Idiopathic Hypertrophic Spinal Pachymeningitis: A Case Report

Idiopathic hypertrophic pachymeningitis (IHP) is a rare, chronic nonspecific and granulomatous inflammatory disorder of the dura with unknown etiology. The diagnosis can be established by open biopsy and exclusion of all other specific granulomatous and infectious diseases. We report a typical case of spinal IHP occurring in a long segment of cervical and thoracic dura from C6 to T8. The patient was a 56-yr-old female, who had been suffering from pain on her upper back and both arms for 3 months and recent onset motor weakness of both legs. During the 9 months of follow-up period, she experienced the improvement of her neurologic symptoms with combined therapy of partial excision and corticosteroid medication. Since early surgical intervention and subsequent pulse steroid therapy are mandatory for this disease to avoid irreversible damage of nervous system, the identification of this unique disease entity is essential on frozen diagnosis. A few cases have been reported in Korean literature.

Key Words: Meningitis; Idiopathic Hypertrophic Pachymeningitis; Spine; Immunohistochemical Study; Chronic Nonspecific Inflammation; Granulomatous Disease, Chronic

INTRODUCTION

Idiopathic hypertrophic pachymeningitis (IHP) is an extremely rare inflammatory and fibrotic disease of the dura (1). According to Mikawa et al., the first case of IHP had been recorded by Amercrombie in 1918 (2). However, before this record, Charcot and Joffroy (3) first described it in 1869 by relation to spinal meninges, and intracranial forms were described some later (4, 5). IHP is commonly occurs in the spinal dura in the 6th and 7th decades and cranial hypertrophic pachymeningitis is rarer than the spinal form, of which involving pattern is either solitary or multiple and diffuse or nodular (1, 2). Early reports were generally attributed to syphilis (4, 5) or tuberculosis (6). However, in 1949, Naffziger and Stern reported an idiopathic case of hypertrophic pachymeningitis involving the dura of the posterior fossa with compression of the underlying brain stem (6). Since then, most reported cases have been idiopathic and their frequency has increased since the advent of CT and MRI.

Spinal IHP commonly involves cervical and thoracic dura (1, 2, 4-10). Mikawa et al. investigated the thickness of the dura with 29 cases of IHP and observed a range of 1.5 to 12 mm, with a mean of 6.9 mm (2). Due to the resulting nerve encasement and ischemia, paraparesis and neuropathies are common clinical manifestations.

The diagnosis of IHP in majority of cases was not made before pathologic examination (1, 3, 11, 12). Pathologically, IHP is characterized by chronic nonspecific and granulomatous inflammation with fibrosis of the dura. The diagnosis can be established by open biopsy and after exclusion of all other granulomatous and infectious diseases. The treatment of choice is early surgical decompression and/or corticosteroid therapy before irreversible damage of the nervous system (13).

We describe clinicopathologic features of a typical case of spinal IHP with a review of literature. From Korean literatures, we found a couple of reports of IHP (14, 15) and a case of hypertrophic pachymeningitis caused by tuberculosis (16).

CASE REPORT

The previously healthy 56-yr-old female had been suffered from pain in her upper back and both arms for 3 months before hospitalization. These symptoms had been progressively aggravated until motor weakness of both legs and voiding difficulty were developed. The family and past histories were noncontributory. On physical examination, her motor system was intact but sensory on both legs was decreased. Deep tendon reflexes were increased. Ankle clonus was positive on both sides. Vital signs and other laboratory data were normal except low hemoglobin level (Hb and Hct, 8.9
g/dL and 35.2%, respectively) and elevated ESR (patient: 120 mm/hr, normal: <25 mm/hr). Serologic tests for HIV antibody and VDRL were negative.

Spine anteroposterior and lateral view revealed straightening of lumbar spine curvature with mild degeneration and mild scoliosis to the right side. Computed tomography (CT) scan revealed the circumferential thickening of the dura with compressed spinal cord. On magnetic resonance imaging (MRI), these thickened dura showed iso- or low signal intensity on T1-weighted image and hypersignal intensity on T2-weighted image with homogeneous and strong enhancement (Fig. 1).

