Several different theories there is an attempt to explain the pathophysiology of syringomyelia associated with the Chiari I malformation. In some of them, mechanisms in which the syrinx expands progressively from an initially small locus of syringomyelia, a "drop" of fluid, to a cavity that is large enough to distend the spinal cord are proposed. This is the mechanism proposed for the theories advocated by Gardner and Angel,4,5 and Williams18–20 in which the persistence of a communication between the fourth ventricle and the central canal at the obex is required, as it is in the suggestion of Milhorat and colleagues13–15 that the syrinx results from obstruction of the central canal by ependymal hyperplasia occluding rostral flow of CSF in the central canal. If these theories are correct, hydromyelia arises from expansion of the central canal by ependymal hyperplasia occluding rostral flow of CSF in the central canal. If these theories are correct, hydromyelia arises from expansion of the central canal, either from forces acting from above or below. On the other hand, proponents of other theories suggest that the pathogenesis of syringomyelia is determined by a mechanism external to the spinal cord and that the fluid comprising the syrinx is CSF that has passed through the surface of the spinal cord along many microscopic paths, enlarging the syrinx from without, not from within. Thus, the theories of Ball and Dayan1 and Oldfield, et al.,16 argue that episodic elevations of either spinal venous pressure or pulse pressure in the subarachnoid space, respectively, propel the CSF through the Virchow–Robin spaces and the extracellular space of the spinal cord and into the syrinx. These latter theories, unlike the former, imply that spinal cord swelling and edema potentially predate the development of a syrinx cavity. However, this stage in the development of a syrinx has not been previously noted in a patient with a Chiari I malformation and syringomyelia.

We present a patient with a Chiari I malformation in whom serial MR images obtained over a 3-month period document swelling and edema of the spinal cord that preceded development of a well-defined syrinx. Spinal cord edema in this case is consistent with transmural passage of CSF from the spinal subarachnoid space into the spinal cord.1,16 The notion that spinal cord edema develops into a syrinx is not explained by theories of syrinx pathogenesis in which it is postulated that syrinx fluid originates from the fourth ventricle via the central canal of the spinal cord4,5,18–20 or results from obstruction of CSF passage within the central canal.13–15 Because treatment with craniocervical decompression and duraplasty in this case, which opened the CSF pathways at the foramen magnum, produced resolution of the spinal cord edema and syringomyelia, the CSF block at the foramen magnum is implicated as the cause of the spinal cord edema. The theory that the cerebellar tonsils act as a piston on a partially enclosed cervical subarachnoid space, creating enlarged cervical subarachnoid pressure...
waves that drive CSF into the spinal cord, is consistent with the association of spinal cord edema and the Chiari I malformation in this case.\textsuperscript{16}

**Case Report**

**History and Examination.** This 48-year-old woman presented with a 10-month history of progressive numbness, paresthesias, and upper-extremity pain. Symptoms were initially noted in her left hand and progressed gradually to involve the other hand, the shoulders, and the arms. Clumsy hand movements eventually slowed her work on the computer keyboard and prevented her from working. She experienced intermittent headaches unrelated to position, coughing, or sneezing. Treatment with corticosteroid and nonsteroidal antiinflammatory medications was ineffective. Neurological examination revealed weakness (4/5) of the right hand movements eventually slowed her work on the computer keyboard and prevented her from working. She experienced intermittent headaches unrelated to position, coughing, or sneezing. Treatment with corticosteroid and nonsteroidal antiinflammatory medications was ineffective. Neurological examination revealed weakness (4/5) of the right hand, paresis, and upper-extremity pain. Follow-up evaluations at 3 months after the initial imaging session. On this study the diffuse signal of intermediate intensity that was noted previously in the upper thoracic portion of the spinal cord was replaced by a syrinx with typical low signal intensity surrounding a rim of spinal cord of normal signal (Fig. 2).

**Operation and Postoperative Course.** The patient underwent a suboccipital craniectomy, laminectomy of C-1, and duraplasty. The arachnoid was translucent, and the absence of subarachnoid scarring was visible through the arachnoid. Thus, the arachnoid was left intact. No attempt was made to drain the syrinx cavity. The patient made an uneventful recovery and was discharged from the hospital with improved upper extremity pain. Follow-up examinations at 3 and 16 months postsurgery demonstrated modest improvement in sensation and motor function.

**Postoperative Imaging Studies.** The syrinx resolved completely by 3 months. There was no evidence of syrinx or spinal cord edema on MR images of the cervical spine obtained 3 and 16 months after surgery (Fig. 3). The cerebellar tonsils had ascended to a normal position, and the cisterna magna was evident dorsal to the cerebellar tonsils.

**Discussion**

In this report we describe a patient who developed spinal cord edema as an intermediate stage in the development of a syrinx. Decompression of the craniocervical junction resulted in expansion of the CSF pathways at the foramen magnum and the cerebellar tonsils assumed a normal morphology and position. The syrinx resolved completely, and clinical progression was arrested.

