Idiopathic Hypertrophic Spinal Pachymeningitis

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Abstract

A 63-year-old man revealed a four-month history of muscle weakness of the lower limbs, hypoesthesia of the L5 and S1 area and ischuria. On MRI, the spinal cord was compressed by an encircled mass, which showed hypointensity on T1- and T2-weighted images with gadolinium enhancement at the Th11 to Th12 vertebra. Because of the rapid progression of myelopathy, posterior decompression was performed and idiopathic hypertrophic spinal pachymeningitis (HSP) was finally diagnosed. The patient’s neurological signs markedly improved with postoperative corticosteroid treatment. Idiopathic HSP is a clinical emergency and early surgical intervention is essential to prevent irreversible damage to the nervous system.

Key words: hypertrophic pachymeningitis, spinal, surgical intervention, improvement, pathology


Introduction

Idiopathic hypertrophic spinal pachymeningitis (HSP) is a rare disease characterized by chronic inflammatory fibrosis of the dura mater with an undetermined pathogenesis (1, 2). The clinical symptoms of HSP gradually develop from local pain to a bedridden state due to compressive myelopathy (1, 3). Diagnosis of HSP is frequently difficult with a radiographic approach such as magnetic resonance imaging (MRI), and most previous cases have required a pathological diagnosis (4). Though the standard treatment for idiopathic HSP is corticosteroid therapy (4-6), idiopathic HSP often causes progressive myelopathy and surgical decompression is required to prevent neurological sequelae. We experienced a patient with idiopathic HSP who experienced a good recovery from the symptoms and signs of thoracic myelopathy following urgent surgical intervention.

Case Report

A 63-year-old man presented with a four-month history of lower back pain, numbness and paresthesia of his lower limbs. He was aware of an abnormal sensation in both feet and muscle weakness of the lower limbs gradually progressed. He showed gait disturbance and advancing paraplegia. On admission, he had no fever and the findings of physical examinations were normal. A neurological examination with the manual muscle test revealed the following weaknesses: right iliopsoas muscle (score=4), left tibialis anterior muscle (score=4), left triceps surae muscle (score=4) and left extensor hallucis longus muscle (score=4). Numbness and hypoesthesia of the L5 and S1 area were also detected. The patient could not maintain a standing position and had lost sensation of the urges to urinate and defecate. Meningeal signs were not detected. On cranial MRI, including T1-, T2-, diffusion- and T1-weighted gadolinium (Gd), the images showed no abnormalities. A thoracic MRI showed that his spinal cord was compressed by an encircled mass in the extra-spinal space at the Th11 to Th12 vertebral level. The lesion showed hypointensity on both the T1- and T2-weighted images and was homogenously enhanced on the T1-weighted Gd (Fig. 1). An intradural extramedullary tumor was initially suspected, but a definitive diagnosis could not be obtained based on the MRI findings. He was transferred to our hospital because the muscle weakness of

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his lower limbs and ischuria were worsening. The blood
tests revealed an elevation of the inflammatory marker C-
reactive protein (CRP) to 2.23 mg/dL, and an erythrocyte
sedimentation rate (ESR) to 100 mm/hour. The serological
testing was negative for myeloperoxidase anti-neutrophil cy-
toplasmic antibodies (ANCA) and proteinase-3-ANCA using
the enzyme immunoassay method. Testing for anti-nuclear
antibody was negative, and the angiotensin-converting en-
zyme and IgG, levels were normal. A serological test for
syphilis, an interferon-γ assay test for Mycobacterium tuber-
culosis, and blood cultures were all negative. The findings
of his cerebrospinal fluid (CSF) were as follows: leukocytes,
20 /mm³ with 18 mononuclear cells and 2 polymorphonu-
clear leukocytes; total protein concentration, 243 mg/100
mL; and glucose, 49 mg/100 mL (plasma glucose was 105
mg/100 mL). Mycobacterium tuberculosis DNA in the CSF
was not detected via nested polymerase chain reaction. The
cytology in the CSF was Class II. Because of the rapid pro-
gression of myelopathy, the patient underwent surgical re-
moval of the lesion with posterior decompression at the
Th10 to L1 vertebral level. Intraoperatively, the dura mater
was found to be diffusely thickened and compressing the
underlying spinal cord. The hypertrophic dural lesion was
partially removed with a laminoplasty to the extent of the
bony decompression. The pathological microscopic findings
showed dense fibrous tissue with an infiltration of mononu-
clear cells such as lymphocytes and plasma cells. Histio-
cytes were also seen with some multinucleated giant cell
formation without findings of vasculitis. Specific staining
for fungi, bacteria, and mycobacteria were negative. Clusters
of CD20 and CD3-positive lymphocytes and IgG-positive
plasma cells were detected, but IgG-positive cells were not
detected (Fig. 2). These histological features were consistent
with the diagnosis of HSP. The patient was finally diag-
nosed with idiopathic HSP after excluding secondary causa-
tive diseases.

