Spontaneous drainage of syringomyelia

Report of two cases

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Two cases are reported of Arnold-Chiari type I malformation associated with syringomyelia, in which magnetic resonance (MR) imaging revealed spontaneous decompression of the syrinx. In one case axial MR imaging sections showed a communication between the syrinx and the spinal subarachnoid space, which supports the hypothesis that fissuring of the cord parenchyma is instrumental in the spontaneous resolution of syringomyelia. The MR imaging changes were not accompanied by variations in the patients' clinical course.

KEY WORDS • Arnold-Chiari malformation • syringomyelia • magnetic resonance imaging

SYRINGOMYELIA is a malformation marked by the presence of a cavity within the spinal cord. In 75% of cases, it is associated with Arnold-Chiari type I malformation. Several theories on the pathogenesis of syringomyelia, have been put forward but those that have gained widest credence are the hydrodynamic theory of Gardner and the cerebrospinal fluid (CSF) dissociation theory of Williams. Diagnostic imaging has made the identification of syringomyelia and malformations of the craniovertebral junction much easier. Magnetic resonance (MR) imaging in particular, being noninvasive and easily repeatable, allows exact assessment of the time course of the syrinx both in surgically treated and in untreated patients.

We report two cases of cervicothoracic syringomyelia associated with Arnold-Chiari type I malformation. The first case was symptomatic, and the second was found incidentally in a patient with a hypophyseal microadenoma, in which we secured MR imaging evidence of spontaneous resolution of the syringomyelic cavity. In one case communication between the cavity and the subarachnoid space was actually visible.

Case Reports

Case 1

This 42-year-old man presented in March, 1991, with a 15-year history of paresthesias and reduced sensation in the upper limbs, gait disorders, and atrophy of the hand muscles of 2 years' duration. Neurological examination revealed markedly increased reflexes in all limbs, but especially the lower, absence of abdominal reflexes, syringomyelic dissociation of sensation from C-2 to T-1 on the left side, horizontal nystagmus, and wasting of the interosseous muscles of both hands.

A craniocervical MR image obtained in February, 1988 (Fig. 1 left) showed a syrinx extending from C-2 to T-1 associated with an Arnold-Chiari type I malformation with minimal compression at the foramen magnum. These findings were confirmed by MR imaging 1 year later. While he was in the hospital, the patient's neurological condition remained stationary. A third MR image (Fig. 1 right) confirmed the Arnold-Chiari type I malformation but showed near disappearance of the cervicothoracic syringomyelia. Despite this, the symptom pattern and neurological status remained unchanged.

Case 2

This 31-year-old woman had suffered menstrual disorders 10 years prior to our evaluation of her condition. Plain x-ray films of the skull were normal and a hormone assay yielded a prolactin level of 120 ng/ml. Computerized tomography (CT) of the head disclosed a hypophyseal microadenoma. Treatment with bromocriptine (1 tablet three times daily) resulted in normalization of the menstrual cycle and of the prolactin level.

In 1987, in the course of a pregnancy, the patient underwent a CT brain scan which confirmed the pres-
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Fig. 1. Magnetic resonance images, sagittal projection, in Case 1. Left: Image obtained in February, 1988, showing an Arnold-Chiari type I malformation and cervicothoracic syringomyelia. Right: Image obtained in March, 1991, showing almost complete decompression of the syringomyelic cavity. The Arnold-Chiari type I malformation is unchanged.

ence of the microadenoma and imaged incidentally a syringomyelic cavity extending from C-1 to T-7 together with an Arnold-Chiari type I malformation.

She exhibited no signs or symptoms of syringomyelia. Having carried another pregnancy to term in 1990, she underwent MR imaging in December, 1991, which confirmed the presence of the Arnold-Chiari malformation but documented the near disappearance of the syrinx. A further MR image obtained in January, 1992, in the axial sections (Fig. 2) revealed a communication between the syringomyelic cavity and the subarachnoid space, into which the cavity presumably drained.

Discussion

Terminology

The terms "hydromyelia" and "syringomyelia" are used to indicate a condition characterized by the presence of a fluid-filled cavity within the spinal cord and, in the past, were often considered synonymous. A hydromyelic cyst is an asymptomatic dilatation of the central canal, covered by ependymal tissue; it is almost always an incidental finding at autopsy. A syringomyelic cyst, however, extends further both longitudinally and transversally, and typically has a gliotic component; it is more often symptomatic. Two types of syringomyelia exist: communicating or noncommunicating. In the first, there is a direct communication between the syringomyelic cavity and the fourth ventricle at the level of the obex.

Pathophysiology

Syringomyelia is a disease with a capricious clinical course and unknown pathogenesis. In 1892, Abbe and Coley performed a syringostomy and withdrew intramedullary fluid. Since that time, several surgical procedures have been used for the treatment of syringomyelia, not always with satisfactory results.

The pathogenesis of the disease is still debated and several hypotheses have been advanced. The two main propositions are the hydrodynamic theory of Gardner and the CSF dissociation theory of Williams. Gardner's theory relates the persistence and enlargement of the central canal to the conservation of a communication between the fourth ventricle and the central canal of the spinal cord through the obex.

In contrast, Williams focused attention on the CSF pressure dissociation that arises between the intracranial and vertebral compartments as a result of the sudden rise in pressure that occurs when the patient coughs, sneezes, and performs the Valsalva maneuver, all of which initially cause a rise in intravertebral pressure which is transmitted to the skull. When the perimedullary venous pressure returns to normal, the mechanism is reversed, with the return of the pressure wave from the intracranial to the spinal compartment. The presence of an obstruction to the drainage of CSF at the foramen magnum, as occurs in Arnold-Chiari type I malformation, gives rise to a pressure dissociation between the intracranial and spinal compartments, the CSF being sucked into the central canal with the formation of a syringomyelic cavity.

Spontaneous Fissuring

The exact mechanism by which the syringomyelic cavity was drained in our two cases is not known. If Williams' theory is correct, it is highly probable that, owing to an undue rise in intracavitary pressure, a fistula formed in the cord substance and the CSF drained from the syrinx into the perimedullary subarachnoid space.

This mechanism was proposed by Jack, et al., in the only reported case of spontaneous decompression of syringomyelia. Using axial MR imaging, these authors showed the presence of hemiatrophy of the cord.
at C-5 and considered that fissuring occurred at that level. The findings in our Case 2 bear out their hypotheses; in fact, the axial MR image clearly demonstrates the existence of a channel between the syringomyelic cavity and the subarachnoid space of the cord.

It is noteworthy that, despite the near disappearance of the syringomyelia in our two patients, there was no change in the clinical course of Case 1 and no symptoms at any time in Case 2. This lack of association suggests that other factors are at work in the pathophysiology of the syringomyelic syndrome and that, until the various aspects of the pathogenesis have been fully elucidated, it will be hard to devise an ideal surgical treatment. The fact that our patients showed no change in clinical pattern while the patient described by Jack and others exhibited some improvement after spontaneous decompression of the syrinx might explain why a percentage of syringomyelic patients who receive surgical treatment show no improvement or even clinical deterioration despite the disappearance of the cavity.

Conclusions

Since the outcome in syringomyelia is not always satisfactory and since patients may long remain neurologically stable even without benefit of surgery and, finally, since there is a chance of spontaneous drainage of the cavity, we favor a wait-and-see policy with periodic clinical and MR imaging examination for neurologically stable patients. With the increasing availability of MR imaging, it is likely that spontaneous drainage of syringomyelic cavities, rarely seen until now, will be observed more frequently.

References


Manuscript received May 8, 1992. Accepted in final form January 12, 1993.
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