Chiari I malformation and acute acquired comitant esotropia

Case report and review of the literature

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The authors describe a rare case of a Chiari I malformation presenting with acute acquired comitant esotropia (AACE) in a 5-year-old boy. A posterior fossa decompression with duraplasty and a C1–2 laminectomy were performed. There was an immediate postoperative improvement in the esotropia, which completely resolved by 7 months following surgery. The pertinent literature is discussed and reasons are presented for recommending posterior fossa decompression in certain patients, rather than strabismus surgery, as the initial treatment for esotropia. The authors suggest that in patients with AACE, even subtle symptoms and signs of Chiari I malformation should prompt imaging of the posterior fossa. Strong consideration should be given to performing posterior fossa decompression in patients with Chiari I malformation and AACE before strabismus surgery because the esotropia may completely resolve with decompression.

KEY WORDS • acute acquired comitant esotropia • Chiari malformation • pediatric surgery

In 1891, Chiari described three types of hindbrain herniation involving the cerebellum and brainstem, of which the Chiari Type I malformation is the most common. He defined this condition as a herniation of the cerebellar tonsils through the foramen magnum into the upper spinal canal causing brainstem and cervical spinal cord compression, obstruction of cerebrospinal fluid flow, and eventually hydromyelia. Its symptoms and signs depend on the age of onset. Children usually present with oropharyngeal dysfunction (for example, aspiration, regurgitation, and dysphagia) followed by head and neck pain and gait disturbances. For adults, suboccipital pain exacerbated by valsalva is the most common presentation. These symptoms, in association with MR imaging findings of tonsillar herniation at least 5 mm below the foramen magnum, are consistent with the diagnosis of Chiari I malformation.

Nystagmus, particularly downbeat nystagmus, is the most frequent neuroophthalmic sign in children, whereas skew deviation, sixth nerve palsy, ocular dysmetria, ocular flutter, anisocoria, spasm of the near reflex, and esotropia are rare.

Esotropia, the inward turning of one or both eyes, is a common ophthalmic finding in children, occurring in 1 to 2% of the general population. Of the various types of esotropia, AACE is an unusual, but well-recognized, presentation of strabismus (misalignment of the eyes) in an older child or adult. Acute acquired comitant esotropia is described as esotropia in which the following factors are present. 1) The same angle of inward turning is maintained in all directions of gaze (that is, comitant, to differentiate it from deviations in which the angle of deviation varies with the direction of gaze, such as with sixth nerve palsies, which are incomitant). 2) The strabismus occurs after a period without esotropia (that is, as distinct from infantile esotropia, which is present at or soon after birth). 3) It occurs with abrupt onset (that is, it is deemed acute, to distinguish it from a slow-onset [subacute or chronic] esotropia).

The most common cause of AACE in more than 99% of cases is a primary ocular weakness that may require corrective surgery of the eye muscles; intracranial pathological conditions are rarely identified or considered. Acute ac-
quired comitant esotropia as the sole presentation of Chiari I malformation is especially unusual. Indeed, only four such cases have been reported, all of which have appeared in the ophthalmology literature. Here we report a fifth case, which we believe is the first one to be reported in the neurosurgical literature. Despite the inclination to treat AACE by local, extraocular muscle surgery, we report that neurosurgical treatment of the Chiari I malformation in this patient resulted in resolution of the AACE. We review the literature and highlight the features of this unusual presentation of Chiari I malformation to increase neurosurgical awareness of the potential for correcting AACE by direct treatment of the malformation, rather than by ocular muscle surgery.

Case Report

History. This 5-year-old boy presented with a 6-month history of esotropia, mainly involving the right eye but also the left. It was intermittent and subtle at first, but gradually became constant over several months. There was no apparent diplopia and no other neurological problems were reported. He had experienced neck pain approximately four or five times a year for 2 years prior to presentation and he had a history of metopic ridging in infancy that resolved without intervention as he grew older.

Examination and Proposed Treatment. The child was examined by a pediatric ophthalmologist who detected mild hypermetropia (far sightedness) and an 18–prism diopter (moderate-sized) comitant esotropia that significantly decreased with near gaze. When the eyes were aligned with prisms, there was no binocular fusion. Interestingly, as part of a routine evaluation, the child was examined with the same tests of binocular fusion approximately 6 months before the onset of esotropia (1 year prior to this ophthalmological testing), and was found to have excellent binocularity. The neuroophthalmological and general neurological examinations were otherwise normal. In particular, there was no nystagmus, and the extraocular movements were full without evidence of sixth nerve palsy.

