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Syphilitic Optic Atrophy

The article “Progressive Visual Loss in Syphilitic Optical Atrophy” by Sacks, Osher, and Elconin in this issue, raises again the question of ocular and neurological involvement in late syphilis. The subject was discussed in an editorial in the June 1982 issue of this journal as well as in a review article by Prof. Pierre Collart in that issue. The article by Sacks et al. emphasizes that this disease is still with us, and also provides evidence that antibody formation was occurring within the central nervous system in that patient, and further emphasizes that the dose of penicillin therapy for late syphilis is really unknown at this time.

The point that I should like to address here, however, is the pathogenesis of syphilitic optic atrophy, and the mechanisms whereby one may encounter a pale disc in this disease. Dr. Sacks and associates emphasize the occurrence of optic atrophy following syphilitic optic neuritis and perineuritis, or as described in the older literature as “secondary” optic atrophy. In such instances, one certainly would agree with the assumption that vision loss would be unusual after the atrophic stage had occurred following an initially swollen disc. However, it is probably pertinent to review some of the older literature with regards to “primary optic atrophy” in syphilis and also some of the statements about “tabetic optic atrophy.” In Dr. Walsh’s second edition, Clinical Neuro-opthalmology (1957), there is a more expanded section of ocular and neurosyphilis than appeared in the later editions, and some important points are made about “tabetic optic atrophy.” The following quotes are relevant at this point. “The visual disturbances usually appear with the onset of atrophy and increase as the atrophy increases, but there are instances in which the discs may be pale for as long as four or five years before the visual functions are affected (Uthoff). The loss of vision progressed with extreme rapidity in some cases. . . . The loss of vision may be slowly progressive as we have observed in many instances.” Moore and his coworkers stated that of 249 untreated patients 70% were blind within three years, 90% within five years, and all within nine years. Their criterion of blindness was anything below 10/200 in the better eye.” The statement was also made that “gradual loss of vision is more frequently observed as it is characterized almost invariably by concentric narrowing of the visual fields.” I would recommend to the reader that an excellent review would be obtained by reading pages 541-601 in Dr. Walsh’s second edition. Two classic papers also to be noted are: L.L. Sloan and A.C. Woods: “Perimetric Studies in Syphilitic Optic Neuropathies.” Arch. Ophthalmol. 20: 201, 1938; and A.C. Woods. “Syphilis of the Eye.” Am. J. Syphil. Gonor. Ven. Dis. 27: 133, 1943. Finally, Syphilitic Optic Atrophy by Walter L. Bruetsch, Charles C Thomas Co., Springfield, Illinois, 1953, offers a complete, thorough review of the subject.

An excellent recent review is the chapter on “Syphilis” by Dr. King K. Holmes, chapter 146, pages 716-726, in Harrison’s Principles of Internal Medicine (9th ed.), McGraw-Hill Book Co., 1980. The importance of this subject to the practitioner needs additional emphasis. According to the United States Department of Health and Human Services, in 1957, there were 6,251 reported cases of infectious syphilis. However in 1977, there were 20,352 reported cases of infectious syphilis. Furthermore, in 1977, in addition to these 20,000 cases of primary and secondary syphilis, an additional 21,297 cases of early latent syphilis were reported. These facts gain additional emphasis when it is recalled that many cases of syphilis seen by practitioners are not reported. Of 43 million blood tests done in the United States in 1977, 1.5 million were reactive (3.5%). Over half of all reported early syphilis cases were detected as a direct result of either contact tracing or serologic testing. Another significant point is that of all men with infectious or early latent syphilis interviewed in the United States between 1977 and 1978, half were known to be homosexual or bisexual.

A helpful mnemonic in syphilis is known as the “rule of sixes.” The Bible speaking of a man known as “the beast,” makes an interesting statement (Revelations 13:17-18) “And that no man might buy or sell, save he that had the mark, or the number of his name. Here is wisdom. Let him that hath understanding count the number of the beast: for it is the number of a man; and his number is Six hundred threescore and six.” If one remembers that number—666—it may be helpful when considering the following points, which were obtained from Dr. Holmes’s excellent article (in general).
Editorial: Syphilitic Optic Atrophy

The Rule of Sixes in Syphilis
1. Treponema pallidum has six spirals
2. It is six microns long
3. It is made of six fibrils
4. Each fibril has six laminas on electron microscopy
5. Within six weeks from exposure, the primary lesion appears
6. The chancre persists up to six weeks
7. Six weeks after healing of the chancre, secondary syphilis appears
8. Secondary stage subsides within six weeks, and patient enters the latent stage.
9. Six point five percent of untreated cases develop symptomatic neurosyphilis
10. Sixteen percent develop benign tertiary syphilis
11. Aortitis is found in up to 60% of untreated cases at autopsy
12. Six indications for serum FTA-ABS test are: light-near dissociation of the pupil, optic atrophy, uveitis, retinitis pigmentosa, interstitial keratitis, and dislocated lens
13. There are six letters in FTA-ABS test

The point to be emphasized here is that syphilis may involve the optic nerve in many different routes. It may produce a primary intraocular optic neuritis, a vascular occlusion picture on the disc, or as in primary optic atrophy, it often begins in the arachnoid spaces around the optic nerve in its intracranial portion, just distal to the chiasm, and then spreads peripherally in the optic nerve sheath. It can also produce syphilitic optochiasmatic arachnoiditis. Patients can have syphilitic primary optic atrophy without presenting clinical signs or symptoms of tabes dorsalis. Bruetsch also points out that syphilitic optic atrophy can be progressing in patients with normal cerebrospinal fluid examinations. The clinician is again reminded of the importance of obtaining a serum FTA-ABS test in any patient with otherwise unexplained optic atrophy.

J. Lawton Smith, M.D.
Progressive Visual Loss in Syphilitic Optic Atrophy

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ROBERT H. OSHER, M.D.
HOWARD ELCONIN, M.D.

Abstract
A 62-year-old man had progressive visual loss from neurosyphilis while his optic disks appeared atrophic. At no time was there evidence of inflammation of the globe or of either optic disk. The presence of an extremely high IgG index and five oligoclonal bands in the cerebrospinal fluid indicated that the central nervous system was synthesizing antibody specific to Treponema pallidum. Recent investigations indicate that much larger doses of penicillin are necessary for adequate treatment of neurosyphilis than have been recommended previously.

In the past, visual loss produced by the optic neuropathy of neurosyphilis has been described as having an abrupt onset associated with swelling and engorgement of the optic disk. Optic atrophy may ensue.

We recently cared for a patient who upon initial presentation had bilateral optic atrophy with no evidence of ocular inflammation. His vision subsequently deteriorated over several months until the diagnosis of neurosyphilis was established. We are unaware of any previous report of progressive visual loss during the atrophic stage of syphilitic optic neuropathy.

Case Report
A 62-year-old man was first seen at the University of Cincinnati in November 1981 because of progressive loss of vision in the left eye that had begun a year earlier. Vision in the right eye deteriorated some time thereafter.

He first sought care about 6 months after visual problems began. At that time, visual acuity was 20/50 in the right eye and 20/400 in the left eye. Both disks were pale. The following month, vision had decreased to 20/70 in the right eye, while the left eye was limited to counting of fingers at 3 ft. The visual fields showed generalized constriction in the right eye in association with an inferior arcuate defect; in the left eye, only a small inferotemporal island remained (Fig. 1). The right optic disk was excavated, with a pale neural rim. The left disk was flat and diffusely pale. The arterioles were moderately attenuated on the surface of each nerve head. Computerized tomography and evaluation by an internist failed to reveal the cause of visual loss.

The patient denied previous serious illness or hospitalization, although questioning disclosed a history of possible venereal disease in 1945.

The social history was unremarkable. After discharge from the armed forces at the end of World War II, the patient lived at home with his wife and worked regularly. He smoked one or two cigarettes a day on and off for many years and was a social drinker. His diet was well-balanced.

At the time of examination in November 1981, vision was reduced to counting fingers at 12 in. in the right eye and questionable perception of light inferotemporally in the left eye. Visual field examination disclosed severe constriction of field in the right eye; no consistent responses were obtainable in the left eye (Fig. 2). The pupils were mid-dilated and reacted weakly only when light was shone into the right eye. The appearance of the optic disks was unchanged. There was no evidence of ocular inflammation or retinal disease. Electroretinography was normal. Fluorescein retinal angiography showed only a paucity of disk capillaries.

Because of the unexplained progressive visual loss, the patient was admitted to the University of Cincinnati Hospital for further evaluation. Routine physical examination on admission disclosed no abnormalities. The neurology consultant discovered peripheral neuropathy in addition to bilateral optic atrophy.

All of the routine hematologic and chemical studies were normal, but the fluorescent treponemal antibody absorption test was positive on two separate occasions, and the rapid protein reagin test was positive at a titer of 1:128 on one occasion and 1:64 on another. Computerized tomography of the brain and orbits was normal.
Figure 1. Visual fields at the first examination. An inferior arcuate scotoma and generalized constriction are present in the right eye. Only an island of vision is present inferotemporally in the left eye.

Figure 2. Visual fields 9 months later. The field in the right eye is markedly constricted. No field was recordable in the left eye.

Examination of the cerebrospinal fluid disclosed a protein concentration of 90 mg/100 ml; glucose, 38 mg/100 ml; red cells, 2080/mm³; and 66 other cells the differential count of which showed 82% lymphocytes, 5% neutrophils, 2% band cells, and 11% pia-arachnoid cells. The VDRL test on spinal fluid was positive at 1:16.

The lumbar puncture was repeated 6 days later and demonstrated less peripheral blood contamination. The IgG was 42.7 mg/100 ml (upper limit
of normal, 3.6 mg/100 ml) and the IgG index was 4.8 (upper limit of normal, 0.58 mg/100 ml). Five abnormal oligoclonal bands were present in the spinal fluid. The proteins in the peripheral blood were all normal.

The patient was treated with penicillin G sodium, 24 million units a day, by intravenous infusion for 14 days. During treatment, the penicillin level in the cerebrospinal fluid was 1.28 μg/ml.

The patient’s laboratory studies were repeated 14 weeks after penicillin therapy. The RPR on serum was positive at 1:128. All of the abnormal values on cerebrospinal fluid improved: the VDL was reactive at 1:4; protein, 50 mg/100 ml; IgG, 25.6 mg/100 ml; and the IgG index, 4.39. Only four oligoclonal bands were present. The cell count was down to 11/μm³, the differential of which was 78% lymphocytes, 10% neutrophils, 5% band cells, 2% eosinophils, and 4% pia-arachnoid cells. Neither the patient’s visual acuity nor visual fields have improved since he was first hospitalized. The appearance of his optic discs is also unchanged.

Discussion

This case report emphasizes three important features of management of neurosyphilis. First, vision continued to deteriorate during the period in which the optic disks were atrophic. Second, the abnormal oligoclonal bands in the spinal fluid and the elevated IgG index strongly suggest the synthesis of specific antibody to T. pallidum in the central nervous system. Third, recent reports indicate that large doses of penicillin may be necessary for adequate treatment of neurosyphilis.

This patient must be considered as having had optic atrophy resulting from neurosyphilis. The inferior arcuate field defect that was initially found in his right eye is definite evidence of primary optic nerve dysfunction. He thinks that he was infected 36 years ago, a time interval which is compatible with development of central nervous system syphilis. The optic nerve involvement is probably a manifestation of meningo-vascular neurosyphilis rather than parenchymatous syphilis, the latter of which is more typical of general paresis and tabes dorsalis.

We have been unable to find any report of a patient with neurosyphilis whose vision continued to worsen after the optic disks became pale. Visual loss usually occurs during an episode of acute papillitis. It is possible that our patient had bilateral disk swelling when the vision first failed, yet a dramatic progressive visual loss was documented despite the stable appearance of the atrophic disks.

We are not aware of any other reported cases in which modern immunohematological analyses have been performed on a patient with syphilitic optic atrophy. The extremely high IgG index points toward the synthesis of immunoglobulins within the central nervous system. The index is calculated as a fraction in which the numerator is the ratio of the IgG in the cerebrospinal fluid to that in the serum and the denominator is the ratio of the albumin in the cerebrospinal fluid to that in the serum. This calculated value corrects for variation of albumin and IgG concentrations in serum and produces a value that correlates positively with the synthesis of immunoglobulin within the central nervous system.

Oligoclonal IgG was identified in five abnormal electrophoretic bands in our patient. Vartdal and coworkers studied three patients with neurosyphilis and concluded that the oligoclonal IgG in the cerebrospinal fluid of patients with neurosyphilis represents a specific antibody directed toward T. pallidum. This is in contrast to the IgG in the spinal fluid of patients with multiple sclerosis, which is also synthesized intracerebrally but is not associated with demonstrable antigenic specificity.

Recent reports suggest that much higher doses of penicillin are required to treat neurosyphilis effectively than was thought previously. Short and coworkers have challenged the adequacy of the commonly recommended penicillin schedules for treatment of neurosyphilis.

Penicillin therapy may not be infallible in syphilis. In 1980, Greene, Miller, and Bynum reported a case in which 7.2 million units of benzathine penicillin G failed to cure neurosyphilis. Ducas and Robson emphasized that conventional dosage schedules failed to produce an acceptably high level of penicillin within the central nervous system in patients with neurosyphilis. The ideal therapeutic concentration of penicillin in spinal fluid for treatment of neurosyphilis is not known, but it is probably in the range of 0.1–0.4 μg/ml.

The therapeutic regimen given to our patient achieved levels of 1.28 μg/ml, which should have been sufficient to eradicate the organism.

Even “appropriate” treatment of primary syphilis may not be adequate to prevent neurosyphilis. Moskovitz and coworkers recently described the development of meningovascular syphilis in a patient whose primary lesion had been treated with 2.4 million units of benzathine penicillin G in accordance with the recommendation of the Centers for Disease Control.

This case emphasizes that adequate treatment with penicillin is essential in order to minimize the risk of subsequent progressive visual loss in patients with syphilitic optic atrophy.

References

Syphilitic Optic Atrophy


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Amaurosis Fugax for a Long Duration*

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Abstract
A 31-year-old Japanese male with unilateral bouts of transient visual loss lasting for variable periods of time has been observed. In spite of the prolonged duration of some of these attacks (the longest attack lasted 3½ hours), the patient sustained a relatively small amount of functional damage to the retina. Although the exact mechanism underlying these bouts of transient visual loss remains unknown, a discussion of the possible causes of such episodes, as well as the reasons for the relative preservation of vision in spite of the length of these attacks, is included.

Permanent retinal damage occurs within a relatively short period of time following a total ischemic event. We recently saw a patient with attacks of unilateral transient visual loss of variable duration. The peculiarity of the attacks was such that his visual function was not impaired severely, in spite of lasting a long period of time.

Case Report
A 31-year-old Japanese male engineer had his first attack of transient unilateral visual loss on October 30, 1977, after three glasses of beer. He noted that during the attack his right eye was blind for 50 minutes, after which time his vision slowly improved but never returned to the level preceding the attack. Since that time he has had similar attacks once or twice per week (Table 1). The patient has been known to suffer from right hemi-icrania since the age of 6 years, but has otherwise been healthy.

The patient was seen in the Department of Ophthalmology at Keio University on January 11, 1978. His best corrected visual acuity was 20/40 in the right eye and 20/15 in the left eye. With the exception of a small central depression in the right visual field, the remainder of the ophthalmologic examination, including ophthalmodynamometry, was normal. The fundus and fluorescein angiograms were normal. No carotid bruits were noted. The neurological examination was normal. Laboratory tests, including a complete blood count, serology, and glucose tolerance test, were normal. Skull and chest x-rays, CT-scan, and EKG were unremarkable.

Subsequent to his visit to our office the patient kept a log of his attacks (Table 1). On February 20, 1978, while in our office, the patient noted the onset of one of his attacks (Table 1). He was examined by one of us. Sludging was noted in one of the veins (Fig. 1, arrow). A fluorescein angiogram was performed, at which time the patient noted improvement in his vision. The angiogram was interpreted as normal (Fig. 2). The lack of venous collapse on the angiogram led us to question our original speculation of decreased flow. Subsequent examinations revealed a relative central scotoma and a decrease in the visual acuity of the right eye to 20/200.

