Surgical Management of Hydrosyringomyelia Related with Chiari I Malformation

Chiari I Malformasyonuna Bağlı Hidrosirengomiyelinin Cerrahi Tedavisi

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Abstract: A retrospective study was conducted on 48 consecutive patients with hydrosyringomyelia. Fortyseven of them had Chiari I malformation and one had stenosis of foramen magnum. The most common symptom was numbness in arms and the most common physical finding was dissociated sensory loss. The patients were operated by syringosubarachnoid shunt (12 cases), syringoperitoneal shunt (3 cases), posterior fossa decompression and syringosubarachnoid shunt(17 cases), posterior fossa decompression alone (13 cases), and ventriculoperitoneal shunting alone (3 cases). Fifteen patients had had previous surgery for their ilness. The majority of previous surgery was found to be syringosubarachnoid shunt or syringoperitoneal shunt. Our results and our literature review suggest that a posterior fossa decompression is the first option for the surgical treatment of hydrosyringomyelia related with Chiari I malformation.

Key Words: Chiari I malformation, hydrosyringomyelia, surgical management

Özet: Hidrosirengomiyelisi olan 48 hasta retrospektif olarak değerlendirildi. Kırkyedi olguda Chiari I malformasyonu, bir olguda ise foramen magnum stenozu vardı. En sık rastlanan yakınma kollarda uyuşukluk, ve en yaygın muayene bulgusu ise dissosiye his kusuru oldu. Hastalar sirengosubaraknoid şant (12 olgu), sirengoperitoneal şant (3 olgu), posterior fossa dekompresyonu ve sirengosubaraknoid şant (17 olgu), yalnız posterior fossa dekompresyonu (13 olgu) ve yalnız ventriküloperitoneal şant (3 olgu) yöntemleri ile tedavi edildiler. Onbeş hasta daha önce rahatsızlıkları sebebi ile ameliyat edilmişlerdi. Önceki ameliyatların çoğunluğunun sirengosubaraknoid şant veya sirengoperitoneal şant olduğu tesbit edildi. Sonuçlarımız ve literatür taramamız kranyovertebral dekompresyonun, Chiari I malformasyonuna bağlı hidrosirengomiyeli olgularında ilk cerrahi tedavi seçenek olduğunu düşündürdü.

Anahtar Sözcükler: Cerrahi tedavi, Chiari I malformasyonu, hidrosirengomiyeli

INTRODUCTION

Chiari I malformation is the main cause of cerebrospinal fluid filled cavitation within the spinal cord (11,12,16,17). It is characterized by a downward herniation of the caudal part of the cerebellum and/or medulla oblongata into the spinal canal. Among the various mechanisms proposed for hydrosyringomyelia, the blockage of cerebrospinal fluid (CSF) flow at the foramen magnum between the intracranial and intraspinal subarachnoid space is always pronounced (15). Although there is still considerable controversy about the methods of surgical treatment in hydrosyringomyelia, the practice of posterior fossa decompression (PFD) has been widely accepted (10,18,19).

PATIENTS AND METHODS

During the period between August 1988 through May 1996, 65 patients with hydrosyringomyelia presented to the Neurosurgery Department of the Cerrahpaşa Medical Faculty. The etiological factor was attributed to Chiari I malformation in 47, and to the foramen magnum stenosis in 1 patient. In the remainder who were excluded from the study, the spinal cord cavitation was due to spinal cord tumor, trauma or arachnoiditis. Twentynine of the 48 patients considered in this study were male. Their ages at operation ranged from 13 to 62 years (mean 39 years).

Clinical information was obtained by review of the patient records and operation reports. Follow-up data could be obtained from 36 (75 %) patients for an average length of 13 months (range 9-42 months).

