Tethered hindbrain

Case report

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The authors present the case of an 11-year-old boy with symptomatic tethering of the hindbrain related to a Chiari Type III malformation repaired at birth. Surgical release of the cord resulted in functional improvement.

KEY WORDS • cerebellum • encephalocele • hindbrain • medulla oblongata • meningocele • tethering

T he tethered spinal cord syndrome is a well-described entity that most commonly involves the lumbosacral portions of the neuraxis. In a few cases, the upper thoracic cord has been affected; the cervical cord is tethered in an even smaller number. Previously reported cases of tethering of the rostral spinal cord have been associated with trauma, split cord malformation, myelomeningocele, or encephalocele. Review of the literature reveals only two examples of tethering rostral to the spinal cord. In the first case, symptomatic tethering of the cerebellar vermis developed in a 25-year-old man who had undergone limited repair of an encephalocele at 8 days of age. Exploration revealed a fibrous stalk tightly attaching the vermis to the overlying dura, and after sectioning of this band, the patient’s symptoms improved. In a very recent report by Liu, et al., the cervicomedullary junction was tethered in a 5-year-old child with a Dandy–Walker cyst treated by cystoperitoneal shunting. Removal of the shunt, fenestration of the cyst membrane, and untethering of the medulla relieved the patient’s headache and cranial nerve deficits. We report a second example of tethering at the cervicomедullary junction in a child with a previously surgically treated craniocervical meningoencephalocele involving the cervicomедullary junction and the cerebellum, which was hypoplastic, a lesion sometimes referred to as the Chiari Type III malformation. Our case and the case of Liu, et al., suggest a correlation between neuroimaging and the clinical phenomena of hindbrain tethering.

Case Report

History. The patient was born with a saclike lesion protruding from the posterior aspect of the craniocervical junction. His mother recalls that the skin over the sac had an abnormal coloration. The patient underwent excision of the sac on the 1st day of life, but further details of his early investigation and treatment are unavailable.

The development of hydrocephalus at 1 month of age required cerebrospinal fluid (CSF) shunting, and the occurrence of a shunt infection 2 weeks later necessitated revision of the shunt. Subsequently the patient’s neurosurgical course stabilized, and until the age of 10 years his developmental progress had been considered satisfactory. His speech was nearly appropriate for his age, he walked with braces, although he preferred a walker or a wheelchair for prolonged distances, and he was able to dress independently and to feed himself.

Between the ages of 10 and 11 years, however, a gradual loss of these basic skills occurred, prompting the boy’s mother to seek evaluation. At his presentation, he was unable to crawl or to use his wheelchair, and in addition, he had great difficulty maintaining a sitting position. His mother also described progressive incontinence, as well as increasing gagging and difficulty speaking. He did not have headaches or neck pain.

Examination. General physical examination was remarkable only for small stature and torticollis; the head was held to the right and tilted downward. There was prolonged horizontal nystagmus in both directions. A right esotropia and tongue fasciculations, with left greater than right, were present. All four extremities were hypotonic, and sensation was otherwise normal. The superficial abdominal reflexes were absent; deep tendon reflexes were absent in the upper extremities and brisk in the lower, with ankle clonus and Babinski reflexes bilaterally.

Imaging Studies. Computerized tomography of the head confirmed satisfactory CSF shunt function. Magnetic res-
onance (MR) imaging of the spine demonstrated marked tethering of the hindbrain with dorsal displacement of the medulla, which appears adherent to the dura. The cervicomedullary junction is also caudally displaced.