Because of the peculiarity of the patient’s complaints and the patient's personality, psychiatric

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<th>Record of Attack Reported by Patient</th>
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<tr>
<td><strong>Visual Loss</strong></td>
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<td>Feb. 20, 1978</td>
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<td>Mar. 1, 1978</td>
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* Computed for the first 30 minutes. Vision gradually returned over the next 1 hour 45 minutes.

* See in text.
Amaurosis Fugax

Figure 1. Right fundus, normal appearance, on May 14, 1978. (Same findings on January 11, 1978 and February 20, 1978.)

Figure 2. Normal fluorescein angiogram, on January 13, 1978.
consultation was obtained. Tranquilizers were prescribed which seemed to reduce the frequency of the attacks. His subsequent visits to our office were sporadic. On September 29, 1978, he experienced his longest attack which lasted for 7 1/2 hours.

On February 18, 1980, we were fortunate to be able to examine his fundus during an attack. At this time the retinal blood flow appeared completely arrested. However, fluorescein angiography revealed extremely slow filling of the vessels. A representative picture of the angiogram 46 seconds after injection demonstrated minimal filling of the arterial tree with choroidal circulation present only temporally (Fig. 3). At 86 seconds (Fig. 4), all of the retinal arteries were filled, but the veins were just beginning to show some laminar flow. The slowed retinal circulation suggested a possible circulatory abnormality, so a cardiovascular angiographic study was obtained. A congenital anomaly of the origin of the ophthalmic artery was found.

In July 1982, the patient was still in the same condition, but his right visual acuity had reduced to 20/400. The cause of the attack, however, remained unknown.

Comment

Where is the site of occlusion? On the fluorescein angiogram both the central artery and the medial posterior ciliary arteries were occluded. According to Hayreh the most common origin of these arteries is initially a single branch from the ophthalmic artery. The site of occlusion in this case is postulated to be at this point.

What is the mechanism of the arterial occlusion? It is still undetermined. Attacks were never precipitated by pressure on the globe or carotid artery. The attack often was accompanied by headache, but the use of antimigraine medications including Migristene (dimebutazone mesilate), Kallikren (kallidinogenase), and Nitrol (isosorbide dinitrate), did not abort the attacks. Three stellate ganglion blocks were also not effective in preventing the attacks. However, the inhalation of carbon dioxide did improve the vision slightly during an attack. Unfortunately, the vision decreased immediately when the patient was allowed to breathe room air.

Is there any relationship between the attack and the congenital vascular anomaly? This type of anomaly of extrudal origin of the ophthalmic artery is an extremely rare congenital anomaly and occurs at the 20-mm embryo stage. The anomaly was recently reported by Vignaud et al. and Dilegge and Ascherl, but has not been noted to have ophthalmologic significance. In any case, the site of the postulated blockage of flow is distal to the site of origin, i.e., the origin of the central retinal artery and medial long posterior ciliary artery.
Amaurosis Fugax

Figure 4. At 86 seconds, all the retinal arteries are filled and some laminar flow is beginning in the vein.

Therefore, the relationship to the congenital vascular anomaly, if any, remains unknown.

How does the retina survive ischemic attacks of such long duration (up to 7½ hours)? Longfellow et al. report a similar case in which an attack lasted 10 hours. It was believed that the tolerance of retinal nerve cells to ischemia was measured in terms of minutes, but it has been shown recently that retinal cells seem to be more tolerant than previously believed. Experimental evidence in primates supports this contention. There is also some supporting evidence in humans. In the present case, it is most likely that the blood flow, although severely decreased, was sufficient for survival of the retina but not sufficient to maintain physiologic function.

References


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An Unusual Presentation of Isolated Optic Nerve Sarcoidosis

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Abstract

A 32-year old diabetic female had unilateral reduced vision (20/30) in an eye having the ophthalmoscopic appearance of papillophlebitis of the optic nerve head. After an apparently benign course, with recovery, the optic disc became edematous again and a central retinal vein occlusion blinded the eye. Thereafter, an avascular bilobed mass progressively protruded from the optic nerve head. Computerized tomography demonstrated uniform thickening of the optic nerve from the globe through the optic canal. The radiographic appearance was consistent with an optic nerve glioma or meningioma. Spinal fluid, chest x-ray, and serum angiotensin converting enzyme activity were normal. Tuberculin skin test was positive. Optic nerve biopsy demonstrated a noncaseating granuloma and a subsequent Kveim test was positive. Isolated optic nerve sarcoidosis may exist without other systemic manifestations. The appearance on computerized tomography may be similar to that of an optic nerve glioma or meningioma.

Introduction

Unilateral optic nerve involvement by a noncaseating granulomatous process has rarely been reported as being the only manifestation of presumed sarcoidosis. In these cases an optic nerve glioma or meningioma was suspected prior to the biopsy. Computerized tomography of the orbits and head was not performed in these patients but it would be hoped that this modality would be of value in differentiating tumor from granuloma. To the contrary, the present report describes a patient in whom computerized tomography appeared to support a preoperative diagnosis of optic nerve glioma.

Case History

A 32-year-old, black woman complained of recent onset of blurred vision in her left eye. Her acuity was 20/20, right eye and 20/30, left eye. The left eye had a moderate afferent pupillary defect to light and ophthalmoscopic examination revealed an edematous optic disc with dilated veins and peripapillary hemorrhages (Fig. 1). Visual fields demonstrated a relative central defect to a 2-mm red test object at 1 meter distance and enlargement of the physiologic blind spot. The right eye was normal in all parameters.

Ten years earlier, she had converted her tuberculin skin test and received a 1-year course of isoniazid. The patient's father was diabetic and the patient was known to have an abnormal glucose tolerance test. Ten months earlier, she had developed a transient left abducens nerve paresis that resolved within 4 weeks and was attributed to diabetes mellitus. Computerized tomography of the head and orbits using intravenous contrast dye was unremarkable at that time and her serum fasting glucose level was mildly elevated, 127 mg %.

Because of the recent loss of left eye vision, the patient was admitted to the hospital. Her fasting serum glucose level was 144 mg %. The following tests were within normal limits: x-rays of the chest (Fig. 2), skull, and optic foramina; repeat computerized tomography of the head and orbits with intravenous contrast dye enhancement; spinal fluid analysis, hemoglobin, hematocrit, white cell count and blood sedimentation rate; latex fixation, antinuclear antibody, LE, and VDRL tests; serum electrolytes, including calcium; and serum protein and globulin electrophoresis. A tentative diagnosis of diabetic papillophlebitis was made and she was placed on oral prednisone, 60 mg/day for 2 weeks. There appeared to be little therapeutic effect so the drug was tapered and discontinued. During the next 2 weeks, the disc edema and hemorrhages resolved, leaving a somewhat pale nerve head. Acuity returned to 20/30. Three weeks later, the left eye vision again decreased to 20/40 and disc...
Optic Nerve Sarcoidosis

Figure 1. Marked unilateral edema and hyperemia of the left optic disc with dilated veins and multiple peripapillary hemorrhages, yet good visual acuity (20/30) was consistent with the diagnosis of diabetic papillophlebitis.

Figure 2. Roentgenogram of the chest without evidence of sarcoidosis.

edema was present especially inferiorly, but without the previous florid venous vasculopathy (Fig. 3). The left eye vision continued to deteriorate over the next week to 20/200 and the patient complained of mild periorcular discomfort on adduction. The eye was neither proptotic nor inflamed. Prednisone 60 mg orally every other day was begun; after 2 weeks without visual improvement, however, the medication was rapidly tapered and discontinued. Two weeks later all vision was suddenly lost in the left eye. Fundus examination revealed an appearance compatible with a central vein occlusion (Fig. 4). Over the next 4 weeks, the venous vasculopathy resolved but a raised (7 diopter) bilobed mass progressively protruded from the inferior disc (Fig. 5) and several small white masses appeared in the adjacent vitreous. Computerized

Figure 3. Recurrent edema of the left optic disc especially inferiorly, but without the previous florid vasculopathy. The acuity was 20/40.

Figure 4. Central retinal vein occlusion producing a blind left eye.
Figure 5. Bilobed mass projecting 7 diopters from the inferior medial aspect of the left optic disc. Multiple small white opacities can be seen in the vitreous.

Figure 6. Computerized tomography demonstrating uniform thickening of the left optic nerve from the globe through the optic canal. (Reprinted by permission from the Journal of Computer Assisted Tomography 6: 614-616, 1982. Raven Press Publishers, New York.)

Figure 7. External appearance of the patient at the time of optic nerve biopsy. The blind left eye is slightly exotropic. The eye movements are full and there is no proptosis. The lids are retracted in the central and inferior photographs to demonstrate that the conjunctiva is normal and without inflammation.

tomography was performed again. A uniform enlargement of the optic nerve was found (Fig. 6). It extended from the sclera through the optic canal. Carotid arteriography indicated a relatively avascular optic nerve lesion. The radiologic diagnosis was optic nerve glioma. The external appearance of the eye and the ocular movements appeared normal (Fig. 7). Mammography and repeat chest x-rays were normal. An optic nerve biopsy was performed using a lateral orbital approach. The tissue diagnosis was noncaseating granuloma consistent with sarcoidosis (Fig. 8). A subsequent Kveim test was positive. Serum angiotensin converting enzyme activity, 14 nmole/minute/ml, was within normal limits. Tuberculin skin test (intermediate strength) was positive. Pulmonary function tests were normal.

Discussion

Optic nerve involvement in sarcoidosis can take many forms. Intraocular granulomas of the nerve head may produce neovascularization or be seen as nodular masses with the ophthalmoscope. Unilateral or bilateral granulomas of the retrobulbar nerve may produce edema of one or both
Increased intracranial pressure may cause papilledema. Compression by suprasellar, hypothalamic, or chiasmal granulomas may produce optic atrophy. However, isolated optic nerve involvement is rare in sarcoidosis. Only 5% of patients have central nervous system involvement, and in these the most commonly impaired cranial nerve is the facial nerve. In addition, only 5% of patients have a normal chest roentgenogram at the time the diagnosis of sarcoidosis is made. The present report may be the first in which a Kveim test confirmed the diagnosis in a patient having no other manifestations of sarcoidosis except optic nerve involvement. There are a few scattered reports of isolated noncaseating granulomas of the optic nerve but Kveim tests were not performed. In this situation some authors have hesitated to make the diagnosis of sarcoidosis while others have not. False-positive Kveim tests occur with a 2-3% incidence. Perhaps the present authors should be more circumspect in accepting this diagnosis in their patient.

It is interesting to speculate on the cause of the transient abducens nerve paresis 10 months before the visual loss. In retrospect, this may have been a manifestation of sarcoidosis and not diabetes mellitus, as was assumed at that time. If so, this too would have been a rare presentation. Obenauf et al. reported only one instance of an isolated abducens muscle paresis in 532 patients with sarcoidosis. Kendall and Tatler reported abducens muscle pareses in five patients who presented with neurosarcoidosis but in all of them other cranial nerve abnormalities were simultaneously present. Direct infiltration of the extraocular muscles has never been documented in sarcoidosis and it is assumed the abducens nerves were infiltrated or compressed. The present patient had an abnormally high fasting blood glucose level, a prior elevated glucose tolerance test, and a family history of diabetes mellitus. Palsies of the oculomotor and abducens nerves have occurred in young diabetics without retinopathy or other neuropathy. Perhaps the initial episode of left optic disc swelling was also a manifestation of diabetes. Her presentation with minimal visual involvement despite a florid vasculopathy of the nerve head (Fig. 1) was compatible with the diagnosis of papillophlebitis, which has been reported in young diabetics. If so, this would explain why the computerized tomography at that time was unremarkable, why the response to corticosteroids was unimpressive, and
why the condition initially resolved in an apparently benign manner. According to this scenario, it was only the second episode of disc edema (Fig. 3), which was not accompanied by vasculopathy (until the central retinal vein occlusion intervened) that was due to sarcoidosis.

The absence of systemic manifestations of sarcoidosis and the presence of normal chest roentgenograms have already been mentioned as contributing to the delay in diagnosing this disease in this patient. In addition, her serum angiotensin converting enzyme activity was within normal limits, her tuberculin skin test remained positive, and the abnormal appearance of the optic nerve on computerized tomography was compatible with an optic nerve glioma or meningioma. Approximately 80% of patients with active sarcoidosis have elevated serum angiotensin converting enzyme activities if they are not receiving corticosteroid therapy.\(^5\) At the time this test was performed, more than 8 weeks had elapsed since discontinuing prednisone. With regard to the tuberculin skin test, Siltzbach and co-workers\(^9\) and James et al.\(^10\) found approximately one-third of sarcoidosis patients demonstrated positive responses. They pointed out that the insensitivity to tuberculin was relative, not absolute. Positive responses tended to be weakly so. Most misleading of all was the apparent optic nerve tumor seen on computerized tomography (Fig. 6). The uniform thickening of the nerve, its poor enhancement after contrast dye injection, and the avascularity demonstrated on arteriography were compatible with this impression. It has been said that computerized tomography is of value in diagnosing neurosarcoidosis both because the granulomas can be seen and because they tend to enhance after contrast dye injection.\(^14\) The relatively rapid rate with which the nerve thickening developed and the intravitreal small white masses (Fig. 5) did suggest that other diagnoses such as lymphoma, metastatic malignancy, tuberculosis, and sarcoidosis be considered. The histologic diagnosis of noncaseating granuloma was, therefore, not entirely unexpected.

The postoperative management of this patient has presented another problem. The process may extend back along the intracranial portion of the left optic nerve toward the chiasm and involve the optic nerve of the remaining useful eye. Monitoring such a progression prior to visual loss in the right eye has proven difficult. The authors have been reluctant to perform a craniotomy and remove the remainder of the left optic nerve in order to prevent contiguous spread. Computerized tomography performed 2 months after the optic nerve biopsy did not show the chiasmal area in sufficient detail to rule in or out a progressive granulomatous infiltration. Perhaps intrathecal injection of contrast material in conjunction with computerized tomography would better delineate the chiasmal area but how often could this be repeated in a patient requiring long-term follow-up? Prophylactic systemic corticosteroids could be administered. It has been stated that this treatment is more effective earlier in the disease.\(^22\) However, there are no parameters by which to judge the appropriate dosage and duration of treatment in this diabetic female. No therapy has been administered in the 13 months following the optic nerve biopsy. The visual acuity and visual field of the right eye continue to be frequently monitored and as yet no deficit has occurred.

References

Optic Nerve Sarcoidosis


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Clinical and Subclinical Oculomotor Findings in the Eaton-Lambert Syndrome

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Abstract
Five patients with the Eaton-Lambert syndrome were examined neuro-ophthalmologically. Three were studied using electro-oculographic saccadic velocity recordings. Four complained of blurred vision and all complained of ptosis during the course of their disease. Clinical examination revealed mild ptosis in three of the five patients. Saccadic velocities before exercise were normal. After saccadic exercise, an increase in velocity of up to 40% was noted in two of the three patients studied.

Introduction
The Eaton-Lambert syndrome, or myasthenic syndrome, is a rare disorder of neuromuscular transmission which can be differentiated from myasthenia gravis on the basis of clinical and electromyographic findings. One of the clinical distinctions between the Eaton-Lambert syndrome and myasthenia gravis is that ocular signs and symptoms are reportedly rare in the Eaton-Lambert syndrome in contrast to their frequent occurrence in myasthenia gravis. This report describes the clinical and the sub-clinical oculomotor findings in patients with the Eaton-Lambert syndrome and reports analysis of the saccadic eye movement velocities before and after exercise in three patients.

Patients and Methods

Patients
Four males and one female patient with the myasthenic syndrome were evaluated (Table 1). The diagnosis of myasthenic syndrome was well established by the clinical neurologic symptoms and characteristic electromyographic abnormalities. The initial evoked muscle response in the hypothenar muscle following stimulation of the ulnar nerve was very small (0.2 mV; 1.6 mV; and 0.9 mV in RK, MR, and WC). In the other two patients (MK and TF), the initial evoked muscle response in the thenar muscle following stimulation of the ulnar nerve was 2.1 and 0.4 mV respectively. With repetitive stimulation at slow frequencies (2/second), a further decline of the first few responses were seen in all patients. At high frequencies (50/second), or following a brief period of exercise (15 seconds), marked facilitation was observed (from 4 to 20 times the initial response). All patients had been fully investigated for the presence of an underlying malignancy but none was found. All patients were participating in an experimental protocol of plasmapheresis and immunosuppressive treatment at the time of their neuro-ophthalmological evaluation. Medications included prednisone and azathioprine. The patients were not on guanidine. Anticholinesterase medication was withheld for at least 10 hours prior to testing.

