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Surgical management of syringomyelia–Chiari complex

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Abstract Great variety exists in the indications and techniques recommended for the surgical treatment of syringomyelia–Chiari complex. More recently, magnetic resonance (MR) imaging has increased the frequency of diagnosis of this pathology and offered a unique opportunity to visualize cavities inside the spinal cord as well as their relationship to the cranio-cervical junction. This report presents 18 consecutive adult symptomatic syringomyelia patients with Chiari malformation who underwent foramen magnum decompression and syringosubarachnoid shunting. The principal indication for the surgery was significant progressive neurological deterioration. All patients underwent preoperative and postoperative MRI scans and were studied clinically and radiologically to assess the changes in the syrinx and their neurological picture after surgical intervention. All patients have been followed up for at least 36 months. No operative mortality was encountered; 88.9% of the patients showed improvement of neurological deficits together with radiological improvement and 11.1% of them revealed collapse of the syrinx cavity but no change in neurological status. None of the patients showed further deterioration of neurological function. The experience obtained from this study demonstrates that foramen magnum decompression to free the cerebro-spinal fluid (CSF) pathways combined with a syringosubarachnoid shunt performed at the same operation succeeds in effectively decompressing the syrinx cavity, and follow-up MR images reveal that this collapse is maintained. In view of these facts, we strongly recommend this technique, which seems to be the most rational surgical procedure in the treatment of syringomyelia–Chiari complex.

Key words Chiari malformation · Foramen magnum decompression · Syringomyelia · Syringosubarachnoid shunt

Introduction

Magnetic resonance imaging (MRI) has facilitated the diagnosis of syringomyelia in recent years, providing sharp delineation of syrinxes and accurate identification of malformations at the cranio-cervical junction. What is the mechanism of origin and maintenance of syringomyelia? Syringomyelia cannot be ascribed to a single pathophysiological mechanism, and its etiology and natural history are quite variable. It occurs in relation to intramedullary tumors, compressive myelopathy, spinal trauma, hydrocephalus and anomalies such as the Chiari malformation. Syringomyelia, is most frequently associated with Chiari malformation, and several hypotheses have been proposed to explain its pathophysiology in this particular manifestation.

One explanation for syrinx formation, which has recently been reported by Oldfield et al. [18], requires no

communication between the fourth ventricle and the upper pole of the syrinx, and is based on pressure acting on the cord and syrinx at the surface of the cord, not from within the cord. The perivascular spaces of the central nervous system act as conduits for drainage of extracellular fluid by bulk flow to the subarachnoid space. Cerebrospinal fluid (CSF) enters the cord through these dilated perivascular spaces to produce syringomyelia by multiple microscopic connections with the subarachnoid space, rather than the single channel from the fourth ventricle. The predominant involvement of the cervical segments of the cord may be related to the fact that maximum pulsatile pressure waves in the spinal subarachnoid space occur in the upper portion of the canal and are dissipated with increasing distances down the canal [5, 21].

This report presents 18 consecutive adult symptomatic syringomyelia patients with Chiari malformation who underwent surgical treatment at our clinic. Our aim in reporting this series was to discuss the effectiveness of the combination of two surgical procedures – foramen magnum decompression and syringosubarachnoid shunting – at the same operation. All patients underwent preoperative and postoperative MRI scans. The clinical features and operative outcome are correlated, and our experience in the surgical management of syringomyelia–Chiari complex is described in this manuscript.

Materials and methods

Eighteen patients with syringomyelia–Chiari complex diagnosed by MR imaging were included in the present study. There were 14 men (71.4%) and 4 women (28.6%), ranging in age from 19 to 55 years (mean 31.4 years). All patients had experienced progressive symptoms over a period of 6 or more months before diagnosis. The mean duration from onset of symptoms to diagnosis was 10 months. Clinical presentation of the 18 symptomatic adult patients with syringomyelia–Chiari complex included: neck pain (89.9%), sensory dysesthesias/numbness (77.7%), upper extremity weakness (44.3%), gait problems (22.1%), lower extremity weakness (11.1%) and headache (11.1%). Neurological findings included: sensory loss (77.2%), weakness (55.9%), cerebellar ataxia (51.5%), muscular atrophy (35.3%), unsteady gait (22.1%), nystagmus (20.2%) and lower cranial nerve dysfunction (9.1%).

