Idiopathic hypertrophic spinal pachymeningitis (IHSP) is a comparatively rare disorder characterized by marked inflammatory hypertrophy of the dura mater, with subsequent neurological deficits resulting from the compression of adjacent structures. We present two cases of IHSP with description of etiologies, diagnoses, managements and clinical outcomes.

Case Report

Case 1

A 55-year-old woman visited to our hospital complaining of back pain, progressive paraparesis, both leg numbness, and voiding difficulty in July, 2009. These symptoms had been start-
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opening dura mater, we found yellowish tumor-like lesion and resected partially for biopsy. Pathologic examination revealed a chronic nonspecific inflammation and fibrosis (Fig. 3). Staining for cytokeratin was negative. These findings were compatible with diagnosis of idiopathic hypertrophic spinal pachymeningitis. The patient was treated with dexamethasone postoperatively for 6 days, and symptoms were gradually improved and laboratory data including ESR, CRP turned to be normal. Two months later after the surgery, follow-up MRI showed decreased dural thickening from C7 to T8 (Fig. 4B). The patient had undergone oral prednisolone and kept improved state for 4 months.

However, 4 months later, the patient was admitted to our hospital again presenting with back pain, both leg numbness, and gait disturbance as much as caine-assisted gait. MRI revealed increase of diffusely thickened dura comparing to the previous MRI. ESR (30 mm/hr) and CRP (7.7 mg/L) levels were mildly elevated. She was treated with steroid (solumedrol) pulse therapy and followed by maintenance treatment with oral prednisolone as well as rehabilitation. One month later, MRI revealed decrease of dural thickening. The patient recovered almost completely from the neurologic symptoms slowly and could walk unaided eventually.

Case 2

A 45-year-old woman presented with fever and quadriplegia in November 2000. One day before, she had a spinal fusion operation due to lumbar spinal stenosis and degenerative disc herniation in another hospital. Because she awoke slowly from the general anesthesia and, showed quadriplegia and respiratory difficulty immediate postoperatively, she was transferred to our hospital. At the time of admission, she had mild fever (37.9°C) and mild leukocytosis (WBC count 9,890/ul). Her ESR and CRP were also slightly elevated. However, ordinary blood culture was negative. Initial MRI showed an extramedullary mass compressing the upper cervical spinal cord posteriorly from foramen magnum to C4 level (Fig. 5). On suspicion of epidural
hematoma, emergent decompressive laminectomy from C1 to C4 was performed. There was no definite epidural and subdural hematoma, but 8 mm-thickened dura were noted. Through dural excision, we achieved the decompression of spinal cord. Subsequently, we took multiple biopsy specimens from the dura. Postoperatively the patient was treated with dexamethasone for 2 weeks. In spite of the surgery, neurological symptoms including weakness showed no improvement.

**DISCUSSION**

Idiopathic hypertrophic pachymeningitis (IHP) is a comparatively rare inflammatory disease characterized by hypertrophic inflammation of the dura mater. This rare disease is mostly found intracranially. The spinal form of IHP is rarer and first described by Charcot and Joffroy with its first name as “Pachymeningitis hypertrophica cervicalis” in 1869. Charcot and Joffroy described three stages; in the first stage, patients experience local and radicular pain. The second stage can present signs of nerve root compression. In addition, in the third stage, patients eventually suffer from spinal cord compression.

Our patients have experienced all stages. The etiology of hypertrophic spinal pachymeningitis (HSP) is still unknown in most cases, although various associated diseases have been recognized, including trauma, metabolic disease, sarcoidosis, infections such as syphilis, tuberculosis, HTLV-1, meningococcal meningitis, granulomatous sinusitis, mastoid sinusitis, and fungal infection. Also, HSP has been associated with various autoimmune disease including Wegener's granulomatosis, rheumatoid arthritis, orbital pseudotumor, multifocal fibrosclerosis, and mixed connective tissue disease. Intra- and extraspinal steroids, spinal anesthesia, and contrast medium for myelography also have been implicated as a cause of HSP. A lot of cases have unknown etiology and are classified as IHSP.

In our cases, we could not find any predisposing illnesses such as infectious diseases or autoimmune diseases in spite of the through investigations.

On MRI, IHSP has been showed as dura-based mass of low signal intensity on T1- and T2-weighted images extending over multiple vertebral levels. In addition, predominantly peripheral enhancement pattern is a highly suggestive of IHSP. This pattern represents more intense enhancement in a peripheral zone of active inflammation than in a central zone of fibrosis. Linear dural enhancement pattern was known to have better response to steroid therapy than those with nodular pattern because of its less fibrosis and more vascularity. In our cases, typical radiologic and pathologic features observed in the literature can be shown.

The management of IHSP is controversial. For IHSP, surgical decompression by laminectomy and excision or incision of the involved dura has been recommended. Recently, some case reports witnessed that biopsy with steroid therapy can decrease the thickness of the dura by serial MRI and can improve neurologic deficit. If decompressive surgery is necessary, laminoplasty have been recommended instead of extensive laminectomy because the former could reduce chronic back discomfort and would enhance spinal stability. Others have emphasized postoperative radiotherapy, immunosuppressive therapy such as azathioprine, and cyclophosphamide. Among our two patients, one recovered from the weakness, the other did not recover from the quadriplegia status. The authors thought that the cause of the latter was acute spinal cord injury maybe from neck extension during induction of anesthesia.
meanwhile, the former from chronic myelopathy.

CONCLUSION

Although IHSP is a rare disorder, the clinical and radiological findings are characteristic. Unaccountable motor weakness and gadolinium-enhancing thickened dura which can be shown by spinal MRI probably lead to a diagnosis. For the management of IHSP, we advocate surgical decompression with subsequent prolonged steroid therapy as our cases had.

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