Under the impression of possible spinal cord tumor or inflammatory lesion of the spinal cord, surgical exploration was done. Although the surgeons tried to decompress long segments as possible as they could with extensive laminectomy from C6 to T8, but partial excision of the lesion was carried out. The frozen diagnosis was reported as chronic inflammation.

Fig. 1. (A) T2-weighted sagittal image shows linear elongated mass-like lesion with dark signal at lower cervical canal from C6 (arrows). (B) Homogeneously enhanced circumferential lesion (arrows) is seen with cord compression in post-gadolinium enhanced T1-weighted axial image. (C, D) Post-gadolinium enhanced T1-weighted sagittal images show a well demarcated, elongated shaped mass (arrows) at cervical (C) and thoracic (arrows) (D) spinal canal.
Pathologic findings

Resected specimens were long segments of thickened dura and epidural adipose tissues which showed reddish discoloration (Fig. 2). The thickness of the dura measured up to 3-4 mm.

Microscopically, the lesion was composed of thickened dura diffusely infiltrated by lymphoplasma cells, some lymphoid follicles with or without germinal centers and frequent granulomas with central necrosis and relatively small giant cells (Fig. 3). The giant cells had two to twenty nuclei and some of them were Touton type. The central necrosis was characteristically blue and karyorrhexis-type. These granulomas were not associated with blood vessels. Fibroblastic and myofibroblastic proliferations were also noted. These inflammatory reactions were also noted in the epidural adipose tissue. Neither neutrophilic nor eosinophilic infiltration was observed.

Neither bacterial nor fungal organisms were detected on Gram, Gomori methenamine silver, periodic acid Schiff and Ziehl-Neelsen stains. On immunohistochemical study, majority of the infiltrated lymphocytes were CD3 immunoreactive T cells with CD68 immunoreactive histiocytes. Rare CD79a-positive B cells and plasma cells were intermixed. Ki67 index was low (less than 1%). Polymerase chain reaction (PCR) study for tuberculosis was negative.

Follow-up

Most neurologic symptoms, such as back pain, lower leg weakness and voiding difficulty were markedly improved after the operation and the patient was subsequently treated with prednisolone for 6 weeks with tapering. Thoracic spine MRI with full contrast enhancement after one month revealed that posterior part of the intradural mass was almost removed from the lower cervical level to the mid-thoracic level (Fig. 4). However, the ventral portion of the dura showed residual enhancing mass. On sagittal image, there was fluid collection in the deep muscle layer at the level of C7 vertebral body.
which had communication with spinal canal, suggesting postoperative pseudomeningocele due to extensive laminectomy.

During the 9 months after the surgery, she was relatively doing well without recurrence of symptoms.

**DISCUSSION**

IHP is a rare disease entity which is characterized by 1) unknown etiology, 2) specific dural involvement, 3) unique radiologic features i.e., diffuse thickening of the dura with encasement of central nervous system, and 4) unique histopathology; i.e., chronic nonspecific and granulomatous inflammation of dura, although extremely rare cases with involvement of adjacent brain parenchyma were reported (17).

In review of literatures, cranial IHP showed that the ratio of male to female was 3:2 and the age ranged from 14 to 75 (9). Most patients were in their sixties (mean age, 61.8 yr) (9). In spinal IHP (2), the patients' gender and range in age were similar (15 to 77 yr) to cranial IHP albeit lower mean age (46.0 yr) (n=52). Spinal IHP commonly caused progressive paraparesis (7, 8, 10), while the most common symptom of the cranial IHP were headache (100%), followed by multiple cranial nerve palsies (78%) (10). Almost all the patients with spinal IHP had motor paralysis (2). Our patient also had motor weakness of extremities along with voiding difficulty and back pain. Kao et al. (10) speculated that the cord atrophy might be caused by the compromising of feeding radicular arteries rather than by direct compression. However, since our case showed markedly reduced cord space by the mass-effect of the lesion, direct cord compression would also be considered as one of the main causes of cord atrophy.