In the respective theories of Gardner and Angel\textsuperscript{4,5} and Williams,\textsuperscript{16–20} it is proposed that development and growth of a syrinx results from progressive expansion of the central canal, with CSF entering the syrinx via the central canal and with the syrinx expanding progressively from the forces transmitted by the ventricular CSF pulse wave\textsuperscript{4,5} or by intraspinal pressure differentials.\textsuperscript{16–20} Milhorat and colleagues\textsuperscript{13–15} have suggested that progressive expansion of the syrinx results from the occlusion of the central canal blocking the egress of CSF from the syrinx. On the other hand, Ball and Dayan\textsuperscript{1} and Oldfield, et al.,\textsuperscript{16} have contended that syrinx development is caused by episodic elevations of spinous venous pressure\textsuperscript{1} or pulse pressure\textsuperscript{16,17} in the subarachnoid space, respectively, increasing transmural passage of CSF from the spinal subarachnoid space to the syrinx. In these latter theories the authors predict that cord edema arises from distention of the extracellular space of the spinal cord and precedes syrinx development and growth, as was noted in our present case.

There is abundant supportive evidence that spinal cord edema and syrinx fluid originate from the CSF within the spinal subarachnoid space: 1) CSF containing ionic and nonionic contrast media has been shown to pass from the subarachnoid space to the syrinx on computerized tomography–myelography studies of patients with syringomyelia.\textsuperscript{12} 2) Transmural flow of CSF from the subarachnoid space to the syrinx has been demonstrated on radioactive tracer studies.\textsuperscript{6} 3) Authors of experimental studies have shown that subarachnoid CSF can enter the spinal cord through the Virchow–Robin spaces, which surround the segmental vascular supply of the spinal cord and that these spaces are enlarged in patients with syringomyelia.\textsuperscript{14,30} Because radiological and pathological studies rarely demonstrate a patent central canal in adult patients with syringomyelia,\textsuperscript{3,10} passage of CSF from the fourth ventricle to the syrinx cannot be an important mechanism of syringomyelia progression in adults. 4) Syrinx fluid in patients with the Chiari I malformation is identical in chemical composition to CSF,\textsuperscript{7} indicating that it originates from CSF and not from vasogenic or cytotoxic spinal cord edema that would produce elevated protein levels within the extracellular space and syrinx. Because the craniocervical decompressive procedures and duraplasty resolved the syringomyelia and cord edema in our case, it appears that opening the CSF pathways at the foramen magnum eliminated the force that drove CSF into the spinal cord.\textsuperscript{3,10} Following surgery, the return of the cerebellar tonsils to a normal position and shape, as well as resolution of the dorsal hump of the medulla at the point of tonsillar impaction, together imply that these arose from a posterior fossa that was too small for its contents rather than a congenital central nervous system malformation.

Syringomyelia has been reported to be a precursor to neurological progression in patients in whom posttraumatic syringomyelia has developed. Three patients with progressive posttraumatic syringomyelia extending from the level of their spinal injuries were reported to have increased...
Syrinx development

FIG. 1. Sagittal and axial MR images of the cervical spine obtained 3 months before surgery. On T₁-weighted images (upper left) the abnormally shaped cerebellar tonsils extend to the upper margin of the arch of C-1, 12 mm below the foramen magnum. A narrow syrinx is present within the center of the unexpanded spinal cord from C-2 to C-7 (arrow) and the thoracic segments of the spinal cord are diffusely enlarged with reduced signal intensity (upper left and right). On the T₂-weighted images the spinal cord is swollen by edema (lower left and right) and the distinction between the syrinx and the edematous spinal cord is clear (arrow in lower left).
produce a major loss of spinal cord substance, neurological recovery in our patient was incomplete. Consideration should be given to early treatment if spinal cord edema is present in a patient with a Chiari I malformation, because subsequent syrinx formation is likely and neurological deficits are more likely to be permanent in the presence of a syrinx. Because decompressive surgery at the foramen magnum, which frees the pulsatile flow of subarachnoid CSF across this level, eliminates the pathophysiological mechanism of syringomyelia in these patients but does not invade the neural tissue, surgical treatment at an earlier stage can be justified.

Conclusions

In previously reported cases of Chiari I malformation spinal cord edema had not been documented to precede syrinx formation. Our case supports the theory that the pathophysiology of syringomyelia in adult patients with the Chiari I malformation requires CSF to pass through the spinal cord from the subarachnoid space to the syrinx. The development of syringomyelia in an area of spinal cord edema supports early intervention in such patients by performing craniocervical decompression and duraplasty to prevent irreversible spinal cord injury due to progression of syringomyelia.

References


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**Fig. 2.** Sagittal T₁-weighted MR image of the cervical spine obtained 1 week before surgery, demonstrating that the cervical syrinx has enlarged and a well-defined syrinx has developed in the thoracic region of the spinal cord at the site of previously documented spinal cord edema. Note that the septum (arrow) separates the syrinx into cervical and thoracic compartments and that the tonsillar impaction is now down to the midportion of the dorsal arch of C-1.

**Fig. 3.** Sagittal T₁-weighted MR image of the cervical spine obtained 16 months after surgery revealing resolution of the syrinx, the ascension of the cerebellum to a normal position above the foramen magnum, normal shaped tonsils, and a visible cisterna magna. Also note the disappearance of the focal dorsal protuberance (“hump”) at the junction of the medulla and cervical spinal cord, a morphological composition consistent with impaction of the tonsils into the foramen magnum and brainstem distortion, that was previously evident in Figs. 1 and 2.
Syrinx development

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