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Resolution of syringohydromyelia with targeted CT-guided epidural blood patching

Case report

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In the scientific literature, syringohydromyelia has only rarely been reported in association with spontaneous intracranial hypotension. Management of the syringohydromyelia in these patients has heretofore involved relatively invasive surgical procedures. The authors report the first case of syringohydromyelia in the setting of intracranial hypotension successfully treated with CT-guided epidural blood patches. This case is important in that it represents a potential minimally invasive treatment strategy. Furthermore, the case also highlights the need to consider spontaneous intracranial hypotension when clinically appropriate as a cause of syrinx in patients with cerebellar tonsillar ectopia, in whom the lesion might otherwise be misclassified as a Chiari I malformation. Finally, the responses to the various attempted treatments offer insight into the pathophysiology of this syringohydromyelia, which may differ from classical models of syrinx formation. *(DOI: 10.3171/2011.3.JNS102164)*

Key Words • **intracranial hypotension** • **blood patch** • **syringohydromyelia**

In the scientific literature, syringohydromyelia has only rarely been reported in association with spontaneous intracranial hypotension. Management of the syringohydromyelia in these patients has heretofore involved relatively invasive surgical procedures. The authors report the first case of syringohydromyelia in the setting of intracranial hypotension successfully treated with CT-guided epidural blood patches.

Case Report

This 53-year-old woman presented to our institution with a history of progressive pain in the right chest and axillary region, with eventual migration of the pain into her right shoulder, arm, and fingers over a 5-month period. She also reported a history of chronic headache over several years, which had recently abated. She had undergone follow-up in the past for a suspected pituitary tumor, which was presumed to be the source of her headaches, given that several brain MR images demonstrated pituitary enlargement and avid enhancement. Spinal cord MR images available at presentation demonstrated extensive cervical and thoracic syringohydromyelia and cerebellar tonsillar ectopia (Fig. 1A and B). Abnormal smooth dural thickening and enhancement was also identified on prior brain MR imaging (Fig. 2A), as was sagging of the midline structures of the brain and pituitary hyperemia, resulting in effacement of the suprasellar cistern and mild mass effect on the optic chiasm. These imaging features are often seen in association with intracranial hypotension, but with the exception of the avid pituitary enhancement, they were not recognized prior to the patient's presentation at our institution.

A syringosubarachnoid shunt connecting the syrinx cavity to the subarachnoid space in the thoracic region was placed with the goal of decompressing the syrinx. Postoperatively, the patient developed worsening symptoms of arm pain and new symptoms of lower-extremity weakness, necessitating the use of a walker to assist with ambulation. Repeated MR imaging of the spine 4 months after surgery demonstrated worsening of the syrinx and persistent cerebellar tonsillar ectopia (Fig. 1C and D). Repeated MR imaging of the brain performed at the same time demonstrated again the diffuse smooth dural enhancement (Fig. 2B). The images were interpreted at this time as consistent with intracranial hypotension, and the patient was referred for CT-guided myelography and a targeted blood patch.

Abbreviation used in this paper: SIH = spontaneous intracranial hypotension.

