Acta Neurochir Suppl (2006) 99: 117–120

Printed in Austria

Idiopathic syringomyelia: case report and review of the literature

J. W. Lin, M. S. Lin, C. M. Lin, C. H. Tseng, S. H. Tsai, I. H. Kan, and **W. T. Chiu**

Division of Neurosurgery, Surgical Department, Municipal Wan-Fang Hospital, Taipei Medical University, Taipei, Taiwan

Summary

Syringomyelia is an uncommon disease that is caused most often by type I Chiari malformation, which develops in the hindbrain, and less frequently by other factors which are not limited to the hindbrain, including trauma, infection, or scoliosis. Idiopathic syringomyelia is rare. We present in this article a patient with idiopathic syringomyelia characterized by hypoesthesia and progressive weakness in the left lower limb. Decompression was attempted by means of laminectomy and a syringoarachnoid shunt. Motor, sensory, and bladder functions were monitored by the change in Japanese Orthopedic Association scores, which increased from 10 points preoperatively to 14 points 30 days postoperatively. This case demonstrates the effectiveness of surgical decompression in a patient with remarkable neurological deficit.

Keywords: Idiopathic syringomyelia; syringoarachnoid shunting; neurological deficit.

Introduction

The pathophysiology of syringomyelia (cavitation within the spinal cord) remains controversial [1–4]. Most cases have been associated with type I Chiari malformation. Others have been associated with typically noncongenital conditions, including scoliosis, arachnoiditis, and trauma. However, idiopathic syringomyelia is rare. Syringomyelia is more easily detected nowadays because of the availability of spinal magnetic resonance imaging (MRI). Before this technique became popular, the nonspecific and highly versatile signs and symptoms of syringomyelia – such as chronic pain, hyperhidrosis, hypertension, limb paresthesia, sensory loss, progressive weakness, and, in some cases, ascending paralysis [3, 5] – resulted in syringomyelia being easily overlooked, especially in patients with minor signs and symptoms. Consequently, most patients were diagnosed so late in the course of the disease that its neurological sequelae were irreversible.

Fortunately, the neurological deficits associated with syringomyelia can be reversed if decompression is carried out earlier in the course of the disease. In reviewing the literature on treatment options for this patient, we found that few articles had been reported about the nature and treatment of syringomyelia in Asia, especially in Taiwan. So, we would like to present a patient with idiopathic syringomyelia, his clinical course, treatment procedure and prognosis.

Case report

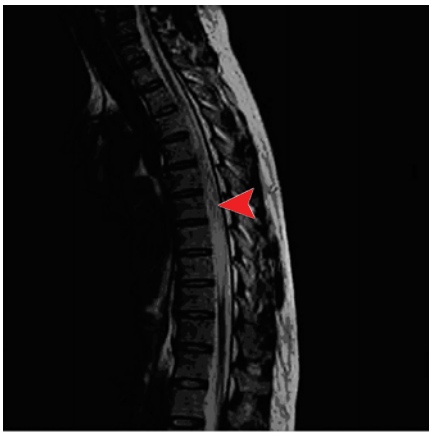
A 35-year-old male dentist had experienced gradually progressive weakness in his left leg since 5 years ago. The weakness exacerbated in the last 2 months and he had to use a cane when walking. A neurological examination revealed weakness in the left leg, especially dorsiflexion of the big toe, and hypoesthesia of pinprick and light touch sensation below the T7 dermatome on the left side of the body. The patient had no history of trauma, spinal tumor, or any evidence of spinal arachnoiditis. MRI studies revealed an inflated cord with a large cavity in the thoracic region of the spinal cord (T2–T9) (Fig. 1). Due to the recent exacerbation of his symptoms, we arranged surgical decompression for him.

Surgical technique

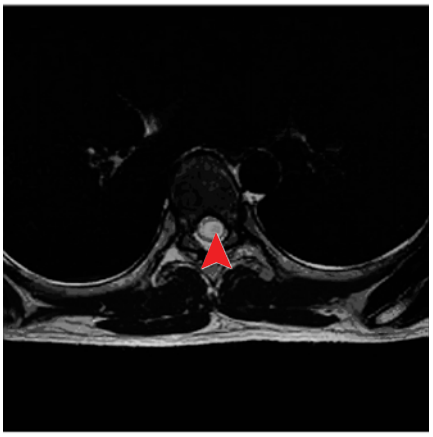
The first surgical procedure was T6–T8 laminectomy to expose the dura mater. The next procedure was to open the dura and a 1-mm midline myelotomy was performed with a microknife to create an opening for the cavitation. The fluid of the cavity was drained out and pressure was relieved, then we inserted an elastic catheter (18-gauge spinal stent: 2 cm in length, 1.24 mm in diameter) through this opening into the cavity in a cephalad direction and the caudal end placed in the dorsolateral subarachnoid space. We secured the tubing to the dura with 6-0 Prolene suture (Fig. 2).

