Neuroimaging Highlight

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A Treatable Mimic of Chiari Malformation with Syringomyelia

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A 19-year-old woman presented with a five month history of headaches. Initially the headaches had been intermittent. For the two months prior to presentation the patient had a constant headache. There was no precipitant at onset. The headache was improved by recumbancy. It was associated with daily vomiting resulting in a 20 pound weight loss but no photophobia or phonophobia. The neurological exam was normal. A computed tomogram scan demonstrated findings consistent with a Chiari malformation.

A magnetic resonance image (MRI) demonstrated cerebellar tonsillar herniation as well as other abnormalities suggestive of spontaneous intracranial hypotension (SIH) (Figure 1) rather than a Chiari malformation. The MRI of the cervical spine was normal.

The patient was initially treated conservatively with bed rest, fluids and caffeine.

A repeat MRI performed one month later demonstrated worsening of the cerebellar tonsillar herniation and new evidence of T2 signal hyperintensity within the cervical spine (Figure 2). There was no evidence of cerebrospinal fluid (CSF) leak.

She underwent a lumber epidural blood patch with improvement in her symptoms. Seven weeks later the headaches recurred. A second lumber epidural blood patch was performed resulting in complete resolution of symptoms. An MRI after clinical symptoms improved revealed decreased tonsillar herniation and resolution of the T2 cord signal abnormality (Figure 3).

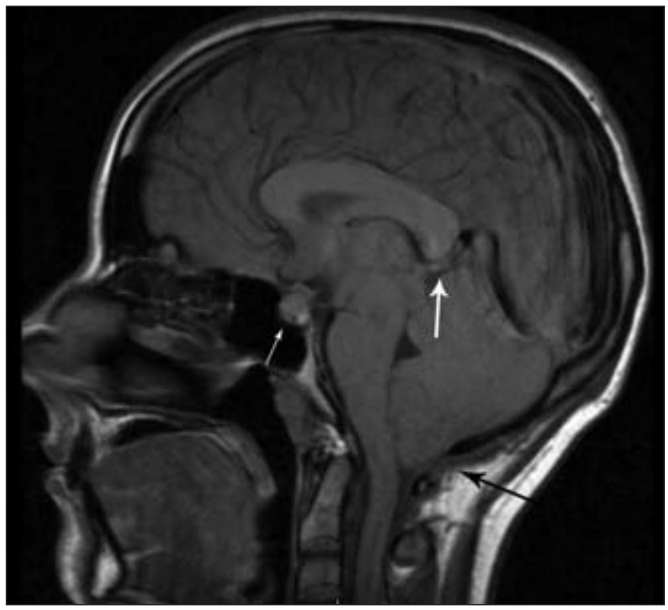

Figure 1: *T1-weighted sagittal MRI image demonstrating findings consistent with intracranial hypotension. There is tonsillar descent (black arrow), descent of the hypothalamus and midbrain structures, flattening of the pons against the clivus, enlargement of the pituitary gland (small arrow), and increased acuity of the angle between the vein of Galen and the internal cerebral veins (large white arrow).*

Discussion

Awareness of the imaging signs of SIH is essential to the diagnosis as tonsillar herniation alone could be misinterpreted as a Chiari malformation. Classic imaging findings include descent of the cerebellar tonsils, enhancement of the pachymeninges, flattening of the pons, pituitary hyperemia, subdural hygromas or engorgement of the superficial and deep venous system[1,2]. On axial imaging, there is crowding of the basal cisterns because of brainstem descent, resulting in crowding of the structures at the tentorial incisura. Brainstem descent also results in a decreased angle between the internal cerebral veins and vein of Galen due to anchoring of the veins at the tentorial incisura[3].