RESULTS

Signs and symptoms: The duration of symptoms varied largely from few months to twelve years. As shown in Table I, the most common symptoms were numbness in arms (28 patients), burning pain (26 patients) and gait abnormalities (26 patients). Interestingly, the presenting symptom was only growth retardation in a 13-year old girl which was attributed to hydrocephalus related to the stenosis of foramen magnum (Figure 1). Thirtyfour patients exhibited the classic dissociated sensory deficit characterized by thermalgesic loss with preservation of the postural sense. The next most common physical

Table I: Symptoms and Physical Signs of 48 Patients with Hydrosyringomyelia.

Symptom	No. (%)	Physical Sign	No. (%)
Numbness	28 (58)	Dissociated sensory loss	34 (71)
Burning pain	26 (54)	Weakness	28 (58)
Difficulty in walking	26 (54)	Hand atrophy	18 (37,5)
Cervical and occipital pain	24 (50)	Babinski sign	14 (29)
Sensory loss	12 (25)	Gait abnormality	11 (23)
Weakness	12 (25)	Scoliosis	10 (21)
Back pain	6 (12,5)	Bowel and bladder dysfunction	8 (17)

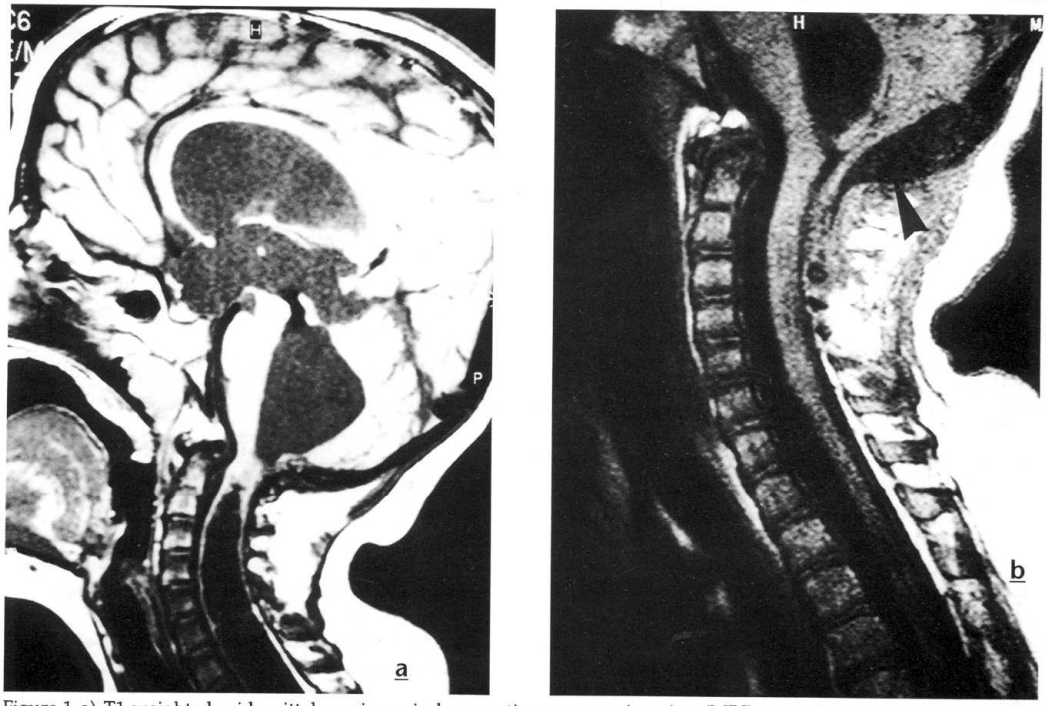

Figure 1, a) T1-weighted midsagittal craniocervical magnetic resonance imaging (MRI) scan showing an enlarged IVth ventricle and the beginning of a holocord hydrosyringomyelia. Note the stenosis at the level of foramen magnum, b) Postoperative T1-weighted MRI scan showing the artificially created cisterna magna by posterior fossa decompression and duraplasty (arrow). The syrinx cavity was completly disappeared.

signs were objective weakness (28 patients), hand atrophy (18 patients), and Babinski sign (14 patients). Thirtyfour patients had more than 1 symptom, and 40 patients had bilateral complaints.