Evaluation and follow-up of neurological function

The Japanese Orthopedic Association (JOA) scoring system was used to evaluate the neurological condition, specifically the peripheral motor activity (upper and lower extremities: 4 points, each), sensory activity



a



b

Fig. 1. Preoperative MRI, sagittal view (a) revealing a syrinx (arrow) with long thoracic extension (T2–T9). Axial view (b) revealing a large syrinx (arrow) with massive compression to cord (flattened appearance)

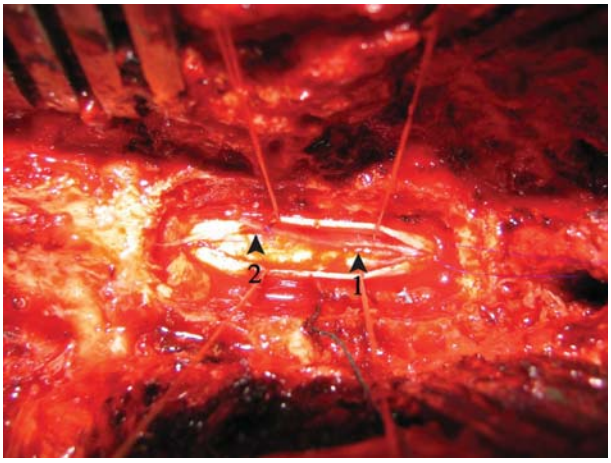


Fig. 2. Intraoperative findings. A bone window was made between T6 lamina and T8 lamina. A midline dural opening was made. Dorsal midline myelotomy was performed to open the syrinx. The shunt tube was placed into the syrinx (*arrow 1*) and the caudal end is of the catheter was secured to the dura with a 6-0 Prolene suture (*arrow 2*) (placed in the dorsolateral subarachnoid space)

Table 1. *Summary of the JOA scale grades for cervical myelopathy [23]**

Variable	Grade
I. Motor function	
a. Upper extremity	
unable to feed oneself	0
unable to handle chopsticks; able to eat with a spoon	1
handle chopsticks with much difficulty	2
handle chopsticks with slight difficulty	3
normal	4
b. Lower extremity	
unable to stand and walk by any means	0
unable to walk with a cane or other support on a level surface	1
walk independently on a level surface but need support on stairs	2
capable of fast walking but clumsy	3
normal	4
II. Sensory function	
a. Upper extremity	
apparent sensory loss	0
minimal sensory loss	1
normal	2
b. Lower extremity	
apparent sensory loss	0
minimal sensory loss	1
normal	2
c. Trunk	
apparent sensory loss	0
minimal sensory loss	1
normal	2
III. Bladder function	
a) Urinary retention and/or incontinence	0
b) Sense of retention and/or thin stream	1
c) Urinary retention and/or pollakiuria	2
d) Normal	3

* Cumulative normal grade in a healthy individual is 17.

(upper and lower extremities: 2 points each; trunk: 2 points), as urinary bladder function (3 points) before operation and on postoperative days (PODs) 7, 14, and 30 (Table 1). The recovery rate was calculated using the following formula:

Recovery rate (%) = (postoperative score − preoperative score) ÷ (17 − preoperative score) × 100.

Results

The JOA score was 10 points before surgery, 11 on POD 7, 12 on POD 14, and 14 on POD 30, and recovery

Table 2. *JOA score and recovery rate: pre-op and post-op*

Parameter	Pre-op	POD 7	POD 14	POD 30
JOA score	10	11	12	14
Muscle power (L foot dorsiflexion)	3	4−	4−	4
Recovery rate	–	14%	28%	57%

rates were 14, 28, and 57%, respectively. Strength in the left extensor hallucis longus muscle rose from grade 3 preoperatively to grade 4 by POD 30 (Table 2).

Discussion

Syringomyelia is characterized by dilation of the central canal in the spinal cord and results in neurological deficits because of gradual compression of the spinal cord. Its clinical presentation includes progressive weakness in the upper and/or lower extremities, diminished sensation, and chronic pain. People with this disorder are frequently misdiagnosed because of vague signs and symptoms. A delay in the diagnosis of this disorder can result in irreversible neurological deficits. These deficits can be reversed, however, by early and effective decompression of the spinal cord.

Current theories about the mechanism for the formation of syringomyelia are controversial. Possible mechanisms include perforation of the foramen of Magendie resulting in the subsequent expansion of the central canal (the Gardner theory) and the ''ball-valve'' effect of obstruction of the foramen magnum associated with type I Chiari malformations (the Williams theory) [6, 7]. Obstruction of the cerebrospinal pathway results in a pressure gradient [3, 5], which is relieved when pressure is dissipated through potential spaces. Eventually, this results in the creation of an intramedullary cavity [3, 5].

Conditions leading to syringomyelia can develop within the hindbrain or elsewhere [3, 8]. The most common hindbrain lesion that results in syringomyelia is type I Chiari malformation, which develops within the foramen magnum [9–13]. Spinal cord trauma, the second leading cause, can also lead to meningeal fibrosis and syringomyelia [3, 8, 14–16]. Other causes of syringomyelia do not necessarily involve the hindbrain such as spinal cord tumor, infection, kyphosis, and a reaction to iophendylate (Pantopaque) [3, 1, 4, 17]. In our patient, syringomyelia was classified as idiopathic after all these potential factors had been excluded.