Because he had mild hypermetropia, corrective lenses were prescribed to rule out an accommodative esotropia. The lenses did not correct the problem. The diagnosis of idiopathic esotropia was made and strabismus surgery involving medial rectus recession was recommended. Because the child’s father is a neurosurgeon, however, an MR image was obtained before this scheduled ocular surgery. This study demonstrated a severe Chiari I malformation without other intracranial or osseous abnormalities (Fig. 1). The T₂-weighted MR images demonstrated herniation of the tonsils to the level of C-3 with associated brainstem and upper cervical spinal cord edema and possibly early hydromyelia. There was no hydrocephalus.

Operation. Posterior fossa decompression was recommended because the malformation was extensive and there was edema and possibly early hydromyelia in the brainstem and upper cervical spinal cord. A decompressive suboccipital craniectomy, C1–2 laminectomies, and a patulous duraplasty were performed. Intraoperatively, there were several tight arachnoidal adhesions binding down the elongated cerebellar tonsils. The compression caused by these adhesions did not lessen until the bands of arachnoid were divided. There were no unexpected intraoperative surgical events, and the patient awoke neurologically intact.

Postoperative Course. By the 2nd postoperative day, an improvement in the esotropia was detected by a bedside examination. The postoperative course was unremarkable, and the child was discharged on the 3rd postoperative day.
The suspected improvement in the esotropia was confirmed 2 weeks after surgery when the examining ophthalmologist detected no unprovoked ocular deviation. There was still a residual 5–prism diopter intermittent esotropia (very mild) that was evident only during the cover–uncover test; recovery of binocular fusion was not evident. A follow-up ophthalmic examination 7 months following surgery did not identify any abnormality: there was complete recovery of binocular fusion and no evidence of esotropia (Table 1). An MR imaging study performed 6 months following surgery demonstrated excellent decompression of the brainstem and upper cervical spinal cord and reconstitution of the cerebrospinal fluid space posteriorly (Fig. 2). One year following surgery the child remained well, without clinical evidence of esotropia or other neurological problems.

**Discussion**

It is unusual for AACE to be the only neurological symptom of Chiari I malformation. Acute acquired comitant esotropia typically presents in the older child or adult. Table 2 provides the definition and pathophysiological findings of each type of AACE. The main diagnostic difficulty is in differentiating Type II (idiopathic type) from the neurological types of esotropia. It is usually not difficult to differentiate Type I from Type III or from the accommodative type by determining whether there is a history of ocular occlusion (Type I), occurrence in adult patients with myopia (near sightedness; Type III), and the presence of significant hypermetropia (accommodative type). It is important to recognize accommodative esotropia because this is easily and effectively treated with corrective lenses in most cases.

Although it has been stated that a truly comitant esotropia is an indication of an extracerebral process, comitant esotropias have been reported in conjunction with brain neoplasms, hydrocephalus, and Chiari I malformation. Several features of our case suggested that an intracranial abnormality might have caused the esotropia. The first was the inability to achieve binocular fusion when the eyes were aligned with prisms preoperatively. This sign has been suggested to indicate intracranial disease, although the validity of this correlation has been questioned. This sign may actually be more suggestive of a compensated eye movement disorder, thus...
to perform a careful physical examination to search for a careful history, including a developmental history, and that children with AACE require the clinician to obtain information. In any case, these features of our patient indicate ever, indicating that several different pathophysiological features of metopic synostosis, although 10 to 25% of patients with compared with 9% of their total patient population. These authors postulated that ridging is within the spectrum of common finding in patients with posterior fossa disease and Chiari I malformation. In our case, however, this symptom occurred so rarely as to be recalled only after an MR imaging study revealed the malformation. Last, the presence of metopic ridging was also a potential clue.

In fact, Tubbs, et al.,37 identified 50 cases of metopic ridging, 30% of which involved Chiari I malformation, compared with 9% of their total patient population. These authors postulated that ridging is within the spectrum of metopic synostosis, although 10 to 25% of patients with metopic ridging have no evidence of synostosis.14,21 It may be that synostosis results in a small anterior fossa, resulting in a compensatory shift of the brain into a normal-sized posterior fossa, further resulting in secondary crowding of the posterior fossa.37 This progression does not seem to be true in most cases of Chiari I malformation in which the posterior fossa is actually smaller than normal.4,6,27,31,33 However, indicating that several different pathophysiological conditions may exist with the end result being Chiari I malformation. In any case, these features of our patient indicate that children with AACE require the clinician to obtain a careful history, including a developmental history, and to perform a careful physical examination to search for any evidence of neurological dysfunction or cranio cervical pain. Any abnormality should prompt brain imaging tests to search for Chiari I malformation.

