Acquired Esotropia
A Manifestation of Chiari I Malformation

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Abstract
A patient with acquired esotropia underwent apparently successful strabismus surgery. Subsequent recurrence of esotropia, associated with square-wave jerks and downbeat nystagmus led to further investigation. Although standard CT scan was normal, rescanning after instillation of metrizamide demonstrated a Chiari I malformation. Posterior fossa decompression alleviated the esotropia. Acquired esotropia has not been recognized as a manifestation of Chiari I malformation. Our case illustrates that a high degree of suspicion is required to make the diagnosis of Chiari I malformation. Specialized techniques, such as metrizamide cisternography, or magnetic resonance imaging may be necessary if routine diagnostic measures are unrevealing.

Introduction
Acquired esotropia, when not associated with systemic or neurologic disease, is often ascribed to a breakdown of fusional mechanisms and treated with strabismus surgery. Convergent strabismus has not previously been recognized as an early sign of Chiari I malformation. We report a patient in whom this association occurred. Strabismus surgery failed, but posterior fossa craniectomy has apparently succeeded in restoring binocular single vision.

Case Report
A 24-year-old saleswoman was referred for neuro-ophthalmic examination in December 1979, because of slowly progressive esotropia over the preceding 8 years. When she was 9 years old, the referring ophthalmologist had noted an asymptomatic esophoria of 4 prism diopters at distance and 8 prism diopters at near fixation. Fusional amplitudes were normal. Subsequent examination of the patient at age 13 disclosed myopia, with no change in muscle balance. Intermittent horizontal diplopia began at age 17, and became increasingly manifest with close work, driving, and alcohol consumption.

Her medical history was unremarkable; however, a brother had strabismus. Corrected visual acuity, pupils, confrontation fields, and fundi were normal. Smooth pursuit and saccades were intact. Abduction was full in both directions of horizontal gaze. An intermittent comitant esotropia was present and variable from 4 to 15 prism diopters. Stereoaucity was 50 seconds of arc. Occasional square-wave jerks were observed only with binocular fixation. Covering either eye abolished this phenomenon. No nystagmus was present.

A diagnosis of decompensated esophoria was made; surgical recession of the left, and 2 months later, the right medial rectus was performed in early 1980. Diplopia returned, and in February 1981, her esotropia measured 20 prism diopters in right gaze, and 10 prism diopters in left gaze.

She was referred for strabismus consultation 4 months later, at which time she had a 14-prism diopter esotropia that practically disappeared on left gaze, but increased on right gaze. At near, the esophoria was 18 prism diopters. Her near point of accommodation was 8.5 cm. (Prior to her surgical procedures, 18 months earlier, her near point of accommodation was noted to be 7 cm.) Vergences were reduced. Wearing prismatic correction, her stereoaucity was only 100 seconds of arc. Although the deterioration of her esophoria could not be satisfactorily explained, orthoptic treatment was suggested, and divergence exercises were subsequently instituted. The orthoptist noted that the patient had 3+ overaction of both superior obliques, yet only a tiny A pattern on extremes of gaze while fixating on a light.

Her personal ophthalmologist noted rotary nystagmus in September 1981, and she was referred for neuro-ophthalmic reexamination in April 1982. A constant esotropia was present and
unaffected by Tensilon. Fine counterclockwise rotary nystagmus was present and worsened in downward gaze to right and left. She denied difficulties walking, but demonstrated an ataxic gait to tandem testing. A standard computerized tomographic scan was normal, but rescanning after instillation of metrizamide in the subarachnoid space disclosed cerebellar tonsillar herniation to the level of the second cervical vertebra (Chiari I malformation) (Figs. 1a and 1b), a finding confirmed at posterior fossa decompression in June 1982 (Fig. 2). Shortly thereafter, the patient became diplopia-free. Examination 5 months after surgery revealed orthophoria and marked dampening of rotary nystagmus. Her gait remained unchanged.

Discussion

Chiari I malformation is the name given an anatomic anomaly, first described by Chiari in 1891, involving displacement of the cerebellar tonsils into the upper cervical canal. It is not associated with overt signs of central nervous system malformation, in contrast to Chiari II (Arnold-Chiari) malformation, which consists of extensive herniation of cerebellum and medullary cord through the foramen magnum in association with spina bifida. Symptoms in patients with Chiari I malformation are vague, with the most frequent being pain (head, neck, extremities), followed by weakness, paresthesias, oscillopsia, and unsteady gait. The average duration of symptoms is from 4.5 to 9 years before diagnosis, with initial misdiagnosis the rule. Ophthalmic signs and symptoms are important as they may be the sole localizing manifestation of a Chiari I malformation. The key localizing nature of downbeat nystagmus is illustrated by our patient. Nystagmus, variably present in 27–60% of affected patients, may be horizontal, rotary, downbeat, or even upbeat. Sixth nerve paresis, skew deviation, spasm of near reflex, Horner's syndrome, ocular dysmetria, and loss of optokinetic nystagmus have also been described.

Acquired comitant esotropia and Chiari I malformation is a rarely recognized association. We know of only one previous report, in which a 54-year-old female with Chiari I malformation had convergent strabismus. She had experienced basilar migraine headaches since age 17, unsteady gait for 10 years, and esotropia for 5 years. Her clinical findings included ataxia, ocular dysmetria, and downbeat nystagmus. As is commonly reported, initial diagnostic studies were normal (including CT scan), and myelography was required to demonstrate the Chiari I malformation.

The mechanism underlying apparently non-paralytic esotropia in Chiari I malformation is
unclear. Downward traction on the abducens nerves due to descent of the brain stem could result in paralytic esotropia, similar to that seen in patients with sixth nerve palsies developing during halo-pelvic traction. We know of one case (personal communication, W. F. Hoyt) involving a 13-year-old girl who presented with comitant esotropia associated with symmetrical abduction weakness and gaze-evoked nystagmus, which recurred after initially successful strabismus surgery. She subsequently developed rotary downbeat nystagmus with square-wave jerks, and had Chiari I malformation diagnosed by vertebral angiography. Some element of sixth nerve paresis could be postulated in this case. Our case lacked typical evidence of sixth nerve weakness or divergence paralysis. The possibility that a disturbance of the fusional mechanism by the Chiari malformation led to decompensation of our patient's preexisting esophoria is a plausible speculation.

A high degree of suspicion is necessary to diagnose Chiari I malformation, since conventional CT scanning is often unremarkable. Metrizamide CT scanning is of special importance, but false-negatives have been reported. Magnetic resonance imaging has been used in the diagnosis of Chiari I malformation, but its exact role remains to be determined.

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