Preoperative MRI scans were carried out in all patients, and both T1- and T2-weighted images were obtained in sagittal and axial planes. In all cases, MR imaging showed an intramedullary syrinx and herniation of the cerebellar tonsils more than 2 mm below a line drawn between the inferior margin of the basion to the opisthion (Chiari-I malformation). An angulated or peg-like configuration of the tonsils was also characteristic of herniation. Cervical cord was primarily involved, with syringomyelic cavities in 12 patients and with cervical/upper thoracic cavities in 6 patients. Isolated thoracic syrinxes were not seen in any of our patients. Septations and loculations were evident on MRI scan in four cases (Fig. 1). The mean length of the syrinx was five vertebral segments (range 3–11). The widest syrinx diameter was most frequently at the C4-C5 vertebral levels (12 of 18 patients).

All the patients underwent both posterior fossa decompression and syringosubarachnoid shunt procedures at the same operation. This procedure consisted of suboccipital craniectomy, removal of the arch of C1 (14 cases), and C2 if necessary (4 cases), opening the dura and arachnoid, microsurgical lysis of arachnoid adherences if present (4 cases) and lateral displacement of both tonsils to leave the exit of the fourth ventricle open. The obex was not plugged and the tonsils were not surgically removed in any of the cases. The dura was closed using a patch graft of autogenous fascia in a watertight fashion in all cases. A single-level laminectomy was then performed at the level of widest syrinx diameter and, after exposure of the dura, the operating microscope was used to facilitate fine dissection. Bleeding from extradural and pial vessels was rigorously controlled to prevent blood from reaching the subarachnoid space and causing meningeal irritation and postoperative arachnoiditis. In all cases, a Silastic T-tube catheter of the Pudenz type (Medtronic PS Medical, Goleta, Calif.) was used and inserted in both directions into the hydromyelic cavity through the dorsal root entry zone (DREZ). We then placed the distal end of the catheter, sliding it over the nearest dentate ligament to the anterior subarachnoid space of the cord without any suture for immobilization. The arachnoid and dura were closed in a single layer.

Results

All patients have been followed up for at least 36 months (range 36–54 months; mean 45 months). No operative mortality was encountered in these 18 patients. Mild temporary neurological deterioration was seen in the early postoperative period in two patients. However, within a few weeks, they had improved to their preoperative state. Six patients suffered aseptic meningitis of 2–5 weeks' duration, and four patients had transient dysesthesias. Transient cardiac or respiratory irregularities were not seen in this series. All patients were reassessed with neurological examination and MR imaging at various intervals after surgery. No patient has suffered recurrent or progressive neurological symptoms or signs and none has shown evidence of recurrence of syringomyelia on MR imaging (Fig. 2). All patients were reviewed in the outpatient clinic postoperatively and in the follow-up period, sixteen patients (88.9%) showed complete recovery of neurological deficits together with collapse of syrinx cavity, and two patients (11.1%) showed only radiological improvement without any change in neurological status. One of these two patients had hyperactive reflexes of the lower extremities, muscular atrophy and weakness of the upper extremities. In the other one, neck and shoulder pain diminished, but nystagmus, weakness of the left lower extremity with gait disturbance and poor dexterity of right hand were unchanged. Sixteen patients who had sensory loss (14 cases), cerebellar ataxia (9 cases), upper extremity weakness (7 cases), lower extremity weakness (1 case) and lower cranial nerve dysfunction (1 case) improved completely, and sensation returned to normal.

Discussion

Gardner and colleagues [6–8] postulated that delayed and incomplete embryonic opening of the outlet of the fourth ventricle, leading to retained communication of the cen-