Fourteen cases of Chiari I malformation associated with comitant esotropias have been reported in the literature (Table 3). Our patient is similar to only four patients in these reported cases because he had no other neurological findings, such as nystagmus, to indicate an intracranial disease process. Lewis, et al.,26 have reported five cases, the largest series in the literature, of AACE in association with Chiari I malformation. All of their patients (age range 17–36 years) had nystagmus, however, with one having classic downbeat nystagmus. Some patients also had other neurological signs, although details of the associated neurological findings were not described. All patients had Chiari I malformation as confirmed on MR imaging. Four patients underwent posterior fossa decompression and the fifth patient refused any treatment. All four of these surgical patients had resolution of their diplopia and reduced nystagmus. These authors also suggested that abducting saccadic velocity could be used to rule out a subtle bilateral sixth nerve palsy, thus providing another potential objective method by which to narrow the differential diagnosis.

Biousse, et al.,8 reported on four patients with Chiari I malformation and esotropia. Three of these cases are similar to the case reported here because the esotropia was the only neurological sign; one patient had mild nystagmus. These patients ranged in age from 5 to 37 years old. Two of the four patients underwent suboccipital decompression with immediate improvement in diplopia: complete resolution of symptoms by 3 months postoperatively in one patient and mild residual symptoms in the other. The other two patients underwent strabismus surgery because of complex cranio cervical junction osseous abnormalities in one and parent preference in the other. Esotropia recurred 1 year following strabismus surgery in the patient with cranio cervical junction abnormalities; the procedure was repeated successfully with a maintained good result 3 months later. A second patient, an 8-year-old boy whose parents opted for strabismus surgery rather than posterior fossa decompression, also had a good result, with straight eyes 2 months following surgery. These authors recommended that the choice between strabismus surgery and posterior fossa decompression be made on an individual basis. They

### Table 2

<table>
<thead>
<tr>
<th>Type*</th>
<th>Features of Esotropia</th>
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<tr>
<td>I (Swan)</td>
<td>follows monocular occlusion or artificial disruption of fusion</td>
</tr>
<tr>
<td>II (idiopathic)</td>
<td>diploria, large deviation, no muscle paresis, normal binocular fusion, intermittent at first then constant, mild hypermetropia</td>
</tr>
<tr>
<td>III (Bielschowsky)</td>
<td>adult, associated w/ variable amounts of myopia</td>
</tr>
<tr>
<td>accommodative (refractive)</td>
<td>majority present when the patient is ~ 2–3 yrs old, may be intermittent or variable at initial onset, hypermetropia, improves or resolves w/ correction of refractive error</td>
</tr>
<tr>
<td>neurological</td>
<td>associated w/ other neurological findings or symptoms, inability to reestablish binocular fusion in some cases</td>
</tr>
</tbody>
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* The breakdown of types of AACE are based on papers by Burian and Miller, Lewis, et al., and Simon, et al.

### Table 3

**Literature review of AACE in association with Chiari I malformation***

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>No. of Patients</th>
<th>Contributing Neurological Factor</th>
<th>Initial Strabismus Surgery</th>
<th>PFD</th>
</tr>
</thead>
<tbody>
<tr>
<td>Passo, et al., 1984</td>
<td>1</td>
<td>square wave jerks</td>
<td>recur after 1 yr</td>
<td>PFD</td>
</tr>
<tr>
<td>Bixenman &amp; Laguna, 1987</td>
<td>1</td>
<td>nystagmus</td>
<td>recur after 3 yrs</td>
<td>PFD</td>
</tr>
<tr>
<td>Lewis, et al., 1996</td>
<td>5</td>
<td>5 nystagmus w/ or w/o ataxia</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>Weeks &amp; Hamed, 1999</td>
<td>2</td>
<td>1 w/ nystagmus; 1 w/o</td>
<td>2 recur (3 mos; 2 yrs)</td>
<td>PFD</td>
</tr>
<tr>
<td>Biousse, et al., 2000</td>
<td>4</td>
<td>1 w/ nystagmus; 3 w/o</td>
<td>1 recur after 1 yr; 1 no eso</td>
<td>PFD</td>
</tr>
</tbody>
</table>

* Eso = esotropia; NA = not applicable; PFD = posterior fossa decompression; recur = recurrence.

