Spontaneous decompression of syringomyelia: magnetic resonance imaging findings

Case report

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The case of a 30-year-old woman with Chiari I malformation and a cervicothoracic syrinx is presented. The patient was followed clinically over a 21/2-year period. Spontaneous and complete resolution of the syrinx, as documented by serial magnetic resonance studies, was accompanied by only a minimal change in objective symptomatology.

KEY WORDS • syringomyelia • Chiari I malformation • magnetic resonance imaging

A significant proportion of patients with Chiari malformation have an associated syrinx of the cervicothoracic spinal cord. Magnetic resonance (MR) imaging is recognized as the optimal method for both diagnosis and follow-up evaluation of patients with Chiari malformations and associated syrinx.4-6 Patients with this condition may be treated conservatively or surgically, depending on the severity of symptoms and whether symptoms are progressive.1,3-8

We report a unique case of a patient with Chiari I malformation and an associated cervicothoracic syrinx in whom spontaneous and complete decompression of the syrinx was documented by serial MR studies. The patient had neurological evaluations at our institution on three separate occasions, and each time she underwent MR imaging of the head and cervicothoracic spine.

Case Report

This 30-year-old woman first presented in August, 1988, with a 2- to 3-year history of paresthesias and decreased pain sensation in the left chest wall and left upper extremity. She had unknowingly burned or cut her left hand on several occasions. She also complained of headaches. Neurological examination revealed absent epigastric, hypogastric, and left upper-extremity reflexes. She had a dissociated sensory loss on the left side from the C2-L1 dermatomes consisting of diminished pinprick, temperature, and deep pain sensation with nearly normal two-point discrimination, touch, and vibratory sensation. Strength was normal. An MR study demonstrated a Chiari I malformation with minimal compression at the foramen magnum but with a large cervicothoracic syrinx (Fig. 1).

FIG. 1. Sagittal magnetic resonance image (TR 500 msec, TE 20 msec) obtained in August, 1988, showing Chiari I malformation and a large cervicothoracic syrinx.
The patient returned in February, 1989, when both the MR image (Fig. 2) and neurological examination were unchanged. However, her headaches, which had been treated with propranolol, were significantly reduced in number. These were presumed to be migraineous in nature.

On her third visit in February, 1990, the patient reported that the left upper-extremity paresthesias had diminished. Physical examination showed persistent absence of the left brachioradialis reflex; the left biceps reflex was obtainable but diminished, while the left triceps reflex was only slightly diminished. Epigastric and hypogastric reflexes remained absent on the left side and diminished on the right. Her left-sided dissociated sensory loss was unchanged except that deep pain sensation was now nearly normal. An MR study at this time showed persistent Chiari I malformation but complete absence of the cervicothoracic syrinx (Fig. 3). The left hemicord was atrophic at the C4—T2 levels. The patient had no surgery, injury, or abrupt change in symptoms between the 1989 and 1990 MR studies.

All T2-weighted MR studies in this patient were performed with motion compensation; therefore, reliable information on the flow characteristics of fluid within the syrinx cavity was not available.

**Discussion**

**Terminology**

The terms “hydromyelia” and “syringomyelia” are both used to describe fluid cavities within the spinal cord, hydromyelia referring to dilation of the central canal and syringomyelia to a longitudinal fluid cavity outside the central canal, typically with gliotic walls. In practice, however, it is difficult to distinguish between the two and many spinal cord cavities have features of both. The more generic term “syringohydromyelia,” or more simply “syrinx,” is therefore often used to describe any longitudinally oriented fluid cavity within the spinal cord.2-5,9

Syringes may be of two types: communicating or noncommunicating. In the former, a direct communication exists between the fourth ventricle and the syrinx at the obex. It is this type which is associated with Chiari malformation. No such communication is present in noncommunicating syringes, which are most commonly associated with spinal cord tumors or cord trauma.2,5,6
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Pathogenesis

