Headache Surgery

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"Chiari I malformation is a hindbrain abnormality consisting of cerebellar tonsillar herniation on MRI >5mm causing impaired CSF circulation. While congenital it can present both in children and adults."

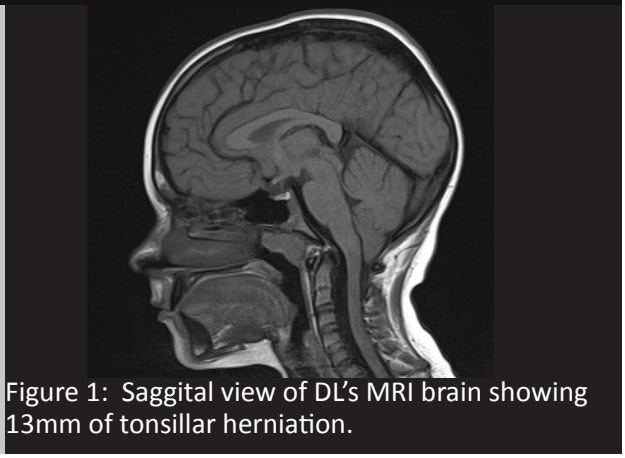

Figure 1: Saggital view of DL's MRI brain showing 13mm of tonsillar herniation.

Case Presentation

DP is a 9-year old school boy who was referred to the outpatient neurosurgery services at St Louis Children's Hospital with a 5-year history of recurrent headaches and "zoning-out" episodes. These grew more severe and frequent over the last 4 months and were associated with physical activity. The headaches were on the top of the head and were only relieved by lying down and resting. They were associated with loss of ability to retain information and focus at school and his results over the last 4 months have gradually fallen from straight As to Ds and Fs. Over the last month there have been associated episodes of a tingling sensation in the right hand as well as blurry vision and episodic mild abdominal pain. There was no difficulty swallowing, no choking or gagging, no apnoea or weakness, no bowel or bladder dysfunction. He was taking ibuprofen and acetaminophen PRN to help relieve the pain.

DP has a history of frequent ear and sinus infections for which he had bilateral myringotomy tubes inserted. He also has a history of sleep apnoea as a baby, which never required treatment, and has had routine dental work under anaesthesia. There is a family history of seizure disorders as well as type I and type II diabetes mellitus. There is no other history of congenital abnormalities or cancer.

The patient lives at home with his parents and four siblings and is well adapted in school. He has no known allergies and is on no other medications.

On physical exam, DP was well nourished and in no acute distress, pleasant and cooperative. He had full range of motion in the cervical spine, moving upper and lower extremities equally and symmetrically. He had normal tone and bulk, and 2+ deep tendon reflexes. Fine touch sensation was intact throughout and there was no clonus present. A cranial nerve exam showed no problems with hearing or smelling, intact extraocular movements, symmetric face, no tongue deviation and pupils were equal and reactive to light.

Investigations and Diagnosis

The patient underwent an MRI brain, which showed 13mm of tonsillar descent with no evidence of a syrinx in the upper cervical cord. His tonsils appeared somewhat peg-like in shape. The MRI brain was otherwise normal. He also underwent an MRI of the cervical, thoracic and lumbar spine without contrast and a CSF flow study. The spine MRI showed no evidence of a syrinx (hyperintense collection of fluid within the spinal cord). The CSF flow study demonstrated that there was no CSF flow along the posterior craniocervical junction with abnormal movement of the cerebellum and brainstem. There was normal CSF flow along the anterior craniocervical junction. These history and imaging findings are consistent with a Chiari Type I malformation and the patient and family were offered a bone-only posterior fossa decompression. After discussing the benefits and risks of this procedure and the expected hospital course, the parents agreed to proceed with treatment.

Treatment and follow-up

The patient was then admitted to St Louis Children's Hospital and sent to theatre for a bone-only posterior fossa decompression. After general anesthesia work-up and intubation and under anesthesia, the patient was placed in a prone position in the operating table with the head positioned on a padded Mayfield horseshoe. The occipital region was then clipped of hair near the hairline and a vertical incision was marked from below the inion down to the palpated C2 spinous process. The region was prepped and draped in the usual sterile fashion and a 15 blade scalpel was used to make the skin incision. Bipolar cautery and Bovie electrocautery were used for hemostasis and for the dissection

through the midline nuchal ligament. At this stage, the occipital bone and foramen magnum were exposed in a subperiosteal fashion. The C1 arch was also exposed centrally and then a self-retaining cerebellar retractor was placed. A high-speed pneumatic drill was used to place two burr holes above the foramen magnum, which were connected using a Kerrison punch and a Leksell rongeur, with special attention not to damage the underlying dura. The foramen magnum, 2cm of occipital bone and the arch of C1 were also removed using a Kerrison punch. After removal of these bony elements, an intra-operative ultrasound assessment of the cerebellar tonsils was made showing scant subarachnoid fluid space posterior to the tonsils and reduced tonsillar motility. The epidural band was then dissected vertically with a curette allowing for relaxation of the craniocervical dura. Another ultrasound assessment showed increased cerebrospinal fluid space dorsal to the tonsils with improved tonsillar motility after the decompression. Bipolar cautery was used for hemostasis and irrigation was placed in the wound, which was closed using interrupted 3-0 Vicryls in the muscle and fascia and interrupted 4-0 Vicryls in the dermis. A layer of Dermabond was then applied to the skin edges. No acute complications were recognised. The patient had a good recovery, was afebrile post-op and had a mild episode of nausea and vomiting. He was discharged at day 1 post-op and is due to follow-up at 2 weeks post-op.

