Chiari I malformation with syringomyelia

Evaluation of surgical therapy by magnetic resonance imaging

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Five patients with a Chiari I-syringomyelia complex of adult onset were evaluated by magnetic resonance (MR) imaging. All patients underwent suboccipital craniotomy with upper cervical (C-1 and part of C-2) laminectomy, arachnoid retraction, and duraplasty. Postoperative MR studies of four patients disclosed collapse of the syringomyelic cavity, even when the cavity extended into the thoracic region. This appeared to be a progressive process taking place over several weeks. Operative complications are noted and physiological implications are discussed.

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

The advent of new imaging techniques in the past decade (in particular, computerized tomographic scanning with injection of water-soluble contrast medium and magnetic resonance (MR) imaging) has greatly facilitated the diagnosis of Chiari I malformation and associated syringomyelia. Indeed, MR imaging has now become the optimal technique, since it is noninvasive and permits simultaneous studies of the brain, craniocervical junction, and spinal cord. As a result, this disease complex, once thought to be quite rare, is now being diagnosed with increased frequency. Surgical treatment of syringomyelia was greatly stimulated by the pioneering work of Gardner, et al., who conceptualized the idea of a hindbrain malformation with cerebrospinal fluid (CSF) being forced into the central canal of the cord to form a syrinx because of obstruction of the outlet foramina of the fourth ventricle. However, the recommended form of treatment — foramen magnum decompression with plugging of the obex — did not always eliminate the syrinx. The concept of dissociation of the cranial and spinal pressure has provided the most useful working hypothesis and basis for current surgical therapy. Williams, who is largely responsible for developing this theory, has summarized current thinking on the subject in an excellent recent review. Stated simply, the obstruction to free communication of CSF flow from the cranial to the spinal subarachnoid space permits development of a pressure gradient between these two fluid compartments. Negative pressure below the level of the block at the foramen magnum is then postulated to draw fluid into the spinal cord via the obex. Pressure gradients are accentuated during sudden Valsalva maneuvers, including coughing and sneezing, and with postural changes. Other excellent reviews are those of Bertrand, Oakes, and Peerless and Durward. Based on concepts similar to those developed by Williams, the present author has treated five patients with the Chiari I-syringomyelia complex. The diagnosis was established by MR imaging. Surgical treatment consisted of hindbrain decompression with particular attention to the arachnoid and insertion of a dural graft. Four patients also had postoperative MR studies.

Summary of Cases

Patient Population

The clinical features of the five patients in this report are summarized in Table 1.

Operative Procedure

The surgical technique employed in these patients consisted of the following essential steps. A suboccipital craniotomy approximately 2.5 x 2.5 cm in area was performed, with removal of the posterior rim of the foramen magnum. The posterior arch of the atlas was resected. Resection of a portion of the posterior arch of C-2 was carried out if preoperative imaging or operative
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FIG. 1. Artist's operative drawings. a: Dissection of the arachnoid away from the cerebellar tonsils. Inset: Closure of the suboccipital wound, showing placement of a dural graft and suture which incorporates the free edge of the retracted arachnoid. b: Separation of the cerebellar tonsils and direction of arachnoid suture in cases where the arachnoid is adherent to the tonsils. c: Placement of a tube from the fourth ventricle to the cervical subarachnoid space in cases where the arachnoid is adherent and the tonsils cannot be widely separated.

Operative Results

The results of treatment in this group of patients are summarized in Table 1. Figures 2 and 3 illustrate Cases 3 and 5, respectively.

Discussion

As stated by Williams, dissociation of cranial and spinal pressure appears to be a fundamental precondi-tion to the associated syrinx formation. However, it is recognized that syringomyelia does not always accompany severe adult Chiari I malformations with brain-stem compression. This is in itself puzzling, since the subarachnoid block (pressure dissociation) at the fora-men magnum level probably need not be complete to permit the development of a syringomyelic cavity; damping of the normal CSF pressure wave at this pathological barrier may be sufficient to permit cyst development.

The mechanism of fluid accumulation within the cord is not clearly understood at this time and more than one process may be operative. The cyst may initially develop and fill from the fourth ventricle via the obex, as suggested by Gardner, et al. Aboulker proposed that an imbalance of fluid pressure forces fluid into the cord parenchyma. Trans-cord fluid migration along Virchow-Robin spaces has been shown to
occur when a cystic cavity is already present.\textsuperscript{3,8} Both fluorescein\textsuperscript{9} and water-soluble contrast material\textsuperscript{2,7} accumulate within cord cysts. Williams\textsuperscript{5} has developed a hypothesis based on dynamic changes in CSF pressure and intracyst pressures that favor progressive enlargement of the cyst in man. The CSF within the syrinx then acts as a fluid dissector, mobile in a cephalo-caudad direction and propelled by transient pressure differences — a mechanism Williams refers to as “slosh.” It has been shown that the pressure in the cyst is greater than the pressure in the subarachnoid space.\textsuperscript{11}

The goals of surgery for Chiari I malformations with syringomyelia are threefold. The first is to relieve the pressure differential which exists above and below the subarachnoid block (partial or total) by restoring the continuity of the subarachnoid space and thereby making it the pathway of least resistance for CSF flow. The second goal is to eliminate the fluid compartment within the cord (that is, the syrinx), thereby thwarting the fluid dissector action. The third goal is to relieve the direct pressure on the brain stem.