Nine patients experienced symptom onset after a Valsalva manoeuvre or sudden physical strain such as violent coughing episode or minor fall.

Radiological findings: Preoperative imaging consisted of plain x-ray, computerized tomography (CT) (39 patients), and gadolinium enhanced magnetic resonance imaging (MRI) (32 patients) of the spine. Additionally, 28 patients underwent also CT scanning of the head. Plain x-ray films showed scoliotic changes in 10 patients. Associated severe basilar impression was found in only one patient. Moderate to sever ventricular enlargement was noted in 16 patients. The syrinx varied widely in length and location. Holocord hydrosyringomyelia was found in 11 patients. There were 10 cervical and 27 cervicothoracic syrinxes.

Surgical treatment: The treatment of the majority of the patients failed into one of the two main groups; drainage procedures or PFD (Table II). For many years, the first treatment of choice for hydrosyringomyelia was syrinx drainage by syringosubarachnoid shunt (SSS) or syringoperitoneal shunt (SPS). Because of high failure rate of both treatment modalities and large contribution of Chiari I malformation to hydrosyringomyelia, we have performed combined PFD and SSS in Chiari I related hydrosyringomyelia since 1991 (Figure 2). Seventeen patients were treated by this technique. Following the evident success of PFD and SSS, we perform now only PFD in patients with Chiari I related hydrosyringomyelia (Figure 3). Although in few cases lyophilized dura was used, we used generally autologous fascia lata for duraplasty. Only 3 patients with minor symptoms and enlarged ventricles were treated with ventriculoperitoneal (VP) shunting alone.

15 patients had had previous surgery for their illness at our institution. They were reoperated because of recurrence of symptoms and/or radiological evidence of continuous enlargement of their syringomyelic cavity. Previous surgery consisted of SSS in 7, SPS shunt in 4, and VP shunt in 4. Eleven patients which were previously treated by SSS or SPS showed initially a clinical improvement of their symptomss but deteriorated slowly 9 to 26 months after surgery. Seven of them were reoperated by PFD. For the remaining 4 patients, we performed decompression of the subarachnoid space by dissection of the arachnoid as well as the scar away from the pial surface of the cord to permit free passage of CSF. The spinal cord was densely adherent to the dura and deformed at the level of SSS or SPS (Figure 2). Pulsations of the spinal cord were generally attenuated at the level of scarring. With dissection of the arachnoid from the spinal cord and dura, normal pulsations of the cord reappeared and a new SSS was inserted. Four patients which were previously treated by VP shunt were reoperated by PFD (Table II).

Surgical results: Thirtysix patients were available for follow-up at an average of 13 months

Table II: Last Surgical Procedures Performed to Treat Hydrosyringomyelia in 48 Patients. The Corresponding Results of the Clinical and Radiological Outcome are Also Summarized.

Procedure	No	Clinical outcome *			Radiological outcome **		
		improved	worsened	unchanged	improved	worsened	unchanged
Shunting of the syrinx							
SSS	8	4	-	2	2	-	3
SPS	3	1		-	-	-	-
PFD							
+ duraplasty	13	10	-	1	8	-	2
+ duraplasty + SSS	17	11	1	3	10	-	3
VP shunt	3	1	-	-	1	-	-
Arachnoid dissection							
+ SSS	4	-	1	1	-	1	-

* only 36 patients were available for follow-up
** only 30 patients were available for follow-up