Discussion

Chiari malformations are hindbrain abnormalities ranging from simple caudal displacement of the cerebellar tonsils into the upper cervical spinal canal (Type I) to cerebellar hypoplasia or aplasia (Type IV)[1]. Type I is the most common, with a study of 22,591 patients who underwent MRI brain showing that 0.775% had tonsillar herniation over 5mm^2, which is often considered as a good borderline limit for identifying Chiari Type I malformations. Its diagnosis is partly one of exclusion since there needs to be no intracranial mass lesion, Dandy-Walker malformation or other causes of hydrocephalus that would explain the tonsillar herniation present. However, it is often associated with other lesions such as syringomyelia and hydrocephalus.

It presents with a variety of symptoms including headaches, usually located in the occipital or upper cervical regions. These are often brought on by neck extension or Valsalva manoeuvre. Other symptoms include weakness or numbness, loss of temperature sensation, and unsteadiness. Often there are associated opthalmologic or otologic symptoms such as blurry vision, nystagmus, extraocular muscle palsies, diplopia, visual field defects, tinnitus, fluctuating hearing loss, vertigo and nausea. Common signs include weakness, hyperreflexia, ataxia and lower cranial nerve dysfunction. This, however, does not mean that there are no asymptomatic cases of Chiari Type I, and while a Chiari Type I malformation present on MRI is probably the cause of these symptoms if no other conditions are present, this is not necessarily the case, and many times there are associated conditions of a medical, surgical or psychiatric nature associated with or partially responsible for the symptoms present[1,3,4].

Hence, while the only treatment available is surgical – posterior fossa

Clinical Points

• It can be asymptomatic or present with recurrent headaches, particularly in the suboccipital region and brought on by neck extension or Valsalva manoeuvre, and focal neurological deficits – especially lower cranial nerve and cerebellar signs and symptoms.

• Differential diagnosis for subacute or chronic recurrent headaches includes any congenital or acquired cause of raised ICP as well as encephalitis, giant cell arteritis, tension headache, migraine, cluster headache, cervical root myelopathy, eye strain headaches or drug-induced headaches.

• Treatment is expectant or surgical – posterior fossa decompression. Indications for surgery include a clear history of symptoms with no other cause found and tonsillar herniation on MRI imaging. However, the criteria for when and how to operate remains controversial.

• Posterior fossa decompression may be bone-only (removing part of the occipital bone and posterior arch of C1) or include incision of the dura with addition of a dural patch as well as extrapial coagulation of one or both of the tonsillar tips.

decompression – the decision on when and how to treat still remains controversial, especially when there are no associated complications such as spinal cord dysfunction or scoliosis due to syringomyelia or focal neurological abnormalities. In this case, the decision to operate was based on a clear and clinically typical history of headaches with no other identified cause, as well as the presence of neurologic and ophthalmologic symptoms – tingling sensation in the right hand and blurry vision. Moreover, 13mm of tonsillar herniation on MRI is a clear value leaving no ambiguity on the diagnosis, while a value of 3–7mm is often considered a grey area as regards the presence of Chiari Type I. Yet the absence of signs and the chronic, rather than acute, history of headaches led to the decision to opt for a bone-only decompression rather than an intradural approach[5-8]. This approach would involve an additional midline incision on the dura, plus the visualization of veils that may be covering the outlets of the 4th ventricle, as well as possibly the visualization of choroid plexus of the 4th ventricle to inspect for the need for extrapial coagulation of one or both of the tonsillar tips in order to restore cerebrospinal fluid flow. An insertion of a dural graft is also sometimes considered due to the presumed protective effect on the neural tissues from chemical contamination, pericranium being a common choice of autologous graft[9].

While in this case the use of intraoperative ultrasound was reassuring of the positive effect of the bone-only decompression on cerebrospinal fluid flow, it is often argued that an extradural approach increases the risk of a need to reoperate based on a lower rate of reduction or elimination of symptoms. However, it is also defended that this approach provides for shorter operative time, reduced blood loss and reduced length of hospital stay, and eliminates the risk of cerebrospinal fluid leak. This trade-off is usually solved by first offering a posterior fossa bone-only decompression and, if symptoms persist, then offering a posterior fossa decompression with duroplasty. While some still advocate an immediate intradural approach, especially in very severe cases (both symptomatically and on imaging), this is the approach recommended by several authors[5-7].

In summary, Chiari Type I malformation is an important differential diagnosis of medically unresponsive headache of subacute or chronic history both in the paediatric and adult populations10. Its diagnosis is made based on a complete history, examination and imaging. Treatment is either expectant or surgical. While some controversy remains as to the role of posterior fossa bone-only decompression, a growing number of neurosurgeons now consider it effective and safe enough to be the best surgical option for the treatment of the vast majority of symptomatic Chiari Type I malformations.

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