Methods
The clinical neuro-ophthalmological examination assessed visual acuity, external appearance, especially the presence of ptosis, pupil function, confrontation visual fields, the ocular motor system including ocular rotations, pursuit and saccades, and the ocular fundi.

Saccadic velocity measurements were obtained in three of the five patients (WC, MK, and TF).
with the Eaton-Lambert syndrome. Each patient was placed supine on a table to minimize head movement. Directly overhead at a distance of 5 ft. was an arc with independently controlled light-emitting diodes as fixation markers at 10° intervals. The skin was prepared with alcohol swabs to insure good contact and Beckman electrodes with Beckman electrode paste were placed at the medial and lateral canthi and above the eye brow of the right eye. Subjects looked at the zero position on the display while any DC offset in the recording system was nulled, and then at a target 20° eccentric while the gain of the system was adjusted to produce a standard output. The DC-EOG signal was electronically differentiated to produce a velocity signal which was passed to a peak detector and "sample and hold" circuitry. The resulting scaled voltage value was displayed as peak saccadic velocity. Simultaneous position and velocity traces were recorded on a Gould Model 220 chart recorder.

The patient was asked to make horizontal saccades of 20° amplitude for baseline saccadic velocity measurement. The patient was then asked to make horizontal saccades of 40° as rapidly as possible (about 2/second) for 20 seconds. Within 5 to 10 seconds after this "exercise," successive 20° saccades were then remeasured. The trial of preexercise velocity, exercise, and postexercise saccadic velocity was repeated twice. Five normal subjects were tested in a similar manner.

### Results

Ptosis was the primary clinical neuro-ophtalmological finding in the group of five patients with the Eaton-Lambert syndrome studied. Three of the five patients demonstrated a unilateral ptosis of no more than 2 to 3 mm. Clinical fluctuation of ptosis was unremarkable during the examination; however, each patient reported an increase in ptosis, usually in the evening. However, at no time was the ptosis marked.

Although four of the five patients complained of intermittent diplopia and blurred vision, on clinical examination no patient demonstrated either diplopia or reduced visual acuity by conventional testing. The blurred vision complained of was primarily at near. One patient, (WC), had reduced near acuity which was optically correctable.

Qualitative examination of the patient's amplitude of horizontal eye movements, saccades, and pursuit appeared normal except in one patient (TF). In this patient the abduction saccades appeared slowed, and the amplitude of abduction was slightly diminished. Table 1 summarizes the clinical symptoms and signs of these five patients.

Quantitative analysis of the horizontal saccades of three of the patients with the Eaton-Lambert syndrome did, however, demonstrate interesting abnormalities. In all the patients the saccadic velocity in the preexercise state was within our normal range for 20° saccades. Following the exercise period with 40° saccades, an increase in saccadic velocity was noted in two of the patients. The increase was about 40% in the first patient and 20% in the second. Figure 1 shows the pre- and postexercise values for the first patient. The increase in saccadic velocity slowly decayed to initial levels after 6 to 8 saccades. For this patient, student's t-test demonstrated significant differences between pre- and postexercise saccadic velocities (p < 0.01). The increase in saccadic velocity was shown for both abduction and adduction saccades. The second patient showed a 20% increase in saccadic velocity after saccadic exercise; this difference was also significant by t-test (p < 0.01). For all of these sets of saccades, obviously hypometric saccades were eliminated from the analysis, so that these changes in saccadic velocity cannot be attributed to increased saccade size after exercise. There were no obvious changes in the size of saccades after exercise. One patient (TF) did not demonstrate an increase in postexercise saccadic velocity. There was no significant change in saccadic velocities in five normal patients tested after similar saccadic exercise.

### Discussion

Although saccadic eye movement studies have been performed on patients with myasthenia
gravis, we know of no oculographic studies conducted on patients with Eaton-Lambert syndrome. Our findings suggest that in patients with the Eaton-Lambert syndrome, there is a subclinical increase in saccadic velocity following "saccadic exercise." In two of the three patients studied (WC and MK), the preexercise saccadic velocities were within normal ranges for 20° horizontal saccades. The third patient (TF) was the only patient with clinically abnormal saccades; he had subnormal horizontal saccadic velocities and did not exhibit a postexercise velocity increase. In the two patients demonstrating an increased velocity with saccadic exercise, the first postexercise saccade velocity was increased. This increase in saccadic velocity was not maintained, but it slowly diminished after 6 to 8 saccades.

The increase in velocity following exercise distinguishes the eye movements of the Eaton-Lambert syndrome from those of myasthenia gravis. In a recent report, Abel et al. emphasized that in myasthenia gravis, saccadic velocity is normal for small amplitude eye movements and that the amplitude limitation is the primary defect.

Clinically, eye movements in our patients appeared neither slowed nor limited in amplitude. This was also the experience at the Mayo Clinic where in 40 patients with Eaton-Lambert syndrome, the only eye findings were ptosis and intermittent blurred vision. Elmqvist concluded that ocular symptoms "...either do not occur or are mild and transient and seldom present at the time of the examination of patients with the Eaton-Lambert syndrome."

Blurred vision, commonly reported in the Mayo Clinic series, was noted intermittently by all but one of our patients. Only one of them specifically stated that the blurred vision was worse at near, and with the appropriate optical correction, his symptom was relieved. Since the Eaton-Lambert syndrome is most common in patients 40 years of age or older, the blurred vision in some cases may be physiologic for age, and not a manifestation of the disease itself.

Our data support the view that there is a paucity of obvious clinical ocular findings in the Eaton-Lambert syndrome. However, in two of the three patients studied the saccadic velocities after exercise increased. This parallels the improvement in strength seen in peripheral muscles which is demonstrated electromyographically as facilitation. This suggests the electrooculographic recording of saccadic velocities is of potential value in diagnosis of complex cases of the Eaton-Lambert syndrome.

References

Eaton–Lambert Syndrome


Acknowledgment

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Occipital Lobe Infarction after Open Heart Surgery

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Abstract
The most common permanent neuro-ophthalmologic complication of cardiopulmonary bypass surgery is visual loss. Bilateral lower altitudinal visual field defects were documented in a patient who noted blurred vision following open heart surgery. A difference of opinion existed as to whether the field defects were due to retina-optic nerve or occipital lobe lesions. Two points are emphasized in this report: 1) the field defects were much easier to define on the tangent screen than on Goldman perimetry, and 2) occipital coronal high resolution CT scan confirmed bilateral upper bank calcarine cortex infarctions in this patient. Occipital coronal, thin-section, high-resolution computed tomographic scans are helpful in studying patients with occipital lobe visual field defects.

Visual loss following coronary artery bypass surgery has received much attention in the literature as the number of such cases has increased. Various mechanisms have been proposed to explain this complication. These have included embolism, cerebral hypoperfusion, ischemic optic neuropathy, and combinations of such lesions. The patient described below noted blurred vision immediately after open heart surgery and presented with unusual visual field defects. Opinions varied as to whether the field defect was due to retina-optic nerve ischemia or was secondary to occipital lobe involvement. Coronal computed tomography of the occipital lobes was helpful in confirming bilateral upper bank calcarine infarctions in this patient. The use of vertical prism glasses in helping patients with this type of altitudinal field defect is also documented.

Case Report
A 56-year-old, right-handed white male was first seen on June 3, 1982, through the courtesy of Dr. T. C. Spoor. The chief complaint was blurred vision since October 1981. He enjoyed good health until March 1981, when he suffered a myocardial infarction 1 day following surgical repair of an anal fistula. Seven months later, he was hospitalized to reevaluate his cardiac status. Cardiac catheterization revealed significant coronary artery disease, and surgical bypass was advised. The patient was operated on October 26, 1981, and had a four-vessel coronary artery bypass and excision of a myocardial aneurysm. He tolerated the surgery uneventfully to his knowledge; postoperatively, however, he noted immediately that something was wrong with his vision.

The patient was not particularly alert after surgery and was sedated for a time; however, while in the intensive care unit he recalled that when he would try to pick up a newspaper and look at it, the entire print would suddenly disappear. He called this a "whiteout." Within a few seconds vision would return, and he described this vividly as "like the tide rolling back in." This pattern recurred for a day or two, and when he mentioned it to his physicians, they became concerned. He had several tests, including an electroencephalogram and a CT scan. He was seen by a neurologist and an ophthalmologist, and apparently there was some difference of opinion as to whether the problem was in his eyes or in his brain.

Within a short time, the blurred vision began to slowly improve. When he alternately covered his eyes with his hands (which he did frequently), the problem appeared to him to be exactly the same in each eye. He noted that as vision returned, he could recognize the first two letters of a six-letter word, and by knowing these letters could then deduce what the word was. The patient was discharged from the hospital in 3 weeks. In January 1982, he was rehospitalized after some palpitations. Thereafter, he considered his visual problem stable. Because of a visually demanding occupation, his difficulty in reading prompted him to seek another opinion.

He also described a slight problem with gait. When walking, at times his right leg might "tingle" and tend to deviate out. The veins grafts had been removed from his left leg, not the right. Also, at times the right arm felt like it was going to sleep, but this too was improving. He had absolutely no other complaints. He was taking Isordil, Inderal, Lanoxin, Valium, and Ascriptine.

From the Bascom Palmer Eye Institute, Department of Ophthalmology, University of Miami School of Medicine, Miami, Florida (J.L.S.) and the Department of Neurology, The Mayo Clinic, Rochester, Minnesota (S.A.C.).
The patient had been examined elsewhere, and Goldmann visual fields were performed (Figs. 1 and 2). These were interpreted as due to a retinal-optic nerve lesion because no categoric vertical hemianopic shift was detected. He was then referred to the Bascom Palmer Eye Institute for another neuro-ophthalmologic opinion.

Examination on June 3, 1982, revealed a corrected vision of 20/25+3 in the right eye, reading slowly letter by letter. The left eye was 20/30-1, again slowly letter by letter or number by number. With +5.50 spheres he could read J-1 print with each eye on a Titmus card at 8½ inches. Although he read slowly, and generally only word by word, with sufficient plus correction he read fairly well. Therefore, there was no apparent organic dyslexia, and his reading difficulty was attributed to the visual field defects. External eye examination was not remarkable. The pupils and ocular motility were intact. Slit lamp and dilated indirect ophthalmoscopic examinations were normal. The applanation tensions were 14 in both eyes, and his blood pressure was 130/90.

The primary point of interest was related to the visual fields. On Amsler grid testing, the patient drew a line across the lower portion of the graph paper and stated he simply did not see below this line with either eye. This was symmetrical in the two eyes. Peripheral fields were normal to 3/330 white and 1/330 white on the Aimark perimeter in both eyes (Fig. 3). The peripheral fields, therefore, to larger isopters were exactly the same as had been plotted on the Goldmann perimeter. On the tangent screen, however, interesting central field defects were seen. There was an absolute scotoma in both eyes—the same to a 5/1000 white projection light pointer as to an 18/1000 white formed Bausch and Lomb target (Fig. 4). The patient fixed very well. In the right eye, the field defect was seen to come out of the blind spot, definitely crossing the midline. At first blush, it looked like an optic nerve type arcuate scotoma. On the tangent screen, however, there was a nasal-temporal step comparable to the midline step previously emphasized by Dr. Max Chamlin. Therefore, the lower altitudinal arcuate scotoma came down about 12° lower on the right half of the field than on the left half, and was congruous. This was not only an absolute scotoma (the same in size to large and small targets), but it was completely congruous. It could be studied more easily by testing him not only at 2 meters, but also at the end of the examination.
Figure 2. Goldmann visual field of left eye, February 23, 1982.

**Visual Fields**

*Peripheral*

Figure 3. Aimark peripheral visual fields performed June 3, 1982.
Figure 4. Tangent screen visual fields performed June 3, 1982. Note the nasal-temporal vertical step at the lower margin of the field defects.

Figure 5. Axial computed tomographic scan, June 4, 1982, showing bilateral hypodensities at occipital poles.

room (about 10 ft. away) with both formed targets, colored Mydriacyl bottle tops, and a projector light. It was then evident that this was a bilateral occipital lobe visual field defect.

The neuroradiologist was called and a specific request made for occipital coronal pictures, as clinically the field defect indicted the upper bank of the calcarine cortex on both sides. A previous CT scan had been reported as negative. Therefore, a careful occipital coronal study was performed, and the clinical impression was confirmed (Figs. 5-9). Bilateral upper bank calcarine hypodensities were seen which spared the lower bank of the calcarine cortex. These softenings were near the tip of the occipital lobes, and, therefore, were consistent with the field defects being near fixation (i.e., towards the macula) and sparing the more anterior occipital cortex (i.e., the peripheral fields). The CT scan also showed that there was some slight tongue of extension into the lower bank, particularly on the left side, which might have indicated an extension of the field larger than the defect plotted. However, the occipital coronal studies established the clinical diagnosis in this patient.

A final interesting point is that the patient had difficulty ambulating because of the bilateral lower altitudinal occipital lobe field defects. He was given a pair of -0.75 spheres (his distance manifest refraction) with 7-diopters base down ground in before both eyes. In addition, he was given a prescription for +5.50 spheres for reading only. The patient later reported that his reading was better with the +5.50 spheres, but he had difficulty
Smith, Cross

Figure 6. Higher cut of axial computed tomographic scan showing occipital lobe involvement.

Figure 7. Coronal occipital computed tomography, June 4, 1982. The large white arrows show bilateral hypodense areas in upper banks of the calcarine cortex. The black arrow shows that the lower bank is intact. "t" = the torcula.

Discussion

As open heart surgery or cardiopulmonary bypass surgery has become more widely practiced in the past decade, the interest in neuro-ophthalmologic complications following such procedures has steadily increased. The pathogenesis of ocular and cerebral complications of open heart surgery has come under increasing scrutiny because of the desire to try to prevent such formidable postoperative sequelae. It appears the visual loss is the most common permanent extracardiac complication following cardiopulmonary bypass surgery.

Gilman\(^1\) examined 35 patients before and after open heart surgery. Twelve patients (34%) had neurological deficits, and five of these died. Another six died either without deficits or before they could be examined. Deficits included gnostic disorders, hemiplegia, visual field defects (right inferior hemianopia with Riddoch phenomenon in the blind quadrants, small bilateral central scotomas resolving into homonymous paracentral defects), and seizures. Etiologies were insufficient cerebral blood flow and embolism.

Tufo et al.\(^2\) did a prospective study of 100 patients. Neurological, psychometric, and behavioral testing was performed before and after open heart surgery. Fifty percent of the patients had neurological damage following recovery from anesthesia, 43% had behavioral abnormalities preceded by focal neurologic damage and depressed intellectual function, and 15% of the survivors had neurological damage at the time of discharge from the hospital.

with the distance pair. The use of vertical prism glasses for bilateral altitudinal field defects is of interest and must be carefully adjusted and ground for each patient. This point is mentioned to help the clinician keep vertical prism glasses in mind for a therapeutic trial in individuals with bilateral altitudinal field defects.
Figure 8. Occipital coronal CT scan only slightly more rostral to Fig. 7, showing that the occipital infarctions were larger near the pole and decreased in size as one went more rostral in occipital lobe. The black arrow shows the upper bank infarction; the small black arrowhead shows intact lower calcarine bank; and the small black arrowheads show the cerebellum.

Figure 9. Enlarged occipital coronal CT scan made June 4, 1982. Small white arrowheads are in the upper bank softenings; T = torcula Hereophili; t = tentorium cerebelli; c = cerebellum.
Cerebral damage was related to increasing age and depression of arterial pressure. Neuropathological changes suggested ischemia.

Williams" examined seven patients before and after surgery. The intriguing part of her study was that she noted intravascular retinal changes occurring during open heart surgery by performing ophthalmoscopy during the procedure. White plugs, some forming in situ, were seen to traverse the retinal circulation. Microinfarcts were documented by fluorescein angiography. Refractile specks were also seen. Necropsy material supported the interpretation that these plugs were formed from blood constituents.

Brennan et al." evaluated five dogs undergoing partial bypass. They observed that bubble oxygenators generate microemboli which injure the brain during bypass. Significant depression of cerebral blood flow and metabolism accompanied even relatively short pump time and was not immediately reversible. The depression was microembolic in origin and could be largely avoided by effective filtration.