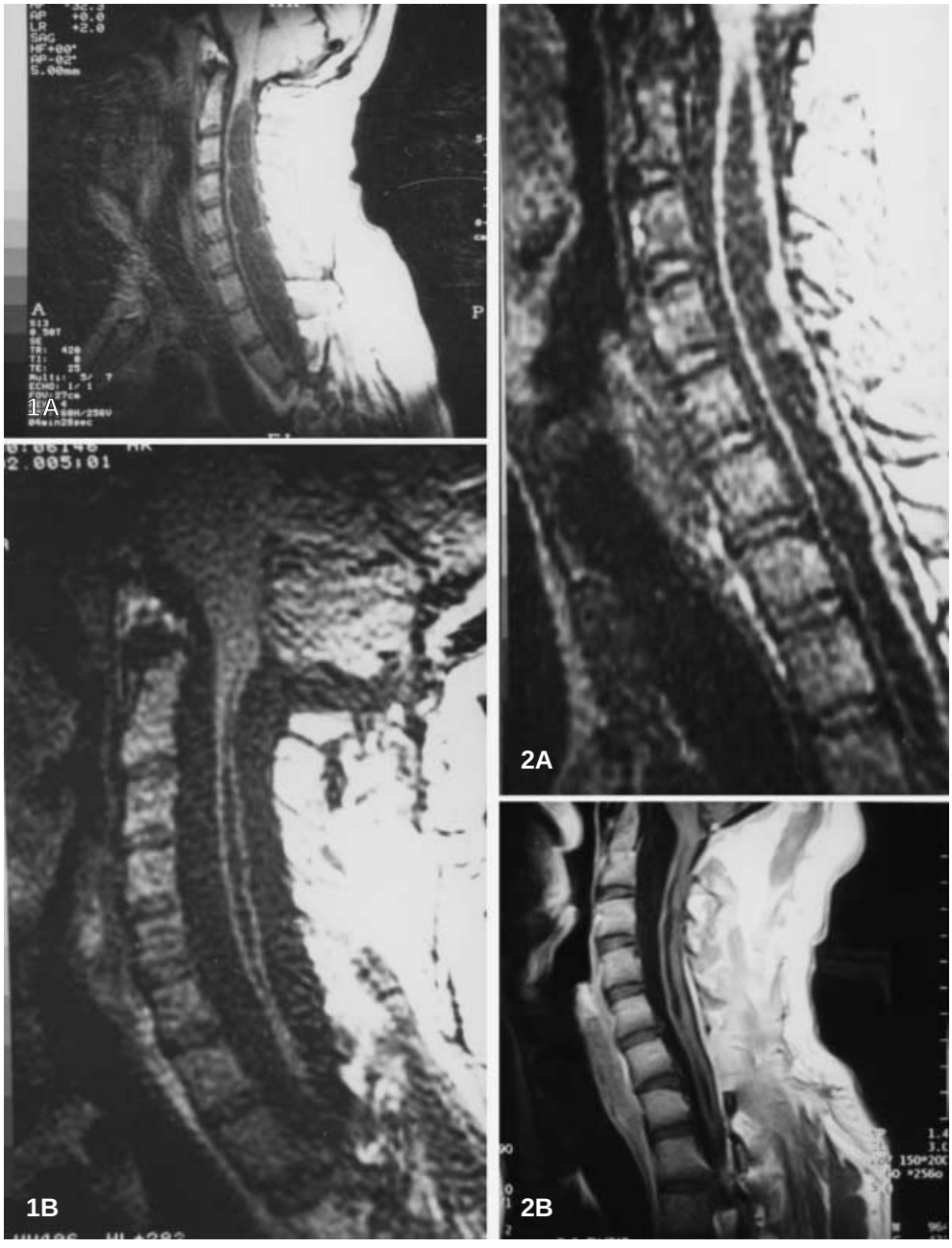

Fig. 1 **A** Sagittal T1-weighted magnetic resonance (MR) image of the cervical spine shows a wide syrinx with multiple septations. **B** Six months postoperatively it demonstrates collapse of the multiloculated syrinx within the cord

Fig. 2 **A** Sagittal T1-weighted MR image reveals a wide syrinx in the cervical region. **B** One year after surgery there is marked resolution of the cystic cavity

tral canal with the fourth ventricle, is the etiology of syringomyelia associated with Chiari malformation. This theory, based on a hydrodynamic mechanism, states that, with continuing partial obstruction of the outflow of CSF from the fourth ventricle through the central canal, each arterial pulse transmits a "water-hammer" effect into the syrinx causing development and progression of cord cavitation.

Williams [28–30] has proposed a modification of the hydrodynamic theory, with venous pressure changes rather than arterial pulsations as the driving force. The "cranial-spinal pressure dissociation theory" argues that intermittent abrupt swings of central venous pressure associated with coughing, sneezing or straining force CSF down an open channel from the ventricular system to the syrinx in patients with Chiari malformation.

More recently, MRI has become the procedure of choice for diagnosis of cranial and spinal malformations [13, 20]. Myelography and myelography assisted with computed tomography (CT), which were the techniques used in detecting syrinxes before MRI, have been the most commonly used radiological methods in the study of

syringomyelia. These have never been entirely reliable in demonstrating the syrinx cavity and its relationship to other intracranial abnormalities. Myelography probably misses approximately one-fourth of syrinxes [16]. High-resolution CT scanning with metrizamide offers an improvement over this, but may still miss some syrinxes [13]. MRI has provided an excellent noninvasive means of studying the anatomy of this malformation in the sagittal and axial planes, both before and after surgery, and has allowed the correlation of these findings with the clinical presentation and response to therapeutic intervention. Further advances in MRI technology, including dynamic imaging, may help elucidate the complex pathophysiology of patients with syringomyelia.