† One patient refused posterior fossa decompression.
suggested that strabismus surgery would be a preference in certain cases, such as in the setting of complicated associated osseous abnormalities.

Weeks, et al.,39 reported on two patients with AACE and Chiari I malformation, one of whom also had nystagmus. Both patients initially underwent strabismus surgery and had experienced recurrent esotropia following surgery: 3 months later in one and 2 years later in the other. Subsequently, these two patients underwent posterior fossa decompression with excellent results and complete resolution of the esotropias. These authors recommended decompression rather than strabismus surgery as the first-line treatment in patients with esotropia and Chiari I malformation. Nevertheless, as they point out, the incidence of undiagnosed malformation and successful strabismus surgery is unknown.

Bixenman and Laguna9 reported one case of AACE and Chiari I malformation in a 13-year-old girl who presented with nystagmus and headache. This patient initially underwent strabismus surgery, and it was not until 3 years later that the diagnosis of Chiari I malformation was made, when she began to experience worsening nystagmus, esotropia, and ataxia. She then underwent posterior fossa decompression and attained normal ocular alignment.

Passo, et al.,34 reported one confirmed case of Chiari I malformation and AACE. This patient, a 24-year-old woman, presented with square-wave jerks as well. She underwent strabismus surgery for a diagnosis of “decompensated esotropia;” cranial imaging was not performed at that time. The diplopia returned 1 year later and was associated with ataxia. At this time, an MR imaging study was performed and revealed Chiari I malformation; subsequently posterior fossa decompression was performed, with resolution of her esotropia. This outcome also supports decompressive surgery rather than strabismus surgery as a first-line therapy for certain patients with esotropia associated with Chiari I malformation.

The pathophysiological mechanisms underlying esotropia in Chiari I malformation are uncertain. Previously, it was thought that hydrocephalus was the underlying cause of the eye-movement disorder;13,19,34 however, it is clear that most patients with Chiari I malformation, particularly in the pediatric population, do not have hydrocephalus, as was the case with our patient.7,15,16,28,29,35 It has been argued that early, mild, bilateral sixth nerve palsies may present in a similar fashion.22 Lewis and colleagues26 measured abducting saccadic velocities in their patients with AACE and Chiari I malformation to rule out subtle bilateral sixth nerve palsies as the cause for the esotropia; the velocities were found to be normal in all four cases. Sixth nerve palsy is an unlikely cause of esotropia in our case because fusion was maintained at near gaze and because the esotropia was noted in all directions of gaze with AACE and not just in lateral gaze, as in sixth nerve palsy.9 A potential explanation for the esotropia seen in Chiari I malformation is dysfunction of the vergence mechanisms in the mesencephalon for patients with extensive hydromyelia/syringomyelia or edema extending into the midbrain.9,25 This theory is supported by primate models in which independent vergence mechanisms have been identified in the midbrain.23 This mechanism, however, does not explain the esotropia in the majority of patients with Chiari I malformation who do not have midbrain involvement. For them, dysfunction of more caudal structures is a necessary explanation, although these vergence structures have not been identified. The rapid improvement in the esotropia following decompression in our patient supports a direct compressive mechanism and the more gradual improvement over a period of weeks supports an edema-associated pathophysiological condition.

Conclusions

On the basis of our one case and the cases in the literature, we recommend that strong consideration be given to posterior fossa decompression, rather than strabismus surgery, as the first-line therapy in patients who have AACE in association with Chiari I malformation, if they are otherwise good surgical candidates. Strabismus surgery treats the symptoms caused by brainstem compression and edema but does not treat the cause itself. Analysis of the available literature tells us that patients with AACE who undergo posterior fossa decompression can expect an immediate beneficial effect with respect to the esotropia, followed by a more gradual improvement and resulting in complete or near-complete resolution of the eye-movement disorder. In our patient, intradural dissection was clearly necessary to divide compressive arachnoidal bands, and we prefer to open the dura mater and perform a duraplasty in all cases. Nevertheless, we cannot prove that division of these bands was a necessary maneuver in the overall treatment of the Chiari I malformation. We believe that neurosurgeons should be aware of this rare presentation of Chiari I malformation so that when referred such a patient, appropriate management decisions can follow.

References


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