Mikawa et al. (2) subdivided IHP into two groups: 1) those with inflammatory signs including fever, increased erythrocyte sedimentation rate, leukocytosis, and increased CRP (P group) and 2) those without inflammatory signs (N group). They analysed and suggested that P group had worse prognosis than N group. Our case had initial elevation of ESR, which was normalized after surgery.

Myelography and gadolinium-enhanced MRI of spinal IHPs commonly demonstrate thickening of the dura and diffuse cord atrophy. If there are the evidences for the spinal root or cord compression in multiple levels just by thickened dura without any bony changes on X-ray, IHP should be suspected (2). On MRI, IHPs usually showed a hyperintense border along a large hypointense lesion in T2-weighted images, attributing the decreased signal to dense fibrous tissue and the hyperintense signal to an inflammatory infiltrate or hypervascularity (13, 17). In addition, Hatano et al. pointed that the responses to corticosteroid treatment varied according to the different types of dural enhancement. They stated that the patients with linear dural enhancement had better response to the corticosteroid treatment than those with nodular enhancement. Nishioka et al. (18) reported that high accumulation of thallium-201 represented inflammatory activity of the IHP on single-photon emission CT (SPECT) scan. The SPECT scan pattern reflects the fluctuation of symptoms and is useful in evaluating the reactivation of the disease after steroid therapy that was not detect-
ed by MRI. Our case showed circumferential thickening of the dura compressing spinal cord from C6 to T8, which showed iso- or low signal intensity on T1-weighted image and hypersignal intensity on T2-weighted image with homogeneous and strong enhancement on MRI. The enhancement was linear along the long axis of the dura, but did not show different intensity to discriminate whether it is center or periphery on T2-weighted image. This radiologic feature suggested diffuse inflammatory infiltration and the histopathologic features coincided with it.

Hatano et al. (1) described that the course of the disease followed one of three patterns, that is, 1) sustained remission, 2) relapse with corticosteroid resistance or 3) relapse with corticosteroid dependence. Pulse corticosteroid therapy usually provided significant relief, and reducing the daily corticosteroid requirement and avoiding side effects were important issues in corticosteroid-dependent relapsing patients.

The etiology of IHP is still unknown, although various associated diseases, such as Wegener’s granulomatosis (20), metabolic diseases (21), sarcoidosis (22), rheumatoid arthritis (23, 24), sinusitis (9, 25, 26), infections such as syphilis (27), tuberculosis (6, 28), fungal (29) and meningococcal meningoencephalitis (20), and autoimmune mechanism (13, 30) have been suggested. In Korea, a case of hypertrophic pachymeningitis (20), tuberculosis (6, 28), fungal (29) and meningococcal meningoencephalitis. Spine 1994; 19: 620-5. However, we could not find any predisposing illness from our patient including any specific infections or autoimmune diseases.

The most valuable diagnostic procedure for IHP is open biopsy and the differential diagnosis can be easily made by histopathologic examination. In our case, tuberculous pachymeningitis was excluded by the absence of caseating granulomas, negative bacilli on Ziehl-Nielsen staining, and negative result of PCR for tuberculosis. The bluish karyorrhectic type necrosis seen in IHP is different from caseation necrosis of the tuberculous type. Wegener’s granulomatosis was excluded by the absence of vasculitis and pulmonary symptoms or signs. Fungal infections were ruled out by absence of evidences for fungal organisms and result of cerebrospinal fluid examination. Rheumatoid pachymeningitis, sarcoidosis, or other infections such as syphilis were excluded by laboratory data as well as clinicopathologic features. As the previous reported cases (32), multiple meningiomas or meningioma en plaque can be differential diagnosis, however, they can be also easily ruled out by biopsy findings.

Decompression surgery usually leads to significant neurological improvement, although thickening of the dura may be improved by subsequent steroid therapy over rather a long period. Surgical resection is essential for making a diagnosis as well as relieving the inflammatory and fibrous encasement and ischemic damage by hypertrophied dura. After partial decorticating operation with corticosteroid therapy, symptoms and signs of our patient were much improved during the 9 months of follow-up.

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