There is still considerable controversy about the indications for surgery and the methods of surgical treatment, so numerous surgical approaches have been advocated for syringomyelia associated with Chiari malformation. Although the etiology and natural history of this disease are variable, surgical treatment has usually been recommended for patients with deteriorating neurological function [23]. There have been several reports of the results of various surgical procedures, including suboccipital craniectomy with or without exposure of the fourth ventricle and plugging of the obex with muscle; upper cervical laminectomy for foramen magnum decompression; shunting from the syrinx to the subarachnoid space, pleura or peritoneum; intermittent percutaneous aspiration of the syrinx; terminal ventriculostomy; and ventriculoperitoneal and lumboperitoneal shunting [3, 9, 10, 18–20, 23–27, 31]. Although posterior fossa decompression remains the traditional surgical approach, performing the additional steps, such as plugging of the obex, shunting of the fourth ventricle, using lumboperitoneal devices or performing terminal ventriculostomy does not enhance the surgical treatment of this disorder [14, 16, 17, 25].

Shunting of syringomyelic cavities was introduced many years ago. After aspiration of the cyst by simple myelotomy, it became evident that any surgical opening into the subarachnoid space would not remain. Currently, with recent progress in microsurgical techniques and improvements in the materials used for shunts, the syringosubarachnoid shunt is receiving attention, with favorable results [11, 12, 23]. Entrance of shunt tube into the syrinx cavity has been advocated through the DREZ region to avoid injury to posterior columns with a midline myelotomy [22]. We used the Pudenz T-tube catheter, made of nonreactive Silastic, which was inserted through the DREZ region of the cord where the cavity was widest in diameter. Both ends of the T-drain tube were sufficiently tensile for dissection in both directions around intramedullary septae, which were encountered in some of the multiloculated syrinxes, yet sufficiently supple to prevent excessive force on cord tissue or intramedullary vessels. However, the principal factor avoiding dislodgement was the suture securing the catheter to pia or arachnoid; we placed the distal end of the tube sliding over the nearest dentate ligament to the anterior subarachnoid space of cord without any suture. Finally, closing the arachnoid and dura in a single layer should be performed, both for maintaining the integrity of the subarachnoid space and for prevention of subsequent arachnoiditis by avoiding the entrance of blood and debris at this site.

It is generally considered that, if there is clear progression of symptoms, especially in young patients, the sensible course would be to choose a surgical procedure designed to decrease the intramedullary tension. However, there are no objective criteria to suggest which procedure will produce the best surgical results in such cases, and comparisons are difficult because of the differences in surgical techniques, indications for operation, and criteria for the evaluation of results, including length of follow-up period. It should be pointed out that in the most successful surgical series, posterior fossa decompression relieves the symptoms of syringomyelia in only 50% of the cases [1, 18, 20]. The other group of authors have advocated syrinx shunting procedures as the initial approach, as they are easy to perform, are generally well tolerated and are associated with results that are easily the equal of those achieved by foramen magnum decompression [2–4]. Nevertheless, it is generally considered that hindbrain malformations have a relative contraindication to shunting procedures of the syrinx as the initial mode of treatment, because of concern that further herniation by spinal cord collapse and compromise of the brain stem will be precipitated [25, 31].

Recognition of septae within the syrinx on MRI, which may hinder drainage of the syrinx, has usually been considered a contraindication for syringosubarachnoid shunting. Although preoperative MR imaging revealed septated syrinxes in our four cases, these cavities collapsed after the shunting procedure (Fig. 1.). Thus, we agree with Lederhaus et al. and Vaquero et al. that free communication within the syrinx can exist despite septations seen on MR imaging [15, 25].

Conclusions

Fortunately, MR imaging is now widely used in daily practice, and we emphasize the importance of early recognition and treatment of this disease. The radiological improvement does not always correlate with the clinical recovery, as seen in our two patients, presumably because of irreversible cord injury having occurred before treatment.

In view of these facts, we strongly recommend combining foramen magnum decompression, to make the CSF pathways free, with a syringosubarachnoid shunt performed at the same operation as the most rational surgical procedure in the treatment of syringomyelia–Chiari complex.

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