopsy</td>
<td>Death 14 d post-op</td>
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<td>Di. visual changes, headaches</td>
<td>Biopsy and partial excision</td>
<td>Death 12 h post-op</td>
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<td>Ghatak et al. (4)</td>
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<td>14/f</td>
<td>Visual changes, hypopituitarism</td>
<td>Biopsy</td>
<td>Death 8 d post-op</td>
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<td>13/f</td>
<td>Hypopituitarism, headache</td>
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<td>Death 3 d after presentation</td>
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<td>14/m</td>
<td>Di. visual changes, headaches</td>
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<td>28/f</td>
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<td>Craniotomy without biopsy, 6,000 rad</td>
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<td>21/m</td>
<td>Di. visual changes, hypopituitarism, headaches</td>
<td>Biopsy and partial excision, 6,000 rad</td>
<td>Surviving 7 yr after presentation</td>
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<td>22/m</td>
<td>Di. visual changes, hypopituitarism, headaches</td>
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<td>15/m</td>
<td>Di. visual changes, hypopituitarism, headaches</td>
<td>Biopsy and 7,500 rad</td>
<td>Death 19 yr later, cause not specified</td>
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<td>28/m</td>
<td>Di. visual changes, hypopituitarism</td>
<td>Biopsy and 1,800 rad</td>
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<td>Biopsy and 840 rad</td>
<td>Death 1 yr post-op</td>
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<td>Case 4</td>
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<td>Visual changes</td>
<td>Biopsy and 3,200 rad</td>
<td>Death 7 mo post-op</td>
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<td>12/m</td>
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<td>Biopsy and 5,850 rad to head, 2,150 rad to spine</td>
<td>Death 6 mo post-op, cause not specified</td>
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<td>Case 6</td>
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<td>17/f</td>
<td>Di. visual changes, hypopituitarism</td>
<td>Biopsy</td>
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<td>13/f</td>
<td>Di. visual changes, hypopituitarism</td>
<td>Biopsy and 8,500 rad</td>
<td>Surviving 9 yr after diagnosis</td>
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<td>16/m</td>
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<td>Biopsy and 3,220 rad to head, 4,000 rad to spine</td>
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<td>Case 10</td>
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<td>18/f</td>
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<td>Death 6 mo post-op, cause not specified</td>
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<td>Case 11</td>
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<td>21/f</td>
<td>Di. visual changes, hypopituitarism</td>
<td>Biopsy, 3,920 rad</td>
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<td>Sung et al. (3)</td>
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<td>Total excision and 4,000 rad</td>
<td>Death 7 mo post-op with spinal metastases</td>
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<td>Biopsy and 5,500 rad</td>
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<td>11/f</td>
<td>Partial excision and 5,000 rad initially, 3,500 rad later with recurrence</td>
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<td>16/m</td>
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<td>Surviving 6 yr after diagnosis and 2 yr after therapy for spinal metastases</td>
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<td>Biopsy and 5,000 rad</td>
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continued
TABLE 1—(Continued)

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<td>Takeuchi et al. (7)</td>
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<td></td>
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<td>Diagnosis by CSF cytology, 3,000 to spine</td>
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<td>Craniotomy without biopsy, 4,500 rad; recurrence treated with chemotherapy</td>
<td>Surviving 12 yr after diagnosis; 1 yr after recurrence</td>
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</table>

* Diabetes insipidus.
* Decreased visual acuity, visual field defects, or diplopia.
* All irradiation therapy is to brain only unless further specified.

FIG. 4. Age and sex incidence of 93 patients with suprasellar germinoma. Black bars, male patients; striped bars, female patients.
FIG. 5. Preoperative signs and symptoms in 64 cases of primary suprasellar germinoma.

series (1969) of 57 craniopharyngiomas, only 8% of the cases were associated with diabetes insipidus (18). It therefore seems appropriate that diabetes insipidus of unexplained etiology should be carefully followed during the first and second decades of life for associated visual disturbances and hypopituitarism.

The optimal approach to diagnosis and treatment of intracranial germinomas has been a much debated topic in the literature. As early as 1950, Rao et al. emphasized the value of irradiation therapy in the treatment of ectopic pinealomas (20). In 1965, Rubin and Kramer published a review of 36 cases of ectopic pinealoma, re-emphasizing the radiocurable characteristics of these tumors (1). The remarkable sensitivity of intracranial germinoma to irradiation therapy, combined with the relatively high morbidity and mortality previously associated with craniotomy and biopsy, has prompted some authors to recommend a radiotherapeutic trial without biopsy when germinoma is suspected (1,2,8,20). Others, however, point out that initial biopsy of these lesions will permit more therapy decisions to be based on tissue diagnoses and reduce the frequency of potentially harmful radiotherapy trials (21). Also, recent advances in microsurgical and stereotactic techniques have significantly decreased the surgical morbidity and mortality previously associated with biopsy or resection of these lesions, and biopsy can usually be obtained without further visual loss (4,21).

The metastatic spread of intracranial germinoma through the spinal fluid to other spinal axis locations has not been well established (1,3,22,23). This has prompted some authors to recommend routine craniospinal radiotherapy for all patients with biopsy-proven germinomas, as well as those with clinical and radiographic evidence suggestive of germinoma (20,23). However, due to the low incidence of spinal metastases and the associated morbidity of irradiation to the spinal axis, most authors do not currently recommend such aggressive treatment (21,24). Camins and Mount recommend that patients be followed regularly with cerebrospinal fluid millipores, one before discharge, and then at yearly intervals. Accordingly, only patients with clinical evidence of subarachnoid metastasis or positive millipores should receive entire neuroaxis irradiation (17).

Currently, the most accepted recommendation for treatment of suprasellar germinoma is irradiation of the tumor field and entire ventricular system with a dose of 4,500–5,000 rad, given over a period of 5–6 weeks (1,3,21,24). Consideration must be given to the postoperative, postradiation management of the patient's endocrine function. Patients may need maintenance with hydrocortisone, testosterone, thyroid extract, as well as pitressin. Replacement therapy and continued management is important so that death from neuro-endocrine complications may be avoided.

The outcome of treatment for intracranial germ cell tumors differs according to the histologic type of tumor and the location, i.e., suprasellar versus pineal. Overall, the radiocurability of germinoma has produced an excellent survival outcome with most authors reporting a 10-year survival rate of 55–72% for all intracranial germinomas (pineal and suprasellar) (4,25). If one narrows this down to only suprasellar lesions, the rate improves to 84.4% at 10 years (26). These survival rates are from data over the past 20 years, with mortality and morbidity of surgical intervention being much higher during the initial 10-year period than in the following 10 years.

CONCLUSION

The inclusion of germinoma in the differential diagnoses of patients presenting with chiasmal field defects is extremely important. The characteristic triad of visual loss consistent with chiasmal compression, diabetes insipidus, and hypopituitarism define this presentation as a distinct diagnostic entity. Review of the literature suggests that diabetes insipidus is usually the initial symptom in suprasellar germinoma. However, in most instances this tumor occurs in the adolescent age
group, in whom symptoms of diabetes insipidus will often go unrecognized. Therefore, it is the ophthalmologist who will often see these patients first, when decreased vision brings them to seek medical attention.

Recognition of the neuroendocrinological triad, along with neuroradiological studies suggestive of an intrinsic chiasmal lesion, should suggest rapid confirmation of the diagnosis by craniotomy and biopsy. Early diagnosis and appropriate irradiation therapy may not only preserve vision, but life as well.

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Papillary Carcinoma of the Sphenoid Sinus Associated with Sphenoid Sinus Abscess Presenting as Cavernous Sinus Syndrome
A Case Report

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Primary carcinoma of the sphenoid sinus is a rare tumor that may present with dramatic neuro-ophthalmological symptoms and signs of which spheno-cavernous syndrome is the best known clinical entity. The most frequently encountered histological types of the sphenoid carcinomas are squamous cell carcinoma and papillary carcinoma, in decreasing order of frequency. In this article, a papillary carcinoma of the sphenoid sinus associated with sphenoid sinus abscess is presented. We are not aware of previously reported papillary carcinoma of the sphenoid sinus associated with sphenoid sinus abscess presenting as a cavernous sinus syndrome.

Key Words: Papillary carcinoma—Spheno-cavernous syndrome—Sphenoid sinus abscess.

Carcinoma of the sphenoid sinus, first described in 1891, is very rare, constituting only 0.3% of all sinus cancers (1–4). When it occurs, it is apt to produce neuro-ophthalmological symptoms and signs that are nonspecific until the sinus wall is penetrated. Specific neurological symptoms and signs of the sphenoid sinus carcinoma are characterized most commonly by the spheno-cavernous syndrome and less frequently by isolated sixth cranial nerve palsy and visual loss on the ipsilateral side (1,4–6).

CASE REPORT

A 31-year-old male patient was admitted to the Department of Neurosurgery with a 2-month history of diplopia and ptosis. He had also suffered from headaches and diminished visual acuity of the right eye.

On neurological examination, right and left pupils were 4 and 3 mm, respectively. The light reflex in the right eye was decreased. Visual acuity was 20/200 in the right eye and 20/20 in the left eye. He had complete ptosis and total ophthalmoplegia on the right side. There was anesthesia in the distribution of the first and second divisions of the right trigeminal nerve. Exophthalmos was found to be 18 mm (base 110) in the right eye. Nasopharyngeal examination was negative, and plain radiograms of the skull revealed a soft tissue density filling the sphenoid sinus and eroding the sella turcica. The other physical signs and laboratory findings were normal. Angiography showed significant displacement of the neighboring vessels. Computerized tomography revealed a massive tumor involvement.
PAPILLARY CARCINOMA OF THE SPHENOID SINUS

of the sphenoid sinus and posterior part of the ethmoid sinus with erosion of the right sphenoid wall and clinoid to spread into the right cavernous sinus. It also showed a hypodense area in the sphenoid sinus resembling an abscess (Fig. 1A and B).

Transnasal sphenoidotomy and biopsy were performed and 8 cc of gray-yellow abscess material was drained. Gram stain of this material showed huge amounts of polymorphonuclear leukocytes (PNL). Cultures were negative. Histopathological examination of the specimen revealed a papillary carcinoma (Fig. 2).

Antibiotic therapy was administered and irradiation of 5,000 rad was performed. Chemotherapy was given. He was alive 14 months after surgery.

DISCUSSION

Malignancies of the paranasal sinuses comprise between 0.2 and 2% of all human cancers (2–4,5,7,8). Frontal and sphenoid sinus carcinomas are quite rare, accounting for <1% of all paranasal sinus malignancies (2,3,5,7,8). The review of the reported cases showed that only four of 127 cases of paranasal sinus malignancies originated from the sphenoid sinus (1,7–9). Thus, carcinoma of the sphenoid sinus appears to constitute only 0.3% of all the paranasal sinus tumors (1,2,4). The cause of the low incidence of this tumor is unknown. It may be related to the paucity of the glandular element in its mucosa and to its location (9). The most frequent histological type is squamous cell carcinoma, followed by transitional cell carcinoma (5). Harbison et al. reported that only one of the 42 cases with sphenoid sinus carcinoma was papillary carcinoma (5).

All patients with sphenoid sinus carcinoma have neuro-ophthalmological symptoms and signs at the same time in their course. Spheno-cavernous syndrome is the most prevalent, consisting of involvement of the 3rd, 4th, 6th, and the 1st and 2nd divisions of the 5th cranial nerves (5,6). In addition, optic nerve palsy may be found frequently. This syndrome would suggest extension to the orbital apex and/or the cavernous sinus.

In our case, diagnosis was reached by direct

FIG. 1. (A and B) Axial enhanced computed tomograms, showing a soft tissue density mass arising the sphenoid sinus and extending into the region of the cavernous sinus, posterior ethmoid sinus, and sella turcica with bony destruction.

FIG. 2. (A and B) Histopathological appearances of the transnasal biopsy, showing the papillary carcinoma with inflammatory cells.
open biopsy, performed by the transnasal approach. It was difficult to diagnose this case because of the additional infection. Sphenoid sinus abscess is a rare condition. It may be the result of the superinfection of the sinus in our case.

We are not aware of previously reported papillary carcinoma of sphenoid sinus associated with sphenoid sinus abscess presenting as a sphenocavernous syndrome. Therefore, it must be noted that some sinus infections may be located on sphenoid sinus carcinomas.

REFERENCES

Oculographic Analysis of Acute Esotropia Secondary to a Thalamic Hemorrhage

Richard W. Hertle, M.D., and Don C. Bienfang, M.D.

We used electrooculography to study the saccadic velocities, smooth pursuit, and vestibular ocular reflex in a patient with an acute thalamic hemorrhage. Our findings confirmed what others have shown in that there were hypometric saccades contralateral to the lesion, impaired smooth pursuit ipsilaterally, and a preserved vestibular ocular reflex. In addition, we demonstrated an asymmetry with the contralateral eye being more affected. It is also shown that the “convergence” movements seen on attempted upgaze are typical of saccades and not vergence movements. A discussion of possible pathophysiologic mechanisms with review of other studies is presented.

Key Words: Thalamic hemorrhage—Saccades—Esotropia.

According to Walsh et al. (1), thalamic hemorrhage accounts for 25–35% of all cerebral hemorrhages diagnosed by computerized tomography. Fisher (2) localized 13% of intracerebral hemorrhages to the thalamus. Castaigne et al. (3) divided them into three categories based on topographical and vascular arrangements: unilateral paramedian, bilateral paramedian thalamic, and paramedian thalamopeduncular. They concluded that paramedian thalamic infarcts were rarely isolated. The mesencephalic gray matter, principal nucleus, and fibers of the third cranial nerve were affected in >75% of cases.

Symptoms from thalamic hemorrhage include altered levels of consciousness from hypersomnia to deep coma, behavioral disturbances, impaired memory, abnormal clonic or atheototic movements, speech and language dysfunction, and oculomotor disturbances (1–3).

Fisher (2) described the oculomotor signs as convergence spasm, paresis of horizontal and vertical gaze, downward deviation, and miotic, poorly reactive pupils. Other signs may include ptosis on the involved side and a skew deviation during the acute stage (3,4). Full vestibularly driven eye movements, decreased convergence and Bell’s phenomenon, and “nystagmoid jerks” are also reported (2). Brigell et al. (4) reported a case with transient opsoclonus, hypometric saccades contralateral to the lesion, and low gain pursuit ipsilaterally. Wall et al. (5) described three cases of rostral midbrain and thalamic infarction with paresis of vertical gaze, absent convergence, anisocoria, variable response to vestibularly driven eye movements vertically, and decreased pupillary responses to both light and near. Ochs et al. (6) found opposed adducting saccades on attempted upgaze in their eye movement recordings of a patient with a thalamic tumor extending to the midbrain causing Paurinaud’s syndrome.
The associated findings of impaired vertical gaze, convergence spasm (acute esotropia), convergence retraction nystagmus with attempted up-gaze, and miotic poorly reactive pupils constitute Parinaud’s syndrome described in 1883 and later elaborated on by Koerber in 1903 (7). Investigations into the pathophysiology of the oculomotor disturbances by Gay et al. (8), using electromyography, showed cocontraction of all muscles tested during retraction. There was an abrupt onset of electromyographic activity consistent with a saccadic discharge (8). Esslen and Papst in 1961 found excessive tonic electromyographic activity resembling spastic innervation from disinhibition of the superior and inferior recti motor nuclei as well as faulty recruitment of motor units in the superior rectus. Daroff and Hoyt (8) found no evidence of horizontal vergence movements during retraction, but a pattern of horizontal dysjunctive saccades of unequal amplitude towards the center.

We recently had the opportunity to perform oculography on a patient with an acute thalamic hemorrhage. The results with subsequent discussion are reported below.

CASE REPORT

An 84-year-old right-handed white woman had a history of coronary artery disease and hypertension. She noticed the acute onset of perioral numbness that spread to both sides of her body. At the same time, she noticed severe right-sided headache, diplopia, dizziness, nausea, and left-sided weakness. She fell while attempting to rise out of a chair and hit her left temple. She had slurred speech while calling her daughter on the phone and was brought to the emergency room.

Past medical history was significant for a myocardial infarction 6 years previously and cataracts.

Physical examination in the emergency room revealed an 84-year-old white woman in no acute distress. Her blood pressure was 180/85, pulse 72 with premature ventricular contractions (PVCs), and respirations 14-18/minute regular and unlabored. There was a fresh 3.0-cm contusion over her left temple. Neurological exam showed orientation to person and place, decreased attention, normal memory, and dysarthria, and cranial nerve examination showed mildly decreased sensation of the left face and central facial weakness. She had a dense flacid left hemiparesis, decreased sensation on the left, and upgoing toes bilaterally. Her oculomotor examination at the time of admission was reported to show miotic poorly reactive pupils to direct light, convergence spasm, limitation of up-gaze with convergence on attempt, and increasing convergence on down-gaze.