Aguilar et al." reported on neuropathologic complications of cardiac surgery, studying 206 patients who died after open heart surgery and eight who died after cardiac bypass procedures. The bypass procedures had twice the morbidity of other open heart procedures. Widely scattered fresh subarachnoid hemorrhage and intracerebral hemorrhages, acute ischemic neuronal necrosis and focal infarcts, and acute neuronal degeneration were noted. The cause was reduced cerebral blood flow and fibrin platelet emboli, fat emboli (reduced by better filters), and polarizable matter (not seen with good filters). The triad of emboli, acute hemorrhage, and focal or diffuse neuronal necrosis represents the postoperative syndrome of neuropathologic complications. The lesions were caused by anoxia, air embolism, hypotension, fibrin, and platelet or antifoam microemboli. Changes were seen in 85% of 206 patients undergoing open heart surgery and eight patients undergoing cardiac bypass.

Patterson et al." studied dogs submitted to cardiopulmonary bypass under conditions generating bubbles and microemboli. Postoperatively, impaired consciousness and ataxia were seen. These deficits cleared within 1 week. Multiple filling defects were seen in the microcirculation of the brain, demonstrated by ink injection before sacrifice. The vascular pattern was normal after 1 week. Neuropathologic findings were restricted to the cerebellum in those studied later.

Muraoka et al." studied 57 children operated on with mild hypothermia and high-flow bypass, and performed CT scanning before and after surgery. There were 27 bubble oxygenator and 18 membrane oxygenator cases. In the bubble group, all 14 with 20 micron filters had normal CT scans. Four of 13 with 40 micron filters or no filters had decreased brain mass on postoperative CT scans. In the membrane group, all postoperative CT scans were normal.

Sweeney et al." described ischemic optic neuropathy in seven of 7685 bypass patients. Blood loss with hypotension was the proximate cause in four of the seven cases. These authors stated that the optic nerve head behaves like an end organ. Hypothermia with decrease in oxygen consumption, also with a decrease of cerebral blood flow of 6 to 7% per degree and increased viscosity and sludging of blood were implicated as well. They proposed that nylon mesh bubble oxygenators might promote activation of complement.

It is evident from these reports that neuroophthalmologic complications 1) frequently follow cardiopulmonary bypass procedures, 2) involve the microcirculation, and 3) fortunately, the postoperative deficits, when documented, usually clear. However, loss of vision or visual field appears to be the most prevalent permanent neuro-ophthalmologic complication following such procedures. It is evident that there are many contributing factors to the development of retinal optic nerve and brain infarcts in such patients. The calcarine cortex is said to be the most sensitive area of the brain to hypoxia, and this may explain the frequency of permanent visual field defects in patients following open heart surgery. It is obvious that retinal and optic nerve involvement also are not infrequently seen, and in our experience a combination of anterior and posterior visual loss mechanisms have been encountered.

The most common cause of a monocular altitudinal visual field defect is optic nerve disease (e.g., ischemic optic neuropathy). The most common cause of binocular altitudinal visual field defects, when not occurring as two different bouts of optic nerve disease separated by a definite time, is either chiasmal or occipital lobe disease. These are usually easy to differentiate in that chiasmal lesions commonly produce asymmetric drops in visual acuity, will lead to optic atrophy in one or both eyes, and usually show diagnostic chiasmal visual field syndromes. Unilateral occipital lobe lesions do not produce a drop in visual acuity, whereas bilateral occipital lobe lesions produce strikingly symmetrical reductions in visual acuity, and the optic discs remain clinically uninvolved. We are excluding the late instances of transsynaptic degeneration.

In any patient complaining of blurred vision following open heart surgery, a careful neuroophthalmologic examination is indicated. The measurement of visual acuity in each eye, a careful examination of the pupils, a confrontation field, and a careful look at the optic disc and retina for cotton-wool spots, bright plaques, or other retinal
emboli is important. An Amsler grid test may be done at bedside. A "history" field is important to see if the defect is notably congruous (i.e., exactly the same in the two eyes). When feasible postoperatively, a high-resolution CT scan of the brain with emphasis on the occipital lobes and specifically occipital coronal views may be invaluable. The near acuity should be measured not only with isolated letters and numbers (as the Rosenbaum near card), but also with text (as the Titmus or Sloan cards) in order to help make a differentiation between acuity drop, visual field defects, and organic dyslexia syndromes. A neurological examination is also obviously indicated.

Finally, the use of hemianopic Fresnel prisms may be considered. A previous report documented the use of these prisms in hemianopias. It should be emphasized that not all patients with hemianopias like hemianopic Fresnel prisms. Those with notable neurologic deficits (e.g., left hemisphere infarcts) usually are not happy with them (as they do not restore their ability to read). Patients with isolated hemianopias (e.g., right occipital lesions) often definitely like them, and they are worth a trial in such cases.

The incidence of permanent loss of vision or visual field defects following open heart surgery must be more precisely defined. Such reports should include meticulous detail as to intraoperative phenomena (cerebral blood flow, hypotensive episodes, size and types of filters employed, serum complement studies, bubble oxygenators, etc.). Further intraoperative careful fundus examinations (as in the study by Dr. Isla Williams) also may be helpful in learning how to decrease the incidence of this problem.

References


Acknowledgments

The authors thank Dr. T. C. Spoor for referring this patient and for providing the Goldmann visual fields of February 23, 1982; and Dr. Robert Quencer and staff for the occipital coronal computed tomography studies.

* A recent communication from Optical Sciences Group. 1331 Commerce Street, Petaluma, California 94952; 1-800-227-2254 established that they will be making some 40-diopter Fresnel prisms. The largest ones now available are 30 diopters. The price quoted for these by Mrs. Astrid Harper was $5.75 each (in lots of 1-23) or $4.31 each (in lots of 24 or more). The price quote was effective August 27, 1982.
Acute Painful Cavernous Sinus Syndrome in Unruptured Intracavernous Aneurysms of the Internal Carotid Artery
Possible Pathogenetic Mechanisms

THOMAS-MARC MARKWALDER, M.D.
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Abstract
Three cases of painful ophthalmoplegia with acute onset due to an unruptured aneurysm of the intracavernous portion of the internal carotid artery are reported. The possible pathogenetic mechanisms responsible for this unusual mode of clinical manifestation are discussed and the neurovascular relationships of the cavernous sinus are analyzed in respect of ischemic versus compressive damage to the intracavernous neural structures.

Most intracranial saccular aneurysms manifest themselves clinically with an acute subarachnoid hemorrhage from rupture. In contrast, aneurysms of the intracavernous portion of the internal carotid artery, which constitute about 2-3% of all intracranial aneurysms,1,2 rupture only rarely. If they do so, they do not bleed into the subarachnoid space, but produce a carotid-cavernous fistula with exophthalmos, congestion of conjunctival and fundus veins, and often a bruit audible over the ipsilateral eye and the skull.3-7

In contrast to these findings, the vast majority of intracavernous saccular aneurysms of the internal carotid artery, however, becomes manifest in either of two characteristic ways:1,5-11 1) A slowly progressive and usually painless loss of function of the third, fourth, fifth, and sixth cranial nerves; or 2) An acute cavernous sinus syndrome with almost simultaneous paroxysmal functional loss of these nerves, accompanied by severe pain. While a progressive compression of the cranial nerves by the aneurysm appears to be an adequate pathogenetic explanation in the first type,11 the rapid and painful onset of symptoms is not completely understood in the second type. In addition, the clinical picture of the second type requires differential diagnostic delimitation against other painful ophthalmoplegias such as the Tolosa-Hunt syndrome, diabetic cranial neuropathy, tumor infiltration of the cavernous sinus, and the paratrigeminal syndrome of Raeder.

Three cases presenting with an acute painful cavernous sinus syndrome due to an intracavernous carotid aneurysm constitute the basis for discussion of possible pathogenetic mechanisms responsible for this unusual mode of manifestation of unruptured aneurysms which previously had remained asymptomatic.

Case Reports

Case 1
A 21-year-old male was admitted on February 6, 1978, because of a sudden onset of severe left frontal headache and double vision. Six weeks ago, he had already suffered from moderate similarly localized headache lasting several days. Neurological examination revealed a complete sixth nerve palsy on the left side, a slight palsy of the left internal rectus muscle, and a minimal ipsilateral mydriasis with preserved light and convergence reactions. In addition, there was a slight to moderate loss of tactile sensation in the left V1 and V2 dermatomes. No proptosis nor any conjunctival or fundal venous congestion were detected. Furthermore, a slight right motor hemiparesis which was only present for some hours was noted. Correspondingly, an intermittent left frontotemporal delta focus was found in the EEG. Plain skull x-rays and a CT scan were normal.

Within the following 6 days, a complete left external ophthalmoplegia developed. The left pupil, however, became only moderately dilated (5 mm in diameter as compared with 3 mm on the right eye) and still contracted promptly to light and
dilated normally in the dark. Carotid angiography demonstrated a saccular aneurysm of the intracavernous portion of the left internal carotid artery which measured about 1.5 cm in diameter and was partially thrombosed. The latter originated from the lateral wall of the juxtasellar segment of the carotid syphon and moderately compressed the presellar segment. Ligation of the internal carotid artery in the neck was performed on February 24, 1978.

Postoperatively, the patient was immediately relieved from his pain and the oculomotor deficits improved quickly. By the fifth postoperative day, the fifth and sixth cranial nerves had already fully recovered. Only a slight adduction and elevation deficit remained from the third nerve palsy while the pupils were equal and normally reacting. The fourth nerve had remained completely paralyzed at that date. The further postoperative course was uneventful except for the transient ischemic attacks which presented as motor hemisyndromes. Within the following weeks all neurological deficits returned to normal.

Case 2

A 70-year-old female was admitted on September 9, 1981, after onset of a paroxysmal right frontal headache and vomiting accompanied by double vision on right lateral gaze. The neurological examination revealed a sensory loss in the right V1 and V2 divisions with a reduced corneal reflex and a complete sixth and partial third nerve palsy with moderate ptosis and an elevation deficit. Additionally, there was a Horner’s syndrome on the right with the pupil only slightly dilating in dim light, while the light and convergence reactions were full and prompt. There was neither proptosis nor venous congestion of the conjunctiva or in the fundus. A CT scan revealed a large, slightly hyperdense space occupying lesion in the right parasellar region with marked enhancement after intravenous contrast infusion. Carotid angiography demonstrated a large, partially thrombosed intracavernous saccular aneurysm whose exact origin could not be determined. It was located laterally to the carotid syphon and was partially compressing the internal carotid artery in its intracavernous course leading to a distinct delay of circulation. Because of the age of the patient no operative intervention was undertaken. Under analgetic drugs the headache subsided during the following weeks, and the neurological deficits remained unchanged.

Case 3

A 58-year-old woman was admitted on July 22, 1981, 3 days after a sudden onset of left retrobulbar pain, double vision, and sensory loss on the left forehead and cheek accompanied by vomiting. Neurological examination disclosed a complete sixth nerve palsy on the left. Oculomotor and trochlear nerve functions were normal. The pupils were equal and reacted fully and promptly to light, convergence, and dark. A sensory loss was present in all divisions of the left trigeminal nerve with absence of the corneal reflex. In addition, the left masseter and pterygoid muscles were partially paralytic. There was no venous congestion. Plain x-rays of the skull were normal. A CT scan demonstrated a left parasellar mass with shell-like calcifications in its lateral aspect and marked enhancement on intravenous contrast infusion (Fig. 1, left). The left carotid angiogram showed a large, partially thrombosed intracavernous saccular aneurysm of the preophthalmic portion of the internal carotid artery. First an expectative attitude was taken but because of persisting heavy facial pain requiring high doses of analgesics, a four-vessel angiography was undertaken to investigate the flow efficiency in the circle of Willis. The latter again demonstrated the aneurysm (Fig. 1, right) and normal patency of the circle of Willis.

This angiographic study was complicated by a right-sided hemiparesis and aphasia which both resolved within 2 days. In view of the high risk of an operative intervention, the patient was thereafter dismissed and drug therapy of the heavy facial pain was continued. The patient was last seen in August 1982. There was still some neuralgic pain in the right V1 and V2 divisions which is now controlled by common analgesics. The right corneal reflex was absent, and third nerve function had returned to normal but there was still a complete sixth nerve palsy.

Discussion

The most conspicuous clinical features in our three patients with an intracavernous aneurysm of the internal carotid artery were the acute appearance of a cavernous sinus syndrome accompanied by severe pain. Although about half of the aneurysms of the intracavernous portion of the internal carotid manifest themselves in this way, the underlying pathogenic mechanisms have not been a point of discussion. Why do such aneurysms, which probably have existed for many years without making any symptom or sign, present themselves one day so dramatically without having ruptured? A sudden simultaneous compression of several cranial nerves from acute dilatation of the aneurysm is hard to imagine. In addition, at the moment such aneurysms become manifest, most of them are partially thrombosed or calcified. In addition, the lateral wall of the cavernous sinus prevents further aneurysmal expansion and acts as a rather firm encasing membrane.

In our opinion, it is conceivable that acute ischemia of the cranial nerves associated with the
The cavernous sinus could produce this symptomatology. This assumption is also consistent with the fact that almost all patients whose intracavernous aneurysm became manifest as described above were more than 50 years old. In our first case (a 21-year-old patient), a transient right-sided hemiparesis with a left frontotemporal EEG-focus, which appeared together with the acute painful cavernous sinus syndrome, indicated an acute thrombotic or embolic event. The carotid angiogram revealed a partially thrombosed aneurysm which moderately compressed the presellar segment of the carotid syphon. Even those patients in whom the intracavernous aneurysm directly compresses the nerves and leads to a slowly progressive and painless loss of function have occasional episodes (of some days or weeks) with severe pain and sometimes worsening of the clinical signs. These patients can possibly develop a sufficient collateral circulation to compensate for the additional ischemic nerve lesion.

An acute onset of eye muscle palsies accompanied by pain closely resembles the clinical picture of diabetic cranial neuropathy. Ischemic lesions of the oculomotor nerve have been demonstrated in the latter disease. An essential feature of ischemic third nerve palsies is sparing of the parasympathetic fibers. Therefore, the light reaction of the pupil remains intact or is only minimally affected in spite of a marked palsy of the external ocular muscles. Meadows, in his analysis of 15 cases of intracavernous carotid artery aneurysms, mentioned that the pupil was usually small. He did not attribute this observation to sparing of the parasympathetic fibers but to additional damage of the sympathetic innervation of the pupil. Unfortunately, he did not examine the reactions of the pupils to light and dark. While his interpretation may be correct in some cases, there are also cases of intracavernous aneurysms with sudden onset of symptoms and fully or relatively preserved pupillary light reactions with respect to the severity of the palsies of the extraocular muscles innervated by the third nerve. Cogan's experience that "The surprising lack of mydriasis despite total paralysis of other functions of the IIIrd nerve in lesions about the superior orbital fissure and cavernous sinus . . . is not always explicable on the basis of coexistent involvement of the sympathetic fibers since the pupillary size and reactions are
Acute Cavernous Sinus Syndrome

often normal," might be explained by the predominance of ischemic over mechanical nerve lesions in this area, in dependence of the particular anatomical conditions in the region of the cavernous sinus.

Most anatomy books omit the intracavernous branches of the carotid artery, although they have been described in large autopsy studies, and sometimes can even be seen on normal carotid angiograms. The meningohypophyseal artery and the inferior cavernous sinus artery supply the third, fourth, and sixth cranial nerves as those vessels pass within the wall of or through the cavernous sinus. In addition, the inferior cavernous sinus artery supplies the Gasserian ganglion (Fig. 2). Occlusion of these vessels, whose origin may be incorporated in the aneurysmal wall, may occur as a result of either compression by the aneurysm or thrombosis and may cause an acute simultaneous ischemic lesion of the cranial nerves. The probability that such a phenomenon occurs in the case of aneurysm of the intracavernous portion of the internal carotid artery is relatively high; these aneurysms typically originate from the "branching off" of the feeding vessels of the cavernous sinus structures. The usually sudden relief from pain and the improvement of the cavernous sinus symptoms after carotid ligation can be well-explained by decompression of the vasa nervorum as by a decompression of the nerves themselves.