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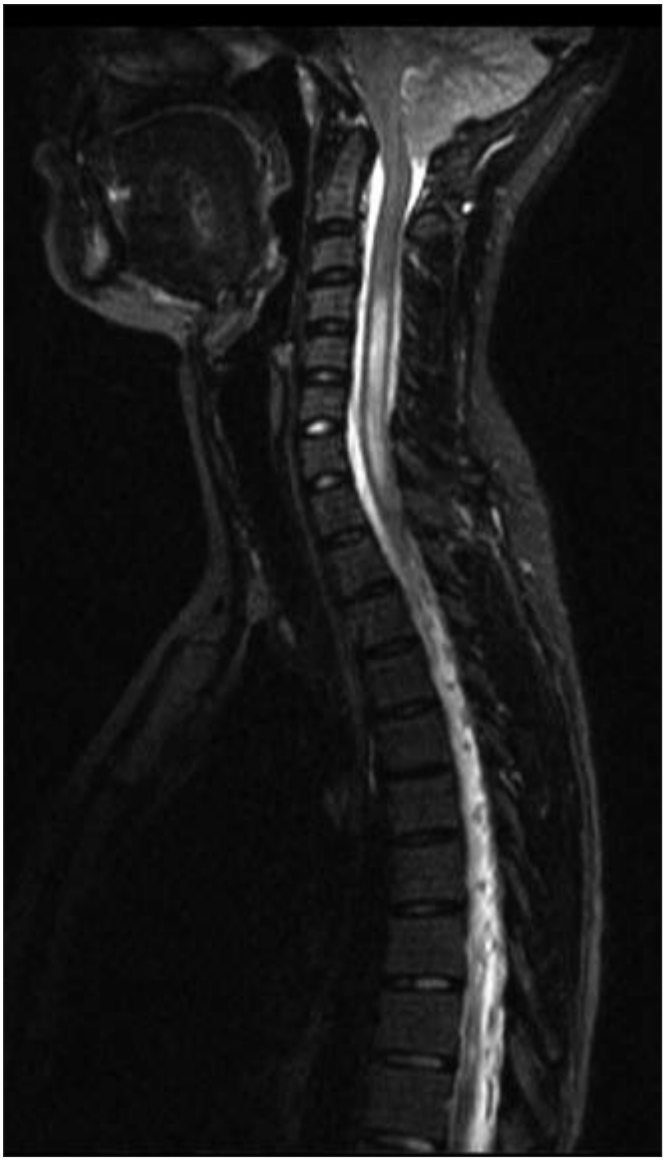

Figure 2: *STIR (short tau inversion recovery) sagittal MRI image demonstrating cerebellar tonsillar herniation and intramedullary hyperintense signal from C3-T1.*