The patient’s medical status prohibited examination in the ophthalmology office at anytime during the hospitalization, and some tests, such as caloric, were felt to be too strenuous for her. Ophthalmologic consultation in the intensive care unit 1 day after admission revealed 2.5-mm pupils poorly reactive to both direct and consensual stimulation. There was a 30-40 prism diopter esotropia, which increased on down-gaze simulating a V-pattern. This was measured with the patient fixing a target at 15 ft while performing both the cover-uncover test and the simultaneous prism-cover test. There seemed to be a preference for the right eye, but she freely alternated fixation. There was limitation of up-gaze to ~10° with “convergence” movements on attempt. The patient responded to commands with limitation of abduction bilaterally. The right abducted to 60% of normal and the left to only 10% of normal. There appeared to be sluggish convergence. Vestibularly driven eye movements showed full horizontal versions. Optokinetic movements using an optokinetic drum showed occasional convergent movements with downgoing stripes. There was also an intermittent, small amplitude, high frequency, rotary nystagmus that increased in intensity on up-gaze. The remainder of her ocular examination was normal except for bilateral cataracts and retinal vascular disease consistent with arteriolosclerosis and hypertension (Fig. 1A–F).

Over the course of her hospital stay, all her physical findings improved slowly and steadily.

Computerized tomography (CT) at the time of admission showed a right midbrain hemorrhage extending rostral to the thalamus and into the mid-brain. There was no midline shift of intraventricular or subarachnoid hemorrhage (Fig. 2A and B).

RECORDING METHOD

A Traucoustics model #BV-275 saccadic velocity recorder that has DC coupling and a bandwidth of 0-75 Hz was used. This unit is portable enough to be used at the bedside, which is where the recordings were done. Electrodes were placed both temporally and nasally as well as a ground on the forehead. The stimulus apparatus was a Traucoustics model #RV-259 Digital Light Bar. This contains light-emitting diodes placed so as to subtend visual angles of 5°, 10°, and 15° both horizontally and vertically 1.5 m from the patient. The patient’s head was held steady as she was asked to look at the test lights. The protocol involved both binocu-
FIG. 1. Oculomotor exam performed one day after the patient’s cerebrovascular accident. Photographs were taken at the patient’s bedside. A: Patient with gaze straight ahead showing both eyes turned in. Acute esotropia. B: Patient asked to look right with limitation of gaze more evident in abducting right eye. C: Patient asked to look left with limitation of gaze more evident in the abducting left eye. D: Patient asked to look up with obvious limitation of vertical gaze. E: Patient asked to look down showing some limitation but less than in upgaze and increasing esotropia. F: Vestibularly driven eye movements to the left showing full versions in abduction. G: Vestibularly driven eye movements to the right showing full versions in abduction.

RESULTS

Electrooculography was performed on the 10th day after the patient’s cerebrovascular accident.

With the right eye, viewing attempts were made between 15° right and 15° left. The first and third abductive saccades had a gain of close to 1. The first and second adductive saccades were hypometric with no corrective saccade made after the first adductive saccade and a small glissadic-like movement followed by a corrective saccade made after the second. Five millimeters equals 2.5° for calibration on the position trace. Average velocities were 244°/s in abduction and 229°/s in adduction calculated for all 15° saccades made during this paradigm (Fig. 3A).

With the left eye viewing center to 15° right (the patient was unable to voluntarily abduct very far past the midline), there was hypometria in both directions but more severe in abduction compared to adduction. The three abducting saccades require more saccades to arrive at the target. The calibration of this position trace is 1.0 mm equals 1.0°. Averages of adding saccadic velocities were 193°/s and abducting velocities were 71°/s calculated for all voluntary or involuntary saccades of 5° made during this paradigm.

With both eyes viewing and attempted upgaze to 10°, bilateral convergence was seen. This consisted of an abducting saccade in both the right and left eyes. The average velocities of these saccades were 45°/s in the left and 42°/s in the right as calculated for all saccades measuring 5° during this paradigm (Fig. 3C).

Because of lack of sensitivity inherent in electrooculography (EOG), the involuntary oscillations are not fully appreciated in the recording, but there can be seen brief periods of the horizontal component of the oscillation on attempted upgaze. They appear pendular with a jerk component. The position calibration is 1.0 mm equals 1.0° (Fig. 3D).

Smooth pursuit showed low gain in both directions. With the right eye viewing, the gain to the right was 0 as evidenced by total saccadic replacement. Gain to the left, although close to 0, did show evidence of some intact pursuit (Fig. 3E). The vestibular ocular reflex was relatively well preserved with the right eye fixating (Fig. 3F).

DISCUSSION

The most obvious disturbance in this patient is the esotropia. In hysterical individuals, conver-
Convergence is usually associated with accommodation (7). Convergence has been described in patients with basilar inflammation following head trauma and as a consequence of hyperopia with a high AC/A ratio (7). Mott and Schafer, in 1960, produced convergence in eyes of monkeys by bilateral stimulation of corresponding areas of frontal lobes or occipital eye fields. Jampel, in 1959, elicited asymmetric convergence movements associated with variable miosis and accommodation with unilateral stimulation of points in the primate’s occipital cortex. In primates, Pasik et al. (9,10) obtained adduction of the ipsilateral eye from an area of the tegmentum lying ventrolaterally at the rostral end of the midbrain. Both convergence and downward deviation of the eyes were obtained by stimulation of the posterior sensory thalamus 13 mm lateral to the midline. In humans, Moster and Hoenig (11) reported a case of intermittent “convergence spasm” secondary to metabolic encephalopathy, and Selhorst (12) reported two patients with acute esotropia secondary to thalamic hemorrhage.

From the preceding animal work and human observation, several areas of cortical and subcortical stimulation could produce convergence. Patients with acquired convergence have lesions that are destructive and not thought to be a cause of excessive stimulation. Selhorst (12) postulated, in his two patients, loss of supranuclear inhibition of convergence to be responsible for the acute esotropia. In studying the convergence system in humans, Schor and Cuiffreda (13) postulates a constant tonic vergence system that operates independently of both accommodative and proximal vergence. It functions to align the eyes from an anatomical position of rest with a possible role in active convergence (13). In our patient, the neural integration between the different types of vergence may be disrupted resulting in an excess of convergence. The associated miosis makes a large contribution from the accommodative component seem highly likely.

We know that there is constant neuromuscular activity in the extraocular muscles that is their “tone” and that the only time no recordable activity is found in an inhibited antagonist muscle palsy manifest as an imbalance of tone or spasticity (14). Because our patient’s esotropia could be overcome by vestibularly driven eye movements, it may be a form of pseudo CN VI palsy (15).

Impaired vertical gaze is seen in a variety of supranuclear, nuclear, and infranuclear disorders. This seems always to be the result of a bilateral lesion, either directly or involving a decussation. The important areas are the posterior commissure, the rostral interstitial nucleus of the medial longitudinal fasciculus, the rostral medial longitudinal fasciculus, and the interstitial nucleus of Cajal (9,10,16,17).

In patients with thalamic hemorrhage, Brigell et al. (4), Hirose et al. (18), and Masdeau et al. (15) reported hypometric saccades contralateral to the side of the lesion. They postulate that interruption of direct synaptic connections from the prefrontal cortex through the thalamus, specifically the “transthalamic bundle,” simulate a decorticate state for saccadic activity. Monkey studies have shown similar abnormalities after decortication (19). It is interesting that we found the contralateral eye to be more affected. This was true in Selhorst’s (12) two cases of acute esotropia secondary to thalamic hemorrhage. One could postulate both contralat-

FIG. 2. CT scan of the head on admission. A: Hemorrhage in the right posterior thalamus. B: Higher magnification in the coronal plane showing hemorrhage in the posterior medial thalamus and midbrain.
FIG. 3. Oculomotor recordings completed 10 days after the patient's cerebrovascular accident. Velocities were either 40°/s/cm or 200°/s/cm as marked on each recording sample. Calibration for position was 5.0 mm equals 2.5° or 3.0° as marked on the photograph and described in the text. Rightward eye movements are up and leftward are down on each of the position traces. A: Right eye (OD) viewing with saccades attempted between 15° right and 15° left showing hypometria in both abduction and adduction but greater in adduction (contralateral to the lesion). See text. B: Left eye (OS) viewing center to 15° right (patient could not voluntarily abduct much beyond midline) again with hypometria in both directions with a more profound difference between abuction and adduction. It is worse in abduction, contralateral to the lesion. C: Saccades made between center and 10° up with representative velocity tracings from both eyes. Dysjunctive movements with saccadic characteristics are evident. These are convergent and correlate with the clinical appearance of convergence on attempted upgaze (arrows). D: Position traces of same saccades between 10° up and center showing bilateral adduction saccades and oscillations. See text for details. E: Right eye smooth pursuit was tested to a hand-held target moving in a sinusoidal pattern at ~5-10°/s with the head held stationary. This shows symmetrical low gain pursuit at almost 0 gain but occasional pursuit was seen to the left (contralateral to the lesion). F: Vestibular ocular reflex with the OD fixating a central target. This shows a well-preserved reflex with almost a gain of 1.0.

Our patient also had impaired smooth pursuit. It was not as easy to see the asymmetry in our patient as it was in the patients reported by Hirose et al. (18) and Brigell et al. (4). Pursuit was more impaired ipsilateral to the lesion. Although not as well understood as the saccadic system, interruption of descending fibers from the inferior parietal lobe or peristriate cortex in and around the thalamus could interfere with smooth pursuit. Leigh and Zee (14) say that low gain pursuit can be attributed to large lesions of the human parietal lobe and the impairment is ipsilateral.

The convergence retraction seen in our patient and those with Parinaud's syndrome is not a true convergence movement but either a cocontraction of all extraocular muscles or impaired saccadic dynamics (6-8).

Hypotheses to explain these movements include a "diffuse spread" of voluntary impulses, in which the medial rectus, being the largest, caused convergence; isolated medial recti activity; and an exaggerated stretch reflex (6-8).

Ochs et al. (6) have shown the fine structure of these eye movements and proposed a more physiologic explanation. These opposed adducting saccades are the result of a normal dynamic overshoot mechanism operating at high gain in the contralateral eye. This explains their observed asynchronous initiation and the convergent movement. The normal lower level brain stem is modified by high-gain signals. In summary, an upward eye movement attempt is followed by a contralateral directed conjugated saccadic impulse that is modified by a supranuclear high-gain instability to the contralateral eye, causing a large dynamic overshoot manifesting clinically as opposed adducting saccades or "convergence" (6).

The variety of oculomotor abnormalities seen with thalamic hemorrhage imply involvement of tracts going through the thalamus, near the thalamus, or coincidental involvement of nearby structures. As Castaigne et al. (3) found in their study of...
28 patients, at least 50% had oculomotor abnormalities. Paramedian thalamic or midbrain infarcts are rarely isolated, and they often involved the mesencephalic grey and third cranial nerve. These patterns are inconstant and dependent on variation in blood supply and etiology of destruction. Oculomotor findings consisted of parts of or the complete Parinaud's syndrome plus variable third nerve involvement. Attention to these deficits can be clues to the diagnosis of thalamic hemorrhage.

REFERENCES

Compensatory Head Tilt in Upbeating Nystagmus

Jorge C. Kattah, M.D., and T. Forcht Dagi, M.D.

Upbeating nystagmus has been described in lesions of the posterior fossa. We report a case of upbeating nystagmus accompanying a focal hemorrhagic lesion of the left brachium conjunctivum, the anterior vermis, and the anterior superior left cerebellar hemisphere. The nystagmus was suppressed by a contralateral head tilt. We postulate that in this instance, acquired central nystagmus was inhibited by the otolith-ocular reflex.

Key Words: Upbeating nystagmus—Focal hemorrhagic lesion—Compensatory head tilt.

The purpose of this article is to describe a patient who displayed primary position upbeat nystagmus that was completely suppressed by rightward head tilt. All other head positions were associated with large-amplitude upbeat nystagmus. He initially appeared with headaches, ataxia, oscillopsia, and positional vomiting, all of sudden onset. A focal hemorrhagic lesion was identified involving the left brachium conjunctivum, the anterior vermis, and the anterior superior cerebellar hemisphere. Recovery followed resection of the lesion. Our case serves as an example of an acquired central nystagmus inhibited by a compensatory head tilt.

CASE REPORT

A 14-year-old boy experienced the acute onset of bifrontal headache, nausea, positional vomiting, inability to stand, left hand clumsiness, oscillopsia, and a spontaneous 30° right head tilt. The neurologic examination demonstrated an inability to stand with the feet together or to perform a tandem gait. In addition, there was marked dysmetria of the left arm and a bilaterally positive Babinski sign. Neuroophthalmological examination revealed primary positional upbeating nystagmus in all head positions except a rightward head tilt, through which it was suppressed. Standard positional maneuvers provoked either vomiting or retching. Additional neuroophthalmological findings included saccadic horizontal and upward pursuit and occasional hypometric horizontal saccades. The patient showed no vertical deviation that could be demonstrated with red glass, screen cover, and Maddox rod testing. The remainder of the examination results were unremarkable.

Multiplanar unenhanced computed tomography demonstrated a round lesion of increased signal density involving the left anterior superior cerebellar hemisphere, part of the superior cerebellar ver-
mis, and the left brachium conjunctivum (Fig. 1). A discrete mass effect was noted, with moderate displacement of the fourth ventricle and the left lateral midbrain. The mass was not enhanced on i.v. contrast injection. Cerebral angiography confirmed the presence of an avascular mass in the left superior cerebellar hemisphere. Surgical exploration of the left cerebellar hemisphere disclosed a hemorrhagic cavity in this location whose contents on microscopic examination consisted of an organizing clot. Following drainage of the clot, the patient’s condition gradually improved, with disappearance of the nystagmus, ataxia, and other abnormalities. The cause of the hemorrhage was not definitively elucidated; we postulate rupture of a small arteriovenous malformation or a cavernous hemangioma in the cerebellar cortex.

**EYE MOVEMENT RECORDINGS**

Conventional electrooculogram recording of horizontal and vertical eye movements was carried out shortly after admission. The testing sequence included visual fixation and effects of static head tilt on fixation, horizontal and vertical saccades, horizontal and vertical pursuit, vestibulo-ocular reflexes, and standard positional testing.

Primary gaze upbeat nystagmus was maintained as long as the head was straight or tilted to the left (Fig. 2) but was suppressed by a rightward head tilt (Table 1). Lateral head tilt angles of 5° or more to the right abolished the nystagmus completely. The nystagmus velocity and frequency were maximal when the patient looked at a near target and the head was straight. Nystagmus was unchanged in the extreme right and left gaze. It decreased in downgaze and increased in amplitude in upgaze and in the supine position. Nystagmus became downbeating in the hanging head (HH) and the left HH (hyperextended) positions and was not present in the right HH position. It disappeared with simultaneous trunk and head hyperflexion, regardless of the angle of lateral head rotation. Body rotation with the head held still had no effect.

**FIG. 1.** Computed tomography scan demonstrates a round lesion of increased signal density involving the left anterior superior cerebellar hemisphere, part of the superior cerebellar vermis, and the left brachium conjunctivum.
FIG. 2. A conventional electro-oculogram recording of horizontal and vertical eye movements shows primary gaze upbeat nystagmus was maintained as long as the head was straight or tilted to the left but was suppressed by a rightward head tilt.

on the nystagmus. Horizontal saccades of $\pm 15^\circ$ amplitude showed normal velocity and latency, but 20% of them were hypometric. Horizontal pursuit was saccadic with a gain of 0.7. While upward pursuit was saccadic, downward pursuit was normal. Rotationally induced nystagmus, with the patient looking at a fixation target, could not be suppressed with either clockwise or counterclockwise directions. The gain of the optokinetic nystagmus was decreased in both the clockwise (0.15) and counterclockwise (0.3) directions (normal value, 0.6). Attempts at caloric testing induced retching and precluded the recording of eye movements.

DISCUSSION

Head tilt occurs in a variety of clinical conditions and with the exclusion of local musculoskeletal conditions involving the neck may be classified as ocular, labyrinthine, or central in origin. Ocularly induced head tilts are by far the most frequent and typically result from superior oblique muscle paresis. Here, the head is tilted in the direction opposite to the paretic muscle to avoid diplopia (1). Compensatory tilts and turns may be induced by other vertical extraocular muscle palsies (2) and may also accompany congenital nystagmus and
Peripheral labyrinthine lesions may be signalled by head tilt ipsilateral to the affected ear (7,8). Both the labyrinth and the brain stem lesions have been imputed in the so-called "ocular tilt reaction" (OTR), a synkinesis characterized by head tilt and vertical divergence and torsion of the eyes (9).

Experimental animal data and clinical observations in man suggest that OTR may result from midbrain lesions at the level of the interstitial nucleus of Cajal (10,11) that disturb the tonic otolithic-vestibular regulatory mechanism that controls vertical ocular alignment and head posture. The OTR may be either paroxysmal or sustained, with a head tilt ipsilateral to the lesion (9-12). Likewise, medullary lesions may be associated with OTR (12). Finally, cerebellar mass lesions also result in head tilt. The direction is unpredictable, and the mechanism is not defined. The head may tilt toward or away from the lesion (8). We concluded that our patient probably had a voluntary compensatory head tilt away from the lesion that curbed the oscillopsia. This head tilt stimulated a normal otolith-ocular reflex with nystagmus suppression. Additional inhibitory signals from the neck could have contributed to nystagmus suppression.

