Isolated Comitant Esotropia and Chiari I Malformation

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- PURPOSE: To report four patients with isolated comitant esotropia and Chiari I malformation and discuss the most appropriate management.
- METHODS: Case reports and literature review.
- RESULTS: All four patients (5, 14, 16, and 37 years of age) presented with an isolated comitant esotropia that led to the diagnosis of Chiari I malformation. The first two patients underwent uncomplicated neurosurgical decompression of their malformation, followed by complete resolution of their esotropia. The third patient underwent strabismus surgery and experienced initial resolution of the esotropia, but eventual recurrence resulted in the strabismus surgery being repeated 5 years later. The fourth patient had strabismus surgery with resolution of the esotropia but only 2 months of follow-up.
- CONCLUSION: Although management of patients with Chiari I malformation and severe neurologic findings typically includes surgical decompression, management is less straightforward in cases with subtle findings or in which ocular findings are isolated. The decision to perform neurosurgical decompression or strabismus surgery should still be made on a case-by-case basis, with the understanding that strabismus surgery may provide only temporary ocular alignment.

Chiari Malformations represent a continuum of hindbrain maldevelopments characterized by downward herniation of the cerebellar tonsils. Chiari I malformation is defined as tonsillar herniation of at least 3 to 5 mm below the foramen magnum (Figure 1). Osseous abnormalities at the craniocervical junction and syringomyelia are common. Chiari I malformation is a disorder of the para-axial mesoderm that is characterized by underdevelopment of the occipital bone and overcrowding of the normally developed hindbrain. The overcrowding of the cerebellum within a small posterior cranial fossa leads to chronic tonsillar herniation. Chiari I malformation usually becomes symptomatic in the second or third decade of life, although it may remain asymptomatic. Chiari I malformation is distinguished from Chiari II and Chiari III malformations, which are present at birth and consist of downward herniation of the lower cerebellum and medulla into the spinal canal, in association with complex anomalies of the brain. Magnetic resonance imaging has revolutionized the diagnosis of Chiari I malformation and has led to the detection of many cases that were previously unrecognized or misdiagnosed. Neuro-ophthalmologic symptoms and signs are common with Chiari I malformation; however, isolated comitant esotropia is rare. The causal relationship of a comitant esotropia with Chiari I malformation as well as the appropriate management of the esotropia is controversial. We report four patients with Chiari I malformation and isolated acquired comitant esotropia and discuss the management of this condition.

CASE REPORTS

- CASE 1: A 14-year-old, previously healthy, myopic girl presented with a 3- to 4-month history of horizontal binocular diplopia. She had no other visual or neurologic symptoms or signs. Her visual acuity was 20/20 in both eyes. Pupils were normal, as were results of biomicroscopy and ophthalmoscopic examinations. Visual fields were full. She had a comitant 35-prism diopter esotropia at distance and near with full versions. There was no nystagmus. A
magnetic resonance image of the brain revealed a type I Chiari malformation (Figure 1, center). The patient subsequently underwent a suboccipital decompression and C1 laminectomy. Postoperatively, she noticed an immediate improvement in her diplopia. Three months after surgery, she was orthotropic at distance and near in primary gaze, but had an esophoria in lateral gaze. Two years later, she was orthophoric in all positions of gaze except in extreme left gaze, where she had a 2-prism diopter esophoria (Figure 2).

**CASE 2**: A 16-year-old, previously healthy boy presented with a 2-year history of intermittent horizontal binocular diplopia that occurred while playing soccer. He had no other visual or neurologic symptoms or signs, even with physical activity. His visual acuity was 20/20 in both eyes. Pupils were normal, as were results of biomicroscopy and ophthalmoscopic examinations. Visual fields were full. He had a comitant 8-prism diopter esotropia. Ductions and versions were full. There was a mild gaze-evoked nystagmus in both right gaze and left gaze. Magnetic resonance imaging of the brain disclosed a type I Chiari malformation (Figure 1, bottom).