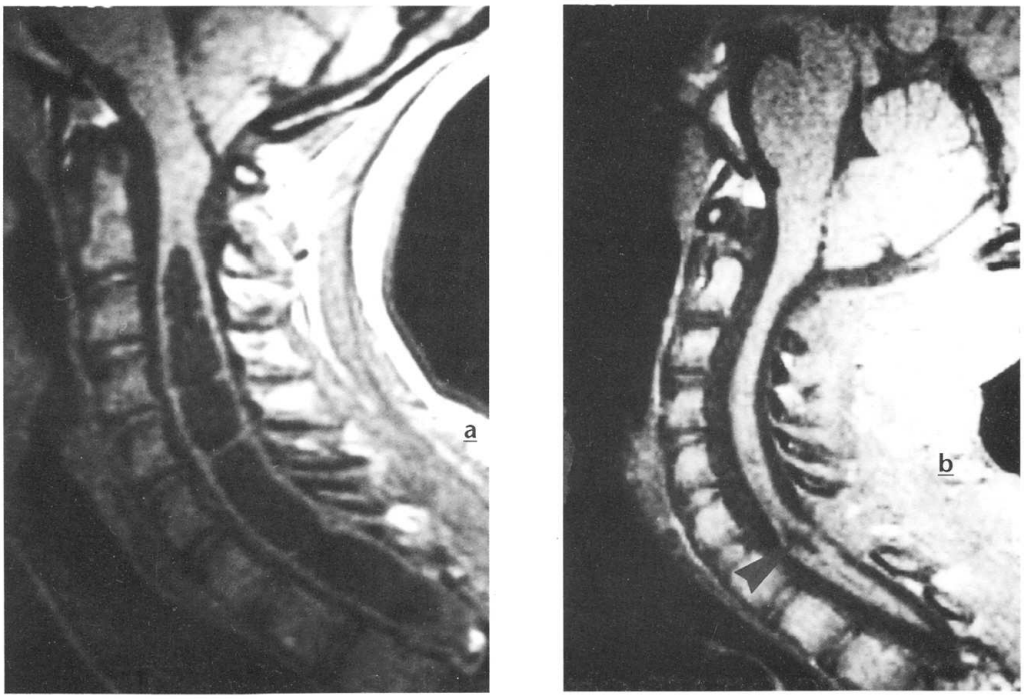

Figure 2, a) Preoperative T1-weighted MRI scans showing hydrosyringomyelia related with Chiari I malformation, b) 1 year after the PFD and SSS, no syringomyelic cavity was seen on T1-weighted MRI scans. The cervical spinal cord is largely deformed at the entry side of the syrinx catheter (arrow).

postoperatively (range 9-42 months). Twentyseven (75 %) of them reported lasting relief of one or more of their symptoms. Postoperative MRI were available in 30 out of 36 patients. The appearance of the syrinx had improved postoperatively in 21 (70 %) of them. Postoperative radiological results did not always correlate with clinical improvement. There were 4 patients with clinical improvement after surgery, but the syrinx appearance did not improve on MRI. As shown in Table II, PFD with or without SSS had the highest success rate on clinical or radiological improvement among all types of surgery performed to treat hydrosyringomyelia. There was no need for reoperation among the patients treated by PFD. Previous surgeries are not figured on Table II.

We have one mortality in a 14 year old boy which was operated on twice for basilar impression. The death happened after the second transoral operation performed to resect the dens. We have no serious complications except five cases of CSF leakage which were treated conservatively.

DISCUSSION

The term syringomyelia has been used to describe a cavitation of the spinal cord partially lined with glial tissue and independent of the central canal. A different entity, hydromyelia, is a dilatation of the central canal covered with ependymal lining. The clinical and therapeutic importance of this differentiation remains uncertain. Now, the term hydrosyringomyelia is widely used to describe all intramedullary cerebrospinal fluid collections corresponding either to a dilatation of the central canal or to an intraparenchymal cavitation (3,18). The accumulation of fluid within the spinal cord is not thought to be the primary manifestation of any disease process. Hydrosyringomyelia is a secondary process with many causes. Although it is known that some pathological conditions such as trauma, infection can be complicated by hydrosyringomyelia, in the majority of patients, hydrosyringomyelia is associated with craniovertebral anomalies and in 75 % of cases, it is associated with Chiari I malformation

