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Visual Diagnosis

Resolution of Chiari-Associated Syringomyelia

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A 12-year-old boy presented with months of worsening left forearm and hand pain, numbness, tingling, and weakness. There was no trauma, central nervous system infection, or scoliosis. Results of magnetic resonance imaging showed a significant Chiari I malformation with foramen magnum crowding and subarachnoid obstruction (Fig A,B). The cerebellar tonsils were triangular. A syrinx was present without tumor or tethered spinal cord.

He underwent suboccipital craniectomy, C1 laminectomy, and duraplasty. The large tonsils were

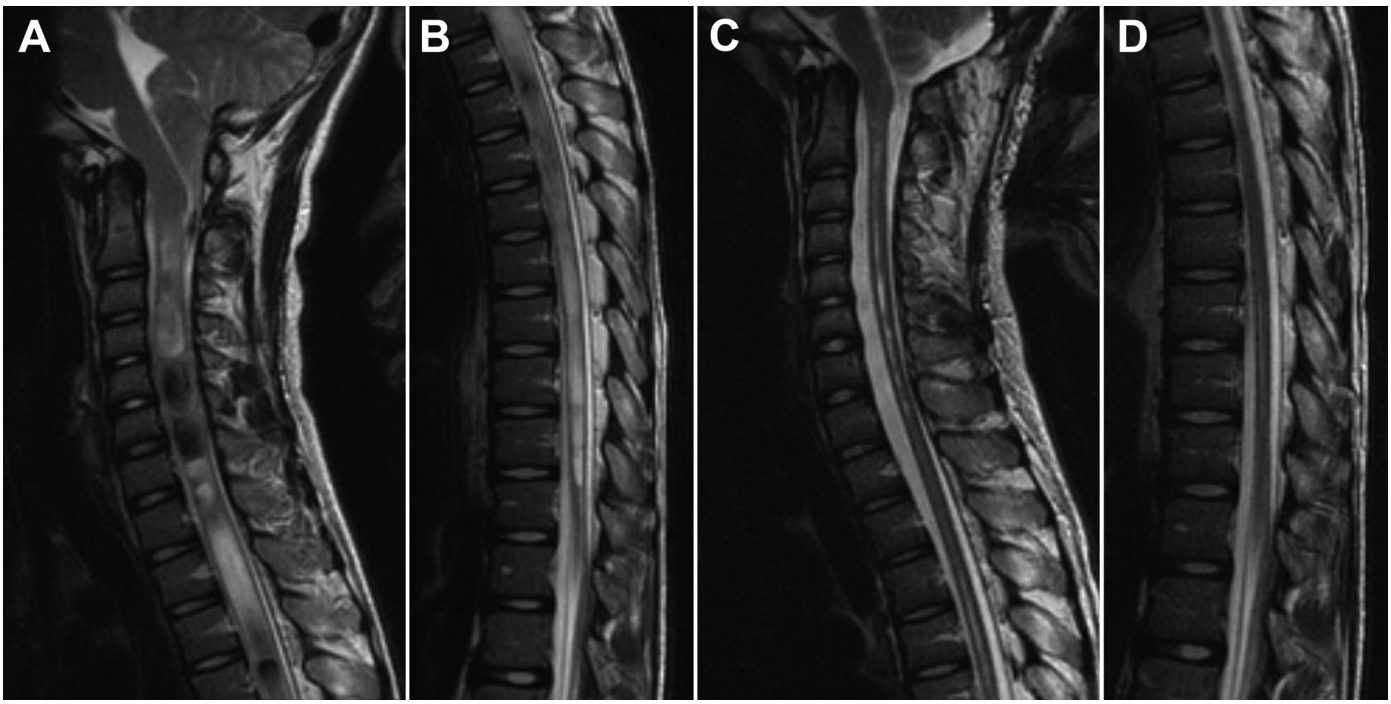

FIGURE.
Preoperative sagittal T2-weighted MRI (A and B) showing a severe Chiari I malformation with associated extensive syrinx. Postoperative sagittal T2 MRI (C and D) showing a resolved Chiari I malformation and collapsed syrinx. MRI, magnetic resonance imaging.

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coagulated, allowing decompression without C2 laminectomy (which increases spinal instability risk). Cerebrospinal fluid (CSF) flowed normally through Magendie after decompression.

Nine months postoperatively, his symptoms were much improved. Magnetic resonance imaging showed resolved foramen magnum compression and restoration of the subarachnoid space (Fig C,D). The tonsils were normal, and the syrinx collapsed.

Chiari I malformations have varied presentations, including tussive occipital headache, syringomyelia, dysphagia, and central sleep apnea.[1] Syringomyelia can diminish pain and temperature sensation from compression of central crossing spinothalamic fibers. Syringomyelia also can cause scoliosis, presumably from asymmetric weakness of paraspinal musculature. Other etiologies of syringomyelia include tethered spinal cord, spinal cord tumor, trauma, and central nervous system infection. The pathogenesis of the latter two is thought to be arachnoiditis causing CSF flow obstruction.

How a Chiari I malformation causes syringomyelia is debated.[2] Some believe compression of Magendie causes fourth-ventricle outflow obstruction, leading to a reopening of the spinal central canal as CSF attempts to find an outlet. Others believe pistoning of the tonsils during the cardiac cycle drives CSF into the spinal cord parenchyma through Virchow-Robin spaces. After decompression, the syrinx should collapse but usually does not completely disappear.

The ideal neurosurgical strategy for Chiari I is also debated. Some neurosurgeons prefer a purely osseous decompression with removal of the C1 lamina and suboccipit at the foramen magnum. It is important to extend the bony removal to the lateral aspect of the thecal sac. Others routinely open the dura and perform a duraplasty. Still others lyse the arachnoid bands at Magendie, coagulate the tonsils, and/or place a silastic stent between the fourth ventricle and cervical subarachnoid space.

CSF flow restoration from the fourth ventricle to the cervical subarachnoid space is paramount, so we recommend a duraplasty and arachnoid lysis. The tonsils are coagulated only if needed to allow decompression without C2 laminectomy, and stents are inserted only if the foramen of Magendie is very scarred and unlikely to remain patent. The best chance for a lasting cure is the first surgery. The goal is to ameliorate the presenting symptoms; a pleasing radiographic result is a bonus.

References

1. Khatwa U, Ramgopal S, Mylavarapu A, et al. MRI findings and sleep apnea in children with Chiari I malformation. *Pediatr Neurol*. 2013;48:299-307.
2. Heiss JD, Patronas N, DeVroom HL, et al. Elucidating the pathophysiology of syringomyelia. *J Neurosurg*. 1999;91:553-562.