Thomas and Yoss, in a study of 102 parasellar syndromes of different etiology, have emphasized that neither the mode of onset of symptoms nor the sequence of evolution or pattern of the neurological deficit is characteristic for the etiology of the underlying lesions. More than two-thirds of their patients had a tumor; 19 patients had an intracavernous aneurysm; and only three had a Tolosa-Hunt syndrome. Neuroradiological examination of patients with an acute painful cavernous sinus syndrome, therefore, is mandatory. One might also argue that in patients with other causes than aneurysms, some phenomena could be explained by ischemic lesions of the intracavernous structures. Recent observations in patients with Tolosa-Hunt syndrome seem to confirm this in part.

References

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Chromograms of Color Normals and Multiple Sclerosis Patients

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Abstract

The Gunkel chromograph was tested on 81 volunteers with normal color vision as indicated by screening with Ishihara plates and the panel D-15. Most of these color normals located their neutral area superior to the geometric center of the chromogram. A minority located their neutral area at the geometric center. Recognition of this variation may prevent misdiagnosis of chromographs of patients suspected of having color vision defects. All 29 eyes with history and VEP findings consistent with optic nerve demyelination had enlargement of the neutral area, even though visual acuity could be corrected to 20/20.

Introduction

The conventional clinical tests used for detecting color vision defects include pseudoisochromatic plates, the Nagel anomaloscope, the Farnsworth dichotomous color 100 hue test, and the less cumbersome panel D-15 test. Unfortunately, these tests fail to provide clinical validity for colors a subject can identify or those colors a subject cannot identify. A new color vision testing device, the Gunkel chromograph, has been used to determine the type and extent of congenital color defects and has been utilized in testing patients with known color vision defects.

The purpose of this study was to determine the location and extent of the neutral area perceived by individuals with normal color vision. In addition, we tested 18 patients with multiple sclerosis and a past history of optic neuritis in at least one eye (29 affected eyes).

Materials and Methods

One hundred volunteers were tested in the Eye Clinic at the Medical College of Ohio during July and August, 1981. Three color vision tests were performed while each volunteer was waiting to receive dilating drops for the ophthalmic examination. The volunteers ranged in ages from 5 to 85 years. Informed consent was obtained from each volunteer or from a parent if the volunteer was a minor. All tests were conducted binocularly.

Twenty-seven patients with definite multiple sclerosis were tested monocularly using the same three color vision tests that were used with the 100 normals. In addition, the visual-evoked potential (VEP) was performed on all 27 patients to determine if demyelination was present. The VEP stimulus was a pattern consisting of checks with a reversal time of 1.88 cycles/second and a check size setting of 16. Scalp electrode placement was CZ (vertex of scalp), OZ (1 cm above inion), and earlobe (ground). The distance between patients and stimulus was 1 meter. One hundred twenty-eight sweeps were computed for each waveform; monocular as well as binocular responses were also recorded.

Each patient or volunteer was asked to identify the numbers on Ishihara plates under a MacBeth Easel lamp and to arrange the caps in the panel D-15 tray in serial order according to hue. The Gunkel chromograph was used under subdued illumination. The volunteer was asked to identify the presence of color and to name the colors as they were produced on the viewing screen. A control handle operated by the examiner altered the colors. The examiner initiated the testing in the neutral area and then proceeded toward the colors in the following manner: neutral to green to neutral to magenta to neutral to turquoise to neutral to red to neutral to yellow to neutral to blue. Each time the subject identified a color (whether right or wrong), the examiner pressed a bulb and a point was indented into the chromogram. The points on
the chromogram were connected together. The area within the connected points was considered the neutral area for that volunteer or patient.

Results

Of 100 volunteers with visual acuity correctable to 20/20, only 81 had both normal fundus examinations and normal color vision screening with the Ishihara plates and the panel D-15. The chromo-

Figures 1A and 1B. Neutral area plotted with the Gunkel chromograph for 81 volunteers with 20/20 visual acuity, normal ophthalmoscopic examinations, and normal results on screening with Ishihara plates and the panel D-15 test. (A) Seventy-two (89%) color normals identified a neutral area superior to the geometric center of the chromogram. (B) Nine (11%) identified the geometric center of the chromogram as neutral.

Figure 2. Mean neutral area for 29 eyes with a past history of optic neuritis from 18 multiple sclerosis patients. All of these eyes had visual acuity correctable to 20/20; their VEPs, however, were consistent with previous demyelinating disease of the optic nerve.

Figure 3. Means and standard deviations of the neutral area plotted with the Gunkel chromograph for 29 eyes with optic nerve demyelination from 18 multiple sclerosis patients.

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grams of these color normals demonstrated two separate neutral areas. The geometric center of the chromograph was perceived as being neutral by only nine (11%) of the 81 color normals. The remaining 72 (89%) color normals said this area appeared distinctly blue, but could locate a neutral area above the geographic center of the chromograph (Fig. 1). Twenty-nine eyes from 18 patients were diagnosed as having optic neuritis by a past history of visual loss with spontaneous recovery to 20/20 over a period of weeks. In each of the 29 eyes, the VEP implicit times were greater than 120 m second, and the waveforms were distorted. Their chromogram neutral areas were enlarged (Fig. 2). Failure of color perception was greatest for yellow, green, and turquoise and less severe for red, magenta, and blue (Fig. 3).

Discussion

Our results indicate that volunteers who are color normals (according to screening with Ishihara plates and the panel D-15 test) may locate a neutral area in the center of the Gunkel chromogram or above the geometric center. Eyes with a previous history of optic neuritis secondary to multiple sclerosis were found to have enlargement of the neutral area of the chromogram, even in the presence of 20/20 vision. In these eyes with 20/20 vision, the diagnosis of a previous optic neuritis was confirmed by visual-evoked potential testing. These findings confirm the importance of color vision testing in patients with suspected optic nerve disease.

References


Acknowledgment

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Acute Recurrent Orbital Myositis

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Abstract

An acute relapsing orbital inflammatory disease predominantly affecting the extraocular muscles was seen in two patients and is reported here as acute recurrent orbital myositis. The association of this disorder with other systemic diseases such as asthma, sinusitis, upper respiratory infection, Crohn's disease, and serum sickness is discussed. The similarities to other forms of acute orbital inflammatory disease such as orbital pseudotumor are noted, and a possible underlying immunologic mechanism is suggested.

Introduction

Acute orbital inflammatory disease is often described under the heading of inflammatory orbital pseudotumor. Included in the numerous case reports are a lesser number of cases of acute recurrent orbital inflammatory disease with extraocular muscle involvement as a predominant feature. We describe two patients with a recurrent form of orbital myositis that was associated with exacerbations of presumed allergic respiratory disorders.

Case Reports

Case 1

A 17-year-old white woman presented in September 1980 with a 3-week history of right frontal headache, diplopia, nausea, and vomiting. Proptosis, hyperemia, and lid swelling of the right eye were reportedly present. She was treated by another physician with prednisone, 60 mg/day, which produced a rapid improvement of the headache. She was referred for further evaluation of persistent diplopia and inflammatory orbital signs. Her past history was significant for a similar episode involving the left orbit 3 years earlier, which resulted in persistent diplopia in downgaze.

On examination on September 3, 1980, visual acuities were 20/15 in both eyes. There was decreased adduction and elevation of the right eye and a 40-prism diopter exotropia. Hertel exophthalmometry measurements were 15 mm on the right and 14 mm on the left. A small, resolving subconjunctival hemorrhage was present in the right eye. The remainder of the neuro-ophthalmic exam including visual fields was completely normal.

Ophthalmic ultrasonography was performed and showed a markedly enlarged right medial rectus (Fig. 1). The prednisone therapy was tapered over 1 week.

Twenty-seven days later, she returned with moderate lid edema, ptosis, conjunctival chemosis, and hyperemia of the left eye. There was pain in upgaze, a 25% limitation of abduction, and a 75% limitation of adduction. There was now a borderline measurable left-sided proptosis of 18 mm versus 16 mm on the right, and ultrasound showed enlargement of the medial and lateral recti of the left eye (Fig. 2).

She was admitted for a diagnostic work-up, which included neurology, rheumatology, and en-
Orbital Myositis

Figure 2. Orbital sonogram showing enlargement of left medial and left lateral rectus muscles (case 1).

Figure 3. High-resolution orbital CT scan showing enlargement of both medial recti and enlargement of the left lateral rectus. (Note: The right orbit is shown on the left; case 1.)

docrinology consults, lumbar puncture, and thyroid function studies. In addition, an extensive battery of immunologic tests were performed, including C1q binding, complement levels, and antinuclear factor, all of which were normal.

Orbital sector CT scan showed thickening of both medial recti, the right inferior rectus, and the left lateral rectus (Fig. 3) along with mucosal thickening of the maxillary and right ethmoid sinuses (Fig. 4).

The patient was discharged and improved gradually. Two weeks after discharge, she had resolution of the swelling and redness, but now had a marked esotropia due to an inability to abduct the left eye. Exophthalmometry was now symmetrical, with readings of 17 mm. Three weeks later, the improvement was marked, and the rotations were full in all directions of gaze. In December 1980, the CT scan showed less thickening of the muscles (Fig. 5) and resolution of the sinus changes (Fig. 6). In September 1981, left-sided headache, redness, and swelling of the left eye suddenly resumed. Immediate treatment with 50 mg/day of prednisone orally resolved the symptoms within a day. The exam was normal 3 days later, except for mild residual pain in the left orbit on up- and right gaze.

Summary. A teenage woman had four separate attacks of orbital myositis over a period of 4 years. Although no immunologic abnormalities were detected, one attack was associated with transient thickening of maxillary and ethmoid sinus mucosae.

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Case 2

A 56-year-old white man developed diplopia and pain on abduction of the right eye on October 3, 1981, which progressed to constant right orbital pain and right frontal headache. He saw his ophthalmologist 5 days later, who found the vision and fields to be normal. Mild right ptosis and limitation of abduction and adduction of the right eye were noted. A CT scan confirmed swelling of the right lateral rectus muscle, polytomograms showed bilateral maxillary and ethmoidal haziness, and a cerebral arteriogram was normal.

The sedimentation rate was slightly elevated at 33 mm/hour. CBC, SMA-18, thyroid function studies, VDRL, and spinal tap were all normal. Immunologic testing was not done. He was placed on 60 mg prednisone daily and began to improve rapidly. When examined by us 3 weeks after the onset of symptoms, the external signs and diplopia were much reduced.

His past history was fascinating; he had six other episodes of orbital inflammation with diplopia since 1959 and had kept exact dates. All episodes occurred during the cooler seasons; none were later than April. Each episode had been preceded 5–7
Figure 6. CT scan showing resolution of sinus mucosal changes described in Fig. 4 (case 1).

Figure 7. Top: Decreased abduction of right eye (case 2). Bottom: Narrowing of interpalpebral fissure on adduction of right eye.

days by an upper respiratory infection. He also had severe allergic asthma, again since 1959, which often required steroids and always resolved in the summer months. The asthma attacks did not coincide directly with the attacks of orbital inflammation, however. The patient also had a history of chronic sinusitis for which he had undergone ethmoidectomy and nasal polypectomy in 1981.

On examination by us, the acuities and fields were normal. There was a 13-prism-diopter esotropia in primary position, which worsened in right gaze (Fig. 7). Upgaze was also mildly limited. On left gaze, the right interpalpebral fissure narrowed (Fig. 7), consistent with a diagnosis of acquired orbital retraction syndrome, which is not infrequently seen with orbital myositis. Other than conjunctival hyperemia over the right lateral rectus insertion, the neuro-ophthalmic exam was normal. All lab studies, including Westergren sedimentation rate, IgE, eosinophil count, and serum protein electrophoresis, were normal. However, the patient was still taking oral steroids when they were performed.

Summary. A man with a history of allergic asthma and chronic sinusitis suffered seven episodes of steroid-responsive orbital myositis over a 22-year period, which were all preceded by symptoms of an upper respiratory tract infection.

Discussion

Acute recurrent orbital myositis commonly presents with pain and headache on the involved side, diplopia, hyperemia of the conjunctiva and eyelids, and mild proptosis. Unilateral and bilateral cases have been described, but bilateral cases usually have several weeks separating the onset of symptoms in either eye.

The clinical course is markedly shortened by steroids, particularly when begun early, as in our first patient whose third attack was aborted. Most patients have no residual, and the ones who do
have permanent effects were usually biopsied or not treated.4, 7 Most patients have normal visual acuities, but visual loss as a result of associated optic neuritis, choroidal folds, or scleritis has been reported.5, 7, 10, 11, 16 Recurrences have been notably seasonal in our patients, and this association has not been previously emphasized.

In reported cases and in our patients, this disorder is often seen in association with other illnesses, which are seemingly unrelated, but can be linked by their immunologic etiologies.

1. One of our patients had allergic asthma (case 2).
2. Two cases of almost identical syndromes following upper respiratory infections were recently reported.16 One had a proven streptococcal pharyngitis, but neither had recurrent episodes. The authors postulated a poststreptococcal vasculitic process or a viral myositis. (Myositis of the lower extremities and back is known to occur in association with influenza and coxsackie viral infections.26-29) Mottow and Jakobiec also described an association with upper respiratory infection in six patients from their pediatric orbital pseudotumor series.5
3. A clinically identical form of acute orbital myositis has been seen in association with Crohn's disease, with the onset of myositis coinciding with increased activity of the disease.17, 23-25 Of even greater interest is the observation in two patients of resolution of orbital inflammation following bowel resection.17, 25
4. Allergic rhinitis and nonpurulent sinusitis have been described in association with pediatric orbital pseudotumor.2, 26 Although the etiology of this form of sinusitis is not fully understood, deposits of immune globulins and complement are found in the hypertrophied sinus mucosae in these patients.27
5. One case of dramatic and severe acute bilateral orbital inflammatory disease closely followed three injections of iron-dextran complex in a 25-year-old woman, accompanied by a clinical picture of serum sickness. After resolution of the inflammation, optic atrophy was present; bilateral orbital biopsies had been performed, however, and steroid therapy was delayed.4
6. A case of recurrent Tolosa–Hunt syndrome was reported in a 56-year-old woman, in whom flare-ups were accompanied by an elevation of the sedimentation and a positive antinuclear factor. She had no other evidence of collagen vascular disease.28 The disorder classified as pediatric orbital pseudotumor is frequently clinically indistinguishable from cases of acute orbital myositis in adults.24 More recently, ophthalmologists treating acute orbital myositis have stressed that the nature of the process is different from the typical adult pseudotumor, and some have suggested eliminating the term pseudotumor altogether.2, 12 The few cases of acute pediatric pseudotumor that have been biopsied have shown a diffuse vasculitis, with polymorphonuclear leukocytes and eosinophils surrounding small arteries and arterioles. Henderson initially termed this a type I pseudotumor,23 and noted its similarity to the Arthus reaction, which is type III, immune-complex-mediated hypersensitivity.23 He now diagnoses this as orbital vasculitis.25 His type II pseudotumor is the more commonly found chronic orbital inflammation seen in the older age groups and is clinically different from our two cases. Of particular interest is the patient with serum sickness,4 as biopsies showed an orbital vasculitis. Acute orbital inflammation with a histologic picture similar to orbital vasculitis has been experimentally produced by injecting bovine serum albumen into the retrobulbar space of previously sensitized rabbits.34 Histology showed more chronic inflammatory cells when biopsy was performed several weeks following the retrobulbar injections.35 Left untreated, the orbital inflammation resolved and did not develop into chronic orbital pseudotumor.

The need to redefine orbital pseudotumor into more precise divisions is apparent, and the use of orbital ultrasonography and CT scanning is allowing more specific diagnoses. This is also causing the literature on the subject to become even more confusing, because the same patient could easily fit into any of four or five different diagnoses, depending upon the doctor's preference and the particular diagnostic tests used.

The distinction between acute orbital myositis and other acute orbital pseudotumors may also prove to be unnecessary, as CT evidence of extraocular muscle involvement may vary at different times during the illness.

It may also be arbitrary to separate pediatric from adult pseudotumor, as this type of acute orbital inflammatory disease does occur in adults, some of whom have long histories of recurrent episodes since youth.

It would seem more appropriate to consider that the same immunologic processes may be occurring in all these patients and to direct attention to that rather than attempting classification by exact localization of the process within the orbit or by arbitrary age limits.

In our cases, blood tests were obtained only after steroids were begun, and this may account for the normal values. However, we recently examined another patient with probable orbital myositis who had a markedly elevated level of IgG and IgM immune complexes by cryoprecipitation. This patient also had a strong seasonal trend to her episodes and a long history of chronic sinusitis.