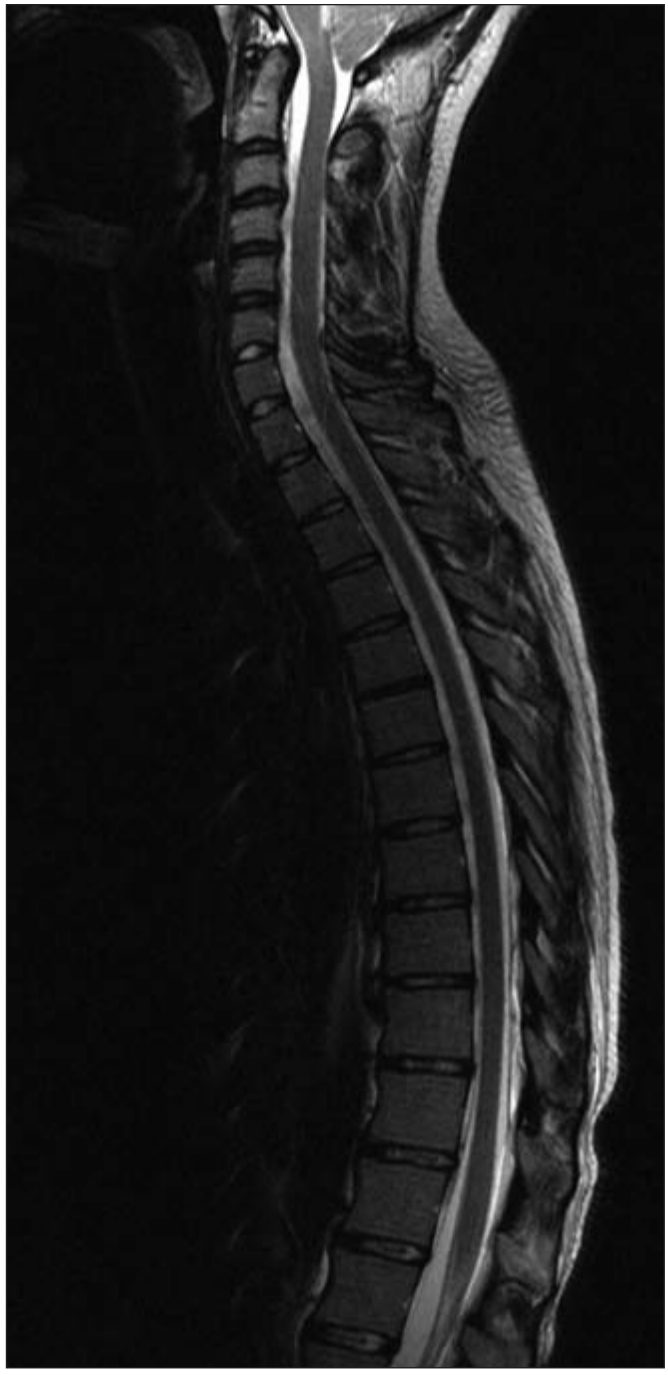

Figure 3: *T2-weighted sagittal MRI image demonstrating improvement in the cerebellar tonsillar herniation and resolution of the intramedullary hyperintense signal.*

Syringomyelia has been previously reported in patients with intracranial hypotension due to lumboperitoneal shunts[4] and due to a post-traumatic fistula between the C8 nerve root and the pleura[5]. Resolution of the cord signal abnormality was achieved in two cases when the lumboperitoneal shunt was changed to a ventriculoperitoneal shunt[4] and after surgical treatment of the nerve root fistula in one case[5]. Only one other case of SIH causing cerebellar tonsillar herniation and syringomyelia has been described[6]. The patient's symptoms, including focal neurological deficits, continued despite two months of conservative treatment. Due to evidence of an epidural fluid collection from the foramen magnum to the C-2 level she was treated with foramen magnum decompression and placement of Gelfoam in the posterior fossa to C-1. Although no dural tear was seen at surgery, she had marked improvement in her symptoms postoperatively.

In our case, the patient's symptoms and the spinal cord signal abnormality resolved with repeat lumbar epidural blood patches

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(EBP). Studies have demonstrated a 56% -77% response rate to ≤ 2 lumbar EBP in patients with SIH[7,8]. There was no significant difference in the rate of response comparing cervical or thoracic leaks to lumbar leaks[8]. Rabin et al report two cases of SIH associated with extradural spinal fluid collections at the cervicothoracic junction that resolved with thoracic or lumbar EBP[9].

This case demonstrates that signal abnormality within the cervical cord can develop acutely in SIH. Levine[10] proposes that cord edema, in the setting of a block at the foramen magnum, results from extravasation of plasma due to alteration of the CSF pressure relative to the venous pressure. With a block at the foramen magnum there is impaired CSF flow between the intracranial and intraspinal compartments. Normally, CSF flows from the intracranial to the intraspinal compartment 1) when intracranial pressure increases during the systolic phase of the cardiac cycle and 2) after periods of high intrathoracic pressure such as coughing. If there is a block at the foramen magnum, less CSF movement to the intraspinal compartment occurs during systole and after coughing. Therefore, CSF pressure caudal to the block is reduced. In the setting of a CSF leak, the CSF pressure is further reduced. The difference between the CSF pressure and the pressure in the epidural veins or capillaries determines the caliber of the vessels. Low CSF pressure leads to dilation of the veins and capillaries. Acute vascular dilation causes disruption of the blood-brain barrier and extravasation of fluid ensues. In our case, we propose this mechanism resulted in cord edema which resolved following treatment.

In conclusion, spinal cord hyperintense T2-signal on MRI suggesting edema can occur in SIH associated with cerebellar tonsillar herniation. This must be differentiated from a Chiari malformation associated with syringomyelia. If recognized early, the symptoms may resolve with non-surgical treatment such as repeated lumbar epidural blood patches, even in the absence of an identified source of CSF leak.

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