The abnormal head postures that develop in torticolis and other dystonic conditions are different. They are distinguished from other attitudinal abnormalities of the head by the rigidity. They are generally held to be primary rather than compensatory. Furthermore, eye movements are generally normal in dystonic conditions with the exception of the oculogyric crises induced by phenothiazines and other neurotropic agents.

Primary position upbeat nystagmus has been described with lesions at different levels of the brain stem. The majority of cases studied pathologically involved the pontomedullary junction (13) and the medullary nuclear complex of a recently described pathway that mediates the vertical vestibulocular reflex, the ventral tegmental pathway, may explain most of the cases with medullary and caudal pontine lesions (15). Lesions affecting the brachium conjunctivum, similar to that seen in our patient, have been described previously (16,17). In one instance, upbeat nystagmus was associated with contrapulsion of the eyes. Finally, the involvement of the anterior superior cerebellar vermis, seen in our case and previously described in isolated lesions in this location (15), may have contributed to the development of upbeat nystagmus. Thus, lesions affecting the two proposed projections from the superior vestibular nucleus, namely, the brachium conjunctivum or the ventral tegmental pathway (16), may be responsible for the generation of this type of nystagmus.

The mechanism of imbalance in ocular position that accounts for upbeat nystagmus has not been completely elucidated. Defective pursuit tone is postulated as a likely possibility (19,20). Alternative explanations include defective input from the anterior semicircular canals resulting in defective upward vestibulo-ocular reflex (17). Because this pathway projects to the oculomotor neurons via the brachium conjunctivum, a lesion in this structure can potentially result in upbeat nystagmus. Although the brachium conjunctivum was partially involved in our patient, there was no component of rotatory nystagmus. This suggests that the operant mechanism involved factors other than the pure interruption of efferents from the anterior semicircular canals (21). The variability in nystagmus velocity, frequency, amplitude, and in some cases direction that we encountered in our patient has been noted previously (22) and suggests that primary upbeat nystagmus may be modified by head position. Static tilt suppression of upbeat nystagmus has been previously reported on only one occasion, to our knowledge (22). While failing to explain the mechanism of upbeat nystagmus, static tilt suppression illustrates how otolithic stimulation may modify primary position central nystagmus. As a compensatory maneuver, static tilt suppression seems to improve visual acuity in some patients with congenital nystagmus and most patients with spasmus nutans and was a valuable spontaneous factor in our patient.

REFERENCES

COMPENSATORY HEAD TILT IN UPBEATING NYSTAGMUS


Congenital Double Elevator Palsy in Identical Twins


A left-sided double elevator palsy in identical twins born prematurely is presented, the first such report in twins. The aetiology of this condition is considered, and given the preservation of Bell's phenomenon and the absence of hypotropia in the primary position, the possibility that this may represent a supranuclear defect is discussed. Key Words: Congenital double elevator palsy—Twins prematurity.

Double elevator palsy, characterised by the limitation of elevation of one eye, is due to weakness of the muscles of elevation and may be congenital or acquired. This diagnosis is made following the exclusion of conditions such as dysthyroid eye disease, cellulitis, orbital blow-out fracture, third nerve palsy, poststrabismus surgery, trauma resulting in disinsertion of an extra ocular muscle, and various conditions associated with rigid muscles, fascia, or adjacent tissues.

CASE REPORT

Identical male white twins B.M. and A.M. were born at 32 weeks gestation and weighed at birth 1,415 g and 1,620 g, respectively. B.M. was a forceps delivery and required resuscitation and ventilation. A.M. was a breech delivery and required oxygen for 2 h. Both were jaundiced and received phototherapy for 3 days, and calcium supplements were given for hypocalcaemia. They were discharged from hospital on the 28th day of life.

Eye examinations began 3 weeks after birth. Both developed stage I acute retinopathy of prematurity, which subsequently resolved. At 18 months of age, limited elevation of the left eye of each child was observed. There was no deviation in the primary position and, in particular, no hypotropia. Other ocular movements were full, and Bell's phenomenon was preserved. A diagnosis of double elevator palsy was made. Neither had an anomalous head position. The eyelids were normal. Pupillary reactions were normal. Cycloplegic refraction at this time was: B.M.—right, −0.50 DS with +1.5 DC at 90 left, −0.50 DS with +1.0 DC at 90; A.M.—right, +0.50 DS, left, 0.00 with 0.50 DC at 90. No retinal abnormality was observed.
CONGENITAL DOUBLE ELEVATOR PALSY

FIG. 1. The upper sequence of twin A.M. and the lower shows twin B.M. (A) On upgaze, there is a limitation of elevation of the left eye. (B) Both eyes are straight in the primary position. (C) Defective elevation in adduction of the left eye. (D) Defective elevation in abduction of the left eye.

DISCUSSION

Double elevator palsy is an uncommon disorder of ocular motility, and this is the first report of its occurrence in twins. It is interesting to note that the twins were identical and that elevation of the left eye was limited in each. Typically, in this condition, the superior rectus and the inferior oblique muscles are variably affected (1), resulting in limitation of elevation above the horizontal plane in all positions of gaze. In addition, in some cases, there may be ptosis due to weakness of the levator muscle or pseudoptosis consequent upon hypertropia (2).

It would appear from the literature that although congenital defects of vertical gaze are uncommon, the most frequent is double elevator palsy. A classification has recently been suggested on the basis of eye movements, whether one or both eyes is affected, anomalous head posture, and binocularity (3), although it had been classified as long ago as 1943 into 3 groups according to binocularity and fixation by the nonparetic or paretic eye (4).

The clinical identification of double elevator palsy is important, as its management is different from other mechanisms of upgaze restriction, such as blow-out fracture, dysthyroid eye disease, and Brown’s syndrome. Clearly, only the last could be considered here. Treatment is generally not indicated, but in the presence of either a vertical deviation in the primary position or an anomalous head posture, the Knapp procedure has been performed (5).

As mentioned, double elevator palsy may be acquired and Jampel and Fells (6) in a series of eight patients, suggested that this could be due to a discrete lesion in the pretectum of the midbrain as a consequence of an occlusion of part of the microvasculature supplying that region of the brain. Lessel (7) reported a patient who developed a double elevator palsy in his left eye and was found at autopsy to have a metastatic tumour in the right side of the pretectum. In Rosner’s opinion (1), the disorder was best explained by a nuclear lesion involving the rostral area of the third nucleus.

Double elevator palsy can be congenital, and this is almost certainly so in our cases. These twins were enrolled in a survey of retinopathy of prema-
turity and had been examined on over eight occasions before the ocular motility problem was identified. Nevertheless, as emphasis had been directed toward the retinal problem in previous examinations, we consider it likely that the condition had existed from birth. The aetiology of congenital double elevator palsy is obscure, especially if one accepts that within the oculomotor nucleus (a) the fibres to the superior rectus are crossed, (b) those to the inferior oblique are not, and (c) that both muscles are innervated by separate branches of the oculomotor nerve (8). The clinical picture of the disorder can be simulated by a long-standing superior rectus palsy, as this muscle can elevate the eye without the aid of the inferior oblique. It has been reported in at least one instance of double elevator palsy that the electromyogram of the inferior oblique has been normal (9). In addition, structural anomalies of the extraocular muscles have been shown to be responsible. McNeer and Jampolsky (10) described an anomalous insertion of an extra band of the inferior rectus muscle, which produced the clinical picture of a double elevator palsy. This was confirmed by a forced duction test and surgical exploration. The presence of Bell's phenomenon in our twins thus would appear to rule out not only a mechanical aetiology, but also nuclear and infranuclear causes. It is interesting to note that neonatal hypoxia has been implicated in the aetiology of double elevator palsy, with four of five patients in one recently reported series having some form of hypoxia (5). This appears to be an unlikely explanation in this case, due to the symmetry of the lesions in each twin.

We have reported the finding of a left-sided double elevator palsy in identical twin boys. In the primary position, there was no ocular deviation, and Bell's phenomenon was present. We conclude that in these two children, the cause of double elevator palsy was supranuclear in origin, and the possibility of an inherited defect in this family cannot be excluded.

REFERENCES

Bilateral Trochlear Nerve Palsies from a Brainstem Hematoma

Hisao Tachibana, M.D., Osamu Mimura, M.D., Mitsuo Shiomi, M.D., and Tadatsugu Oono, M.D.

We present a case of bilateral superior oblique palsies after a spontaneous brainstem hematoma. A computerized tomographic scan of the brain revealed a high-density mass lesion consistent with bleeding in the area caudal to the inferior colliculi, where the trochlear nerves decussate and exit the dorsal brainstem. Subsequent studies showed resolution of the density and persistent failure to enhance. Bilateral trochlear nerve palsies due to the nontraumatic brainstem bleeding are extremely rare.

Key Words: Bilateral trochlear nerve palsies—CT scan—Brainstem bleeding.

Bilateral superior oblique palsies are usually congenital or the consequence of closed head trauma (1–7). Other causes are extremely rare. We describe a patient with bilateral trochlear nerve palsies after bleeding into the lower midbrain.

CASE REPORT

A 60-year-old man suddenly collapsed while working outdoors and then lost consciousness. He was taken to an emergency hospital. Computerized tomographic (CT) scan of the brain without contrast (Fig. 1) revealed bilateral high-density lesions in the midbrain tegmentum, predominantly on the left side, at the level of the inferior colliculi. The patient was treated conservatively. His level of consciousness gradually improved during 2 h and he began to complain of double vision.

Twelve days after onset he was transferred to Tanaka Hospital for detailed examination and treatment. He had neither a previous history of neurological disease nor risk factors for stroke, such as hypertension, diabetes mellitus, or coronary arterial disease.

On admission, his blood pressure was 140/72 mm Hg and the results of a general examination were unremarkable. Neurological examination revealed mild dysarthria, mild left-sided hemiparesis with normoactive stretch reflexes, left-sided hemiataxia, and an ataxic gait. The patient had a right esotropia and reported torsional and vertical diplopia, which became worse in downgaze. Ductions and versions were not limited in any direction.

His ocular motility examination was remarkable for a 24-prism-diopter esotropia and 4-prism-diopter left hypertropia in the primary position with an amblyoscope. Excyclotorsion exceeding 30° was noted using vertically linear fusion.
targets. The degree of exocycletorsion increased in a downgaze. A Hess screen confirmed a V-pattern esotropia in excess of 25 prism diopters (Fig. 2).

Investigations including erythrocyte sedimentation rate, complete blood count, renal function and electrolytes, plasma glucose and lipid level, tests of bleeding and clotting, urinalysis, occult blood for stool, electrocardiogram, and skull and chest X-ray films showed no abnormalities. Liver function tests initially showed moderate abnormalities, although 1 month later liver function was normal.

A repeat CT scan on day 12 showed complete resolution of the high-density mass and with contrast showed no contrast enhancement. Subsequent studies showed persistent failure to enhance.

With conservative treatment, his double vision and other neurological signs and symptoms grad-

FIG. 1. Brain CT scan demonstrating a high density mass in the midbrain tegmentum at the level of the inferior colliculi with extension to the left ventral side, and ventricular enlargement.

FIG. 2. Results of Hess screen examination demonstrating a V-pattern esotropia.
ually improved and ceased to be evident by 6 months.

**DISCUSSION**

The patient suddenly experienced brainstem bleeding although he had neither a history of hypertension nor hemorrhagic tendency.

Recently, Mangiardi and Epstein (8) classified brainstem bleeding into two forms and stressed the distinction between brainstem "hematoma" and "hemorrhage." Subependymal hematoma is a focal, compressive lesion that displaces rather than destroys brain tissue. It occurs in the younger age group and causes neurological deficits that are often partially reversible. On the other hand, hypertensive brainstem hemorrhage usually causes a diffuse lesion occurring in old age and is most often associated with profound, irreversible neurological deficits that are often fatal. On the basis of the clinical findings, our patient's lesion may have been a brainstem hematoma. These often result from bleeding from a cryptic brainstem vascular malformation, usually telangiectasia (8). However, the patient was older than cases reported in the literature (8). Therefore, the etiology of his bleeding remains unclear.

The incidence of trochlear nerve palsy is relatively low. In a retrospective analysis of 1,000 cases of paralysis of cranial nerves III, IV, and VI, there were 172 patients (17.2%) with trochlear palsy (4). Of these, only 13 (7.6%) were bilateral. Furthermore, the incidence of bilateral trochlear nerve palsies in other studies has been noted to be between 7.7 and 17.5% of cases with only trochlear nerve palsy (1-3,6). Most of the causes are head injury, as in traffic accidents. Some authors have reported myasthenia gravis (5,7), polyneuropathy (7), tumor (9), and neurosurgical complications (10) as causes. However, there are few reports of bilateral trochlear nerve palsies after nontraumatic brainstem bleeding, although unilateral trochlear nerve palsy from midbrain hemorrhage has been reported (11,12).

The nucleus of the trochlear nerve is located at the level of the interior colliculus and is ventral to the cerebral aqueduct. The fibers leave the nucleus and pass caudally and superiorly to the anterior medullary decussation just above the fourth ventricle. At this site, a single lesion may impair both nerve functions.

The most common cause of bilateral trochlear nerve palsies is a contrecoup injury to the decussation region after frontal head injury (2,3).

Murray and Ajax (9) reported a case with bilateral trochlear nerve palsies due to a metastatic adenocarcinoma localized in the anterior cerebellar vermis. They speculated that the lesion compressed the area caudal to the inferior colliculi where the fourth nerves decussate and exit the dorsal brainstem.

In our case, the hematoma was located in the dorsal brainstem, the area of the trochlear nerve nucleus and decussation, and directly caused bilateral trochlear nerve injury.

**REFERENCES**

Dissociated Vertical Deviation in a Patient with Duane’s Retraction Syndrome

Steve Rimmer, M.D., and Barrett Katz, M.D.

Dissociated vertical deviation is a not uncommon strabismic syndrome characterized by upward deviation of an eye when occluded, with downward movement of the eye when occlusion is removed. Associated findings include latent nystagmus and horizontal strabismus. Duane’s retraction syndrome is the clinical declaration of anomalous cranial nerve innervation characterized by a marked limitation or absence of abduction, variable limitation of adduction, narrowing of the palpebral fissure, and apparent globe retraction on attempted adduction. We report a patient with both dissociated vertical deviation and bilateral Duane’s retraction syndrome, demonstrating that dissociated vertical deviation can occur with the anomalous neuroanatomic substrate present in Duane’s retraction syndrome.

Key Words: Duane’s retraction syndrome—Dissociated vertical deviation—Cranial nerves—Strabismus—Brainstem anatomy—Eye movements.

Dissociated vertical deviation was first described in 1895 by Stevens (1) as an alternating vertical strabismus. Dissociated vertical deviation is characterized by an upward deviation of an eye when occluded (or spontaneously during periods of inattention), with downward movement of the eye when occlusion is removed (or when a refixation stimulus occurs). Associated findings may include latent nystagmus, horizontal strabismus (usually an esodeviation), downward movement of the occluded eye when filters of increasing density are placed before the fixing eye (Bielschowsky’s phenomenon), inferior oblique overaction, deficient fusion, excyclotorsion with supraduction during occlusion, with incyclotorsion and infraduction after the occlusion is removed (2). The vertical dissociation may be related to inattention or visual deprivation. Though the occluded eye undergoes supraduction, both eyes move concomitantly in horizontal and vertical meridians. While several theories have been proposed to explain the cause of dissociated vertical deviation (3–9), its etiology remains unknown.

Duane’s retraction syndrome is characterized by a marked limitation or absence of abduction, variable limitation of adduction, narrowing of the palpebral fissure and apparent globe retraction on attempted adduction, and vertical adventitious movements on adduction. Duane’s syndrome is more common in women and on the left side, although bilateral cases occur regularly (10). Electromyographic and pathologic studies have shown that Duane’s retraction syndrome is a neurogenic disorder in which the abducens nuclei and nerves are absent from the brainstem, and the lateral rectus muscle is innervated by branches of the oculomotor nerve (11,12).

We describe a patient with both dissociated vertical deviation and Duane’s retraction syndrome. To the best of our knowledge the association of
these two syndromes has not been previously recorded.

CASE REPORT

This 18-year-old right-handed woman first presented in the spring of 1984, when she was referred because of a recognized ocular motility disturbance. Family members noted problems with her eye movements that had been present since birth. When first examined by us (at the age of 18) she was noted to have normal central vision, normal color vision, and normal pupillary function, bilaterally. Her external examination demonstrated narrowing of each palpebral fissure on attempted adduction (Fig. 1), a small angle V-pattern esotropia, and minimal bilateral overaction of the inferior oblique muscles with upshooting of the adducted eye. The patient preferred to fixate with her right eye. Cover-uncover testing revealed a small angle left hypertropia in primary position. There was decreased abduction bilaterally, with each eye just crossing midline (Fig. 1). Alternate cover testing revealed that each eye would elevate in abduction under cover, though the right would elevate more than the left (Fig. 2). As each eye would elevate, it would also extort. A vertical seesaw effect was observed during alternate cover testing, that is, the covered eye moved up as the uncovered eye moved down to fixate. A latent nystagmus was present with fast phase toward the uncovered eye. Her examination was otherwise unremarkable, both ophthalmologically and neurologically; cycloplegic refraction revealed her to be 0.75 diopters hyperopic, bilaterally.