After surgical intervention, the patient’s clinical symp-
toms markedly improved, and 1 mg/kg/day of prednisolone
(PSL) was orally administered to control the residual lesion,
particularly that located in the ventral space. After postop-
erative PSL treatment, the CRP level and ESR of the blood
returned to normal levels, and the CSF protein and cell
counts also decreased. On the follow-up thoracic MRI four
weeks after beginning corticosteroid treatment, the enhanced
lesion on T1-Gd that was located in the ventral space of the
removal region showed improvement. At the 8-week follow-
up, the patient’s limb weakness, sensory loss, and bladder
and rectal function had improved, and he was able to walk
independently.

**Discussion**

HSP is an uncommon cause of nerve root and spinal cord
compression and typically involves the cervical and thoracic
spinal cord levels (4, 7, 8). The signs and symptoms of HSP gradually develop from those of nerve root compression to spinal cord compression (1, 7), and almost all patients that suffer from HSP experience progressive paraparesis (4). Our patient also exhibited weakness of the lower extremities and bladder dysfunction due to compression of the spinal cord at the Th11 to 12 vertebral levels. Mikawa et al. (4) reported that the average interval from when the patient first suffers pain or numbness to the development of motor paralysis ranges from 3 days to 5 years with a mean of 10 months. Therefore, the clinical course of our patient was regarded as rapid progression because he experienced paraplegia and ischuria 4 months after the onset of the other symptoms.

The etiology of HSP is unknown in most cases and is thus classified as idiopathic HSP (9). In our patient, we could not identify any predisposing illnesses, such as infectious disease or autoimmune diseases, despite our thorough investigations. MRI is the most useful diagnostic modality for idiopathic HSP (10-12). The lesion is characteristically hypointense on T2-weighted images with contrast enhancement on T1-Gd (11, 12). However, differentiating HSP from an epidural abscess or a spinal tumor is difficult based on imaging alone without a dural biopsy or removal surgery (4, 13). Pai et al. (12) also reported that the MRI features of HSP are characterized by a long extramedullary mass of low T2 signal intensity with peripheral enhancement of the lesion. MRI of our patient showed circumferential thickening of the dura compressing the spinal cord, which showed hypointense signals on T1- and T2-weighted images and linear homogenous enhancement on post-contrast images. These MRI findings are relatively rare for patients with HSP (12). Therefore, differentiating HSP from other critical diseases, including a spinal tumor located in the subdural space, was difficult in the present case based on the MRI findings alone.

Treatment of HSP relies on surgical decompression and immunosuppressant therapies (4, 5, 8). Surgical decompression of the spinal cord is recommended for HSP to alleviate

Figure 2. Pathological findings of the surgically removed specimens. Hematoxylin and Eosin staining sections revealed abundant fibrotic tissues infiltrated mainly by mononuclear cells such as lymphocytes as well as some plasma cells and histiocytes (A, B). Some multinucleated giant cell formations were also seen (B, arrow). Clusters of CD20-positive cells (C), CD3-positive cells (D), and IgG-positive plasma cells (E) were seen, whereas IgG4-positive cells (F) were rarely observed (all original magnifications ×100).
neurological symptoms and sequelae (2, 5, 10). The surgical removal of the posterior surface of the dura beyond the apparent limit of the lesion is useful for decreasing recurrences (7, 14). Urgent surgical decompression and duroplasty were essential in our patient to prevent irreversible damage to the nervous system, because our patient exhibited rapidly advanced signs of myelopathy and the etiology was not defined. We removed as much hypertrophic dura as surgically possible, but excising the lesion from the ventral portion of the spinal cord was difficult. Moreover, pathological findings of the removed specimens revealed abundant infiltration of various inflammatory cells including multinucleated giant cell formation.

Immunosuppressant therapy has recently been regarded as the mainstay treatment to alleviate the sequelae and recurrence even after early decompression surgery (5, 14). Postoperative corticosteroid therapy is also effective for controlling the residual hypertrophic lesion, particularly when the lesion is located in the ventral space as in our case. Corticosteroids therapy was initially administered at a dose of 1 mg/kg/day that was carefully tapered. However, no standard protocols for the dose and duration of corticosteroids have been proposed for use in HSP (15). Nonetheless, initial combined treatments have been reported to be successful, however, recurrent patients with corticosteroid resistance or even dependence have also been reported (6, 8, 16). Interestingly, Mikawa et al. (4) and Ito et al. (14) reported that patients with active inflammatory signs, such as fever, increased white blood cells, increased CRP levels, and increased ESR, before surgical intervention had a notably higher recurrence rate and a worse prognosis. In our patient, the CRP and ESR levels were elevated at the time of deterioration before surgery. We needed to control the inflammation of the residual pachymeningitis that was located above and below the removed lesion, and we are currently monitoring the patient for possible signs of recurrence.

In conclusion, we experienced a patient with idiopathic HSP who recovered from rapidly progressing myelopathy following urgent surgical intervention. We suggest that surgical intervention is crucial in cases with rapid deterioration of compressive myelopathy to prevent irreversible damage to the nervous system.

The authors state that they have no Conflict of Interest (COI).

References