Long-term outcome of idiopathic hypertrophic thoracic pachymeningitis

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Abstract Idiopathic hypertrophic spinal pachymeningitis (IHSP) is a comparatively rare disease characterized by hypertrophic inflammation of the dura mater and clinical symptoms that progress from local pain to myelopathy. We report a case of IHSP followed up for 20 years in a 46-year-old man. Expansive laminoplasty was performed in 1991, and this case has been previously reported by a co-author. After 17 years, the patient's gait disturbance returned. Physical examination and imaging confirmed IHSP that had developed into syringomyelia at the T2–L1 conus level. This case was diagnosed as adhesive spinal arachnoiditis due to pachymeningitis caused by syringomyelia. T1–T4 laminectomy, a syringo-subarachnoid shunt (S–S shunt), and L2–L3 laminectomy were performed. The patient again developed dysesthesia and gait disturbance 3 years after the second operation. Most reports of IHSP have limited their focus to short-term follow-up after initial treatment with no long-term results. At present, there are only five reports referring to long-term results of greater than 5 years. All but one case needed additional surgery. To the best of our knowledge, this is the first case in which syringomyelia occurred in a patient with IHSP. It is

important to note that syringomyelia may be a cause of symptom recrudescence during long-term follow-up in IHSP patients.

Keywords Hypertrophic pachymeningitis · Long-term outcome · Syringomyelia · Thoracic spine

Introduction

The concept of hypertrophic pachymeningitis was introduced in 1869 by Charcot et al. [3]. Idiopathic hypertrophic pachymeningitis is a chronic, progressive, diffuse, inflammatory fibrosis of the dura mater [13–15, 22]. Idiopathic hypertrophic spinal pachymeningitis (IHSP) is a rare inflammatory disorder that leads to spinal cord compression. However, the long-term evolution of this form of pachymeningitis is unclear. One of the co-authors has previously reported this case [8]. Here, the subsequent long-term outcome of this case is reported.

Case report

First examination, operation, and outcome

The initial details of this case have been previously reported [8]. Briefly, a 28-year-old male presented with a greater than 2-month history of numbness of the lower extremities. He experienced a general onset of spastic gait and mild back pain in May 1991. His symptom completely subsided in response to intravenous steroids (methylprednisolone sodium succinate, 1,000 mg × 3 days). However, he was hospitalized again because of increasing dysesthesia of the trunk and the lower extremities, spastic gait, and

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Fig. 1 MRI findings. **a** Sagittal T2-weighted image shows the high signal area revealing syringomyelia at the T3–L1 level. Expansive laminoplasty has been performed at T5–L2. **b** Sagittal T2-weighted image shows the high signal area revealing syringomyelia to the

conus level. The spinal stenosis at the L2–L3 level is shown. **c** Axial T2-weighted image at the T5–T6 level. The low signal area around the high signal area is shown. The low signal area is thickened dura and thinning spinal cord

urinary retention. Expansive laminoplasty was performed at T5–L2 in November 1991 [16].

Second examination, operation, and outcome

He showed significant improvement in muscle strength and dysesthesia. He exhibited no spasticity 5 years after surgery, but he sometimes felt numbness of the lower extremities. His symptoms subsided completely in response to intravenous steroids at that time.

In September 2008, he was hospitalized again because of increasing dysesthesia of the trunk and lower extremities, spastic gait, and urinary retention. Neurologic examination revealed spastic paraplegia and an L1 sensory level with diminished position and vibration sensation. The muscle power was approximately grade 3/5 in the lower extremities.

Magnetic resonance imaging (MRI) showed high-intensity lesions from T2 to the conus level of the spinal cord on T2-weighted imaging (Fig. 1a, b). A low signal area around the high signal area was shown on axial T2-weighted imaging at the T5–T6 level (Fig. 1c). Moreover, postoperative spinal stenosis was also seen at the level adjacent to the earlier expansive laminoplasty (Fig. 1a) [11].

Therefore, a syngo-subarachnoid shunt (S–S shunt) with T1–T4 laminectomy and L2–L3 laminectomy were performed in November 2008. An S–S shunt tube (Birthel, Create Medic Co., Yokohama, Japan) was placed from the syringomyelia at the T4 level to the subarachnoid space at the T2 level.

The histopathological appearance of the biopsied dura mater showed hypertrophic change with slight infiltration

Fig. 2 Histopathological appearance. Histopathological examination shows the thickened dura with slight inflammatory infiltration consisting of neutrophils (HE stain). Scale bar 100 μ m

of mononuclear inflammatory cells (Fig. 2). In comparison to the histopathological findings at the first operation, the hypertrophic dura mater remained. However, the infiltration of mononuclear inflammatory cells decreased.

