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Surgical Treatment of Craniocervical Decompression without Chiari Malformation in Syringomyelia

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ABSTRACT

We aimed to describe a patient without Chiari malformation who was treated via craniocervical decompression and by creating cisterna manga with an autologous fascia graft, and who displayed a clinical and radiological improvement in the post-operative period. Syringomyelia is a chronic and progressive disease with cavitation and gliosis in the spinal cord. It is more common in adulthood and often involves the cervical region. Due to the fact that craniocervical decompression therapy is successful in syringomyelia with Chiari malformation, this surgical treatment is currently controversial in syringomyelia without Chiari malformation. A 33-year-old male applied to our clinic with numbness and weakness in his left hand that had lasted for 2 years, but his condition had worsened in recent months, with neck pain before over previous the 8 months and a walking disturbance that had appeared 7 months before. Syringomyelia was found between the C2 and T8 vertebrae levels on magnetic resonance imaging (MRI) scans. The patient was diagnosed with syringomyelia and underwent suboccipital decompression. Cisterna magna was created by duraplasty with a fascia lata graft.

Consequently, craniocervical decompression might be accepted as a treatment method of choice in patients with syringomyelia without Chiari malformation.

Key words: *Syringomyelia, Craniocervical decompression, Cisterna magna*

The term syringomyelia was derived from the word "syrinx" which in ancient Greece meant "tube". It was first used in 1824 by d'Angers. Several views exist on the mechanism of syringomyelia. The most widely accepted views are Williams' pressure gradient theory, Gardner's hydrodynamic theory and the theory of Oldfield, which holds holding that piston movements of cerebrospinal fluid (CSF) during the systole and diastole cause syringomyelia[2,3,7]. The first surgical operation for syringomyelia was myelotomy, reported in 1891 by Abbe and Coley. New approaches to the treatment of syringomyelia began with posterior fossa decompression and obstructing the obex, which were performed by Gardner in 1965[1,8]. Syringomyelia is a chronic and progressive disease with cavitation and gliosis in the spinal cord. It is more common in adulthood. It involves the cervical region in most patients[4,5,13,19]. Craniovertebral decompression and creating cisterna magna using dural graft has been accepted as a treatment in syringomyelia of the communicating type. The superiority of craniocervical decompression is being criticized, however in communicating syringomyelia without Chiari malformation and hydrocephalus.

CASE REPORT

A 33-year-old male applied to our clinic with numbness and weakness in his left hand which had lasted for 2 years, but whose condition had worsened in recent months. He had had neck pain over the previous 8 months and a walking disturbance had appeared 7 months before. The strength of the left limb was 5 and muscular atrophies were observed during the neurological examination. Additional findings during the examination were hypoesthesia with a cape-like distribution, an alteration of hot-cold discrimination in both limbs, enhanced bone-veter reflexes in the lower extremity, bilateral Babinski sign and ataxia. Syringomyelia was found between the C2 and T8 vertebrae levels on a magnetic resonance imaging (MRI) scan. The patient was diagnosed as syringomyelia and underwent suboccipital decompression. Total laminectomy was performed on the posterior edge of the foramen magnum and C1 vertebra. The dura was incised in a shape of "Y" and arachnoidal adhesions and fibrous bands were observed there. After these structures were

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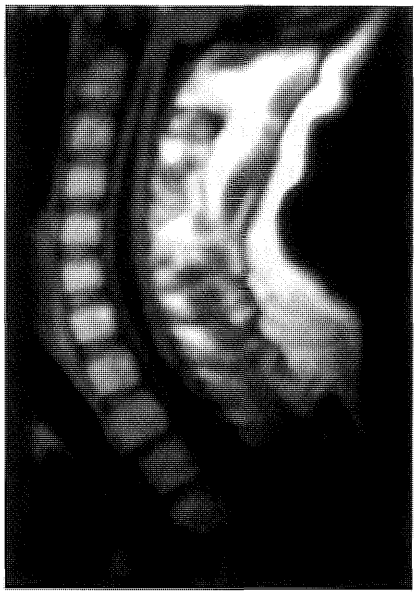

Fig. 1. Preoperative cervical MRI shows severe syringomyelia expanding to brainstem.