Magnetic resonance imaging has proved definitive in establishing the diagnosis of Chiari I malformation with syringomyelia. Prompt employment of this tool allows the correct diagnosis to be made much earlier, improving the outlook for patients.\textsuperscript{6} This imaging technique enables objective evaluation of surgical results, a capability not previously available.

The operative technique described not only decompresses the subarachnoid space by relieving the extradural compression from the bone rim of the foramen magnum and C-1 arch, but it deliberately retracts the arachnoid laterally. We agree with Williams\textsuperscript{5} that careful attention to the arachnoid is an essential step in this procedure so as to minimize new arachnoid adhesions. Williams stated that the arachnoid should be excised over the exposed area of the cerebellar tonsils; we have found it simpler to dissect the arachnoid and retract it (whenever possible, as an intact layer) to the dural edge, incorporating it into the suture line as described above. In this technique (Fig. 1a), the cerebellar tonsils are lifted gently to establish their mobility and then replaced. This is the preferred means of establishing continuity of the subarachnoid space and was employed in the five patients described in this report. When the arachnoid adheres to the cerebellar tonsils with scarring, but the tonsils can be separated with direct visualization of the fourth ventricle, the adherent arachnoid at the medial edge of the tonsil can be sutured to the dural edge, thereby maintaining the opening into the fourth ventricle (Fig. 1b). When the arachnoid overlying the tonsils is so scarred that the tonsils cannot be readily separated to reveal the fourth ventricle, communication between ventricular and subarachnoid fluid compartments can be established by means of a Silastic tube from the fourth ventricle to the cervical subarachnoid

### TABLE 1

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs)</th>
<th>Symptoms &amp; Signs</th>
<th>Prior Treatment</th>
<th>Extent of Syrinx</th>
<th>Clinical Course</th>
<th>Follow-up MR Study</th>
<th>Complications</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>43</td>
<td>UE weakness, atrophy; brisk LE reflexes, extensor plantar responses; clonus</td>
<td>none</td>
<td>FM (\Rightarrow) T-5</td>
<td>UE unchanged; long-tract signs much improved (F/U 22 mos)</td>
<td>no</td>
<td>pseudo-meningocele</td>
</tr>
<tr>
<td>2</td>
<td>26</td>
<td>vertigo; ↓ visual function; ↓ sensation &amp; dysesthesias of face, chest, leg on rt; cramping rt hand</td>
<td>C7-T1 syrinx to subarachnoid shunt 5 mos prior</td>
<td>FM (?) (\Rightarrow) T-1</td>
<td>vertigo improved; residual mild dysesthesias rt arm (F/U 12 mos)</td>
<td>yes, 5\th mos postop</td>
<td>pseudo-meningocele</td>
</tr>
<tr>
<td>3</td>
<td>38</td>
<td>suboccipital headache; hyperesthesia both UE; ↓ sensation rt hand</td>
<td>none</td>
<td>FM (?) (\Rightarrow) C-7</td>
<td>headache relieved; residual mild UE dysesthesia (F/U 9 mos)</td>
<td>yes, 1 yr postop</td>
<td>none</td>
</tr>
<tr>
<td>4</td>
<td>58</td>
<td>progressive gait disturbance; pain cervical, shoulder &amp; waist areas; weakness both UE &amp; LE (rt worse); sensation ↓ rt UE &amp; LE; ↓ extensor plantar</td>
<td>marsupialization of cyst at C5-7 22 years prior</td>
<td>FM (\Rightarrow) T-10</td>
<td>improved gait &amp; UE function (F/U 9 mos)</td>
<td>yes, 16 mos postop</td>
<td>none</td>
</tr>
<tr>
<td>5</td>
<td>15</td>
<td>burning dysesthesias lt trunk, arm, hand, neck; numbness lt face; weakness both hands (rt worse)</td>
<td>none</td>
<td>FM (\Rightarrow) T-10</td>
<td>sensation improved; residual mild dysesthesias lt UE (F/U 7 mos)</td>
<td>yes, 11 mos postop</td>
<td>none</td>
</tr>
</tbody>
</table>

*UE = upper extremity; LE = lower extremity; FM = foramen magnum; FM (?) = extension to foramen magnum assumed, but not clearly shown; F/U = clinical follow-up time; MR = magnetic resonance imaging; ↓ = decreased.*
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space (Fig. 1c). All three of these techniques are believed to satisfy the goal of eliminating the presumed partial or complete subarachnoid block. The use of a dural graft would appear to be essential if one is to avoid adhesion of the suboccipital musculature to the cerebellum and brain stem, with the risk of establishing a new subarachnoid barrier to CSF flow. The occurrence of postoperative pseudomeningoceles in two of our patients has led us to change to autologous fascia lata grafts in place of cadaveric grafts in the hope that the fresh tissue will seal the dural defect more quickly and effectively. We do not agree with Williams that closure of the dura with a fascial graft following bone decompression reimposes the risk of tonsillar jamming and "continuing suck leading to slump."