The syringomyelia was decreased in size on MRI 3 months after the second operation. Over the next 3 years, the patient's dysesthesia and gait disturbance returned.

Discussion

The IHSP is extremely rare, with few described cases, though the first report by Charcot [3] dates back to 1869. Recently, since the advent of MRI, the frequency of IHSP

Table 1 Clinical characteristic of IHSP patients followed up more than 5 years

Case	Reference	Age/sex	First operation	Other operation (duration ^a)	Cause of subsequent operation	Outcome	FU (years)
1	Guidetti et al. [5]	15/M	T9–T11 laminectomy/durotomy	C4–T3 laminectomy/durotomy (3 years)	Recurrence	Recovered	14
2	Adler et al. [1]	47/F	T8–T11 laminectomy/duraplasty	C3–C7 laminectomy T1–T7 laminectomy (4 years)	Recurrence	Recovered	7
3	Khadilkar et al. [12]	42/F	C1–C4 laminectomy/duraplasty	None	Not applicable	Fluctuating	5
4	Ito et al. [7]	67/M	T6–T8 laminectomy/duraplasty	T3–T4 laminectomy/duraplasty (5 years) C6–T1 laminoplasty/duraplasty (7 years)	Recurrence	Recovered	10
5	Present case	28/M	T5–L2 laminoplasty	T1–T4 laminectomy/L1–L3 laminectomy/S–S shunt (17 years)	Syringomyelia	Recovered	20

FU follow-up period, M male, F female, C cervical, T thoracic, L lumbar S–S syringo-subarachnoid, N/A not available

^a The time from the first operation to the subsequent operation

has increased. Patients display a wide age range, from 15 to 77 years (median 46.0 years) [17]. Males are affected more frequently than females (3:2). It commonly involves the cervical and thoracic dura [1, 5, 7, 12] and presents initially as progressive radicular symptoms, with muscle weakness and atrophy as the second stage, and paraplegia, loss of bladder function, bowel disturbance, and respiratory distress caused by intercostal and diaphragmatic denervation as the third stage [2].

The IHSP is a chronic inflammatory disease and thus is usually treated medically. Steroid therapy represents the first choice and is often effective for IHSP [12, 18, 19]. Besides steroid treatment, radiotherapy [3], azathioprine therapy [1, 12, 15], and cyclophosphamide therapy [15] have been used. Although surgery is not curative, marked improvements in neurological symptoms can be expected [8]. In the present case, the symptoms improved after both the first and second operations.

Most reports of IHSP have limited their focus to short-term follow-up after initial treatment with no long-term results. The natural history of cranial hypertrophic pachymeningitis has been well documented. The course of the disease follows one of the three patterns: (1) sustained remission, (2) relapse with corticosteroid resistance, or (3) relapse with corticosteroid dependence [6, 21]. In contrast, the course of the spinal form has been less studied, since it is uncommon, but it is believed to run a progressive course. Of the 96 patients with IHSP gathered from the literature by Ito et al. [7], the recurrence rate was 11% over a mean follow-up of 1 year and 4 months, and the factors for recurrence were at least one positive inflammatory sign before surgery and the duration of the mean follow-up period. Therefore, long-term follow-up is extremely important to clarify the prognosis in IHSP. To

the best of our knowledge, there are only five cases, including our case, with long-term results of greater than 5 years (Table 1). In the process of the spread of the lesion, most cases needed additional operations. In only one case (Case 4) did symptom exacerbation subside with medication.

Symptom recrudescence is often caused by IHSP recurrence during follow-up. In the present case, however, syringomyelia caused the symptom recrudescence 17 years after the first operation, and a second operation was needed. Syringomyelia is a rare disease in which a syrinx develops in the spinal cord. Arnold-Chiari malformation, spinal trauma, spinal surgery, spinal cord tumor, and arachnoiditis are known underlying causes of syringomyelia. Although we have performed many laminoplasties, we have yet to see syringomyelia after laminoplasty [9, 10, 16]. In addition, chronic slight inflammation of the dura persisted, as seen on the histopathological findings. Therefore, this case was diagnosed as adhesive spinal arachnoiditis due to pachymeningitis caused by syringomyelia. Adhesive spinal arachnoiditis is a chronic inflammatory process in the pia mater of the spinal cord. It was reported that the peripheral margin in a case of pachymeningitis was enhanced on MRI and was unusually close to the highly vascularized arachnoid mater [4]. Oonishi et al. [20] found that this disease process was not just confined to the dura, but also involved the arachnoid mater and pia mater (trigeminitis). These findings suggest that the location of inflammation in IHSP may be the dura, as well as the arachnoid mater. Therefore, during long-term follow-up, IHSP appears to cause the development of syringomyelia. Further IHSP cases are necessary to resolve questions regarding the causes of symptom recrudescence and the clinical course.

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