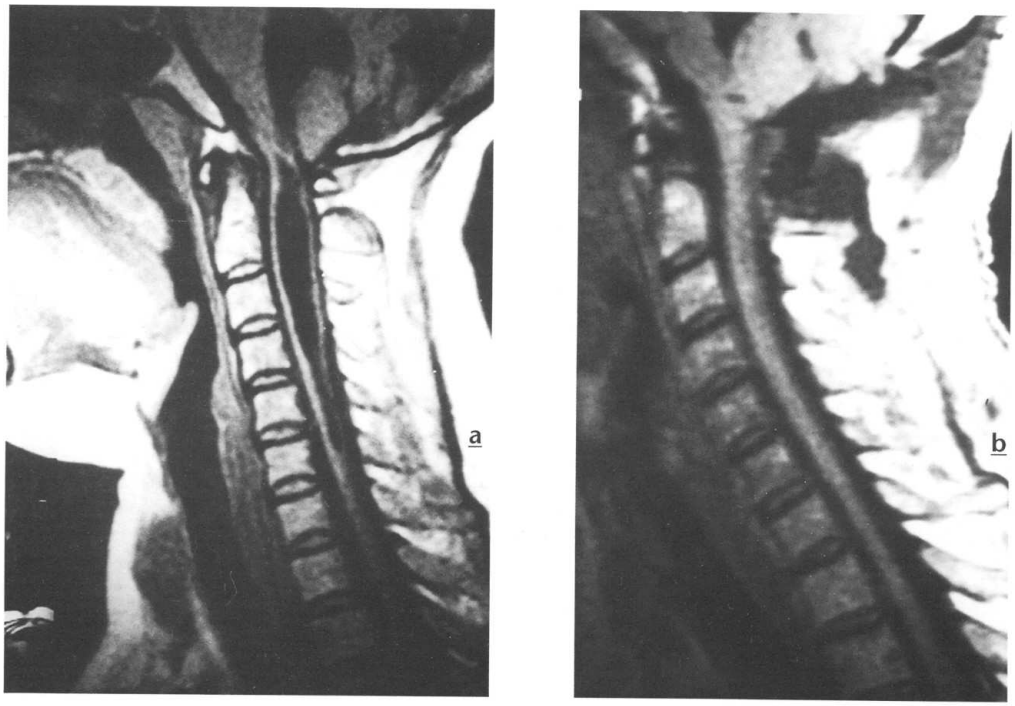

Figure 3, a) Preoperative T1-weighted MRI scans showing cervical hydrosyringomyelia related with Chiari I malformation, b) no syringomyelic cavitation was seen on T1-weighted sagittal MRI scan 18 months after the PFD.

(11,12,16). Pillay et al. (17) founded that 57 % of patients with Chiari I malformation had also hydrosyringomyelia. This was similar in our studies. Seventytwo percent of our patients had Chiari I malformation.

Several theories exists to explain the pathogenesis of this condition (2,7,22,23). A rational explanation for hydrosyringomyelia formation was made by Raftopoulos et al. (18).They classified different theories about hydrosyringomyelia formation into two groups: One based on the CSF entering the central canal from the fourth ventricle (7,22), and another based on formation of the hydrosyringomyelia by transparenchymal passage of CSF into the central canal from the subarachnoid space surrounding the spinal cord itself (1,2). They suggested that these theories might function simultaneously in association with a valvular mechanism. Recently, Nishikawa et al. (15) proposed, by a morphometric study of the posterior cranial fossa, that Chiari I malformation is most likely produced primarily by underdevelopment of the occipital enchondrium, possibly due to underdevelopment of the occipital somite originating from the paraaxial mesoderm. They suggested that overcrowding in the posterior cranial fossa due to a normal-sized hindbrain in the underdeveloped occipital enchondrium secondarily induces a downward herniation of the brain (15).