Several theories have been proposed to explain the formation and extension of communicating syringes in patients with Chiari malformation. Review of the literature would indicate that the unifying theories of Williams' regarding formation and extension of syringes in these patients are the most widely accepted at this time. It is Williams' view that the Chiari malformation acts as a ball-valve obstruction at the foramen magnum. Driven by expansion of the epidural veins, this obstruction creates a pressure dissociation between the intracranial cavity and the spinal subarachnoid space which tends to "suck" cerebrospinal fluid (CSF) into the central canal, thereby initially forming a hydromyelic cavity. When this cavity reaches a critical size, extension is brought about by what is termed a "slosh mechanism." During Valsalva maneuvers, the spinal epidural venous plexus will expand, forcing the fluid within the syrinx in a cephalad direction. Upon release of the Valsalva, the fluid is forced downward. This up-and-down "sloshing" action can be quite forceful with vigorous Valsalva maneuvers and is responsible for extending the syrinx, either by expansion of the central canal (hydromyelia), dissection into the central gray matter (syringomyelia), or both.

The mechanism by which an initial spinal cord cavity is formed in patients with a noncommunicating syrinx is different. Once formed, however, extension of a fluid cavity within the cord may occur by the same "slosh" mechanism described above, regardless of the mode of origin. Several different means by which cavities may initially form in the spinal cord after trauma have been proposed. A traumatic hematomyelia with subsequent breakdown of blood products will form a cavity. An area of myelomalacia which subsequently breaks down into a necrotic cavity may be formed either by ischemia or by the direct action of destructive cellular enzymes (such as lysozymes) released at the time of trauma. It has also been postulated that adhesions at the site of a traumatic cord fissure may form a one-way valve from the subarachnoid space into the cord substance, through which CSF can be forced by CSF pulsations.

Spinal cord tumors may themselves cavitate. This condition is to be differentiated from non-neoplastic syrinx cavities which may form in association with either intra- or extramedullary tumors in patients with or without surgery. Proposed mechanisms for cavity formation are similar to those in trauma; hematomyelia, mechanical compression, or adhesion formation.

Decompression of Syrinx

The actual mechanism by which the syrinx in our patient became decompressed is not known. The most plausible explanation, however, would seem to be that a tear was formed in the cord, creating a communication between the syrinx cavity and the spinal subarachnoid space. Vigorous Valsalva-driven propulsion of the fluid within the syrinx cavity, as described by Williams, could explain such an occurrence. Sudden spinal cord deficits have been reported in patients with obstructing foramen magnum lesions (including Chiari I malformation) and cervicothoracic syringomyelia following a forceful Valsalva maneuver (such as rope climbing or heavy lifting). Acute dissection of the syrinx into the cord with exertion (Valsalva maneuver) has been cited as the most plausible explanation for the sudden deficits observed in these patients.

Of particular interest is the fact that despite complete collapse of an extensive syrinx cavity, our patient experienced little objective improvement in her symptoms. Compression at the foramen was not particularly striking (Fig. 3). It seems unlikely, therefore, that either cervicomedullary compression or the presence of the cervicothoracic syrinx cavity alone caused her symptoms. An axial image (Fig. 3 lower left) demonstrated atrophy of the left hemicord from C4—T2. It appears reasonable to postulate that the syrinx cavity had dissected into the left hemicord in this region, producing permanent structural damage in this portion of the cord and an irreversible neurological deficit which was not affected by decompression of the syrinx. Such a scenario may explain why some patients fail to improve symptomatically despite successful surgical decompression of syrinx cavities, as documented on postoperative imaging studies.

Surgical Indications

Symptoms in Chiari patients with syrinx have been attributed to compression of the cord by the syrinx, mechanical compression of the brain stem/cervical cord by the hind-brain deformity, or both. Numerous surgical interventions have been devised for the treatment of syringes in Chiari patients. Response to surgery has been variable; patients may experience mild improvement, remain stable, or deteriorate postoperatively. Patients may also remain stable without surgery. For these reasons, surgical intervention is rarely undertaken in these patients upon initial presentation to our institution. Serial neurological and MR examinations are used to monitor the progression of neurological deficits. Surgical management is considered when a measurable increase in neurological deficit occurs. As long as the deficit noted on the original examination is static, continued observation is advised.

References
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