The patient underwent suboccipital decompression associated with C1 laminectomy without complications. Postoperatively, he noticed an immediate improvement in his diplopia. Three months after surgery, the diplopia with exercise had resolved and he was orthophoric at distance and near in all positions of gaze. Three years later, he was still orthophoric.

**CASE 3**: A 37-year-old woman presented with a 4-year history of progressively worsening horizontal binocular diplopia. Her medical history was remarkable for petit mal epilepsy and intermittent headaches diagnosed as migraine. She had no other visual or neurologic symptoms or signs. Her visual acuity was 20/20 in both eyes. Pupils were normal, as were results of biomicroscopy and ophthalmoscopic examinations. Visual fields were full. She had a
comitant 20-prism diopter esotropia with full ductions and versions. There was no nystagmus. Magnetic resonance imaging of the brain showed a type I Chiari malformation associated with severe basilar impression and hypoplastic clivus and odontoid process. The patient and the neurosurgeon elected not to intervene neurosurgically because of the complexity of the craniocervical junction abnormality, and the patient initially underwent correction with prisms. However, because of progressive worsening of her esotropia, she subsequently underwent strabismus surgery.

The patient was orthophoric in all positions of gaze for approximately 1 year after strabismus surgery, but her esotropia subsequently recurred and progressively worsened. Five years after surgery, she had a 30-prism diopter comitant esotropia in the distance with full ductions and versions. Neurosurgical treatment was again deferred because of the complexity of the craniocervical malformation. Strabismus surgery was repeated, and 3 months later she was asymptomatic with a 2-prism diopter esophoria in the distance.

**CASE 4**: A 5-year-old, previously healthy boy presented with a 13-month history of horizontal binocular diplopia. He had no other visual or neurologic symptoms or signs. His visual acuity was 20/20 in both eyes. Pupils were normal, as were results of biomicroscopy and ophthalmoscopic examinations. Visual fields were full. He had a comitant 8-prism diopter esotropia at distance with full versions; at near, he was orthophoric. There was no nystagmus. Magnetic resonance imaging of the brain disclosed a type I Chiari malformation. The possibility of strabismus surgery compared with suboccipital decompression was debated, and his parents decided on strabismus surgery. Two months later, the patient was asymptomatic and his eyes were straight.

**DISCUSSION**

CLINICAL DESCRIPTIONS OF CHIARI I MALFORMATION have undergone continuous revision since the original report by Chiari of tonsillar herniation in patients dying as a result of hydrocephalus.\(^\text{21}\) In recent years, Chiari I malformation has been defined radiologically as a tonsillar herniation of at least 3 mm\(^\text{8}\) or at least 5 mm\(^\text{7}\) below the foramen magnum (Figure 1). However, this definition is limited to a single criterion and makes no reference to clinical symptoms or the presence of associated findings such as syringomyelia. Because tonsillar herniation of at least 5 mm can be encountered as an incidental finding among asymptomatic patients,\(^\text{3}\) the causal relationship between Chiari I malformation seen on the magnetic resonance image and neurologic or ophthalmologic symptoms is difficult to assess. Indeed, the symptoms reported by most patients with Chiari I malformation are typically nonspecific and nonlocalizing, such as headache, retroorbital pressure, floaters or flashing lights, blurred vision, photophobia, intermittent diplopia, dizziness, disequilibrium, dysphagia, hoarseness, chronic cough, sleep apnea, palpitations, poor coordination, numbness, pressure in the ears, tinnitus, decreased hearing or hyperacusis, vertigo, and oscillopsia. Objective findings such as nystagmus, cerebellar signs, and cranial nerve deficits occur in only a minority of patients.\(^\text{1–6}\) Common misdiagnoses include migraine, fibromyalgia, and multiple sclerosis. Indeed, by the time of definitive diagnosis, approximately 50% of patients have been told that they suffer from a psychogenic disorder.\(^\text{1}\)