In summary, patients with acute orbital inflammatory disease, with or without myositis, deserve a thorough immunologic evaluation in addition to
computerized tomography and ultrasound examinations.

Circulating immune complexes can be detected by total hemolytic complement (CH50) and C1q binding determinations as well as by testing blood for the presence of cryoglobulins. If biopsy material is obtained, it also should be studied for immune complex deposition.

Inflammatory orbital pseudotumor has been a heterogeneous diagnosis for many years. Recent technological advances, which allow more careful dissection of underlying immunologic processes, promise to simplify the diagnosis and pathogenesis of related orbital inflammatory diseases.

References


Journal of Clinical Neuro-ophthalmology

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Monocular Rotary Nystagmus

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Abstract

A patient with transient monocular rotary-vertical nystagmus demonstrated decreased gain of vertical pursuit and normal vestibulo-ocular reflex on electro-oculogram. A supranuclear brain stem lesion, resulting in lack of monocular inhibition of oculomotor neurons, is postulated on the basis of these findings.

A 62-year-old man with a history of hypertension, myocardial infarction, and late latent lues noted sudden onset of blurring of vision and a pulling sensation behind the right eye. He also complained of light-headedness and gait instability and was admitted to the hospital for further evaluation.

While the general physical examination was unremarkable, the neurologic examination demonstrated a slightly wide-based gait and mild right-sided ataxia. Neuro-ophthalmologic examination showed intermittent monocular, intorting, rotatory-vertical movement of the right eye. It was present in all directions of gaze but most prominently looking down and to the left where it became primarily vertical. In addition, he had a decreased blinking rate. Vertical optokinetic nystagmus and vestibulo-ocular reflex were normal. The neuro-ophthalmologic examination was otherwise normal.

The ataxia and gait instability cleared within the first day of hospitalization, but the abnormal movements of the right eye persisted for 2 more days. Neurologic workup included a normal CT scan of the head. Cerebrospinal fluid was obtained under normal pressure and revealed no cells, a glucose of 80 mg%, and a protein of 60 mg%. Cerebrospinal fluid serology was nonreactive. On the fifth day of hospitalization, the patient was discharged with improvement of his neurologic deficit and remains symptom-free to date.

Methods and Results

Conventional electro-oculogram recording of horizontal and vertical eye movements was obtained the night of admission. The equipment used have been described previously. Vestibulo-ocular reflex was tested in the dark. The patient was asked to fixate on a distant static target as his chair followed a sinusoidal oscillation. The oblique vestibulo-ocular reflex gain was obtained by asking the patient to tilt the head 45° to the right and left from midposition, as the chair oscillated.

In the primary position of gaze, the patient showed primary position nonrhythmic, pendular rotatory, and vertical monocular oscillations in the right eye with an average frequency of 2-3 Hz and an amplitude in the vertical and horizontal planes of 6°, respectively. When the patient fixated on an eccentric target, the oscillation became strictly vertical with an amplitude of 12°, and the nystagmus components in this position were pendular, with an average velocity of 35°/second (Fig. 1).

Horizontal saccades were tested following a sequence described previously. The saccades were normometric and had normal velocity and latency. While the gain of horizontal pursuit was 1.0, vertical pursuit showed decreased gain (0.6). The gains of horizontal and oblique vestibulo-ocular reflex were normal. The rest of the electro-oculogram was unremarkable.

Discussion

Dissociated nystagmus refers to a group of disorders in which the nystagmoid movements of the two eyes differ in amplitude and/or direction. This definition includes patients with monocular nystagmus. A large variety of clinical conditions have been associated with monocular nystagmus. Spasmus nutans is probably the most common cause. Chiasmal and optic nerve gliomas and other suprasellar mass lesions have been reported in association with rotatory, vertical, or horizontal monocular nystagmus. In most of these cases, decreased visual acuity in the affected eye was noted in addition to significant local mass effect. Monocular visual loss without other known abnormalities has been reported in association with horizontal or
vertical monocular nystagmus. In these cases, a supranuclear system for monocular control of fixation dependent on visual cues initiated in the same eye has been postulated. Yee believes that this supranuclear control system could involve the vergence system, the only oculomotor control system known to produce monocular movements. This possibility, however, seems unlikely, since there is no evidence at present that the vergence system is involved in the generation or control of vertical eye movements.

Horizontal monocular nystagmus has recently been reported as an ictal phenomenon in a patient with photosensitive epilepsy. Animal experiments have demonstrated brain stem sites where electrical stimuli lead to monocular nystagmus. Brain stem lesions in humans can also cause monocular nystagmus. Duane described two patients with monocular rotatory nystagmus similar to ours. One of them with an apparent brain stem lesion, had a diagnosis of general paresis. Initially, this patient had been in coma for 3 weeks. Delirium, gait ataxia, and dysarthria were noted as the patient regained consciousness; 1 year later, monocular rotatory nystagmus was found. The second patient had a long-standing history of intermittent vertical diplopia and had been aware of involuntary, periodic monocular oscillations for several years. With the exception of monocular rotatory nystagmus, no other abnormalities were found. Circumduction nystagmus has been reported in patients with multiple sclerosis. These cases are of interest because the nystagmus necessarily involved vertical as well as horizontal monocular movements. In none of these reports was a precise location for the causative lesions apparent.

Monocular eye movements attributable to the action of the superior oblique muscle have been described as superior oblique myokymia. Patients with superior oblique myokymia tend to have chronic courses of intermittent oscillopsia and resting hyperphoria in the affected eye. Characteristically the movements are of high frequency and low amplitude, often requiring ophthalmoscopic examination to be identified. Electromyographic studies revealed few abnormal interference pattern units in the superior oblique muscle which were
consistent with a lesion in the region of the fourth nerve nucleus. Simultaneous recordings in the inferior oblique muscle were normal.\textsuperscript{15}

In our case, the monocular rotatory nystagmus was similar in character to that described in superior oblique myokymia, but differs in the greater amplitude of the eye movements and their nearly continuous occurrence during the acute episode. The normal gains on the horizontal and oblique vestibulo-ocular response suggest that a supranuclear lesion was responsible for the production of the nystagmus seen in our patient. Maintenance of steady fixation is the result of burst neuron inhibition by pause cells located in the medial pontine reticular formation.\textsuperscript{20} Lesions involving this group of cells could result in unsteady fixation and involuntary saccadic horizontal, vertical, and oblique eye movements.\textsuperscript{20} Although this inhibitory influence is likely to be binocular in nature, monocular interruption of this prenuclear inhibitory mechanism could result in monocular nystagmus. Finally, considering the brief duration and complete reversibility of the monocular nystagmus in this case, it is likely that transient ischemia was the main etiologic factor.

References


Acknowledgment

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The Wyburn-Mason Syndrome
Concomitant Chiasmal and Fundus Vascular Malformations

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Abstract
We report a 56-year-old female with a 50-year history of a progressive chiasmal syndrome who was found to have a suprasellar arteriovenous malformation involving the chiasm and both optic nerves associated with an unusual fundus picture consistent with the Wyburn-Mason syndrome. A review of the literature of this rare syndrome is also presented.

Introduction
The coexistence of arteriovenous malformations of the fundus and brain is known as the Wyburn-Mason syndrome.1 We recently evaluated a patient with a 50-year history of a progressive chiasmal syndrome secondary to a suprasellar arteriovenous malformation involving the chiasm and intracranial optic nerves, associated with an unusual fundus picture consistent with the Wyburn-Mason syndrome.

Case Report
A 56-year-old, right-handed white woman was referred to the Bascom Palmer Eye Institute with a complaint of loss of the temporal visual field in her right eye. Her history was particularly interesting—for as long as she could remember she was only able to see "half of objects" with the left eye. However, the left eye vision became progressively worse, so that she eventually lost all light perception in that eye between 20 and 30 years of age. She had no problems with the right eye to her knowledge for the next 25 years.

Approximately 8 years prior to presentation, her visual field was checked during a driver's license examination, and she was found to have some difficulty in the right temporal field. She was not aware of any change in the field thereafter until approximately 6 months prior to presentation. The patient then noted that she was missing objects in the right temporal field that she would normally have seen. Of interest was a history of severe vascular headaches since childhood that centered over the left eye and gradually lessened in severity and frequency as she became older. Except for a 40-lb. weight loss without a change in eating habits during the past 2 years, she has had no other complaints. Family history revealed that one daughter was operated for a pituitary tumor at age 26 and another daughter had a suboccipital mass excised at 1 year of age, but was doing well otherwise at age 31. The family history was otherwise negative for vascular headaches, aneurysms, or subarachnoid hemorrhages.

The patient presented to an ophthalmologist for the first time in her life in March 1982, and a temporal field defect was found in her right eye. A computed tomographic scan was obtained, and she was referred to the Neurosurgical Service at the University of Michigan, where a transfemoral selective arteriogram was performed.

Neuro-ophtalmological examination in Miami on August 26, 1982, revealed that the best-corrected acuity was 20/15+1 and I-I in the right eye and no light perception in the left eye. The right pupil reacted crisply to light, and the left pupil was amaurotic. Perimetry demonstrated a dense temporal hemianopia in the right eye (Fig. 1). Ocular motility was full. The patient had slow vertical ocular pendular oscillations of the left eye that were classic for the Heimann-Bielschowsky phenomenon.2 Slit lamp examination was normal, but for subtle, slightly dilated vessels on the plica, caruncle, and bulbar conjunctiva of both eyes. No ocular or cranial bruits were heard. Ophthalmoscopy revealed a normal fundus in the right eye (Fig. 2). The left fundus, however, was strikingly abnormal (Fig. 3). The left disc was pale, elevated,

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Figure 1. Peripheral visual field (3/330 white) showing temporal hemianopic defect in right eye.

Figure 2. Right eye, normal fundus.

Figure 3. Left fundus is markedly abnormal with pale, elevated disc, arterial narrowing, and dilated, sclerosed retinal veins.

blurred, and showed an optociliary shunt vessel at 5 o’clock (Fig. 4). The retinal arteries were narrow in that eye; however, the veins were distended, tortuous, and had a striking white appearance to them (Fig. 5). The peripheral retina was avascular with thinning of the retinal pigment epithelium. There was peripapillary pigment migration into the retina.

Fluorescein angiography demonstrated delayed filling of the retinal blood vessels with leakage and
staining of the vessels and optic disc. There was marked loss of capillarity and loss of all peripheral retinal vasculature. This was in striking contrast to the abnormal vessels on the nerve head (Figs. 6-8).

The computed tomographic scan demonstrated an enhancing lesion involving the chiasm and extending from the left to the right optic canal (Figs. 9 and 10). The transfemoral selective arteriogram demonstrated a moderately small arteriovenous malformation arising from the supraclinoid portions of both carotids (Figs. 11-16).

Discussion

Arteriovenous communications of the retina are a rare disorder that was first described by Magnus.
in 1874. The association of retinal arteriovenous communications with more extensive vascular malformations of the face and brain was first recognized as a clinical entity by Bonnet et al. in 1937. This grouping was further characterized by Wyburn-Mason in 1943 and coexistent arteriovenous malformations of retina, optic nerve, and brain have since been referred to as the Wyburn-Mason syndrome. The arteriovenous communication represents a congenital abnormality that is not felt to be hereditary.

Archer et al. divided retinal arteriovenous communications into three groups. Group 1 consists of those cases in which there is an arteriolar or abnormal capillary plexus interposed between the artery and vein. These patients usually show no
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Figure 10. Postcontrast axial CT scan of arteriovenous malformation (arrow) seen at the level of the optic chiasm.

Figure 11. Lateral view of the right common carotid arteriogram demonstrating a vascular mass (arrow) arising from the supraclinoid portion of the right carotid artery.

signs of vascular decompensation, have normal vision, and are not associated with arteriovenous malformations of the face and brain. In group 2, there is a direct arteriovenous communication. A hyperdynamic flow pattern develops and may result in alterations of the neighboring microvasculature consisting of capillary nonperfusion and microaneurysms. There may be vascular decompensation ranging from mild edema to exudation and hemorrhage. This group is only rarely associated with cerebral vascular malformations. Only one patient from their series was categorized as belong-
Figure 12. Selective left lateral internal carotid injection demonstrates abnormal vascularity (arrowhead) in supraclinoid portion of the left internal carotid artery.

Figure 13. Left posterior anterior common carotid arteriogram identifies multiple small feeder vessels arising from the supraclinoid portion of the left internal carotid artery to feed a highly vascular midline mass (arrowheads).

Figure 14. Left vertebral posterior anterior projection is normal.
Figure 15. Capillary phase shows the arteriovenous malformation (arrow) draining into the basal vein of Rosenthal (white arrow).

Figure 16. The arteriovenous malformation (white arrow) is seen draining into the basal vein of Rosenthal (black arrowheads) to the vein of Galen (black arrow) with subsequent drainage into the sigmoid sinus.
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ing to group 3, which consists of large caliber intertwined anastomosing channels in which it is difficult to distinguish the artery from the vein. These vessels undergo sheathing and sclerosis, and the retina may show exudation, hemorrhage, and pigmented migration. The visual prognosis is poor. These patients (of which ours is an excellent example), have associated cerebral vascular malformations and fall into the category described by Wyburn-Mason.

Theron et al.\(^6\) reviewed 25 cases of documented vascular malformations of the retina and brain. Decreased vision was present in 16 of 21 patients. The most frequent visual field abnormality was a homonymous hemianopia. Orbital involvement was present in the vast majority of patients. The intracerebral vascular malformation was always unilateral, ipsilateral to the retinal lesion, deeply located, and often related to the optic pathway. Four patients had cerebral or subarachnoid hemorrhage. Other neurologic symptoms were mental disturbance, hemiparesis or hemiplegia, cerebellar dysfunction, and Parinaud's syndrome. Seizures were present in only one patient, and four patients were neurologically asymptomatic. Angiomas of the face were present in approximately half of the patients. In contradiction to Archer's classification, three patients with normal vision and visual fields were found to have intracerebral arteriovenous malformations with retinal lesions that would have been classified in groups 1 or 2.

The neurologic presentation of patients with Wyburn-Mason, therefore, differs significantly from patients with cerebral vascular malformations in general. According to Mackenzie,\(^7\) the presenting complaint of 50 patients with cerebral arteriovenous malformations was epilepsy 32%, hemorrhage 30%, and hemiparesis 12%. In the study by Theron et al.,\(^6\) the incidence of epilepsy was less than 5%, hemorrhage 12%, and hemiparesis 50%. The incidence of seizure activity is probably much less because of the deep location of the vascular malformation in the Wyburn-Mason syndrome. The eventual risk of cerebral hemorrhage is unknown in these patients.

Many authors claim that the retinal lesions do not progress.\(^8,9\) Our case, as well as many others,\(^10\) would refute that premise. Patients may present with either acute or gradual loss of vision. The former occurs secondary to spontaneous hemorrhage, and the latter is due to loss of nerve fibers from mechanical compression of the optic nerve, chiasm, or optic tract. Retinal arteriovenous communications have been shown to enlarge in size\(^11\) or undergo spontaneous thrombosis with resolution of the abnormal communication.\(^12\) Augsburger et al.\(^13\) describe a patient who developed sclerosis of a maculopapillary arteriovenous communication associated with enlargement of a superonasal malformation and decrease in vision. Archer et al.\(^5\) feel that moderate to large-sized arteriovenous communications may result in progressive retinal microvascular alterations that could represent yet another mechanism for progressive visual loss.

Pathologic specimens\(^16,17\) show that the vascular malformation may extend from the retina to the optic nerve, chiasm and tract, and to the midbrain and cerebellum. Histopathologically, the abnormal vessels may occupy the entire thickness of the retina; it may not be possible to distinguish arteries from veins. The blood vessels may develop fibromuscular medial casts. The retina may be attenuated and undergo cystic degeneration with migration of pigment from the retinal pigment epithelium into the stroma. The optic nerve may show marked compression from the large vascular channels associated with loss of nerve fibers and a decrease in the number of ganglion cells. Absence of the choriocapillaris with cystic degeneration of the choroid has been reported in a rhesus monkey with a retinal arteriovenous malformation.\(^18\)

The true incidence of cerebral vascular malformations associated with retinal arteriovenous communications is unknown. Wyburn-Mason originally felt 80% of patients with retinal arteriovenous communications had associated cerebral vascular malformations. However, the incidence of cerebral vascular malformation documented by angiography, surgery, or autopsy was only 17% in his series. Bech and Jensen\(^19\) reviewed the world literature on racemose aneurysms of the brain and found associated retinal vascular malformations in 8% of cases. In their own series,\(^20\) of 52 cases of cerebral vascular malformations, none were associated with retinal vascular malformations. They also found an incidence of approximately 23% of cases of racemose aneurysms of the retina associated with intracranial vascular malformations.\(^19\) Theron et al.\(^6\) felt that of the more than 80 cases of retinal arteriovenous malformations reported by 1974, 25 had documented cerebral vascular malformations, an incidence of approximately 30%.