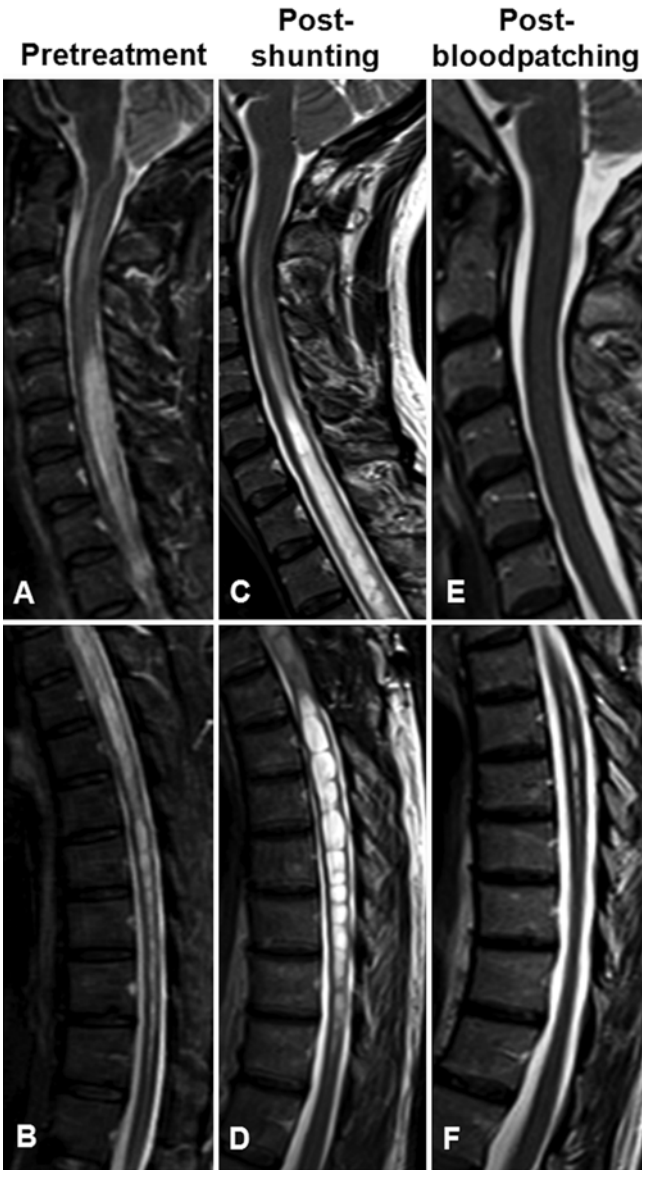

Fig. 1. Sagittal T2-weighted MR images of the cervical and thoracic spinal cord. **A and B:** Extensive syringohydromyelia seen at presentation. **C and D:** Repeated imaging after placement of a syringosubarachnoid shunt demonstrating worsening of the syrinx. **E and F:** After CT-guided blood patching, there is a marked decrease in the size of the syrinx and elevation of the cerebellar tonsils.

A CT myelogram demonstrated numerous small irregular meningeal diverticula involving the nerve roots at multiple levels throughout the thoracic spine. Although no direct leakage of contrast was seen, these irregular diverticula were chosen for empirical targeted blood patches because of the known association between such irregular diverticula and sites of CSF leakage.[12] A total of 7 irregular diverticula were targeted and patched using 3 ml autologous blood at each site. We obtained 2 subsequent myelograms and performed CT-guided targeted blood patching, with 9 and 11 diverticula treated during each procedure. No other interval therapy was instituted.

The patient improved clinically following blood patching, and follow-up imaging at 9 months revealed near-complete resolution of the syrinx (Fig. 1E and F) and resolution of the MR imaging–documented findings of intracranial hypotension (Fig. 2C). She experienced significant decrease in her arm pain, with significant increase in her lower-extremity strength such that she was once again able to ambulate without the assistance of her walker.

Discussion

We demonstrate resolution of an extensive syrinx associated with SIH, which had been unresponsive to initial syringosubarachnoid shunting, using targeted CT-guided blood patches of multilevel meningeal diverticula of the thoracic spine. To the best of our knowledge, this represents the first reported case of syringohydromyelia treated in this manner. A limited number of cases of syringohydromyelia have been reported previously in association with intracranial hypotension.[7,10,14] The patients in these cases underwent surgical repair of the dura mater, shunting, or suboccipital craniectomy. This case is significant in that it offers a minimally invasive alternative therapy to patients who might otherwise undergo much more invasive surgical procedures.

We have previously described the rationale and technique for performing multiple blood patches targeted at meningeal diverticula when no high-flow leak is seen.[8] In brief, most spontaneous CSF leaks are thought to originate in the thoracic spine.[6] Numerous cases of directly observed leakage from enlarged diverticula of spinal nerve roots have implicated these structures as potential weak spots prone to spontaneous leakage.[3,13] When evidence of SIH is demonstrated but no clear leak is localized, the authors have developed the practice of empirically placing blood patches adjacent to such enlarged thoracic diverticula to increase the chance of occluding slow or intermittent leaks not visible on imaging.

In many patients with SIH the condition is initially misdiagnosed, leading to delay in appropriate therapy.[11] This is likely in part due to lack of awareness of the typical clinical and imaging features of the disease but also may be compounded by the frequency of atypical presentations.[9,12] In our patient, headache was not, in fact, the primary complaint, although she had suffered from chronic headaches in the past. Knowledge of typical and variant presentations of SIH is therefore important in establishing the correct diagnosis.

This case also highlights how imaging features of SIH overlap with those of other disease processes. In particular, cerebellar tonsillar ectopia is seen with SIH due to loss of buoyancy of the brain in the setting of CSF leakage, resulting in downward sagging of the cranial structures, and may be accompanied by other signs of sagging including effacement of the suprasellar cistern and descent of the floor of the third ventricle.[12] In contrast, true Chiari I malformations are thought to be congenital abnormalities, the result of an inappropriately small posterior fossa.[2] The term "acquired Chiari malformation" is sometimes applied to cerebellar tonsillar ectopia in the setting of CSF hypovolemia (such as in the setting of