DISCUSSION

Several theories have been proposed to explain the etiology of dissociated vertical deviation. These include superior rectus hypofunction (3), "insufficient retinal stimulation causing elevation as a return to the position of absolute rest" (4), superior oblique hypofunction (5), anomalous excitations of subcortical vertical divergence centers (13), aberration of extraocular muscle tonus (6), bilateral paresis of the depressor muscles (7), and a defect of optomotor impulses from the lower nasal quadrants (8,9). Helveston (2) proposed that a center for vertical vergence may influence ocular motility, and that this center may be overridden by the relative visual experience of the two eyes. He suggested dissociated vertical deviation to be a sensory neurogenic disorder because he recognized it to occur in some normals, after prolonged occlusion.

Duane's retraction syndrome is now recognized to be the clinical expression of anomalous extraocular muscle innervation. The lateral rectus muscle is (presumably) innervated by fibers from the inferior branch of the oculomotor nerve (the same ramus that innervates the inferior oblique) (11,12). Patients with Duane's syndrome may exhibit other brainstem dysfunction (14), as well as congenital defects involving ocular, skeletal, and neural structures (15-27). Jay and Hoyt (14) found abnormal brainstem auditory-evoked potentials in nine of 14 patients with Duane's syndrome. The specific abnormality was a delay in wave III (thought to be generated in the superior olivary complex of the pons). In a study of 186 patients with Duane's retraction syndrome, Pfaffenbach et al. (19) found neurologic abnormalities that included hearing loss, seizures, generalized hypotonia, congenital facial palsy, and microcephaly. Maruo et al. (23) found a significant number of patients with Duane's retraction syndrome to have the gustolacrimal reflex (tear production while masticating). Isenberg and Blechman (26) described a patient

FIG. 1. Decreased abduction with associated narrowing of contralateral palpebral fissures and apparent enophthalmos of the adducting eye in gaze right (A) and gaze left (B).

FIG. 2. Supraduction of the right eye under cover as the left eye fixates (gaze directed to the right, minimizing actions of the right inferior oblique).
with Duane's retraction syndrome and Marcus Gunn jaw winking.

If dissociated vertical deviation is a neurogenic phenomenon, it is interesting to note its occurrence with the anomalous neuroanatomic substrate present in Duane's retraction syndrome. Certainly, most patients with dissociated vertical deviation do not have coexistent Duane's retraction syndrome, yet the concurrence of both syndromes implies that a clinically common strabismic entity can occur with differing underlying neuro-anatomy.

Acknowledgment: We acknowledge the support of The Pacific Vision Foundation and Research to Prevent Blindness Inc., New York, which made this work possible. (Grants to BK.)

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Progressive Cranial Polyneuropathy Caused by Primary Central Nervous System Melanoma

Larry A. Fish, M.D., Ph.D., Deborah I. Friedman, M.D., and Alfredo A. Sadun, M.D., Ph.D.

Primary malignant melanoma of the central nervous system (CNS) is exceedingly rare. The earliest description by Virchow in 1859 has been followed by ~50 autopsy-proven cases reported in the literature. These tumors are considered to arise from leptomeningeal melanocytes whose embryonic origin is neural crest tissue. Given the rarity of primary CNS melanoma, the diagnosis requires a thorough search to exclude a dermatologic, ocular, or visceral site of tumor origin. We report an unusual case of primary CNS melanoma in a patient with painful, progressive cranial polyneuropathy that eluded ante-mortem diagnosis despite extensive clinical, radiographical, and laboratory investigations.

**Key Words:** Progressive cranial polyneuropathy—Primary central nervous system melanoma.

CASE REPORT

In July 1986, a 44-year-old white man presented with a 1-year history of progressive painful ophthalmoplegia of the left eye. He initially experienced severe burning pain in the left upper face that progressed to involve the entire left face. He developed left facial and oral cavity numbness associated with a 2-month history of left facial weakness and diplopia. Initial neuroophthalmic examination revealed the following: Cranial nerve I was normal. Cranial nerve II had a visual acuity of 20/20 in each eye with recognition of 8/8 American Optical color plates. There was 4 mm of proptosis on the left; orbital repositus was normal bilaterally. The right pupil was 4 mm, round, and briskly reactive. The left pupil was 7 mm with no light response and no afferent pupillary defect. Intraocular pressure measured 14 mm Hg in each eye. Amsler grid and tangent screen perimetry were normal for both eyes, as were slit-lamp biomicroscopy and dilated fundus exam. There was a 60 prism diopter exotropia in primary gaze and complete III and IV cranial nerve palsies on the left. Motility was normal on the right. Cranial nerve V had decreased sensation to pinprick in V1 and V2 on the left. Cranial nerve VI appeared normal. Cranial nerve VII had left hemifacial weakness. Cranial nerve VIII had subjective decreased hearing on the left. Cranial nerves IX–XII were normal as was the remainder of the neurologic and physical examinations.

Computed tomography of the head and orbits suggested a subtle diffuse mass lesion along the left orbital apex with evidence of ipsilateral cavernous sinus enlargement (Fig. 1). Prednisone (60 mg daily) was begun with symptomatic pain relief, but no improvement of the ophthalmoplegia.

Craniotomy performed in July 1986 revealed only slight thickening of the arachnoid in the cli-
FIG. 1. Contrast-enhanced computed tomographic scan suggestive of diffuse mass in left orbital apex, extending to left cavernous sinus (arrowheads).

vus region; histopathological examination of biopsy tissue from this region showed reactive meningeal hyperplasia. Protein S-100 staining of this tissue was negative. The posterior orbital apex specimen revealed chronic inflammatory changes. Magnetic resonance imaging (MRI) performed 1 month postoperatively was normal.

The facial pain worsened over the next several months. A left VI nerve palsy developed, along with left corneal numbness and right facial numbness. In addition, left posterior pharyngeal numbness occurred. There was no dysphagia. Repeat MRI scan failed to disclose any abnormality. Lumbar puncture revealed a mildly elevated protein (54 mg/ml) and no malignant cells on cytospin. Cerebrospinal fluid bacteriological, viral, acid fast bacilli, and fungal cultures were negative on several occasions. Antinuclear antibody was 1:80 (nuclear) and Wintrobe sedimentation rate was 19. Syphilis serology and Lyme disease antibody were negative. Serum chemistries and complete blood counts were normal except for a persistently elevated white blood cell count (range, 18,900–41,400). Epstein–Barr virus (EBV) titers were elevated (Epstein–Barr nuclear antigen, 1:16; EBV antibody, 1:160), suggesting a recent infection. On findings from thorough systemic and oncologic evaluations, including chest roentgenogram, liver function tests, computed tomography of the abdomen, bone marrow biopsy, and gallium scan were normal.

Six months after surgery, a neurotropic corneal ulcer developed on the left, despite complete ptosis. The ulcer resolved with intensive topical antibacterial therapy. However, visual acuity in that eye decreased to hand motions coincident with the appearance of a marked afferent pupillary defect and left papillitis. The patient was admitted to an outside hospital in June 1987 with altered mental status. He suffered cardiopulmonary arrest during intubation for ventricular shunt placement. He died on July 7, 1987, following a protracted hospital course complicated by seizures and pneumonia.

RESULTS: HISTOPATHOLOGIC FINDINGS

The brain exhibited small foci of parenchymal cystic degeneration infiltrated by lymphocytes, few plasma cells, and polymorphonuclear leukocytes. Bordering these areas were zones of reactive gliosis and capillary proliferation. Perivascular cuffing by lymphocytes was seen.

Sections of the medulla at the level of the olive exhibited diffuse infiltration of the leptomeninges, nerves, and blood vessels by tumor cells and lymphocytes (Fig. 2). Some tumor cells demonstrated enlarged bizarre nuclei while others were oval to spindle-shaped with eosinophilic nuclei. Cytoplasm was abundant and pale pink. Sections at the level of the pons showed meningeal perivascular and perineural infiltration by tumor cells. The optic nerves were unavailable for examination. The optic tracts were examined using the paraphenylenediamine (PPD) technique that stains degenerated axons (1). The left optic tract showed marked degeneration in fascicles representing uncrossed fibers originating from the left optic nerve.
confirming the patient’s clinical optic neuropathy (Fig. 3). Pathologic diagnosis was primary malignant CNS melanoma deriving from the meninges. Exhaustive gross and histopathologic examination failed to reveal evidence of cutaneous or visceral tumor.

**COMMENT**

Primary melanoma of the central nervous system (CNS) typically exhibits one of two patterns of growth and development. Tumors may take the form of a discrete lesion or alternatively may arise as a diffusely infiltrative melanosis that shows a predilection for cranial nerve roots and perivascular spaces (2–4). Extracranial extension occurs rarely (5).

Discrete tumor masses occur predominantly in the spinal cord (6). Solitary lesions have also been described as arising in the pituitary, choroid plexus, and dura (7–10). The majority of intracranial cases are diffuse lesions. However, Bojesen-Moller described a series of six cases in which four patients were found to have solitary brain tumors. These patients had a relatively long survival as compared to other reported cases (4).

In their extensive review of CNS melanoma, Savitz and Anderson compiled autopsy results from a number of published case reports (2). Based on histopathologic sectioning, most cases demonstrated extensive infiltration of the subarachnoid space with perivascular cuffing and cranial nerve root involvement. Most cases presented in the third or fourth decades of life, and the majority died of complications from CNS melanoma within 1–2 years of presentation. Although it is now well accepted that melanoma can originate in the CNS, such was not always the case. In 1897, Ribbert [cited by Savitz and Anderson (2)] maintained that all pigmented tumors of the brain, spinal cord, and meninges were metastatic. Lubarsch [cited by Savitz and Anderson (2)] in 1920 likewise felt that the presence of even one pigmented cutaneous nevus precluded the diagnosis of primary CNS melanoma. In 1948, Rawles studied the migration of neural crest cells to the epidermis, iris and choroid of the eye, and leptomeninges. She conclusively demonstrated that, when neural crest was deleted from transplants, skin and hair were structurally normal but were devoid of pigment (11). This conclusively established the embryologic origin of melanocytes and thus permitted the diagnostic possibility of primary CNS melanoma. The neural crest origin of melanocytes may account for the reported clinical association of CNS melanoma, a neuroectodermal dysplasia, with a variety of phacomatoses, including neurofibromatosis and hemangiomatosis (2).

Therapeutic intervention in CNS melanoma has been disappointing. With the exception of localized spinal cord tumors, surgical resection is unsuccessful, and these tumors are resistant to both radio- and chemotherapy (2,4,6). Novel therapeutic efforts are aimed at in vitro enhancement of cytotoxic activity of autologous lymphocytes isolated from a tumor specimen, with subsequent reintroduction of these activated cells into the host (12).

Our patient manifested two frequently seen clinical features of primary CNS melanoma: a relentless deteriorating course and failure to establish antemortem diagnosis. His clinical presentation with unilateral involvement of cranial nerves III, IV, V, and VI suggested a cavernous sinus syn-
drome or orbital apex lesion. Failure to improve dramatically on steroids mitigated against Tolosa-Hunt syndrome, and tissue biopsy performed at orbitotomy was nondiagnostic. Progressive involvement of cranial nerves VII (bilateral), VIII, and IX suggested brainstem involvement; however, high-quality radiographic studies and numerous lumbar punctures failed to disclose any abnormality. Oncologic consultation and workup were unrevealing.

In the present case, it appears that a primary leptomeningeal melanoma arose at the level of the pons and affected both an ascending course that eventually involved the left optic tract via the cavernous sinus, and a descending course encompassing the medulla. In addition to rapidly progressive cranial nerve dysfunction, the disease course was characterized by severe pain. These clinical features correspond with the diffusely infiltrative tumor growth pattern found on histopathologic examination, in which parenchymal, perivascular, and perineural invasion was seen. Unfortunately, our orbital biopsy failed to detect malignancy, although a tissue diagnosis at that time would not have altered the clinical course, management, or outcome.

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Relative Afferent Pupillary Defect in the "Better" Eye

John D. Bullock, M.D., M.S., F.A.C.S.

A relative afferent pupillary defect usually occurs in an eye with unilateral or asymmetric optic nerve or extensive retinal disease. In general, the eye with poorer visual acuity has the afferent pupillary defect. Twenty-five patients are reported, however, in whom an afferent pupillary defect occurred in the eye with better visual acuity. These eyes had optic nerve or retinal dysfunction. The eyes with worse visual acuity but no afferent pupillary defect had an abnormality of the ocular media (corneal opacity, hyphema, anterior segment membrane, cataract, or vitreous opacity), amblyopia, refractive error, age-related macular degeneration, or cystoid macular edema. An afferent pupillary defect does not necessarily occur in the eye with poorer visual acuity.

Key Words: Afferent pupillary defect—Pupil—Marcus Gunn pupil.

In patients with unilateral or asymmetric optic nerve or extensive retinal lesions, differences in the pupillary light responses between the two eyes occur. This difference in pupillary light reactivity is known as an afferent pupillary defect (APD). The concept of an APD was first described by Hirschberg (1) in 1884, but other authors have contributed additional information in clarifying this entity (2). Kestenbaum (3) credited Gunn (4, 5) with the discovery, and it is now also known as the "Marcus Gunn pupil." Further modifications in the diagnostic technique were made by Levatin (6, 7), Thompson (8), and Gruber and Lessel (9).

Thompson et al. (10) have described a method to quantitate the APD using neutral density photographic filters of progressive logarithmic values. These are placed over the normal eye until the APD is eliminated in the involved eye. This technique has been used to quantitate relative afferent pupillary defects (11, 12). In the present study, however, no attempt was made to quantitate the APD.

As the test is now performed, subtle differences in pupillary reactivity can be exaggerated by quickly alternating a light from one eye to the other. As the light shifts from the normal to the affected eye, the direct stimulus is no longer capable of maintaining the previously evoked degree of pupillary constriction; thus, both pupils redilate. The magnitude of this redilation is proportional to the severity of the conduction or visual field defect in the affected eye. In general, this phenomenon is easily observed and, when the test is performed properly, it reveals very subtle differences in the photomotor input from each of the two eyes (13).

An APD also can be detected in patients with a fixed or unseen pupil (as from a hyphema, corneal scar, or anterior segment membrane). If a patient has a fixed or unseen pupil and normal retinal and optic nerve function in that eye and the other eye has an optic nerve or extensive retinal lesion, then
when light is directed into the fixed or unseen pupil, the other pupil constricts, and when the light is directed into the other eye, that pupil dilates. If a patient has a fixed or unseen pupil and an optic nerve or extensive retinal lesion in the same eye, and a normal other eye, then when light is directed into the normal other eye its pupil constricts; when light is directed quickly into the eye with the fixed or unseen pupil, the pupil of the normal other eye dilates.

An APD usually occurs in an eye with unilateral or asymmetric optic nerve or extensive retinal disease, which is usually the eye with poorer visual acuity. Thompson et al. (14) demonstrated, however, that an afferent pupillary defect is not proportional to visual acuity loss, but is proportional to visual field loss. Their finding was recently verified by Johnson et al. (15). The present communication substantiates not only a lack of correlation between visual acuity and pupillary function, but reports that, in fact, an APD can occur in the eye with better visual acuity. This phenomenon of pupillary response–acuity dissociation is recognized by neuro-ophthalmologists, but is less well known to general ophthalmologists, neurologists, or neurosurgeons. Its occurrence is reported in 25 patients.

**CASE REPORTS**

**Case 1**

A 48-year-old woman was first seen complaining of "trouble with my good eye." The patient had a long history of poor left eye vision and, as a child, she had undergone strabismus surgery on the left eye.
RELATIVE AFFERENT PUPILLARY DEFECT

FIG. 3. (A) Computerized tomographic scan, axial view (case 1), shows a high-density fusiform mass in the right posteromedial orbit, attached to the optic nerve. (B) Coronal view (case 1).

eye. Her corrected visual acuity measured 20/30² in the right eye and 20/200 in the left eye.

The external examination showed 1 mm of right proptosis. In addition, there was resistance to retropulsion of the right globe. An APD was noted on the right (Fig. 1A and B). The right globe elevated poorly, and diplopia was noted on upgaze.

The right fundus examination revealed slight optic atrophy, chronic papilledema, and choroidal folds in the macular area (Fig. 2A). The fundus examination on the left was normal (Fig. 2B). Goldmann visual field testing of the right eye showed marked peripheral and central constriction, especially inferiorly, and a full field on the left. An orbital roentgenogram revealed a calcified mass in the right orbit. A computerized tomographic (CT) scan of the orbits showed a high-density fusiform mass in the right posteromedial orbit attached to the optic nerve (Fig. 3A and B). Optic foramina views showed no enlargement. Clinical diagnoses of a right perioptic meningioma and left amblyopia were made.