The treatment strategy for patients with this disorder varies with the extent of disease progression. Some patients show no signs or symptoms during disease progression for many years; such patients may be treated conservatively [6, 7, 18–21]. Syringomyelia has been reported to resolve spontaneously with conservative treatment in a few cases [1, 6, 18, 21, 22], but some patients deteriorate progressively with that approach. Such patients may better be treated with surgical decompression, comprising myelotomy, syringosubarachnoid or syringopleural shunt, and spinal cord transaction [3]. In our patient, severe spinal cord compression induced several symptoms of syringomyelia, i.e. left foot drop and left-sided hypoesthesia, which worsened over 1 month. We performed a shunting procedure, which was followed by improvement in muscle strength in his left leg and improved sensation on the left side of his body. It has been reported that once neurological deficits develop in patients with syringomyelia, they cannot be completely reversed with surgery [1, 22]. However, our experience supports the concept that surgical decompression may be of some help for neurological deficits in patients with syringomyelia.

References

1. Kyoshima K, Bogdanov EI (2003) Spontaneous resolution of syringomyelia: report of two cases and review of the literature. J Neurosurg 53: 762–769
2. Iwasaki Y, Hida K, Koyanagi I, Abe H (2000) Reevaluation of syringosubarachnoid shunt for syringomyelia with Chiari malformation. J Neurosurg 46: 407–415
3. Goldstein JH, Kaptain GJ, Huy MD *et al* (1998) CT-guided percutaneous drainage of syringomyelia. J Comput Assist Tomogr 22: 984–988
4. Hilton EL, Henderson LJ (2003) Neurosurgical considerations in posttraumatic syringomyelia. AORN 77: 135–156
5. Sgouros S, Williams B (1996) Management and outcome of posttraumatic syringomyelia. J Neurosurg 85: 197–205
6. Andreas K, Thomas N, Helge T (2001) Spontaneous resolution of idiopathic syringomyelia. Neurology 57: 1519–1520
7. Sherman JL, Barkovich AJ, Citrin CM (1987) The MR appearance of syringomyelia: new observations. AJR Am J Roentgenol 148: 381–391
8. Sgouros S, Williams B (1995) A critical appraisal of drainage of syringomyelia. J Neurosurg 82: 1–10
9. Ball MJ, Dayan AD (1972) Pathogenesis of syringomyelia. Lancet 2: 799–801
10. Gardner WJ (1965) Hydrodynamic mechanism of syringomyelia: its relationship to myelocele. J Neurol Neurosurg Psychiatry 28: 247–259
11. Gardner WJ, Angel J (1959) The mechanism of syringomyelia and its surgical correction. Clin Neurosurg 6: 131–140
12. Williams B (1970) Current concepts of syringomyelia. Br J Hosp Med 4: 331–342
13. Williams B (1980) On the pathogenesis of syringomyelia: a review. J R Soc Med 73: 798–806
14. Caplan LR, Norohna AB, Amico LL (1990) Syringomyelia and arachnoiditis. J Neurol Neurosurg Psychiatry 53: 106–113
15. Stoodley MA, Gutschmidt B, Jones NR (1999) Cerebrospinal fluid flow in an animal model of non-communicating syringomyelia. J Neurosurg 44: 1065–1076
16. Schurch B, Wichmann W, Rossier AB (1996) Post-traumatic syringomyelia (cystic myelopathy): a prospective study of 449 patients with spinal cord injury. J Neurol Neurosurg Psychiatry 60: 61–67
17. Lee TT, Alameda GJ, Camilo E *et al* (2001) Surgical treatment of post-traumatic myelopathy associated with syringomyelia. Spine 26: 119–127

18. Sun JC, Steinbok P, Cochrane DD (2000) Spontaneous resolution and recurrence of a Chiari I malformation and associated syringomyelia: case report. J Neurosurg 92 [Suppl] 2: 207–210
19. Jack CRJ, Kokmen E, Onofrio BM (1991) Spontaneous decompression of syringomyelia: magnetic resonance imaging findings. Case report. J Neurosurg 74: 283–286
20. Santoro A, Delfini R, Innocenzi G *et al* (1993) Spontaneous drainage of syringomyelia: report of two cases. J Neurosurg 79: 132–134
21. Yeager BA, Lusser MA (1992) Spontaneous resolution of idiopathic syringomyelia: MR features. J Comput Assist Tomogr 16: 323–324
22. Bindal AK, Dunsker SB, Tew JM Jr (1995) Chiari I malformation: classification and management. J Neurosurg 37: 1069–1074
23. John KR, Paul RC (2003) Cervical laminoplasty: a critical review. J Neurosurg 98: 230–238

Correspondence: Wen-Ta Chiu, Division of Neurosurgery, Surgical Department, Municipal Wan-Fang Hospital, Taipei Medical University, No. 111, Section 3, Hsing-Long Rd, Taipei, Taiwan. e-mail: wtchiu@tmu.edu.tw