Therefore, the ophthalmologist may be faced with the question of whether to pursue a search for an intracranial arteriovenous malformation should a retinal arteriovenous communication be discovered. In the presence of neurologic symptoms and/or visual field changes, an intracerebral malformation should be highly suspected. The diagnosis is based upon radiologic demonstration of the vascular lesion. Skull films may demonstrate whorl or circular calcifications, dilation, and tortuosity of vascular channels and an enlarged optic foramen. Postcontrast computed tomography is recommended as a screening procedure in these patients; if a vascular lesion is suspected, intraocular digital subtraction angiography may be used as a confirmatory study. If the lesion appears
to be in a surgically accessible location, cerebral arteriography would then be necessary as a preoperative study in order to define more clearly the vascularity of the mass.

It is important to differentiate the retinal arteriovenous malformation from other vascular anomalies. The benign capillary angioma seen in von Hippel-Lindau disease is a hamartoma that could be confused with the retinal changes of Wyburn-Mason syndrome. It progressively enlarges from a small nodule and develops a tortuous, dilated feeding arteriole and draining vein. The capillaries become incompetent and leak, requiring oblitative therapy because of the high incidence of exudative retinal detachments. It has an autosomal dominant inheritance pattern. Approximately 50% of cases affect both eyes, and 25% of cases are associated with central nervous system hemangio- blastomas. There is also an association with renal cell carcinoma, pheochromocytoma, and cysts of the pancreas, lung, epididymis, and kidney. Retinal telangiectasias consist of focal areas of dilated, irregular leaking vessels that vary in size and symptomatology. The disorder is usually unilateral and is not associated with intracranial vascular anomalies. Venous stasis may have enlarged tortuous veins, but should be easily distinguished from an arteriovenous malformation.

Our patient is unusual in several respects. Most patients present before the age of 30. Although she has had significant symptoms for her entire life, she did not present for evaluation until the age of 54 and is the oldest patient, to our knowledge, with the Wyburn-Mason syndrome. We were able to find reference to only one other patient with such severe fundus change. Archer et al. describe a patient who was blind since birth in one eye with a temporal hemianopia in the contralateral eye. The fundus had tortuous, obliterated veins with aneurysmal dilations. There were areas of pigmentation and lipid deposition with accumulation of fibrous tissue. Interestingly, their patient also had a suprasellar arteriovenous malformation involving the chiasm and both optic nerves.

Asymptomatic retinal arteriovenous communications do not require therapy. If the lesion undergoes vascular degeneration with hemorrhage or lipid exudation, photocoagulation may be of benefit. The cerebral arteriovenous malformations are usually not accessible to surgical intervention because of their deep location. Tamaki et al. reported a patient with a large vascular malformation of the posterior fossa who had symptomatic improvement of her cerebellar ataxia and headache following a course of cobalt radiation therapy. Schlieter et al. described a patient with the Wyburn-Mason syndrome with an inoperable vascular malformation of the left temporomedial region that involved the basal ganglia that was reducted in size by partial embolization. To our knowledge, the suprasellar and chiasmal arteriovenous malformation in the patient here reported is inoperable without placing the visual field of the only remaining eye in jeopardy. We have elected to follow her conservatively at this time.

References
Wyburn-Mason Syndrome


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Complete Bilateral Internal Carotid Artery Occlusion in a Young Man

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Abstract

Partial or complete occlusion of the internal carotid artery is a familiar consequence of severe atherosclerosis seen in the elderly. Complete obstruction of both internal carotids is rare, particularly in the young or middle-aged. The rapid onset of bilateral internal carotid occlusion would be expected to produce devastating neurological sequelae and probably not be compatible with survival. We present a case of a young man with complete obstruction of both internal carotid arteries whose presenting symptoms were those of a visual field cut. The history suggests that the carotid occlusion occurred as a result of blunt trauma. The patient had no known predisposition to vascular abnormalities (no history of hypertension, hyperlipidemia, signs of systematic arteriosclerosis or vasculitis, and an unremarkable family history for vascular abnormalities). Computerized tomography revealed an infarct in his right parietal lobe. Angiography demonstrated complete occlusion of both internal carotid arteries and the right posterior communicating artery and failed to disclose the development of extensive collaterals, adding further evidence to the acuteness of the occlusion. The patient was followed by noninvasive studies and in the subsequent year showed marked neurological and ophthalmological improvement.

Case Report

R.B., a 39-year-old, right-handed white man, presented to Massachusetts Eye and Ear Infirmary with the chief complaint of blurred vision in the left eye of 4 days duration. While driving his cab he noted difficulty judging distances. He also complained of dull right frontal headaches of 5 days duration exacerbated by alcohol consumption. He denied any other neurological, visual, or constitutional symptoms. The family history was negative for diabetes, hypertension, or stroke.

Past medical history was remarkable for a motor vehicle accident 4 months previously with blunt trauma to the left lower face and left shoulder that left him with left-hand clumsiness and paresthesias, felt by a neurologist to be due to damage of the left brachial plexus.

On physical examination he was an alert, cooperative, well-developed, well-nourished white man in no apparent distress. BP was 142/100 left arm and 138/100 right arm. Visual acuity was 20/25 in both eyes, though he seemed to ignore letters to the left. Color vision was intact in both eyes. Extraocular movements including smooth pursuit and voluntary saccades and pupil responses were normal in both eyes. Optokinetic nystagmus testing demonstrated a slight difficulty in making saccades to the left. Slit lamp exam, tonometry, and fundus exams were all normal. Visual field testing by Goldmann perimetry revealed incongruous homonymous left field defects most pronounced in the left inferior quadrants (Figs. 1A and 1B).

Neurological exam was significant for inabilities to reproduce spatial figures and left arm weakness and hypoesthesia.

A right parietal lobe lesion was suspected and subsequently confirmed by computerized tomography (CT). As seen in Figure 1B, there was an area of reduced enhancement in the right parietal lobe consistent with encephalomalacia. Skull x-rays, Chest x-ray, CBC with differential, lipid profile, and ESR were all normal.

The patient was admitted to the Massachusetts General Hospital with suspicions of a parietal lobe astrocytoma and underwent cerebral angiography. Injections into the right and left common carotid arteries demonstrated complete occlusion of both internal carotid arteries just beyond their bifurcation sites (Figs. 2A and 2B). Posterior circulation injections demonstrated occlusion of the P1 segment of the right posterior cerebral artery, with no filling of the right posterior communicating artery (Fig. 2C). The left posterior communicating artery filled and supplied the left middle and anterior cerebral arteries. Doppler ultrasonography studies of the neck, 6 and 12 months after the onset of symptoms, confirmed the bilateral internal carotid occlusion without evidence of recanalization. Digital subtraction films (Fig. 2D) 1 year later also showed persistent complete internal carotid artery occlusion. Examination of the anterior segment by
slit lamp exam and of the fundus by indirect exam as well as fluorescein angiography failed to disclose any ocular abnormalities.

In summary, an ostensibly healthy, relatively young man presented with uniocular blurred vision and was found to have visual field defects referable to a parietal lobe lesion. Computerized tomography demonstrated encephalomalacia of the right parietal lobe. Angiographic evidence of markedly advanced cerebral arterial stenosis led to the conclusion that the parietal lobe lesion was due to old cerebral infarct. The widespread nature of the disease was felt to be inoperable; the patient was placed on aspirin, advised to change vocations, and has been followed with noninvasive studies of his cerebral blood flow.

Discussion

One of the first clinical-pathological studies of a case shown at autopsy to have complete occlusion of one internal carotid artery was described by Virchow (1859). The ophthalmic and central retinal arteries were patent, yet the patient had lost all vision in the eye ipsilateral to the thrombosed carotid artery.

In 1893, Elshnig noted frequent findings of complete internal carotid artery occlusion at necropsy in patients without any visual symptoms. He performed injection studies on cadavers and demonstrated the profuse collateral supply of the ophthalmic artery. Elshnig concluded that an occlusion of the internal carotid artery would have to be sudden to produce ocular symptoms. In 1905, Chiari noted that mural thrombi or atheromatous plaques in the internal carotid arteries of the elderly could be a source of emboli which produced lesions more peripherally.

In 1951, C. M. Fisher described 200 cases of middle cerebral artery cerebral symptoms which were shown at necropsy to be due to the occlusion of the internal carotid artery. He concluded that internal carotid artery stenosis was often unrecognized, unsuspected, and, therefore, far more frequent than previously suggested.

The clinical picture of carotid artery disease was addressed by Ramsy Hunt (1914), who mentioned intermittent attacks and Denny-Brown (1951), who described transient ischemic attacks as a symptom of carotid artery or circle of Willis disease. The symptomatology of such transient ischemic attacks consisted of contralateral cerebral or ipsilateral ocular (amaurosis fugax) dysfunction. The most common cerebral symptoms were transient hemiparesis or limb weakness, fleeting somatosensory disturbances on one side of the body, and mental confusion. Transient homonymous hemianopsia was uncommon in cases of carotid system disease.

Complete occlusion of the internal carotid artery may be neurologically indistinguishable from middle cerebral or anterior cerebral thrombosis. The middle cerebral picture is characterized by hemiplegia, hemianaesthesia, and hemianopia. Hollenhorst noted visual field defects in 45 of 235 cases of carotid artery occlusions. The homonymous hemianopsia was moderately incongruous and always seen with hemiplegia.

It is interesting to note that in the present report, the patient did present with incongruous homonym-
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Figure 2A. Right carotid angiogram. The right common carotid was filled revealing complete blockage of the right internal carotid artery (arrow) near the site of bifurcation. Note the vertebral artery filling by reflux (double arrow).

Figure 2B. Left carotid angiogram. The left common carotid was injected revealing complete blockage of the left internal carotid artery near the site of bifurcation (arrow).

Figure 2C. Left vertebral angiogram, lateral view. The left vertebral artery was injected revealing filling the basilar artery (arrow) and filling of the intracranial internal carotid artery circulation via the posterior communicating arteries with subsequent opacification of the middle and anterior cerebral arteries (double arrows).

Figure 2D. Digital subtraction angiogram following venous injection of contrast material, 1 year after presentation. Note continued complete occlusion of right (arrow) and left (double arrows) internal carotid arteries. Compare with Figs. 2A and 2B.

Hughes and Brownell" describe two cases of blunt trauma which produced a dissecting aneurysm of the internal carotid artery. In each case, the patient developed neurological symptoms and died within 48 hours from distal emboli. The fact that our patient remained alive and relatively intact is striking and suggests that either the dissection...
occurred slowly or that a thrombus did not immediately develop after the blunt trauma.\textsuperscript{10,11}

Fisher et al.\textsuperscript{12} describe a "string sign" which can be seen on angiography of an internal carotid artery occluded by spontaneous dissection. They saw this angiographic pattern in about half of their cases which were ultimately diagnosed in surgery or pathology as dissecting aneurysms. They noted, however, that late after the dissection, the angiographic appearance is often that of a complete occlusion. Our patient's angiograms did not show a "string sign," suggesting complete occlusion of the lumen by extension of the aneurysm or retrograde thrombosis.

There were no ocular symptoms or signs; this is probably due to the rich collateral supply to the ophthalmic artery. However, ophthalmic testing revealed cerebral deficits produced by the abnormal circulatory pattern. It is evident that the ophthalmologist or neuro-ophthalmologist often is in position to suggest the diagnosis of internal carotid artery stenosis. This case points out the sensitivity of visual field testing, the power of angiography for definitive description of the cerebral hemodynamics, and also of the value of noninvasive studies such as ultrasound-doppler and digital subtraction, for the long-term follow-up of a patient with vascular abnormalities.

References


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Orbital Myositis

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Abstract

We report five cases of presumed orbital myositis mimicking extraocular muscle motility disturbances and manifesting clinical signs of active inflammation over the involved muscles. Computed tomographic evidence for extraocular muscle enlargement is helpful in confirming the diagnosis. If not present or atypical, another etiology should be sought. All patients responded rapidly and dramatically to systemic corticosteroids. Anterior inflammation may be accompanied by iritis and respond to topical corticosteroids. We believe the diagnosis of orbital myositis may be made on clinical grounds with confirmation by computed tomographic evidence for extraocular muscle enlargement and clinical response to corticosteroids. Biopsy is unnecessary except in atypical cases.

Introduction

The diagnosis of orbital myositis implies orbital inflammation confined to one or two extraocular muscles and may represent a distinct clinical entity mimicking a paretic extraocular muscle of acute onset, with restricted ductions. It is responsive to systemic corticosteroids. We report five patients with acute extraocular motility dysfunction presumably due to orbital myositis. The diagnosis, based upon suspicion, clinical presentation, and computed tomographic evidence, was confirmed by rapid resolution after treatment with systemic corticosteroids.

Case 1

A 37-year-old woman presented with a swollen, painful right eye and horizontal diplopia. Her symptoms had increased for 3 weeks prior to referral. She denied any antecedent upper respiratory infection or history of sinusitis. Examination revealed 20/15 acuity in each eye. The right eye was restricted in both abduction and adduction (Fig. 1). The conjunctiva over the medial rectus was inflamed (Fig. 2). Six millimeters of proptosis were present in the right eye. Slit lamp examination and intraocular pressures were normal as was funduscopic. B-scan ultrasonography revealed a markedly enlarged medial rectus, better demonstrated by computed tomography (Fig. 3a). There was no other evidence for orbital pathology or involvement of the adjacent sinuses (Fig. 3b). She was treated with 80 mg prednisone daily with a rapid resolution of both pain and proptosis. Steroids were tapered over 8 weeks. Three weeks later, while taking 40 mg prednisone per day, she was asymptomatic except for mild restriction of abduction of the right eye. Examination was otherwise negative. Two weeks later, while taking prednisone 10 mg per day, there was still limitation of ocular abduction. Eight weeks after presentation, she was asymptomatic and extraocular motility was normal. Prednisone was discontinued and she has remained asymptomatic for the past 6 months.

Case 2

A 16-year-old boy was referred with a diagnosis of orbital cellulitis. He related a 2-day history of pain and swelling of the right periorbital region. Afferent visual function was normal. Examination revealed 4 mm of right proptosis, swelling, and erythema of the right orbit, and mild limitation of abduction and adduction of the right eye (Fig. 4). Neuro-ophthalmologic examination was otherwise normal. B-scan ultrasonogram demonstrated enlargement of the right medial rectus. Computed tomography (Figs. 5a and 5b) demonstrated an enlarged medial rectus and a diffuse right pansinusitis involving the right frontal, ethmoid, and maxillary sinuses. He was treated with 80 mg of prednisone daily with resolution of orbital and sinus abnormalities over 48 hours. Steroids were rapidly tapered and decreased over a 2-week period. He has remained asymptomatic. Computed tomography 4 months later documented normal orbits and paranasal sinuses.

Case 3

A 53-year-old woman was referred with a 1-week history of pain in the left eye increasing with abduction. She was treated by her family physician with intramuscular penicillin and oral erythromycin for a "blocked tear duct." When she failed to improve, she was referred for evaluation. No other significant history was elicited. Visual acuity was...
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Figure 1. Restriction of both ocular abduction and adduction due to enlarged, myositis, right medial rectus.

Figure 2. Localized conjunctival inflammation overlying medial rectus acutely inflamed by orbital myositis.

There were no localizing pupillary signs. The left orbit was exquisitely tender to palpation. A partial left ptosis was present. Extraocular motility was restricted in elevation and abduction (Fig. 6). There was obvious inflammation over the superomedial portion of her left globe. Two millimeters of left proptosis were present. Slit lamp examination, intraocular pressures and funduscopy were normal.

B-scan ultrasonography revealed a mass in the superomedial orbit. Computed tomography demonstrated a well-circumscribed mass in the superomedial orbit adjacent to and contiguous with the medial rectus (Fig. 7a); after injection of contrast, ring enhancement was evident (Fig. 7b). There was no computed tomographic or x-ray evidence for bony destruction or involvement of the adjacent sinus.