Esotropia is an objective finding that can be observed in patients with Chiari I malformation. It is sometimes referred to as divergence insufficiency when comitant or as unilateral or bilateral sixth nerve palsies when incomi-
In almost all cases, however, it is associated with typical symptoms and signs of Chiari I malformation, such as Valsalva maneuver–induced headaches, downbeat nystagmus, a cerebellar syndrome, and papilledema from hydrocephalus. Isolated comitant esotropia is rare in Chiari I malformation, and only a few cases have been reported. Indeed, acute acquired comitant esotropia is a rare cause of strabismus in any patient. It occurs most commonly with uncorrected refractive errors or after ocular occlusion. Its occurrence as the only sign of intracranial neoplasm and Chiari malformation is so rare that some authors have advocated that neuroimaging studies be performed only when other neuro-ophthalmic symptoms and signs are present.

Comitant esotropia in the Arnold-Chiari syndrome was initially attributed to the coexisting hydrocephalus; however, most patients with symptomatic Chiari I malformation do not have hydrocephalus. It is very unlikely that comitant esotropia is related primarily to abducens nerve dysfunction in light of the comitancy, and elevated intracranial pressure certainly cannot account for all the cases of comitant esotropia in Chiari I malformation. Lennerstrand and Gallo suggested that vergence mechanisms in the mesencephalon may be important in the genesis of comitant esotropia, with compression of this region in Chiari malformation. Milhorat and associates recently suggested that some symptoms and signs of Chiari I malformation may be related to changes in the cerebrospinal fluid flow rather than to direct compression of the brain stem. Indeed, in their recent series of 364 symptomatic patients with Chiari I malformation, Milhorat and associates observed that 9% of patients had tonsillar herniation of less than 5 mm. They suggested that the most constant feature of Chiari I malformation is not tonsillar herniation but a volumetrically small posterior cranial fossa that predisposes patients to hindbrain overcrowding and subsequent abnormalities in the cerebrospinal fluid dynamic as demonstrated by flow magnetic resonance imaging.

Although management of patients with Chiari I malformation and associated neurologic findings, such as a severe cerebellar syndrome or hydrocephalus, is usually straightforward, management is much less clear among the cases with subtle findings or in which ocular findings are isolated.

Nystagmus, especially downbeat nystagmus, is a common sign of Chiari I malformation. Surgical treatment of Chiari I malformation has been performed in patients with isolated oscillopsia from nystagmus. In most cases, the nystagmus improves dramatically postoperatively.

Suboccipital decompression has also been performed in rare cases of Chiari I malformation with isolated esotropia. In all cases, including our first two patients, the esotropia completely resolved and this result was maintained after surgery. Because suboccipital decompression associated with CI laminectomy is an invasive procedure, strabismus surgery is sometimes preferred in patients with Chiari I malformation and isolated esotropia. However, as illustrated by Case 3, because strabismus surgery does not correct the underlying process, the esotropia may recur. The neurosurgeon was reluctant to perform an invasive procedure in this patient because of the associated osseous abnormalities, which increased the complexity of the procedure. Without early neurosurgical treatment of the malformation, other ocular or neurologic symptoms may develop with time. Bixenman and Laguna described a young patient with comitant esotropia associated with Chiari I malformation who experienced recurrence of her esotropia 6 months after strabismus surgery. Because of new-onset downbeat nystagmus associated with headaches 3 years later, she underwent neurosurgical decompression of her Chiari I malformation, with resolution of all her symptoms postoperatively. Passo and associates observed a similar case. Weeks and Hamed recently reported two patients with acute comitant esotropia and Chiari I malformation who were initially treated with strabismus surgery alone. After initially successful surgical realignment, both patients developed recurrent esotropia, which resolved on suboccipital decompression. The authors suggested that the sequence of treatment should be suboccipital decompression and then strabismus surgery only if realignment does not occur.

However, this decision should still be made on a case-by-case basis, with the understanding that strabismus surgery may provide only temporary realignment but that neurosurgical intervention is a more invasive procedure.

REFERENCES


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