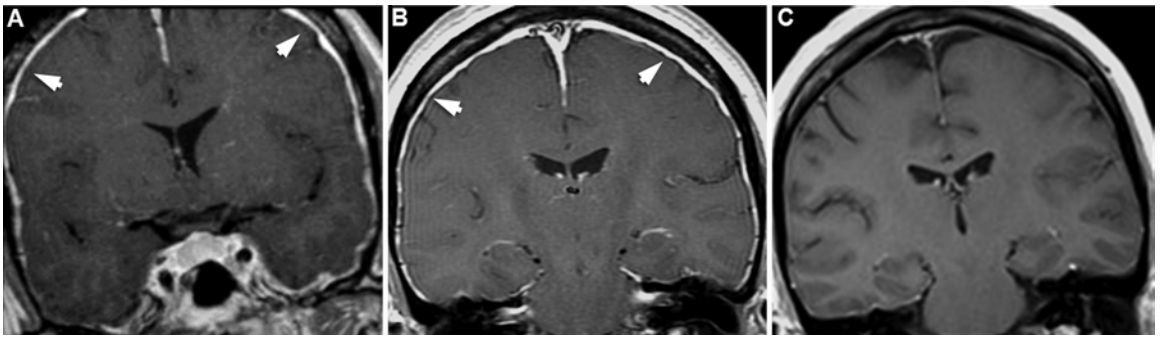

Fig. 2. Coronal T1-weighted postcontast MR images obtained prior to treatment **(A)**, after syringosubarachnoid shunting **(B)**, and after CT-guided blood patching **(C)**. Smooth dural enhancement *(arrows)* is seen on pretreatment images. Note also the hyperemia of the pituitary gland and effacement of the suprasellar cistern. The dural enhancement *(arrows)* persists following syringosubarachnoid shunting **(B)**. Following CT-guided blood patching, there is resolution of dural enhancement **(C)**.

shunting or lumbar drainage of CSF) or intracranial hypotension. This terminology is potentially confusing and should not be mistaken for a true Chiari I malformation, as the pathogenesis and treatment are different. Because the appearance of the cerebellar tonsils is similar in both Chiari I malformations and SIH, it is imperative to maintain a high level of suspicion in the appropriate clinical setting to correctly select and interpret diagnostic imaging. In our case, abnormal dural enhancement suggestive of SIH had not been appreciated in the past, but it was a key finding in establishing the correct diagnosis. Pituitary hyperemia, another well-described imaging manifestation of intracranial hypotension, had been misinterpreted in this case as a pituitary tumor, leading to a misdiagnosis of the cause of the patient's headaches. The distinction between SIH and Chiari I malformation is also important in the selection of therapy. Posterior fossa decompression, used to treat Chiari I malformations, will not treat the underlying cause of SIH and may not result in relief of all symptoms in patients with intracranial hypotension.[11]

The worsening of the syrinx following placement of a spinal syringosubarachnoid shunt raises interesting questions about the pathogenesis of the syrinx in this case. In general, theories of syrinx pathogenesis can be divided into two categories: those in which CSF is driven or sucked into the spinal cord from above the foramen magnum through the aperture of the central canal, and those in which CSF below the foramen magnum is driven into the cord. The former group includes theories such as the hydrodynamic theory (that is, the water-hammer effect)[4] and craniospinal pressure dissociation theory.[15] The latter theories propose that CSF is produced by the spinal cord parenchyma and is directed into the spinal cord along perivascular spaces, either due to excess CSF volume in the subarachnoid space or a pistonlike effect of the cerebellar tonsils.[1,5] In our case, there are two possible explanations for the progression of the syrinx after syringosubarachnoid shunting. First, it is possible that placement of the syringosubarachnoid shunt facilitated flow of CSF from above the foramen magnum along the syrinx cavity and into the spinal subarachnoid space, where leakage along the spinal nerve roots ultimately exacerbated CSF hypovolemia. Alternatively, a pistonlike effect of the tonsils could have driven CSF in a retrograde fashion through the shunt into the syrinx cavity, thereby expanding the syrinx. In either scenario, this case raises questions about the utility of syringosubarachnoid shunt placement in syringes caused by intracranial hypotension.

In summary, we have described a case of syringohydromyelia in the setting of intracranial hypotension treated with CT-guided blood patches. Intracranial hypotension should be considered as a potential cause for syringes associated with cerebellar tonsillar ectopia, and brain MR imaging may help to suggest the diagnosis. Treating potential leak sites with image-guided blood patching offers a targeted therapeutic strategy that can be used to avoid the morbidity associated with surgery and simultaneously address potential leaks at more than one site.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Kranz, Gray. Acquisition of data: Gray. Analysis and interpretation of data: Gray. Drafting the article: all authors. Critically revising the article: Kranz, Gray. Approved the final version of the paper on behalf of all authors: Kranz.

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