Signs and symptoms in hydrosyringomyelia-Chiari I complex are caused by the syrinx or by the Chiari I malformation. In our series, the majority of symptoms and signs were with a long duration and were related to the syrinx. The well-recognized dissociated sensory loss was found in 71 % of patients and it is related to the disruption of crossing spinothalamic fibers. For the burning pain which was encountered in 54 % of our cases, Milhorat et al. (13) suggested that it could be caused by a disturbance of pain-modulating centers in the dorsolateral quadrant of the spinal cord. They also founded that the substance P, which is a putative neurotransmitter

and pain related peptide, was abnormally at high levels in the spinal cord of syringomyelic patients (14).

Variability in the pathogenesis of hydrosyringomyelia has generated much controversy regarding its treatment. In surgical management, posterior fossa decompression, posterior fossa decompression with plugging of obex (Gardner's operation), SSS, SPS, thecoperitoneal shunt, excision of the filum terminale have been undertaken (5,6,8,9,21). The favorable results varies from 53,5 % to 75 % in different series (6,12,20). At our institution, for many years, we performed SSS or SPS for treatment of hydrosyringomyelia. Our results with these technics were obviously less favorable then the previously reported series. At reoperation, arachnoiditis caused by the shunt itself was a severe handicap for establishment of a normal CSF circulation which is undoubtedly one of the factor contributing to the syrinx formation (24). Tethering and deformation of the spinal cord at the syrinx catheter entry side was another complication of SSS or SPS. Especially for SSS, it is a well known entity that if the syrinx CSF pressure is lower than the subarachnoid space pressure, the shunt does not work, and there is no objective method for testing if SSS or SPS are patent. Now, the practice of PFD in the treatment of hydrosyringomyelia has been widely accepted. The goal of PFD is the restoration of a normal CSF circulation at the foramen magnum level (15). To avoid postoperative arachnoiditis and for preservation of artificially created cisterna magna, many varieties of this procedure have been described (10,18,19). Batzdorf (4) recommended to place a silastic shunt tube between the fourth ventricle and the subarachnoid space to create a communication from the fourth ventricle into the cervical subarachnoid space. Raftopoulos et al. (18) have obtained excellent results with hollowing of the cerebellar tonsils. The aim of both techniques is to create a free communication between the IVth ventricle and the subarachnoid space. In our practice of PFD, we perform a suboccipital craniectomy, with removal of the posterior rim of the foramen magnum and resection of the posterior lamina of C1, and when necessary of C2. The dura mater is then opened in a Y shaped pattern, with the arachnoid being left intact. The dural opening expose entirely the cerebellar tonsils in the cervical spinal canal. Generally, the free circulation of the CSF in the subarachnoid space is observed. But, if no CSF circulation is seen, the arachnoid is opened and dissected to the lateral margins of the dural opening. The cerebellar tonsils are gently lifted to provide a view of the floor of the fourth ventricle to ascertain that the CSF pathways are open. In these cases, arachnoid is not closed. For the dural graft, autologous fascia lata is generally used. Plugging the obex of the IVth ventricle which was proposed by Gardner was subsequently rejected by many authors. For Raftopoulos et al. (18), a free communication between the central canal and the fourth ventricle is essential for the restoration of normal CSF pathways. VP shunting is a simple alternative for hydrosyringomyelia (9). We have no broader experience with VP shunting in the treatment of hydrosyringomyelia related with Chiari I malformation. We have only one documented case with clinical and radiological improvement after insertion of a VP shunt.

Our study is not a prospective study for any chosen surgical treatment of hydrosyringomyelia. Here, we presented our experience about the surgical treatment of hydrosyringomyelia in Chiari I malformation. We conclude that shunting of the syrinx is a palliative therapy with high rate of recurrence. It can be associated with PFD as an adjoin. PFD is a valuable method for the treatment of this complex anomaly. If possible, preservation of the integrity of arachnoid layer is recommended to avoid further attachments. Better understanding of the pathophysiology of this anomaly is still needed for a perfect surgical strategy.

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