During the next 5 months the visual acuity in the right eye deteriorated to 20/200. A biopsy and decompression of the right optic nerve was then performed. Histopathological examination showed a psammomatous meningioma of the right optic nerve (Fig. 4).

Postoperatively, the visual acuity in the right eye stabilized at 20/400 and the papilledema resolved. The right optic nerve remained atrophic (Fig. 5). Four years postoperatively, the visual acuity measured counting fingers in the right eye and 20/100 in the left eye. The right APD persisted. Ten years postoperatively, the best corrected visual acuity measured no light perception in the right eye and 20/80⁺¹ in the left eye.

Case 6

A 46-year-old woman with left optic neuritis was upholstering a chair when a spring shattered her
flu.

FIG. 5. Fundus photograph, right eye (case 1) after biopsy and decompression of the right optic nerve. Optic atrophy and resolution of the papilledema are noted.

glasses, severely lacerating the previously normal right eye. An examination in the emergency room revealed an uncorrected visual acuity of light perception in the right eye and “counting fingers” in the left eye. An extensive corneal–scleral laceration with uveal prolapse was noted (Fig. 6). The laceration was repaired surgically. Two months later (Fig. 7) an intraocular glass foreign body was removed from the right eye and an anterior segment membranectomy and cataract extraction procedures were performed. One year later the visual acuity measured “hand movements” in the right eye and 20/200 in the left eye. When a light was directed into the right eye, the left pupil con-

FIG. 6. External photograph (case 6) shows extensive inferior corneal–scleral laceration with uveal prolapse.

stricted and when the light was directed quickly into the left eye, the left pupil dilated (Fig. 8A and B). These findings suggested that the right optic nerve and retina were functional. Slit-lamp examination of the right eye showed bullous keratopathy, corneal fibrosis, and slight ruberosis of the iris (Fig. 9). Contact B-scan ultrasonography of the right eye showed a normal posterior segment with a clear vitreous cavity (Fig. 10). Fundus examination demonstrated left optic atrophy (Fig. 11). Several months later the patient developed sudden decreased vision in the left eye. Examination at that time revealed a visual acuity of “hand movements” in the right eye and no light perception in the left eye. The patient was treated with oral prednisone, 60 mg per day. A CT scan of the brain and orbits was normal. One week later the visual acuity measured “hand movements” in each eye. The prednisone dosage was eventually tapered during a 3-month period. One year later, the visual acuity measured “hand movements” in each eye. The patient then underwent a right penetrating keratoplasty, a right anterior segment membranectomy, and a right anterior vitrectomy. The patient did well postoperatively (Fig. 12) and 6 weeks later, the corrected aphakic visual acuity measured 20/80−2 in the right eye and “hand movements” in the left eye. The right retina was completely attached, and the posterior pole appeared normal (Fig. 13). A repeat corneal transplant was performed 18 months later due to opacification of the cornea. The patient subsequently developed a retinal detachment. She was lost to follow-up; when she returned 7 years later her visual acuity measured no light perception in each eye. The right eye was phthisical. A cranial CT scan was otherwise normal.
FIG. 6. (A) External photograph (case 6) shows left pupillary constriction when the handlight is directed into the right eye. (B) External photograph (case 6) shows left pupillary dilation when the handlight is directed into the left eye. (A and B document a left afferent pupillary defect.)

FIG. 9. External photograph, right eye (case 6) shows corneal opacification from bullous keratopathy and corneal fibrosis.

FIG. 10. Contact B-scan ultrasonogram (case 6) shows a normal posterior segment with an acoustically clear vitreous cavity.

FIG. 11. Fundus photograph, left eye (case 6) shows optic atrophy.

FIG. 12. External photograph (case 6) 6 weeks after right penetrating keratoplasty, anterior segment membranectomy, and anterior vitrectomy, shows a clear central cornea.
Fig. 13. Fundus photograph (case 6) taken 6 weeks after surgery, showing normal disc and macula.

Discussion

Reduced vision can occur by a variety of mechanisms; some affect pupillary function and others do not. If a patient with bilateral loss of vision has a different condition in each eye, then different pupillary responses may occur when each eye is stimulated with light. It is therefore possible to have an APD occur in an eye with a lesser degree of visual acuity loss, and, thus, pupillary reaction–acuity dissociation will be present.

Table 1 summarizes the diagnoses and visual acuities of 25 such patients. The listed visual acuities are those that were present at the time that pupillary response–acuity dissociation was first noted; in many cases, the acuities in each eye later changed, as illustrated by the case reports. The eyes with the APD all had optic nerve or retinal dysfunction: optic nerve meningioma, retinal de-
tachment, optic neuritis or atrophy, peripapillary choroidal rupture, glaucomatous cupping, panretinal photocoagulation, ischemic optic neuropathy, and orbital malignancy. The fellow eyes had visual loss (greater in each case than the eye with the pupillary abnormality) due to amblyopia, a refractive error, corneal scarring, hyphema, anterior segment membrane, cataract, vitreous hemorrhage, age-related macular degeneration, or cystoid macular edema.

In the eyes with amblyopia and refractive errors the pupillary reactions are usually normal. Several authors, however, have reported an APD in eyes with amblyopia, but noted a lack of correlation between pupillary response and visual acuity (16,17). An abnormality of the ocular media (corneal opacity, hyphema, anterior segment membrane, cataract, or vitreous hemorrhage) is an optical barrier to light, but does not affect processing of the visual stimulus or transmission of the electrical impulse. If the optical barrier produces light reduction, however, rather than light diffusion, then an APD can occur by the same mechanism as that which occurs with a neutral density filter (18). Age-related macular degeneration and cystoid macular edema affect the macula but, in general, not enough neural elements are destroyed to produce an APD. Macular lesions have been reported as a cause of an APD, but the macular lesion must be extensive (6,19–21).

An APD is present when a relative difference in pupillary light reactivity exists. If one eye is perfectly normal and the other eye has an optic nerve or retinal lesion, amblyopia, an extensive macular lesion, a total hyphema, or, perhaps, a vitreous hemorrhage, then an afferent pupillary sign can occur in the affected eye with worse visual acuity. However, an APD can also occur in an eye with better visual acuity, affected by a retinal or optic nerve abnormality, if the vision in the other eye is affected to a greater degree by a refractive error, amblyopia, an abnormality of the ocular media, or limited macular disease.

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Corneal Permeability in Patients with Tonic Pupil
A Reevaluation of Its Cholinergic Supersensitivity

Takashi Utsumi, M.D.

Cholinergic supersensitivity of the iris sphincter helps to make the diagnosis of "tonic pupil." Because uncertain responses sometimes occur with mecholyl 2.5%, dilute solutions of pilocarpine are often used. There has always been some question as to whether individual variation in corneal permeability was one of the factors contributing to the variability in pupillary constriction to topical cholinergics in these patients. In this study, the degree of intraocular penetration of topical fluorescein was compared with the degree of pilocarpine-induced miosis in six tonic pupils and in seven age-matched normal controls. A noncontact fluorophotometer and a computerized videopupillogram were used. The patients showed a significant cholinergic supersensitivity in their affected tonic pupils and had normal intraocular penetration of fluorescein through the cornea in both eyes. It is concluded that the cholinergic supersensitivity of the iris in tonic pupils is not the result of increased corneal permeability but instead results from an increased sensitivity of the cholinergic receptors in iris sphincter muscles.

Key Words: Tonic pupil—Cholinergic supersensitivity—Corneal permeability—Fluorophotometry.

Adler and Scheie (1) recommended methacholine 2.5% (mecholyl, Merck) to demonstrate cholinergic supersensitivity in patients with tonic pupil, but it was soon recognized that some tonic pupils failed to constrict to mecholyl 2.5%. In 1966, Loewenfeld and Oono (2) showed that large interindividual variations in mecholyl sensitivity existed in normal subjects and suggested that this might be due to variations in corneal permeability. In 1977, Thompson (3) reported that the mecholyl 2.5% test was positive in only about two-thirds of patients with Adie's syndrome.

In 1977, Purcell and colleagues (4) reported focal corneal insensitivity in patients with tonic pupils. Thompson (3) suggested in 1977 that despite the patchy numbness of the cornea in patients with Adie's syndrome, the corneal penetration of topical drugs appeared to be normal. He directly showed that adrenergic mydriatics and anticholinergic mydriatics dilated Adie's pupils without supersensitivity. Because of these uncertainties, I measured the corneal permeability in eyes with tonic pupils using a recently developed fluorophotometer technique (5).

SUBJECTS AND METHODS

Six patients aged 40-60 years (mean 49 years) with unilateral tonic pupil were included in the study (Table 1). Although their pupillary cholinergic supersensitivity was detected by topical instillation of a dilute solution of pilocarpine in the manner recommended by Pilley and Thompson (6), a weaker concentration was used (0.0313%) (7). This concentration was a little stronger than the minimum effective dose (0.022%) for Japanese irides (8). The patients had no neurologic disorder except tonic pupil. Seven age-matched normal
CORNEAL PERMEABILITY IN TONIC PUPIL

TABLE 1. Data of tonic pupil patients

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age (yr)/sex</th>
<th>Affected side</th>
<th>Affected eye</th>
<th>Sound eye</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>40/M</td>
<td>R</td>
<td>-5.82</td>
<td>-0.98</td>
</tr>
<tr>
<td>2</td>
<td>44/F</td>
<td>L</td>
<td>-3.38</td>
<td>-0.81</td>
</tr>
<tr>
<td>3</td>
<td>46/F</td>
<td>L</td>
<td>-2.20</td>
<td>-0.20</td>
</tr>
<tr>
<td>4</td>
<td>47/F</td>
<td>R</td>
<td>-2.96</td>
<td>-0.79</td>
</tr>
<tr>
<td>5</td>
<td>57/M</td>
<td>L</td>
<td>-6.66</td>
<td>-1.02</td>
</tr>
<tr>
<td>6</td>
<td>60/F</td>
<td>R</td>
<td>-2.66</td>
<td>-0.25</td>
</tr>
</tbody>
</table>

subjects aged 34–64 years (mean 50.1 years) were examined as controls (Table 2).

We used a noncontact fluorophotometer (Coherent FM-1, Fluorotron Master, Gherent, Palo Alto, CA) capable of automatically measuring the concentration of fluorescein in various parts of the eye by the strength of the emitted fluorescence. Before and 150 min after topical instillation of two 20-μl drops of a 10% sodium fluorescein solution into the cul-de-sac of both eyes, the strength of the emitted fluorescence was measured by scanning through the eye. The maximum value in the cornea and the minimum value in the anterior chamber were obtained. The tissue autofluorescence before topical application of fluorescein was subtracted from the later measurements, and the concentration of fluorescein in the aqueous was determined.

A degree of miosis caused by topical instillation of pilocarpine 0.0313% was obtained from the following equation as decibels (dB): degree of miosis = 20 log (PA post/PA pre), where PA is the pupil area. Pupil area was measured by a computerized infrared videopupillogram (HTV C-301) which provided a digital output in square millimeters.

After the fluorescein and pilocarpine tests, we estimated corneal sensitivity by touching the cornea with the twisted tip of cotton gauze and measured the basal tear volume by a phenol red-impregnated cotton thread, the crimped end of which was placed in the cul-de-sac for 15 s (9).

TABLE 2. Data of normal subjects

<table>
<thead>
<tr>
<th>Subject</th>
<th>Age (yr)/sex</th>
<th>Pilocarpine miosis (dB)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Right eye</td>
<td>Left eye</td>
</tr>
<tr>
<td>1</td>
<td>34/F</td>
<td>-1.15</td>
</tr>
<tr>
<td>2</td>
<td>40/F</td>
<td>-0.43</td>
</tr>
<tr>
<td>3</td>
<td>46/M</td>
<td>-0.08</td>
</tr>
<tr>
<td>4</td>
<td>52/F</td>
<td>-0.46</td>
</tr>
<tr>
<td>5</td>
<td>57/M</td>
<td>-6.22</td>
</tr>
<tr>
<td>6</td>
<td>58/M</td>
<td>-1.56</td>
</tr>
<tr>
<td>7</td>
<td>64/F</td>
<td>-5.22</td>
</tr>
</tbody>
</table>

RESULTS

The concentrations of penetrated fluorescein in the anterior chamber and in the cornea in patients with tonic pupil and in normal subjects are shown in Tables 3 and 4, respectively. Values were higher in the cornea than in the anterior chamber in both groups. Means and SD were computed for the patients’ affected eyes, for the patients’ unaffected eyes, and for the normal subjects. Mean values of the concentrations of each group were 1.68, 1.72, and 1.84 log ng/ml in anterior chamber, and 2.35, 2.40 and 2.49 log ng/ml in cornea, respectively (Table 5). Because no statistically significant differences were found between the groups either in the anterior chamber or in the cornea, we concluded that both eyes of patients with unilateral tonic pupil had normal permeability.

Correlation between the fluorescein concentration in the anterior chamber and in the cornea is shown in Fig. 1. Because all eyes in this study had the same concentration of fluorescein in the anterior chamber and another concentration in the cornea, all anterior chamber values were considered in a single group and all cornea values were considered in another group. The two groups are compared in Fig. 1. The significance of the linear correlation is evident.

Cholinergic sensitivity to topical pilocarpine

TABLE 3. Log concentration of penetrated fluorescein in patients with tonic pupil

<table>
<thead>
<tr>
<th>Patient</th>
<th>Anterior chamber (ng/ml)</th>
<th>Cornea (ng/ml)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Affected eye</td>
<td>Sound eye</td>
</tr>
<tr>
<td>1</td>
<td>1.27</td>
<td>1.40</td>
</tr>
<tr>
<td>2</td>
<td>1.73</td>
<td>1.79</td>
</tr>
<tr>
<td>3</td>
<td>1.92</td>
<td>2.00</td>
</tr>
<tr>
<td>4</td>
<td>1.33</td>
<td>1.32</td>
</tr>
<tr>
<td>5</td>
<td>1.91</td>
<td>2.11</td>
</tr>
<tr>
<td>6</td>
<td>1.91</td>
<td>1.71</td>
</tr>
</tbody>
</table>

TABLE 4. Log concentration of penetrated fluorescein in normal subjects

<table>
<thead>
<tr>
<th>Subject</th>
<th>Anterior chamber (ng/ml)</th>
<th>Cornea (ng/ml)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Right eye</td>
<td>Left eye</td>
</tr>
<tr>
<td>1</td>
<td>1.44</td>
<td>1.42</td>
</tr>
<tr>
<td>2</td>
<td>1.50</td>
<td>1.27</td>
</tr>
<tr>
<td>3</td>
<td>1.53</td>
<td>1.62</td>
</tr>
<tr>
<td>4</td>
<td>1.96</td>
<td>1.81</td>
</tr>
<tr>
<td>5</td>
<td>1.88</td>
<td>1.96</td>
</tr>
<tr>
<td>6</td>
<td>2.37</td>
<td>2.20</td>
</tr>
<tr>
<td>7</td>
<td>2.53</td>
<td>2.29</td>
</tr>
</tbody>
</table>
DISCUSSION

Opinion has varied regarding corneal permeability in tonic pupils (2,3). This study revealed that although patients with unilateral tonic pupil had a clearly higher sensitivity to topical pilocarpine than did normal controls in the affected eyes, penetration of fluorescein through the cornea appeared to be the same in both eyes. Figure 2 clearly shows that there was no statistically significant correlation between the concentration of penetrated fluorescein in the anterior chamber and the degree of miosis caused by topical pilocarpine. Corneal sensation was generally normal to touch, and basal tear volume was in the normal range (10–25 mm/15 s) in both eyes of all subjects.

The strong linear correlation between the concentration of fluorescein in the anterior chamber and in the cornea suggests that once the drugs penetrate the corneal epithelium they merely pass through the stroma and endothelium and are diffused into the aqueous humor in the anterior chamber according to the slope of the concentration.

Abnormal corneal sensitivity in tonic pupils was carefully studied by Purcell and associates (4). These abnormalities suggest the possibility of an abnormality of corneal penetration. However, results obtained in this study suggest otherwise. On the other hand, because I did not map corneal sensitivity with the attention to detail used in the study by Purcell and associates (4), I may have missed some regional lowered sensitivity.

### TABLE 5. Comparison of log concentration

<table>
<thead>
<tr>
<th>Subjects (no. of eyes)</th>
<th>Anterior chamber (ng/ml)</th>
<th>Cornea (ng/ml)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tonic pupils</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Affected eye (6)</td>
<td>1.66 0.30</td>
<td>2.35 0.18</td>
</tr>
<tr>
<td>Sound eye (6)</td>
<td>1.72 0.32</td>
<td>2.40 0.19</td>
</tr>
<tr>
<td>Normal controls (14)</td>
<td>1.84 0.40</td>
<td>2.49 0.26</td>
</tr>
</tbody>
</table>

0.313% as represented by the degree of miosis was obviously greater in the affected eyes than in the sound eyes in the tonic pupil patients (Table 1), and the normal subjects had matched cholinergic supersensitivity in the two eyes. Figure 2 clearly shows that there was no statistically significant correlation between the concentration of penetrated fluorescein in the anterior chamber and the degree of miosis caused by topical pilocarpine. Corneal sensation was generally normal to touch, and basal tear volume was in the normal range (10–25 mm/15 s) in both eyes of all subjects.