Bertrand and Williams both recommend amputation of the tips of the cerebellar tonsils to achieve opening of the CSF pathways. This step has not been found necessary in any of the patients described in the present report, but it is recognized that it may apply in some instances. Evidence from MR imaging that the subarachnoid space is opened is both direct and indirect. The fluid equilibrium state existing prior to surgery accounts for the existence of a relatively normal subarachnoid space around the cervical spinal cord in these patients. Widening of the subarachnoid space, however, can be visualized on postoperative MR studies (Fig. 2 right). Elimination of the dissociation of cranial and spinal pressure exerts greater fluid pressure against the pial surface of the spinal cord, which apparently gradually and progressively eliminates the fluid compartment within the cord over a period of time up to several weeks (Fig. 3a), the most caudal portion of the syrinx being the last to clear. Magnetic resonance imaging may not always demonstrate a connection between the syrinx and the fourth ventricle. In some patients there may be no such connection; in some the small channel may be beyond the capacity for resolution of the imaging technique, and in some the communication may be off the midline or oblique in direction, and thus difficult to visualize.

When a communication exists between the syrinx and the fourth ventricle, it appears plausible that the cyst fluid is gradually expelled through the opening at the obex. This might explain why in some instances successful plugging of the obex was not followed by resolution of the syrinx: although the completeness of the obex seal in Gardner's patients is not known, it is possible, at least theoretically, that the procedure of hindbrain decompression and plugging of the obex succeeded in eliminating the syrinx in those patients whose obex seal was incomplete, while creating an isolated syringomyelic cavity when the seal was effective. Reversal of trans-cord fluid migration (see above) could explain the gradual collapse of the syrinx in those patients with noncommunicating syrinx cavities.

Fig. 2. Case 3. Left: Preoperative cervical magnetic resonance image (TE 38 msec) showing a septated cervical syrinx and portions of a presumed communication with the fourth ventricle. Right: Image obtained 6 weeks postoperatively (TE 39 msec) showing collapse of the syrinx following suboccipital and C-1 decompression with duraplasty as described.

Fig. 3. Case 5. Magnetic resonance images (TE 28 msec) of the cervical region preoperatively (a) and 10 days (b) and 4 1/2 weeks (c) after suboccipital decompression with duraplasty. Progressive collapse of the cervical syrinx over the 4 1/2 week period is evident.
In four patients in the present series, postoperative MR studies demonstrated that the cyst had collapsed following hindbrain decompression alone, even to the low thoracic level, without the additional step of direct cyst shunting, thereby avoiding additional risks: the added step of myelotomy is eliminated and implantation of a tube into the cord becomes unnecessary. Improvement in the fifth patient was based only on clinical criteria. This single-step procedure thus offers several advantages; it is hoped that more extensive use of this technique and longer follow-up care will further validate this approach. One patient had undergone a cervical cyst-to-subarachnoid shunt five months earlier; further collapse of the syrinx was demonstrable on MR studies following hindbrain decompression. Presence of a cyst-to-subarachnoid shunt tube in this patient may account for very focal widening of the collapsed cyst at the level of the shunt. Probably in this situation the tubing acts as a stent holding the cavity walls apart.

The third goal of surgery, relief of direct pressure on the brain stem, is difficult to validate by imaging techniques, but relief of brain-stem symptoms and signs often is quite dramatic in patients with Chiari I malformations (for example, our Case 2), both with and without associated syringomyelia. It is important to carry the laminectomy to the point of lowest tonsillar descent, which in many instances implies removal of at least a portion of the C-2 lamina.

The demonstration by MR imaging of collapse of even extensive syringomyelic cavities extending as low as the lower thoracic cord, following suboccipital decompression and duraplasty validates the general concepts of Williams. Diversionary shunting of the syrinx at the initial operation as advocated by Williams would seem unnecessary in many of these patients with Chiari I malformation, however, and should be reserved for those individuals whose syringomyelic cavity fails to collapse following the suboccipital decompression procedure. The observation that even successful hindbrain decompression and cyst collapse may not eliminate dysesthetic pain, particularly in regions exhibiting reduced pain sensation prior to surgery, is again confirmed, and remains an ongoing challenge in the care of these patients.

Since the initial submission of this manuscript, an additional patient has been treated who had a partial membranous fourth ventricle obstruction in association with a cervical syrinx, but without cerebellar ectopia. This patient's syrinx collapsed following fenestration of the fourth ventricle into the cisterna magna, using the technique depicted in Fig. 1b.

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References


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