Operation. Because of the marked neurological decline and the examination consistent with dysfunction at the level of the tethering, the patient underwent exploration for untethering. A laminectomy was performed at the level below the lesion, and normal dura was opened at this level. As the dural opening was extended superiorly, a fibrous epidural cicatrix was found confluent with the dorsal surface of the cervicomedullary junction. Sharp dissection separated the neural tissue from the fibrous scar. Superiorly there was a veil of thick, opaque arachnoid obscuring the medulla that was opened sharply as well to expose the hypoplastic cerebellum. The lower cranial nerves were seen ascending at a steep angle to enter the skull. Further lateral dissection through thickened arachnoid completely mobilized the hindbrain, allowing it to relax into a large ventral CSF cistern. At this point the hindbrain became pulsatile. The dura was reconstructed with a large graft of pericranium.

Postoperative Course. His mother noted postoperative improvement in his use of the left upper extremity and in his ability to sit, so that he was able to use his wheelchair again. He began crawling, and his speech returned to baseline. Two years later, however, he has not regained the ability to walk, and he still requires assistance with dressing and eating. Follow-up MR imaging has shown ventral relaxation of the hindbrain without any suggestion of recurrent tethering (Fig. 2).

Discussion

In this patient, progressive neurological deterioration correlating with signs on neuroimaging prompted us to make the diagnosis of hindbrain tethering. Surgical release of this tethering led to functional and imaging improvement. However, the cause of the tethering remains obscure. As previously mentioned, tethering of the rostral neuraxis is often associated with myelomeningocele (and in one patient, encephalocele.) Technically speaking, the Chiari Type III malformation is a different entity, defined as “caudal displacement of the cerebellum and brainstem into a high cervical meningocele.” However, because the herniation of neural tissue into a meningocele is common to all of these anomalies, a similar pathogenetic mechanism may be responsible.

The formation of postoperative adhesions at the site of the original meningocele repair is one possible explanation. However, surgery in this region is not uncommon; many patients have undergone decompression of Chiari I or II malformations or excision of tumors in this region. Although these patients also develop postoperative adhesions, symptomatic tethering has never been reported among them.

To explain the development of symptoms from tethering of the more caudal neuraxis, several authors have blamed the traction that results from repetitive flexion and extension of the spine, and they propose that the resultant mechanical distortion leads to ischemia. Evidence that tethering interferes with oxidative metabolism certainly supports such an explanation. This traction theory can be easily applied to explain symptomatic tethering of the cervical cord, as repeated neck flexion is known to cause stretching of the cervical spinal cord; the traction is greater in the lower cervical region and around an area of tethering. The degree to which neck movement induces stretching of the medulla is less well defined. However, a Chiari malformation was present in our patient, and the resultant caudal displacement of the hindbrain into the

Fig. 1. Midline preoperative sagittal T₁-weighted magnetic resonance image of the cervical spine, showing hindbrain tethering. There is extreme posterior displacement and angulation of the medulla, which appears adherent to the dura. The cervicomedullary junction is also caudally displaced.

Fig. 2. Postoperative magnetic resonance image at the same level as seen in Fig. 1. Although this image had been obtained after administration of gadolinium, the precontrast study was identical. The ventral relaxation of the hindbrain is evident. A small piece of tissue is present immediately superior to the point of previous tethering; this area represents a portion of dysplastic cerebellum.
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neck may have subjected the medulla to repetitive mechanical distortion with neck movement.

Our case shares with the case of Liu, et al.,\textsuperscript{8} two dramatic neuroimaging signs: 1) dorsal displacement of the cervicomedullary junction; and 2) elongation and attenuation of the medulla. In both cases postoperative clinical improvement correlated with resolution of the neuroimaging abnormalities.

Tethering of neural elements is not isolated to the spinal cord. Although rare, it may occur at the brainstem level. In the primary repair of craniocervical meningoceles and encephaloceles, some consideration must be given to the prevention of secondary tethering; congenital tethering lesions must be sought and taken down, and the dura must be reconstructed in a capacious fashion. In the setting of previous intradural surgery at the craniocervical junction, if progressive clinical deterioration correlates anatomically with findings on MR imaging and particularly if MR imaging demonstrates dorsal displacement of the cervicomедullary junction and elongation of the medulla, then operative exploration for untethering may be indicated.

References


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