![FIG. 1. Correlation between the fluorescein concentration in the anterior chamber and in the cornea. A linear regression curve is shown. Eyes of normal controls (open circles); sound eyes of patients with tonic pupil (open triangles); affected eyes of patients with tonic pupil (closed triangles).](image)
**Corneal Permeability in Tonic Pupil**

**FIG. 2.** Relationship between the fluorescein concentration in the anterior chamber and the degree of miosis caused by topical pilocarpine. Statistically significant correlation is absent. Eyes of normal controls (open circles); sound eyes of patients with tonic pupil (open triangles); affected eyes of patients with tonic pupil (closed triangles).

**Acknowledgment:** I thank Dr. H. Stanley Thompson of the Department of Ophthalmology of the University of Iowa for reviewing a draft of this paper, and Dr. Shigetoshi Nagataki of the Department of Ophthalmology of the Ryukyu University for technical advice. I also thank Ritsuko Miyachi for secretarial assistance.

**REFERENCES**

Transient Partial Oculomotor Nerve Paresis with Posterior Communicating Artery Aneurysm
A Case Report

Beverly N. Greenspan, Ph.D., M.D., and Alexander G. Reeves, M.D.

A 38-year-old woman is described who developed a partial right oculomotor paresis which cleared spontaneously prior to clipping of an associated nonhemorrhagic bilobate right anterior communicating artery aneurysm. Key Words: Reversible oculomotor paresis—Cerebral aneurysm.

Aneurysms of the posterior communicating artery produce oculomotor nerve palsy in ~30-40% of cases, and about half of these are without concomitant subarachnoid hemorrhage (1,2). Aneurysm of the internal carotid or distal basilar arteries also may present as painful oculomotor palsy, sometimes with only minimal signs (3). We report a case in which the oculomotor paresis associated with an aneurysm resolved spontaneously.

CASE REPORT

A 38-year-old woman presented with a complaint of a severe right retroorbital throbbing headache that had been constant for 2 weeks. She also complained of blurred vision in the right eye, beginning a few days after the onset of the headache. She had intermittent nausea, vomiting, anorexia, and photophobia during the headache. Examination revealed no meningismus. Visual acuity was 20/25-1 in the left eye and 20/40-1 (without improvement with pinhole) in the right eye at distance. There was slight ptosis of the right eye. The pupils were 4 mm and equal and reactive to light and accommodation, and extraocular movements were intact. The examination was otherwise unremarkable.

Lumbar puncture produced clear, colorless cerebrospinal fluid with no white cells, 24 red cells, a protein of 34 mg/dL and glucose of 55 mg/dL. A tapering course of prednisone was prescribed.

One week later she had no change in her headache, blurred vision, or intermittent nausea and vomiting, and she complained of vertical diplopia on upgaze. Examination showed more pronounced ptosis of the right eye and evidence of a...
right superior rectus palsy. The right pupil was larger than the left by <1 mm, and both pupils reacted briskly to light. Visual acuity was unchanged. The remainder of the examination, including fundoscopic exam, corneal reflex testing, and test of afferent pupillary defect, was unremarkable. A contrast-enhanced computed tomography (CT) scan of the head with 3-mm cuts through the base of the brain did not show any abnormalities.

A telephone conversation with the patient 10 days after she was first seen elicited that her headache and visual symptoms were without change. When next seen 5 days after this, she said her blurred vision and vertical diplopia were gone, although the headache was unchanged. Examination revealed complete resolution of previously observed signs. The next day cerebral angiography revealed a bilobate posteriorly directed 6-8-mm aneurysm of the right posterior communicating artery (PCoA) projecting posteriorly from the junction of the PCoA and the supraclinoid portion of the internal carotid (Fig. 1). Uneventful surgical obliteration of the aneurysm was performed.

**COMMENT**

Partial oculomotor palsy due to unruptured intracranial aneurysm of the internal carotid artery near the takeoff of the PCoA or of the basilar artery has been reported (3), but except for one report of transient mild ptosis with an unruptured internal carotid-PCoA junction aneurysm (4), we know of no report of oculomotor palsy of this etiology that resolved over days or weeks. In the case mentioned (4) there was also progressive mydriasis of the affected eye. Gale and Crockard (5) reported a case of transient unilateral mydriasis with a basilar artery bifurcation aneurysm directed toward the relevant third nerve. This case differs from ours in that their patient had definite hemorrhage from the aneurysm with seizures, meningeal signs, papilledema, and blood in the ventricles shown by CT scan of the brain. The mydriasis was present for only 1 h at 16-17 h after the hemorrhage, and possibly represented an early transient sign of transtentorial herniation, as the patient clearly had dilated ventricles and increased intracranial pressure at that time. Walsh and Hoyt (6) state "some oculomotor palsies caused by aneurysms will disappear without treatment," but they cite no examples. A few untreated patients included in old series showed recovery of oculomotor palsy over years, often with signs of aberrant regeneration of the third cranial nerve (7–9). Small PCoA aneurysms can also readily cause oculomotor palsy since they lie close to the course of the nerve, and are said in a recent review (10) to be the most common single cause of isolated oculomotor palsy. Ptosis is typically the first symptom of an initially incomplete paresis and may be preceded by ipsilateral headache (1), as in this case. The levator palpebrae is also the first muscle to recover function after surgical treatment of aneurysms, but the superior rectus shows the least postoperative recovery (2,7). In our patient both these muscles were clinically involved and both recovered before treatment. More commonly, the initially partial oculomotor palsy due to aneurysm becomes complete 2-3 days after onset (1,10,11). The pupil is usually involved but may be spared (11–15). PCoA aneurysms that produce oculomotor palsy may be large, and as in our case, multiloculated (2).

Possibly the resolution of the deficits in our patient was due to the pressure of the aneurysm causing a conduction block rather than axon degeneration, as postulated by Hamer (1) to account for postoperative resolution. The aneurysm may have been expanding and may have shifted slightly in position as it grew, pressing only transiently on the third cranial nerve.

As stressed by Bartleson et al. (3) and Hamer (1), oculomotor palsy when caused by aneurysm may be an important warning sign of hemorrhage, which in Okawara’s (16) six patients with aneurysmal “extraocular muscle impairment,” occurred at a mean of 29.6 days after onset of this symptom. His data came from retrospective studies of pa-
tients who had hemorrhage, however, and so do not represent the incidence of subarachnoid hemorrhage in patients who present with oculomotor palsy. Nonetheless, our case shows that the possibility of aneurysm as a cause of oculomotor palsy and ipsilateral headache must be considered even when the ocular symptoms resolve over the next weeks. In 1946 Jefferson (7) wrote "The difference between [ophthalmoplegic migraine] and aneurysmal compression is that in the periodic palsies the nerve usually recovers so completely that an entirely fresh palsy is possible and may be repeated time after time. In the aneurysms the recovery after a single episode is never complete enough for so strikingly fresh a palsy to present itself." Our patient initially was treated with prednisone as for migraine "status," but it is difficult to attribute the resolution of her ocular findings to this, since at the time she completed a 10-day course of prednisone her symptoms were unchanged. Her case demonstrates that oculomotor deficits secondary to aneurysm may not only be incomplete (3), but may be transient.

REFERENCES

Ptosis as the Sole Manifestation of Compression of the Oculomotor Nerve by an Aneurysm of the Posterior Communicating Artery

Edward F. Good, M.D.

Oculomotor palsy secondary to a berry aneurysm is usually present with pupillary dilatation, followed by other signs of third cranial nerve dysfunction, including oculomotor paresis and ptosis. Partial paralysis of the nerve with pupil sparing has been observed, but ptosis as the sole sign of oculomotor paralysis has not previously been reported until now.

Key Words: Berry aneurysm—Oculomotor Palsy—Ptosis.

Compression of the oculomotor nerve (third cranial nerve) in the subarachnoid space almost always presents with pupil dilatation and other signs of third nerve dysfunction, including ptosis and external ophthalmoparesis. If due to an aneurysm, facial and orbital pain are also present. Recent reviews point out the frequent occurrence of partial and minimal palsies, particularly with pupil sparing (1,2). I report a patient who developed a headache and ptosis as the sole manifestation of compression of the oculomotor nerve by a saccular aneurysm of the posterior communicating artery.

CASE REPORT

A 42-year-old woman had the insidious onset of a left frontal and retroorbital headache that was progressive. After 1 week, the pain became so severe that she ceased working. At that time, she noted drooping of the left upper lid. A computed tomography (CT) scan of the head was performed, which was normal. Two weeks later, she consulted an ophthalmologist. There was no diplopia reported with a red lens test. Cover–uncover testing was orthophoric for distance, with a small degree of exophoria at near. Versions were full. The pupils were 3 mm and reacted briskly, directly, and consensually. Palpebral fissures measured 8 mm OD and 6.5 mm OS. A diagnosis of acquired ptosis was made. A neurological consultation was requested because of the severe headaches. The patient denied any double vision or other neurological symptoms. Internal and external function of the oculomotor nerve again was normal with the exception of a left-sided ptosis. A review of old photographs (Figs. 1 and 2) did not reveal any evidence of ptosis.
Because of the intractable headache, an angiogram was performed. The study revealed a large bilobed aneurysm of the posterior communicating artery. At operation, the oculomotor nerve was found to be compressed by the aneurysm, with fresh clot in the dome. There was evidence of old bleeding by the presence of hemosiderin pigment and xanthochromia. The third nerve was encased by an old hematoma and fresh thrombus. The patient made an uneventful recovery, but her ptosis persisted (Fig. 3). Several weeks postoperatively, the patient noted double vision on extreme right gaze and downgaze. A red lens test at that time revealed an exophoria on right gaze and a left hyperphoria on downgaze. The pupils remained equal.

**COMMENT**

The cardinal signs of an oculomotor palsy are ptosis, mydriasis, and extraocular motion paralysis. Bartleson et al. (2) reported 12 cases of minimal oculomotor paralysis secondary to unruptured intracranial aneurysms. In each of these cases, at least one element of oculomotor dysfunction was absent. Mydriasis and ptosis were equally common as signs of third nerve paralysis followed by dysfunction of the extraocular muscles. Mydriasis alone occurred in one patient. Mydriasis is generally the most common first sign of oculomotor compression and is generally considered to occur because of the peripheral dorsomedial location of the pupillomotor fibers within the nerve. Walsh and Hoyt (3) state that pressure anywhere along the basilar segment of the third nerve produces pupillary dilatation, and this effect is not dependent on the topography of the pupillary axons within the nerve. Pupil sparing of the oculomotor nerve has been reported along any portion of the nerve, but is most common in the cavernous sinus (1,4).

Partial third nerve paralysis of an external type implies ptosis and decreased range of motion in the appropriate direction of the muscles innervated by the third nerve with or without deviation from the primary position. In several recent reviews of ptosis and third cranial nerve lesions, it has been stated that an isolated unilateral ptosis should not be regarded as a sign of an oculomotor paralysis (5–7). To my knowledge, there are no reported cases of isolated ptosis attributed to the compression of the oculomotor nerve.

Isolated ptosis without pain is probably rarely compressive; however, the association of ipsilateral headache with acquired ptosis, as in other partial third cranial nerve palsies, should alert one of the possibility of an intracranial aneurysm and should prompt careful follow-up and possible use of other diagnostic modalities. CT scanning reportedly may miss up to a third or more of these lesions (2). (It is uncertain what percentage would be picked up by MRI, and angiography remains as the only definitive test to elucidate this lesion.) It is difficult to explain the late, i.e., postoperative, onset of double vision. In 6 of 12 of Bartelson’s patients, third nerve dysfunction was worse postoperatively. Faulty clip placement, clip slippage, operative dissection, and hemorrhages have been reported as possible causes (8).

**Acknowledgment:** I would like to thank Victor J. Basso, M.D., for evaluation and referral of this patient.
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Disorders of the Visual System in Alzheimer's Disease

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Alzheimer's disease (AD) is associated with disturbances in basic visual, complex visual, and oculomotor functions. The broad range of visual system disorders in AD may result from the concentration of neuropathology in visual association cortex and optic nerves in this disease. AD patients and their caregivers frequently report visuospatial difficulties in these patients. Examination of the visual system in AD may reveal visual field deficits, prolonged visual evoked potentials, depressed contrast sensitivities, and abnormal eye movement recordings. Complex visual disturbances include constructive and visuoperceptual abnormalities, spatial agnosia and Balint's syndrome, environmental disorientation, visual agnosia, facial identification problems, and visual hallucinations. The purpose of this article is to review the spectrum of visual system disturbances found in AD and, in particular, to describe the methods used to screen for complex visual abnormalities in these patients.

Dementia established by clinical exam and documented by mental status questionnaire or neuropsychological testing (1), (2)

No disturbance of consciousness

Onset between the ages of 40 and 90

Absence of other potentially causative disorder(s), such as systemic disorders or brain disease, that could account for dementia

TABLE 1. NINCDS-ADRDA criteria for clinically probable Alzheimer's disease (2)

| 1. Dementia established by clinical exam and documented by mental status questionnaire or neuropsychological testing |
| 2. Deficits in two or more areas of cognition |
| 3. Progressive worsening |
| 4. No disturbance of consciousness |
| 5. Onset between the ages of 40 and 90 |
| 6. Absence of other potentially causative disorder(s), such as systemic disorders or brain disease, that could account for dementia |

Visual symptoms in Alzheimer's disease

Spatial agnosia: Five patients had prominent difficulties finding objects, looking at them, and reaching for them. These patients were unable to walk without bumping into things and could not judge distances.

Visual localization problems: Seven additional patients had less severe difficulties visually finding objects.

Environmental disorientation: Five patients had an isolated problem finding their way in familiar surroundings that was not due to a general spatial agnosia.

Spatial alexia: Three patients had reading difficulties not due to language disturbances or a general spatial agnosia. On reading, these patients easily lost their place.

Possible mild optic ataxia: Two patients had difficulties with fine hand-eye coordination tasks that was not due to general spatial agnosia.

Visual agnosia: Three patients had difficulty recognizing common objects when visually presented.

Facial identification problems: One patient had prosopagnosia.

Visual hallucinations: Three patients had formed hallucinations that were not otherwise explainable by medications or confusional states.

TABLE 2. Visual symptoms in 30 patients with Alzheimer's disease (4)

Visualization of the visual system in AD patients reveals a broad range of disturbances. All visual functions are not uniformly affected in AD; there are specific patterns of involvement. Frequently impaired basic visual functions include peripheral vision, visual evoked potentials (VEPs), and contrast sensitivities (4,6,24,25,27,28). Difficulties with constructions, figure-ground discrimination, and visual synthesis are present in the majority of patients with AD and are the most common findings potentially attributable to complex visual dysfunction in this disorder (4,29). Other common complex visual abnormalities involve visuospatial abilities (4–9) and visual object and face recognition (4–9). Finally, patients with AD have abnormal oculomotor functions, such as increased saccadic latency and an inability to inhibit anticipatory saccades (30–35). The rest of this article discusses specific visual system abnormalities in AD and visual system testing in demented patients, particularly the screening tests used for complex visual disturbances (see Table 3).

CLINICAL EVALUATION

The most common visual complaints in AD are problems in visuospatial functioning (4–8). In a study of 30 community-based patients with clinically probable AD, almost half (43%) had visual symptoms, and these were predominantly visuospatial (4) (see Table 2). Visuospatial difficulties occur in general spatial orientation (e.g., walking without bumping into things), in visual localization (e.g., finding door handles or other common objects), in environmental orientation (e.g., finding their way in their surroundings), in reading (e.g., locating the next word or line of print), and in performing fine hand-eye coordination activities (e.g., sewing) (4–9) (see Table 2). AD patients and their caregivers complain less frequently of difficulties in visually identifying objects, scenes, or faces (5) and of visual hallucinations (26).