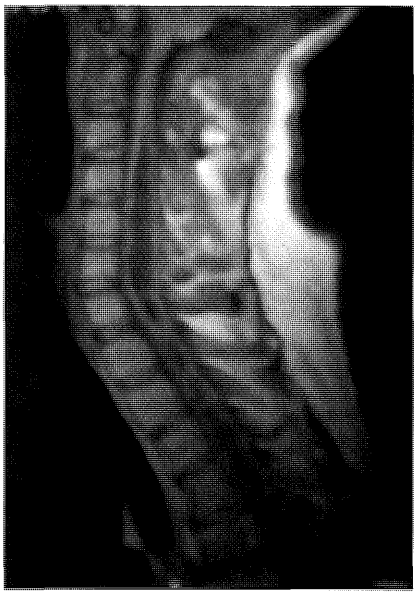

Fig. 2. Postoperative cervical MRI shows decreasing of the cavity.
Floor of the fourth ventricle was reached through retracted tonsils.

opened, tonsils were observed not herniated from the foramen magnum. The floor of the fourth ventricle was reached through retracted tonsils. Fibrous adhesions on the floor of the fourth ventricle were opened and it was observed that the obex was open. Cisterna magna was created by duraplasty with a fascia lata graft. No objective change was observed in the patient who described an improvement in the numbness in his left arm in the early postoperative period. An improvement was observed over a sequence of time in the neurological findings and the complaints of the patient. The complaints of the patient disappeared by the 6th postoperative month and by this time, a partial improvement was observed in the muscular atrophy. In addition, an improvement was observed radiologically in the syrinx cavity in parallel wit the clinical improvement.

DISCUSSION

Syringomyelia, as it relates to Chiari malformations, presumably develops as a result of obstructed CSF flow at the level of the craniocervical junction. The surgical restoration of CSF flow should then result in a reduction in the size of the syringomyelia. If the syringomyelia remains unchanged after posterior fossa decompression, it would seem that CSF flow has not been restored and consideration of more aggressive surgery is warranted (e.g., tonsillar coagulation, ventriculosubarachnoid stent placement). Clearly, all surgical decisions need to be made in the context of the clinical course of each patient. Syringomyelia may be detected with a careful neurological examination and its diagnosis can be confirmed by MRI scans. The central canal is part of CSF circulation path at the intra-uterine stage. Postnatally, however, the obex, which provides a communication between the fourth ventricle and the central canal, is closed. The obex may lead to syringomyelia if it is exposed to anomalous flows and pressures following different pathologies. Water hammer, which is also known as Gardner's hydrodynamic theory, has been widely accepted on this issue[10,12,14,17]. Another theory, Williams' pressure gradient theory, also explains the development of syringomyelia. Oldfield suggested that piston pressure originating from the heart during the systole and diastole through the foramen magnum into the cervical channel might cause syringomyelia[15]. Lee demonstrated that de-compressive operations were more effective in the restoration of CSF flow dynamics between the syrinx and subarachnoidal cavity than simple drainage of the syrinx cavity[11]. Sindou observed that craniocervical operations performed by protecting the arachnoidal membrane in patients with Chiari malformation type I yielded the best results with minimal complications[16]. Xie J. and colleagues used posterior fossa reconstruction and crania-vertebral decompression as a treatment modality in their series including 37 patients with Chiari malformation/Syringomyelia complex (CM-SM complex)[18]. This procedure consists of suboccipital craniectomy and C1 laminectomy (as well as C2 laminectomy, if necessary) and resection of arachnoidal adhesions. Dural defect was used to artificially reconstruct the cisterna magna. No shunt was applied to the syrinx. The follow-up period ranged between 6 months and 4.5 years. They observed symptomatic improvement in 31 patients (83.8%), stabilization of the symptoms in 5 patients (13.5%) and a worsening clinical condi-

tion in 1 patient at the end of the follow-up period. Findings observed on MRI scans were shrinkage of the syrinx, upward shift of the hindbrain and an artificial cisterna magna formation. Cranio-vertebral decompression and posterior fossa reconstruction were advised for restoration of the cranio-vertebral junction in the treatment of CM-SM complex according to the study by Xie[18].

Iskandar described 5 syringomyelia patients without hindbrain herniation and performed posterior fossa decompression in these patients. The symptoms disappeared in 4 of the 5 patients and syrinx dimensions shrank in all 5 patients. Thus, Iskandar concluded that craniocervical decompression might be beneficial for patients with syringomyelia resulting from Chiari-like physiopathological mechanisms without hindbrain herniation[9]. According to David two operative approaches are useful in the treatment of syringomyelia. They are posterior fossa decompression and shunting the syrinx (subarachnoidal, pleural and peritoneal shunts). He suggested that posterior fossa decompression be accepted as a preferred procedure for syringomyelia patients with Chiari malformation[6]. Syringomyelia may develop in a number of pathologies affecting CSF circulation, especially in Chiari malformation. Cranio-cervical decompression has been successful in syringomyelia patients with Chiari malformation. Given the fact that the underlying pathology with similar mechanisms in cases without Chiari malformation is the same as in cases with Chiari malformation, cranio-cervical decompression has been thought to be more effective. Howewer, no study in the literature has been found by us.

The patient treated in our clinic by artificial cisterna magna with an autologous fascia graft and craniocervical decompression displayed a clinical and radiological improvement in the postoperative period. As a consequence, in our opinion craniocervical decompression might be accepted as a treatment method of choice in patients with syringomyelia without Chiari malformation.

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