She was treated with 80 mg prednisone per day. Within 24 hours she was pain-free; after 72 hours she was asymptomatic. Prednisone was decreased...
to 40 mg per day. Two weeks later, she was totally asymptomatic and her ophthalmic examination was normal. B-scan ultrasonography revealed the acoustically homogeneous medial orbital mass to be markedly decreased in size. Prednisone was rapidly tapered and discontinued over a 6-week period. She remained asymptomatic. A repeat computed tomography was obtained 6 weeks after her initial scan and demonstrated a marked reduc-

Figure 4. Periorbital swelling and mild limitation of ocular abduction and adduction, right eye.

Figures 5a and 5b. Computed tomograms (axial and coronal reconstruction) demonstrating enlargement of the right medial rectus and adjacent ethmoid sinusitis.
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of the globe was inflamed. Forced ductions were positive in the right eye confirming the presence of a pseudo-Brown's syndrome. The remainder of the ocular and orbital exam was normal. There was no x-ray evidence for sinusitis. A diagnosis of superior oblique myositis was made. She was treated with 80 mg prednisone per day with rapid resolution of her symptoms. Four days later, examination was normal. Steroids were rapidly tapered over 2 weeks. She was asymptomatic for 7 months. She then presented with mild iritis and a recurrent pseudo-Brown's syndrome. Both responded to topical 1% prednisolone acetate over a 2-week period. Three months later, her superior oblique myositis recurred accompanied by superior periorbital edema. Orbital computed tomography was negative, except for soft tissue swelling in the vicinity of the trochlea. She again responded to topical prednisolone acetate and is presently asymptomatic on a maintenance dose of topical prednisolone acetate twice daily.

Case 5

A 10-year-old girl was referred for evaluation of ptosis. Three weeks prior to evaluation, she awoke with a ptotic upper lid. Over the next 2 weeks, mild swelling and tenderness developed. After a negative laboratory and neuroradiologic evaluation, she was hospitalized and treated with intravenous ampicillin for presumed orbital cellulitis. When the swelling and erythema increased, she was referred for neuro-ophthalmologic consultation.

No significant additional history was elicited. Visual acuity and color vision were equal and normal. The left upper lid was ptotic with no levator function evident. The lid was mildly erythematous and swollen with a palpable mass extending the orbital septum. A left hypotropia was present and left superior rectus dysfunction was evident (Fig. 10). Two millimeters of left proptosis were present. The superior conjunctiva was inflamed. The remainder of the anterior segment and fundus examination was normal. B-scan ultrasonography demonstrated enlargement of the left superior rectus. X-rays of the sinuses were normal. Computed tomography, obtained prior to referral, demonstrated soft tissue swelling in the anterior and superior orbit. She had not been febrile and her white blood count was normal. After discontinuing the ampicillin, she was treated with 60 mg of prednisone daily. In 24 hours, her ptosis and superior rectus paresis had partially resolved as did her subjective symptoms. One week later, she was asymptomatic except for mild residual left ptosis. Extraocular motility was normal. Steroids were tapered and discontinued over 3 weeks. She has remained asymptomatic.
Discussion

The first two cases represent orbital myositis confined to the medial rectus. The first patient had neither an adjacent sinusitis or an antecedent upper respiratory infection as described previously by others.\(^3\)

The second patient demonstrated orbital myositis of the medial rectus associated with a diffuse pansinusitis, mimicking orbital cellulitis. Eshaghian and Anderson\(^4\) reported inflammatory orbital masses with adjacent sinus involvement mimicking malignant neoplasia of the sinus, invading the orbit through apparent erosion of the medial orbital wall. No such pseudoerosion was present in our patient. Our patient was referred with the diagnosis of orbital cellulitis. Since he was only mildly febrile (100°F) and had a normal white blood count, we suspected inflammatory orbital disease. Computed tomographic evidence of an enlarged medial rectus confirmed our diagnosis. Both these patients had
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Figure 8a. Follow-up computed tomogram demonstrating decreased size of orbital mass and loss of ring enhancement.

Figure 8b. Coronal reconstruction demonstrating mass to be superior to and adjacent to the left medial rectus.

the rapid and dramatic response to systemic corticosteroids reported by others. They also demonstrated that medial rectus myositis may occur with or without adjacent sinus involvement and without a significant antecedent upper respiratory infection. Follow-up computed tomography (case 2) documents total resolution of the inflammatory process in both the orbit and sinuses.

Our third case represented what we believe to be a variant of orbital myositis. She presented with a painful ophthalmoplegia thought secondary to orbital myositis and her symptoms responded dramatically to systemic corticosteroids within 24 hours. However, her computed tomography (Figs. 7a and 7b) was atypical, demonstrating an enhancing medial orbital mass that failed to resolve totally on repeat scan 6 weeks later (Figs. 8a and 8b).

Evaluation of coronal reconstructions obtained during her follow-up scan revealed the mass to be contiguous with the medial rectus (Fig. 8b). These scans are obviously different from those demonstrating obvious medial rectus enlargement. It has been suggested that this picture may represent the lymphocytic variant of idiopathic orbital inflammation. Shortly after the second computed tomography, extensive metastatic melanoma from a cutaneous primary was documented.

Although we have no histopathologic evidence, we wonder whether this orbital mass may also represent metastatic melanoma with a surrounding inflammatory response which responded dramatically to systemic corticosteroids. A discrete metastasis to an extraocular muscle from silent small cell carcinoma has been recently described. Although mimicking orbital myositis, this patient did not respond to systemic corticosteroids. However, our patient's orbital symptoms did not recur after discontinuing the corticosteroids. A follow-up scan was negative, suggesting an atypical form of orbital
myositis. We urge clinicians to be suspicious of the diagnosis of orbital myositis if the computed tomography picture is atypical in spite of a good clinical response to systemic corticosteroids, or if the presumed myositis fails to respond to corticosteroids.

Case 4 represented orbital myositis presenting as a pseudo-Brown's syndrome. Again, external signs of inflammation were present over the involved area. Computed tomography failed to demonstrate extraocular muscle enlargement, but we suspected a trochleitis or myositis based upon clinical findings (point tenderness, restriction of motion, and overlying inflammation). A concomitant anterior uveitis has been reported previously to accompany orbital inflammation and might be expected with an anterior site of inflammation. This patient also demonstrated that idiopathic orbital inflammation, if anterior, may be controlled with topical corticosteroids.

Case 5 represented involvement of levator and superior rectus by orbital inflammation in a child. Again, there was evidence for active inflammation (warmth, erythema, a palpable tender mass over the involved lid, and injection of vessels over the involved superior rectus). Computed tomography was nonspecific as obtained on an outmoded unit, demonstrating only swelling of the lid, compatible with a preseptal cellulitis. The diagnosis here was made on clinical grounds—levator and superior rectus dysfunction accompanied by evidence of inflammation and confirmed by the rapid response to corticosteroids. High-resolution computed tomography with appropriate coronal reconstructions might well have documented enlargement of the levator and/or superior rectus.

Orbital myositis represents a distinct clinical entity that may mimic many disorders of extraocular motility. Evidence for local orbital inflammation should obviate against a neurologic etiology for these motility disturbances. Orbital myositis must be differentiated from other causes of orbital inflammation with motility disturbances. We believe this can be done by clinical examination, laboratory data, and computed tomography.

The most important entity to differentiate is orbital cellulitis. In our experience, these patients are febrile greater than 102°F, have a polymorphonuclear leukocytosis greater than 15,000, exhibit extraocular motility dysfunction, and often have a concomitant sinusitis or history of trauma. Our patients with orbital myositis have been otherwise well, essentially afebrile, and have normal white blood cell counts.

Partially treated orbital cellulitis may be confused with orbital myositis. Case 5 was referred...
after her orbital swelling failed to respond to intravenous and then oral antibiotics. In this situation, without diagnostic, computed tomographic evidence, we suggest discontinuing antibiotics and observation for 24–48 hours. If the clinical status does not worsen or remains stable, the patient is treated with systemic corticosteroids expecting a prompt, dramatic response.

The patient with orbital myositis has an exquisitely tender orbit to palpation and an obvious motility defect often limited to one or two extraocular muscles. Vision has been unimpaired in all of our cases.

We believe the diagnosis of orbital myositis can be made on clinical grounds and confirmed by computed tomographic evidence of an enlarged extraocular muscle and rapid response to systemic corticosteroids. Clinical findings included limitation of extraocular motility in the field of action of the affected muscle or in the opposite field, coupled with evidence of overlying inflammation and exquisitely tender to palpation. Proposis is often present, but may be minimal with more anterior lesions.

Computed tomography of the orbit may document markedly enlarged extraocular muscles thought to be distinguishable from enlargement due to Graves’ orbitopathy. The tendon is spared in a muscle enlarged secondarily to Graves’ orbitopathy, whereas both muscle and tendon are enlarged (Figs. 3a, 3b, 5a, and 5b) when involved by myositis. Other causes of extraocular muscle enlargement on computed tomography such as metastatic tumors, carotic cavernous fistulae, and trauma should be discernable by history, examination, and failure to respond to corticosteroids. Concomitant sinusitis may also be documented by computed tomography (case 2, Figs. 5a and 5b).

We agree with Purcell and Taulbee that orbital myositis is a distinct clinical entity. However, we find no consistent relationships to antecedent viral or streptococcal upper respiratory infections. Adjacent sinusitis may or may not be present. When present (case 2), it responds as dramatically as the orbital involvement to systemic corticosteroids. Eshaghian and Anderson have demonstrated similar histopathology in idiopathic orbital and adjacent sinus inflammation.

We do not postulate an etiology for orbital myositis but urge its recognition as a diagnostic entity by the clinician. Such recognition obviates the need for biopsying these inflamed orbits with their higher incidence of postoperative complications. We also recognize that some tumors can respond favorably to corticosteroids for a short time, but rarely as rapidly and completely as orbital myositis. We urge biopsy of patients presenting in an atypical fashion, possibly by fine-needle aspiration under sonographic guidance as described previously.

Patients with orbital myositis may be extremely uncomfortable and, in a referral practice, far from home. Their true clinical picture may be obscured by previous antibiotic treatment or surgery. Accordingly, our treatment regimen consists of hospitalization for observation and to expedite obtaining computed tomography. Treatment consists of 50 mg prednisone on admission followed by 80 mg daily in four divided doses. A dramatic subjective and objective improvement is usually evident in the first 12–24 hours. Corticosteroids are rapidly tapered over 3–4 weeks. Recurrences may be treated again with systemic corticosteroids. One patient (case 4) is successfully maintained on low-dose topical steroids after refusing further systemic prednisone. Patients who fail to respond to this regimen, or have an atypical clinical picture, subsequently undergo either open or fine-needle aspiration biopsies as their clinical condition dictates.

In essence, orbital myositis is a clinical diagnosis based upon suspicion, signs of extraocular muscle dysfunction with concomitant overlying inflammation, and a rapid response to systemic corticosteroids. Computed tomographic evidence for enlarged extraocular muscles is helpful; however, if the computed tomographic picture is atypical, another etiology should be considered and a histopathologic diagnosis obtained.

References

5. Personal communications with S. Trokel.
The article in this issue entitled “Amaurosis Fugax for a Long Duration,” by T. Fujino, S. Akiya, S. Takagi, and H. Shiga was published because it describes in detail the universally poorly recognized syndrome of oculal migraine. The syndrome of migraine has been recognized for over 2000 years. Migraine is best described in terms of recurrent attacks of headache, commonly unilateral in onset, associated with anorexia, nausea, and vomiting. These attacks may be preceded by or associated with conspicuous neurologic and mood disturbances. There are many types of migraine, but the ophthalmologist generally relates the expected symptomatology to that of classic migraine which occurs in only about 10% of patients. There is an acute onset of a prodromal phase which lasts 10–30 minutes and is classically visual in nature. The hemianopic scintillating scotoma was best described by Sir William Gowers in 1895. These scotomas usually begin just eccentric to fixation. They are sharply defined, white or multicolored, picket-like lines that may have a shimmering quality and, for the most part, are relatively localized. These scintillations then begin to slowly expand simultaneously moving peripherally. The center of the fortification specter is variously described as being blurred, grayed-out, or blank. As the scintillating scotoma begins to fade the patient notes the onset of the headache phase.

A much less common and much less publicized variety of migraine is the so-called “ocular migraine.” Ocular migraine may be defined as a transient or permanent monocular visual disturbance occurring in an individual with a strong history of migraine episodes. In this unique form of migraine, vasospasms of the retinal vessels have been postulated to be the cause of the unilateral visual loss experienced by the patient. As reviewed by Kline and Kelly, a variety of ocular manifestations have been reported including retinal and vitreous hemorrhages, ischemic optic neuropathy, branch and central retinal artery and vein occlusions, central serous retinopathy, and unexplained bouts of transient visual loss. These attacks of transient visual loss last for minutes to hours and are rarely accompanied by a headache phase. With repeat episodes the patients may develop permanent visual loss.

Kline and Kelly had the opportunity to document the fundus findings in such a patient with fundus photographs and fluorescein angiography. They demonstrated narrowing of the retinal veins during the attack similar to the findings reported by Wolter and Burchfield. There was also delay in appearance of fluorescein dye in the branches of the central retinal artery, but no delay in choroidal filling. They postulated that the visual loss in their patient was due to reduced arteriolar flow into the retinal circulation, possibly due to arteriolar spasm between the branching of the ciliary vessels and the surface of the optic disc. A marked variation in the amplitude, but not in the latency, of the visual-evoked response in this patient was mentioned by Kline and Kelly and subsequently discussed in detail by Kline and Glaser. The variation in amplitude paralleled the recovery of visual function. They suggest that the loss of the visual evoked response signal reflects the loss of inner retinal function, the inner retina being supplied by the central retinal artery. With the resumption of central retinal artery perfusion, there is a return of inner retinal activity and a gradual and progressive return of the visual evoked response signal. Since there was no permanent damage to the myelinated ganglion cells in this patient, there was no change in latency.

The parallel between the patient reported in this issue by Fujino et al. and the one reported by Kline and Kelly and Kline and Glaser is remarkable. As in the previous reports, the major change is in the caliber of the veins, not the artery (see Fig. 1, Fujino et al.). There is a suggestion of a relative delay in the venous return during this attack as demonstrated by the differences in laminar flow between various vessels (see Fig. 2, Fujino et al.), although this type of sequence is also occasionally noted in normal individuals. In the patient reported by Fujino et al. there was a general delay in the filling of the arteries and a more profound delay, at least during one episode, in the filling of the choroid (see Fig. 3, Fujino et al.). These findings suggest the occurrence of a vasospastic phenomenon occurring proximal to the branching of the long posterior ciliary artery somewhere in the course of the ophthalmic artery.

We concur, therefore, with the postulated site of occlusion as proposed by Fujino et al. We believe the mechanism of the “occlusion” is vasospasm secondary to a migraine diathesis. Since Kline and Kelly reported the prompt cessation of visual symptoms in their patient with the use of propranolol, we believe that such a trial is warranted in this patient. We do not believe that the congenital abnormality noted on angiography is of significance.

Fujino et al. pose a last question with respect to how the retina survives ischemic attacks of such long duration. It is highly likely that the ischemia
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is relative—severe but not total occlusion occurring with arteriolar vasospasm. The ischemia induces a physiologic block of retinal ganglion cell function producing a loss of vision reflected electrophysiologically by the loss of the visual-evoked response signal. If the spasm is severe enough or recurrent in the same distribution, permanent damage may be induced insidiously, as in the patient reported by Fujino et al., or cataclysmically in those patients who develop true branch9 or central10,11 retinal artery occlusions or ischemic optic neuropathy.12

The points to remember are: 1) migraine can affect the anterior visual system, and 2) these attacks may last for minutes to hours in contrast to the fixed pattern of 15–40 minutes for the fortification specter of classic migraine.

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References


This superimposed photograph of the left eye during full upward and downward gaze illustrates the action of the superior oblique muscle in the presence of a complete (pupil-sparing) third nerve palsy. An iris freckle dramatizes the intorsional and downward movements. A white line connecting the top of the pupil and the iris freckle has been added to show the intorsion.

The patient was a 79-year-old man with systolic hypertension and mild diabetes in whom a 48-hour evolution of a third nerve palsy was preceded by 3 days of supraorbital pain.

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