Examination of the visual system in AD patients reveals a broad range of disturbances. All visual functions are not uniformly affected in AD; there are specific patterns of involvement. Frequently impaired basic visual functions include peripheral vision, visual evoked potentials (VEPs), and contrast sensitivities (4,6,24,25,27,28). Difficulties with constructions, figure-ground discrimination, and visual synthesis are present in the majority of patients with AD and are the most common findings potentially attributable to complex visual dysfunction in this disorder (4,29). Other common complex visual abnormalities involve visuospatial abilities (4–9) and visual object and face recognition (4–9). Finally, patients with AD have abnormal oculomotor functions, such as increased saccadic latency and an inability to inhibit anticipatory saccades (30–35). The rest of this article discusses specific visual system abnormalities in AD and visual system testing in demented patients, particularly the screening tests used for complex visual disturbances (see Table 3).
TABLE 3. Screening test for complex visual functions

| Construction: Two-dimensional design or cube, three-dimensional cube or open box, complex design, e.g., clock face |
| Perception: Tests of figure-ground discrimination (overlapping, cross-hatched, or hidden figures) and visual synthesis tasks (completion or visual closure tasks) |
| Visuospatial: Tests of inattention or neglect (line bisections), localization (dot circling or picture searching), simultanagnosia ("linked" versus "unlinked" figures or embedded figures), oculomotor apraxia (eye movements to visual stimuli), and optic ataxia (hand movements to visual stimuli) |
| Environmental orientation: Tests of topographic amnesia and topographagnosia (follow, describe, draw, and learn route) and geographical orientation (map reading or placing landmarks on map) |
| Object recognition: Visual naming, describe or demonstrate use, tactile naming, and matching (actual objects, pictures of common objects, and drawings of common objects) |
| Face identification: Tests of prosopagnosia (identification of pictures of famous faces) and facial discrimination (matching unfamiliar faces) |

There are two special problems involved in the visual evaluation of AD patients. First of all, basic visual deficits may affect tests of complex visual functions. The evaluation of basic visual functions must precede the administration of complex visual tests. Secondly, other cognitive impairments, such as deficits in general comprehension, language, or memory, can interfere with performance on visual tasks. The testing methods can partially compensate for deficits in other areas of cognition by keeping the task and instructions as simple as possible, by allowing as much time as needed for a response, and by allowing the patients to respond by description or demonstration as well as by a specific verbal answer. Ultimately, clinicians must interpret critically the results of visual system testing in demented patients, as abnormal performance may be due to a combination of several visual and cognitive disturbances.

BASIC VISUAL FUNCTIONS

Investigations of basic visual functions in AD have found abnormalities particularly in peripheral vision and in VEPs (6,24,27,28,36). Sadun et al. (6) reported binasal field loss, inferior field loss, or constriction of the visual fields in 3 of 12 AD patients. Others have also found constriction of the visual fields (27) or left homonymous deficits (36). On VEPs, some investigators have found that flash stimuli, but not pattern reversal stimuli, may result in an increased latency and a loss of amplitude (19,24). This pattern of VEP findings suggests that the main problems in the visual system in AD are not in the visual pathways up to the primary visual cortex, but in the visual association areas (28). However, late in the course of AD, some patients have optic nerve pallor, afferent pupillary reflexes, mild diminutions in visual acuity, and dyschromatopsia, particularly for blue-yellow (6,23,25).

The most common basic visual dysfunction in AD may be abnormal contrast sensitivity functions. Several studies show a general depression across all spatial frequencies in AD (37,38) (see Fig. 1), and a subgroup of AD patients with more prominent visuospatial problems have a greater depression at the lower spatial frequencies (36,38). Others may have failed to find contrast sensitivity difference between dementia and normal aging.

FIG. 1. Contrast sensitivity function curves for 19 patients with moderately advanced clinically probable Alzheimer’s disease and 19 normal age-matched controls.
possibly because of methodological differences (10). Furthermore, in a recent study of 19 AD patients compared to 19 normal elderly controls, lower contrast sensitivities at a low spatial frequency (2 cycles per degree [cpd] alternating at 7.5 Hz) correctly predicted the presence of AD in 89.5% of the patients (37). The decrease in contrast sensitivities may be the source of much of the visual impairment found in AD patients and may be a consequence of the pathology in both the optic nerves and the visual association cortex.

**COMPLEX VISUAL FUNCTIONS**

**Constructional Disturbances**

Visual system dysfunction contributes to the constructional disturbances in drawing, copying, and block assembly that are common in dementia and widely used to screen for AD (29). However, constructional disturbances are nonspecific and may reflect not only visual disturbances, but also problems with motor praxis, conceptual abilities, executive functions, or some combination of these (39). Constructional disturbances occur in AD, particularly with decreased metabolism in the posterior right hemisphere (18); however, these disturbances may also result from lesions in different parts of the brain (39).

Screening tests for constructional difficulties most frequently involve drawing or copying simple two-dimensional objects, such as a simple design or a cross, and simple three-dimensional objects, such as a cube or an open box (see Fig. 2). Drawings of more complex spatial figures, such as the face of a clock, are frequently abnormal in AD, but can result from disturbances in several areas of cognition. Copying or assembling blocks are also good tests for constructional difficulties (40). Constructional errors on these tests include impoverishment (omission of essential features), fragmentation (loss of spatial relationships and faulty orientation), cramping (smaller with perseveration across items), and, on copying, a "closing-in phenomenon" (the copy overlaps the model) (29). These elements may reflect a disturbance in both hemispheres, with impoverishment related more to the left hemisphere and fragmentation more to the right posterior hemisphere (29).

**Perceptual Disturbances**

Several visuoperceptual processes are commonly disturbed in AD (4). Abnormal figure-ground discrimination occurs in most AD patients, and abnormal visual synthesis, the ability to conjoin different parts of a stimulus, is frequently present (4). AD patients also have difficulties on complex visual form discrimination tasks, although, when administered untimed, they may be able to successfully perform these tasks in a slow, serial feature-by-feature fashion (4,7,41). Other areas of visual perception in AD need further exploration; however, there is some evidence for declines in depth perception (6,19,42), the discrimination of line orientation (43), backward pattern masking (10), and possibly, visual memory, visual imagery, and the detection of movement.

Screening tests for visual perceptual difficulties in AD most commonly include tests of figure-ground discrimination (overlapping, cross-hatched, or hidden figures) and visual synthesis (visual completion or closure tasks) (39,44,45) (see Fig. 3). More detailed assessment of visual perception involves the administration of neuropsychological tests (44).

**Spatial Agnosia and Balint's Syndrome**

Patients with AD frequently have difficulties locating objects in space (4-9). They have lost the
sense of "whereness" (9) and are clumsy in their attempts to reach for objects or avoid bumping into them. This "spatial agnosia" is responsible for the most common visual complaints in AD, such as visual localization difficulties and spatial reading problems ("spatial alexia") (5,6) (see Table 2). Spatial agnosia is associated with abnormal spatial attention or the ability to focus on some spatial location to the exclusion of others. What little work has been done suggests that AD patients have a decreased capacity of spatial attention, difficulties in moving their focus of attention to a new focus in the periphery, and, even when their visual fields are normal, have a smaller, constricted field of view (8,45,46).

The spatial agnosia may result in abnormalities in scanning, searching, and hand–eye coordination severe enough to constitute Balint's syndrome in up to 20% of AD patients (4). These patients "see," but may have to be led as if blind because of the degree of visuospatial impairment (8,47). Patients with Balint's syndrome can only attend to a single visual object at a time ("simultanagnosia"), have inaccurate eye movements to visuospatial locations (oculomotor apraxia), and cannot accurately direct hand or other movements by visuospatial relationships (53-55). Simultanagnosia may result in part from a relative weakness of the peripheral fields (49,50) with suppression of extrafoveal images (51,52), or from the interferences of low spatial frequencies necessary for the spatial integration of images (36). The visuomotor disturbances of oculomotor apraxia and optic ataxia may result in an inability to explore the peripheral fields due to disruption of saccadic and fixation neurons in the posterior parietal lobe (53-55). Finally, investigators have questioned whether or not demented patients with full-blown Balint's syndrome constitute a separate degenerative disorder; however, it is likely that these patients are a subpopulation of AD, consistent with the broad spectrum of visual disturbances that occur in this disorder (4,8,47).

Screening tests for spatial agnosia and Balint's syndrome include tests for spatial inattention, spatial localization, simultanagnosia, oculomotor praxis, and optic ataxia. Line-crossing tasks, where various lines on a paper are presented for bisection, are tests for visual hemi-inattention or neglect of a part of the visual field. Spatial localization tests include finding random dots in a visual field or searching for specific items in a picture. Simultanagnosia tests include distinguishing the larger picture when there are "unlinked" items (e.g., two unconnected circles versus two circles "linked" to make a pair of glasses) or when there are smaller embedded figures (e.g., smaller letters combined to form a larger letter). Oculomotor praxis tests involve moving the eyes to visual stimuli, particularly into the peripheral fields, and optic ataxia tests involve moving the hands under visual guidance in reaching for objects or through openings in a box (55).

Environmental Disorientation

Disorders of environmental orientation are common in AD and are operationally defined as difficulty with following a route (5,56,57). Patients with environmental disorientation are unable to find their way around unfamiliar surroundings, a hospital ward, or even their homes, and may have a tendency to wander (56-58). The inability to follow a route may be due to a primary memory disorder for spatial concepts ("topographic amnesia") or to a primary disorder in the visual recognition of landmarks and spatial relationships ("topographicalagnosia"). Both of these forms of environmental disorientation occur in AD. In addition, a related disturbance that is common in AD is geographical disorientation—the inability to read a map.
Environmental orientation is tested by following, describing, drawing, or learning a route. With topographic amnesia, the patients are unable to describe or draw a previously familiar route and are unable to learn a new route. When topographic agnosia, patients may be able to describe or draw a previously familiar route or even learn the map of a new route, but they cannot recognize the route itself or its landmarks. Geographical disorientation tests involve placing well-known cities or landmarks on a map.

Visual Agnosia

Visual object recognition problems or “agnosia” are present in almost half of AD patients and can be a major source of disability from the dementia (4,6). Visual agnosias are deficits in the recognition of visually presented material in the absence of basic visual or general cognitive deficits sufficiently severe to otherwise account for the recognition problems (41,45,59). There are traditionally two forms of visual agnosia: an “apperceptive” form, where the inability to recognize an object is associated with significant impairments in visual perception, and an “associative” form, where perception is sufficiently intact, as indicated by the ability to visually match objects, but the object’s visual representation is isolated from its other cognitive associations. The existence of a true visual agnosia in AD has been controversial, as demented patients may fail to recognize an object from failure to understand or cognitively interpret the visual representation, especially if it was altered by an abnormality in basic visual functions (52,60). However, some AD patients have had visual recognition difficulties in the presence of normal basic visual functions and only mildly impaired general cognition (4,6,61). In sum, the visual recognition problems in AD represent a true visual agnosia and may be either of the “apperceptive” or of the “associative” form.

Screening tests for visual agnosia in AD involve the identification of actual common objects, simple photographs of common objects, and simple drawings of common objects. Actual objects are less difficult to recognize than photographs or line drawings. Testing for agnosia involves excluding language difficulty, causing inability to name the objects, excluding global recognition difficulties not restricted to the visual sphere, and assessing the apperceptive-associative dichotomy. In order to exclude the effects of language difficulties, patients must identify visually missed objects, unseen, by touch or sound. Finally, in order to assess the apperceptive-associative dichotomy, patients must identify missed objects by visual matching with a second series of identical objects (or, alternatively, correctly copy unrecognized drawings).

Facial Identification Problems

There are several forms of facial identification problems in AD. Prosopagnosia, the inability to recognize famous faces, is a perceptual disturbance usually associated with other perceptual discrimination difficulties (62) and occurs to some degree in most moderately advanced AD patients (5). True prosopagnosia is distinct from an inability to remember the specific faces tested and, when extreme, demented patients may not recognize themselves in the mirror (the “mirror sign”). Testing involves the presentation of photographs of famous individuals that cannot otherwise be distinguished on the basis of salient features, such as beards or glasses. Patients with AD also have difficulty discriminating unfamiliar faces, as tested by the Benton Facial Recognition Test (40). Finally, many AD patients suffer from Capgras syndrome, a related form of misidentification of others (26). In Capgras syndrome, the AD patient denies the identity of a person and claims that the person has been replaced by a look-alike, often with a malevolent intent. The Capgras syndrome appears to be tied to paranoid thinking, but complex visual processing may contribute to this phenomena.

Hallucinations

Hallucinations may occur in up to 20% of patients with AD, and 80–90% are in the visual sphere (26). Visual hallucinations in AD are usually formed, animate, changing, and often in color, illusory, and frightening (26). The hallucinations are not associated with clear evidence of eye pathology or basic visual impairment; however, the combination of visual perceptual disturbances and paranoid thinking in AD may predispose to paranoid hallucinatory experiences. Other positive phenomena, such as illusions or palinopsia, are much rarer in AD.

Oculomotor Functions

Eye movement recordings in AD patients show saccadic pursuit (33), increased latency and hypometric saccades (30,32), fixation instability (30,35),
inability to inhibit anticipatory saccades (30,31), disorganized scanpaths (34), and, as previously discussed, oculomotor apraxia (8) (see Table 4). In addition to nonspecific disturbances in pursuit eye-tracking, patients with AD show a prolongation of saccadic latency, are likely to undershoot a small target displacement by 10–30%, and may have a decreased peak velocity for large amplitude saccades (40°) but not for small ones (30,31). The cortical neuronal loss in AD may also impair the ability to maintain or release fixation, and saccadic intrusions, such as large square-wave jerks, may intrude into fixation (30–35). There are difficulties in suppressing anticipatory saccades, which, in the extreme, may constitute a "visual grasp reflex" with inability to inhibit a saccade to an extraneous visual stimuli (30,31). Patients with AD also have abnormal scanpaths with inability to concentrate on high information areas of a picture or scene (34). In sum, the prolonged saccadic latency and the inability to suppress anticipatory saccades may be the most characteristic oculomotor findings in AD (31).

CONCLUSIONS

A range of disturbances of the visual system is common in AD. The most common complaints involve difficulties in visuospatial orientation, but these patients also have visual recognition difficulties, such as visual agnosia and abnormal facial identification. On examination, AD patients may have abnormal visual fields, VEPs, contrast sensitivity functions, and eye movement recordings, as well as tests of complex visual processing. The evaluation of complex visual functions requires special attention to the effects of basic visual problems and other cognitive deficits on complex visual tasks. Difficulties with constructions, figure-ground discrimination, and visual synthesis are almost universally present in moderately advanced AD. Visual agnosia, although less common, may contribute disproportionately to the disability from AD. Finally, some demented patients develop spatial agnosia sufficiently to constitute Balint's syndrome. The prevalence of these visual changes in moderately advanced AD patients suggests overlapping subpopulations with abnormalities in contrast sensitivity functions, constructions, figure-ground discrimination, and saccadic latency in nearly 100%, visuospatial difficulties in about two-thirds, abnormalities in visual recognition in about one-half, visual hallucinations in about 20%, and the full triad of Balint's syndrome also in as many as 20%.

Further research in AD should establish management strategies, such as the use of color coding and card-hole and other visuospatial guidance tools (6), and the relationship of these visual system disturbances to the concentration of neuropathology in visual association cortex and the optic neuropathy found in AD.

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TABLE 4. Oculomotor disturbances in Alzheimer's disease

| Abnormal pursuit: Saccadic pursuit, catch-up saccades |
| Abnormal saccades: Prolonged latency, hypometric/dysmetric, decreased velocity for long jumps |
| Abnormal fixation: "Sticky" or "wandering"; saccadic intrusions |
| Inability to inhibit anticipatory saccades; "visual grasp reflex" |
| Abnormal visual search and scanpaths |
| Abnormal visually guided eye movements (oculomotor praxis) |

Literature Abstracts

Acute Macular Neuroretinopathy. Miller MH, Spalton DJ, Fitzke FW, Bird AC. *Ophthalmology* 1989;96:265–9 (Feb). [Reprint requests to Dr. M. H. Miller, Moorfields Eye Hospital, City Road, London EC1V 2PD, England.]

The authors report five cases of this rare disorder that appears mainly in young women and causes paracentral scotomas corresponding to irregular dark areas in the maculae. This syndrome seems to occur with or after systemic, probably viral, illness and generally the field defects improve with time.

_Lyn A. Sedwick, M.D._

Isolated Metastasis to Optic Nerve from Medulloblastoma. Garrity JA, Herman DC, Dinapoli RP, Waller RR, Campbell RJ. *Ophthalmology* 1989;96:207–10 (Feb). [Reprint requests to Dr. J. A. Garrity, Mayo Clinic, 200 First Street SW, Rochester, MN 55905.]

A 19-year-old man was found to have a cerebellar medulloblastoma that was treated with gross total resection and radiation therapy. Unilateral visual loss occurred 28 months later secondary to a biopsied optic nerve sheath metastasis. He died of metastatic disease 20 months later.

_Lyn A. Sedwick, M.D._


The authors report a 68-year-old woman with gradual bilateral loss of vision, whose radiologic studies showed thickened optic nerves in the posterior orbit. Biopsy of one nerve revealed a necrobiotic, granulomatous inflammation with vasculitis. No other lesions were found. The patient had no clinical or laboratory evidence of any other disease known to affect the optic nerves. Thus, a diagnosis of idiopathic inflammatory syndrome of the orbit was made.

_Lyn A. Sedwick, M.D._

Visual Outcome After Surgical Removal of Craniopharyngiomas. Repka MX, Miller NR, Miller M. *Ophthalmology* 1989;96:195–9 (Feb). [Reprint requests to Dr. M. X. Repka, Bi-35 Wilmer, Johns Hopkins Hospital, Baltimore, MD 21205.]

Thirty patients with craniopharyngioma, 12 less than 18 years of age and 18 more than 18 years of age, were examined pre- and postoperatively at Johns Hopkins Neuro-ophthalmic Unit between 1975 and 1987. Patients younger than 18 years had slightly worse visual dysfunction on presentation than those who were older, and the authors found no improvement in visual function over that achieved in the first month postoperatively. Also, the visual improvement postoperatively with these tumors is less than that with pituitary adenomas or suprasellar meningiomas.

_Lyn A. Sedwick, M.D._

Macular Abnormalities in Papilledema from Pseudotumor Cerebri. Gittinger JW Jr., Asdourian GK. *Ophthalmology* 1989;96:192–4 (Feb). [Reprint requests to Dr. J. W. Gittinger, Jr., University of Massachusetts Medical Center, 55 Lake Avenue North, Worcester, MA 01655.]

Three patients with pseudotumor cerebri developed mottled macular pigmentation and choroidal folds. Two initially had macular star figures in addition to papilledema. The authors advise that "previous papilledema should be considered in the differential diagnosis of macular pigmentation."

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Lyn A. Sedwick, M.D.
Primary Liposarcoma of the Orbit: Problems in the Diagnosis and Management of Five Cases. Jakobiec FA, Rini F, Char D, Orcutt J, Rootman J, Baylis H, Flanagan J. Ophthalmology 1989;96:180-91 (Feb). [Reprint requests to Dr. F. A. Jakobiec, Department of Ophthalmology, Massachusetts Eye and Ear Infirmary, 243 Charles Street, Boston, MA 02114.]

Five cases of primary liposarcoma of the orbit are reviewed, with attention to clinicopathological features and diagnostic studies and treatment. Computerized tomography was done in all, ultrasound and magnetic resonance imaging in some. The authors recommend excision and postoperative radiation therapy routinely and orbital exenteration only for postsurgical failures or when there is concern about direct spread from the orbit. The pathology of these lesions is well discussed.

Lyn A. Sedwick, M.D.


An immunocompetent 24-year-old man developed bilateral visual loss attributed to optic neuritis that was unsuccessfully treated with corticosteroids after an initially negative result from a computerized tomographic scan. He was subsequently found to have sinusitis with orbital extension that yielded the fungus *Bipolaris hawaiiensis*. His rocky clinical course included relapse after amphotericin treatment and he became totally blind in both eyes. This fungus has been previously reported to cause sinusitis with orbital extension in another immunocompetent host and the authors warn that "ophthalmologists must be aware of the virulence of these darkly pigmented fungi and their ability to devastate orbital structures from a contiguous sinusitis."

Lyn A. Sedwick, M.D.


A 33-year-old woman with migraine and a previous central retinal artery occlusion, possibly ophthalmic artery occlusion, in her left eye at age 25 presented with a central retinal artery occlusion in the right eye. The authors postulate "the combination of bilateral [disk] drusen and migraine may have contributed to the occurrence of bilateral sequential central retinal artery occlusion."

Lyn A. Sedwick, M.D.


A 10-year-old boy with migraine headaches had two episodes of a small-angle comitant esotropia and diplopia witnessed by the author after a headache. Examination was otherwise normal and all findings were normal after the episodes.

Lyn A. Sedwick, M.D.

Intracranial compressive optic neuropathy, at the opening of the optic canal, can be a challenging clinical and radiologic diagnosis at best. Drs. Unsold and Seeger provide a very nicely illustrated text describing their experience with optic neuropathy secondary to compressive meningiomas and dolichoectasia of the carotid artery, as well as optic canal compression from pneumosinus dilatans of the sphenoid bone. There are 10 chapters, the first of which is a very nice anatomical review of the anterior visual pathways with cadaver dissection. There are several chapters concerning the clinical manifestation in patients who have compressive optic neuropathy, including differential diagnosis and physical findings. Perhaps the greatest emphasis of this text, as well as the majority of the case presentations, is spent on the recognition of dolichoectasia of the internal carotid artery with its encroachment on the optic nerve in the opening of the intracranial portion of the optic canal. The possible influence that pneumosinus dilatans has on the further luxury compression of the internal carotid artery on the optic nerve is also emphasized. Most of the case reports have beautiful computed tomography scan images outlining in detail the exact location of the optic nerve compression by either the narrowed optic canal or the internal carotid artery. There is a very nice discussion of the neurosurgical approach to decompression of the intracranial portion of the optic canal by Dr. H. R. Egger. Photographic evidence of compression of the optic nerve by the superior dural fold, as the optic nerve enters the cranial vault, is quite impressive.

Drs. Unsold and Seeger have reemphasized an often forgotten pathological process that certainly causes optic nerve compression, albeit somewhat uncommonly. The practicing neuro-ophthalmologist cannot help but be more attuned to this process, as well as have his neuroradiologic diagnostic skills improved from reading this text.

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It is quite frustrating, indeed, to expect an updated and "fresh" approach to all of the various aspects of the field of neuro-ophthalmology today. Drs. Smith and Katz have fortunately provided us with an excellent weapon with which to attack these complexities! They have managed to accumulate an excellent cross-section of the leading neuro-ophthalmologists, neurologists, and internists in the country, to provide us with an excellent review and update of many problem areas in neuro-ophthalmology.

It would be difficult to list each specific chapter with its topic and author, as there are a total of 37 chapters. Good review articles include blepharospasm, as well as its treatment, congenital nystagmus and its associated visual problems, opsinclonus, as well as other forms of nystagmus, opsoclonus, as well as see-saw nystagmus. There are also nice updates on ocular myasthenia, as well as the ophthalmic manifestations of neurofibromatosis. Intraocular and intracranial infectious diseases are reviewed nicely, including the Lyme/AIDS/syphilis chapter by Dr. Smith, as well as review of fungal infections in the brain by Dr. Joseph C. Parker et al.

Various causes of papilledema, including papillophlebitis, papilledema from arteriovenous malformation, pseudotumor cerebri, intracranial tu-
mors, including craniopharyngiomas, gliomas, and orbital tumors are reviewed in detail. The chapter by Dr. Roger J. Packer concerning the chemotherapeutic approach to chiasmatic gliomas is of particular interest.

Retinal problems, including the Hippel-Lindau syndrome, retinitis pigmentosa, and ocular albinism are reviewed also in detail.

Additional topics that are nicely reviewed include neuroimaging of the cavernous sinus by Dr. Jerome J. Sheldon et al. and Donald W. Chakeres et al., as well as neuroimaging of horizontal gaze disorders by Dr. Robert F. Saul.

The neurologist will be happy to see discussions by Dr. Jonathon Trobe on visual problems in Alzheimer's disease, as well as Dr. Charles Manner's discussion on visual system dysfunction in Parkinson's disease. All readers should greatly benefit from the hypertension update by Drs. Barry J. Masterson and Joseph A. Asch.

Perhaps the most valuable segments of this text are the frequent anecdotal comments by Dr. Smith, relating his own personal experience from his neuro-ophthalmological practice to that of each author. There are numerous “practice pearls” to be garnered here.

In summary, Drs. Smith and Katz have done a very nice job in bringing together various experts in their respective fields across the country for basic reviews of topics of particular importance to the practicing neuro-ophthalmologist. I know of no other single source that would get such a nice review in these various fields. For the price, this text should be required reading for all neuro-ophthalmologists.

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Dr. Roy has once again provided us with an excellent reference text for differential diagnosis. I doubt that any ophthalmology resident in the United States is unaware of the usefulness of this book, particularly before weekly grand rounds. Since the first edition in 1972, Dr. Roy has provided the ophthalmology resident with an invaluable source of differential diagnosis based not only on specific ocular findings, but clinical presentation in specific diagnosis as well.

In this latest edition, Dr. Roy has not only expanded his lists of differential diagnoses, but also added a diagnostic decision table, which includes symptoms typically or rarely found in most major disease processes, as well as clinical findings of the eye and orbit. I found these to be quite helpful after some initial practice time. The chapters are arranged according to anatomical structure, such as orbits, lids, lacrimal system, extraocular muscles, etc. There are specific chapters on visual field defects, visual disturbance, including transient or permanent loss of vision, visual complaints, and head position. These last chapters should be of particular interest to the practicing neuroophthalmologist.

In summary, Dr. Roy continues to provide us with an excellent updated addition of ocular differential diagnosis that is now beginning to evolve into a true clinically oriented “handbook.” I think it should be in all ophthalmologists’ library and certainly within immediate arm’s distance of all ophthalmology residents!

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As Dr. Reinecke states in the preface of this text, the author's goal is to give the reader an “updated, comprehensive, but understandable, report on subjects of interest to most ophthalmologists.” There are a total of 12 chapters, which review all fields of ophthalmology.

Dr. Soham Hayreh begins the text with a nice chapter on arterial hypertension and its ophthalmic complications, including both hypertensive retinopathy and optic neuropathy. There are excellent fluorescein photographs, which demonstrate the retinal pathological changes in a nice way.

Dr. Clifford Terry provides us with a nice review of postoperative astigmatism from cataract surgery, with tips on both how to prevent it and how to treat it. Surgical techniques to reduce corneal astigmatism are also discussed.
BOOK REVIEWS

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Dr. Peter Laibson provides us with an excellent review on corneal erosions, as well as their diagnosis and treatment. This not only provides us with an excellent photographic review of the pathology, but also excellent suggestions for office and surgical management.

Drs. Shirley Wray and Georgina Scholl provide a nice review of optic neuritis and its relation to multiple sclerosis. Current controversies in the association of these two diseases are discussed. Also interesting to the practicing neuroophthalmologist is the excellent review of magnetic resonance imaging (MRI) use in ophthalmology by Dr. Mahmood Mafee. Dr. Mafee once again attempts to educate us in the basic principals of MRI physiology in a relatively basic approach. Orbital and ocular pathology is also discussed in detail. Also of interest is the chapter on the clinical use of the visual evoked cortical potential by Dr. Michael Fendick. The advantages and disadvantages of this technique are discussed in detail.

Other interesting chapters include an excellent review of central serous chorioretinopathy by Drs. G. Robert Hampton and Peter B. Hay, and ophthalmic anesthesia by Dr. W. Sanderson Grizzard. The ophthalmic surgeon will enjoy reviewing the various techniques for retrobulbar and periorbital blocks that Dr. Grizzard discusses. The last chapter by Dr. Stephen L. Bosniak is a comprehensive review of the history, pathogenesis, and management of blowout orbital floor fractures. Although Dr. Bosniak tends to be an “early treater,” he reviews the risks and benefits of all approaches in an objective way.

In summary, Dr. Reinecke has provided us with an excellent review of general ophthalmology that is, indeed, “welcome down-to-earth reading for the busy practitioner.”

Bradley K. Farris, M.D.
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Letters to the Editor

Bedside Monocular Indirect Ophthalmoscopy

To the Editor:

Neuro-ophthalmologists frequently perform fundus examinations at the bedside. Occasionally, I am unable to see fundus details with the direct ophthalmoscope. This is due either to hazy media or to high refractive errors. In these cases I have found it very useful to use a method of monocular indirect ophthalmoscopy. The Welch Allyn Finoff transilluminator with fiberoptic light source provides bright and uniform illumination. I use this light source and a 20-diopter aspheric lens to perform monocular indirect ophthalmoscopy, as follows: Hold the transilluminator with the right hand and shine the light into the pharmacologically dilated pupil. With the left hand, hold the lens (Fig. 1) about 10–15 cm from the patient’s eye. Then move the lens backward and forward until one can see clear fundus details.

I have found this method very helpful, especially in cases with corneal opacity such as a corneal leukoma or due to recently applied ocular ointment, cataracts, or vitreous hemorrhage that would not allow good fundus examination with the direct ophthalmoscope. Naturally this method serves only as a quick check, and if an abnormal finding is seen I would perform conventional binocular indirect ophthalmoscopy.

The principle of monocular indirect ophthalmoscopy is well known, and any bright beam could serve as a light source. My experience with this simple method is excellent, and I would like to recommend it to colleagues.

Benjamin Hartmann, M.D.
Neuro-ophthalmology Service
Department of Ophthalmology
Henry Ford Hospital
Detroit, Michigan

Hysterical Bitemporal Hemianopia "Cured" with Contact Lenses

To the Editor:

Recently, an 18-year-old woman had sudden onset of decreased peripheral vision in both eyes. Her past medical history was unremarkable. The patient had had one previous episode of "tunnel vision" 2 years previously. Complete physical and neurological examination results were normal. Corrected visual acuity was 20/20 OU. Refractive error was OD, −0.50 sphere; OS, −0.75 + 0.25 × 88. The remainder of the ophthalmologic examination, including pupils, extraocular movements, and funduscopic examination, was normal. Automated perimetry demonstrated a bitemporal hemianopia (Fig. 1). Goldmann perimetry and tangent screen testing confirmed these findings but showed inconsistencies in the borders of the defects along the vertical meridian (Fig. 2.) Cranial computed tomography and pattern reversal visual evoked responses were normal. The patient expressed interest in contact lenses and was fitted.

FIG. 1. Hold the transilluminator with the right hand and shine the light into the pharmacologically dilated pupil. With the left hand, hold the lens about 10–15 cm from the eye. Move it until clear fundus details are visible.
Letters to the Editor

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with daily-wear soft lenses. She claimed return of full visual fields "within minutes" of inserting the lenses. Repeat automated perimetry was normal.

A variety of visual field defects have been associated with malingering and hysteria (1). These commonly include generalized constriction of the visual field with no expansion to increasing test distance ("tunnel vision"), spiraling or crossing of isopters, and monocular temporal hemianopia (2). Although hysterical bitemporal hemianopia has been reported (3), it is exceedingly uncommon. Automated perimetry documented a bitemporal hemianopia in our patient, yet manual forms of field testing failed to confirm a reproducible defect. Smith and Baker (4) have reported that current automated techniques cannot differentiate functional from organic visual loss. Our findings support this contention. This case emphasizes the need to include functional visual disorders in the differential diagnosis of bitemporal field loss. It is also unique

FIG. 1. Automated threshold perimetry of the central 60° demonstrating bitemporal hemianopia.

FIG. 2. Goldmann perimetry of the same patient demonstrating incomplete bitemporal hemianopia. Note the inconsistencies along the vertical meridian.
in that field abnormalities resolved with contact lenses.

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REFERENCES

Announcement

NEW PROSPECTIVES IN OTOLARYNGOLOGY,
HEAD AND NECK SURGERY

A seminar jointly sponsored by the Department of Otology and Laryngology at Harvard Medical School, the Joint Center for Otolaryngology, Beth Israel and Brigham & Women’s Hospital, and the Service D’Oto-Rhino-Laryngologie et de Chirurgie Cervico-Faciale, Universite Rene Descartes, Faculte de Medecine, Necker Enfant Malades, Hospital Laennec, will be held in Paris, June 26-29, 1990. The international symposium will bring together outstanding faculty from the United States and Europe to explore the newest development in otolaryngology—head and neck surgery. The symposium will cover the entire gamut of this specialty. For more information, contact Harvard Medical School, Department of Continuing Education, Boston, Massachusetts 02115. Tel: (617)732-1525 (Monday–Friday 10:00 a.m.–4:00 p.m. eastern time). (617)732-1562.
Bitemporal Hemianopia Associated with Dolichoectasia of the Intracranial Carotid Arteries

Michael L. Slavin, M.D.

Slowly progressive visual loss with bitemporal hemianopia is the hallmark of chiasmal compressive syndromes. Specific causes of the chiasmal syndrome include pituitary adenoma with suprasellar extension, suprasellar meningioma, craniopharyngioma, and, less frequently, aneurysm of the supraclinoid carotid artery. I recently saw a case in which a progressive chiasmal visual field defect was associated with dolichoectasia of the intracranial carotid arteries.

A 73-year-old hypertensive woman noticed progressive visual loss in her right eye, nonspecific

**FIG. 1.** Perimetry (Goldmann) reveals bitemporal hemianopia typical of optic chiasmal dysfunction.
intermittent frontal headaches, and difficulty with a sense of smell over 9 months. On neuroophthalmic examination, visual acuities were 20/50 right and 20/30 left with normal color vision. A 1+ right afferent pupil defect was seen. Visual fields (Goldmann) revealed a pattern of bitemporal hemianopia (Fig. 1). Fundi were normal except for subtle temporal pallor of the right optic disc. Her sense of smell was diminished to tea leaves and cologne.

Computed tomography (CT) of the suprasellar area showed atherosclerotic enlargement of the intracranial carotid arteries (dolichoectasia) (Fig. 2). The basilar artery as well as the middle cerebral arteries were also abnormal. Conservative treatment was recommended, and vision was stable 6 months after.

Ectasia of cerebral arteries is a rare cause of the chiasmal syndrome (1,2). It is unclear whether the visual disturbance is due to a compressive effect (as most likely is the cause with neoplasia and aneurysm) or to ischemia from thrombosis of branches of the abnormal parent arteries (3). Dolichoectasia (or fusiform aneurysm) should be distinguished from focal arterial enlargement due to saccular (or berry) aneurysms. Diagnosis of the former may often be made with CT or magnetic resonance imaging without angiographic confirmation.

REFERENCES