ever, associated osteolytic lesion adjacent to IHSP has barely been described previously. We report a case of IHSP involving cervical spine with an osteolytic lesion.

CASE REPORT

A 58-year-old female visited our institution with progressive weakness of both lower extremities and clumsiness of both hands. These symptoms had been developed 3 years ago and worsened gradually. The patient received a hysterectomy 10 years ago because of benign myoma and total thyroidectomy 2 years ago because of follicular thyroid carcinoma. There was neither noticeable history of trauma nor contributable family medical history.

On physical examination, there was slight motor power weakness (grade 4+/5) in both lower extremities. The deep tendon reflexes were normal and the ankle clonus was absent. There was no fever or leukocytosis. However, the erythrocyte sedimentation rate was slightly elevated. The patient was not taking any medications that could cause neurologic symptoms.

MRI scans revealed an epidural lesion involving the C6-C7 level with low signal intensity on T1 and T2 weighted images and non-enhancement on T1 weighted-enhanced images. The lesion extended over multiple levels and was thought to be compressing the spinal cord. A differential diagnosis was considered, including neoplastic disease, infection, and autoimmune disorders.

The lesion was surgically excised, and histopathological examination revealed thickened fibrous tissue with minimal inflammation. The diagnosis of idiopathic hypertrophic spinal pachymeningitis was confirmed.

After surgery, the patient's neurologic symptoms improved partially. The motor power grade of both lower extremities returned to normal, but mild weakness in the right lower extremity persisted. The patient reported no significant improvement in her symptoms.

In conclusion, idiopathic hypertrophic spinal pachymeningitis is a rare disorder that can present with various neurologic symptoms depending on the location of the lesion. Surgical intervention is necessary to relieve neurologic compression and prevent further deterioration of symptoms.

Key Words: Idiopathic hypertrophic spinal pachymeningitis · Osteolytic lesion · Mimicking giant cell tumor · Decompressive surgery · Differential diagnosis.
Idiopathic Hypertrophic Spinal Pachymeningitis with an Osteolytic Lesion

TK Jee, et al.

ESR was slightly elevated to 38 mm/hr (reference range: 0–27 mm/hr). Initial computed tomography (CT) scan showed an osteolytic mass-like lesion involving base of the C6 spinous process, which caused spinal canal stenosis. Radiologic report suggested neoplastic disease such as giant cell tumor, which has the characteristics of an osteolytic neoplasm (Fig. 1). MRI was carried out after CT scan and revealed an epidural mass involving the dorsal aspect of cervical spinal canal from C5 to C7 level, with low signal intensity on T1 and T2 weighted images and non-enhancement on T1 weighted-enhanced images. There was abnormally high signal intensity of the spinal cord on T2 weighted image at C6 level, suggesting myelopathy (Fig. 2). The patient underwent surgical exploration, with a complete hemilaminectomy of C5, C6, and C7 with left side approach. There was yellow-colored, thickened fibrous tissue over the dura mater. The mass was grossly totally removed, and decompression of spinal cord was achieved. Dexamethasone was prescribed temporarily to reduce inflammatory reactions. Five days after surgery, patient discharged without complications related with surgery and the symptoms were improved but not completely recovered. Pathologic examination revealed fibrotic pachymeninges with no significant inflammation, and these findings were compatible with diagnosis of IHSP (Fig. 3). Six months after operation, the patients visited an out-patient clinic, and the symptoms were stationary compared with at discharge. A follow-up MRI was conducted and revealed no recurrence of dura mater thickening.

**DISCUSSION**

IHSP is a rare condition, but possibly under-recognized because of its indolent nature and non-specific symptoms. Since accessibility of the diagnostic imaging technique including spinal MRI has improved recently, IHSP has been diagnosed and reported more frequently.

According to etiology of the disease, hypertrophic pachymeningitis is classified as idiopathic and secondary. Some infectious diseases including syphilis, and diseases caused by HTLV-1, mycobacterium, and fungi are reportedly associated with hypertrophic pachymeningitis. Various autoimmune disorders, such as rheumatoid arthritis, Wegener's granulomatosis, multifocal fibrosclerosis, sclerosing cholangitis, and sarcoidosis are also regarded as etiologic factors in particular cases. The diagnosis of IHSP depends on exclusion of these possible causative factors. In this case, we tried to reveal possible causative factors but failed. Although ESR was slightly increased, it was normalized 2 months after the operation and seemed to be a non-specific finding rather than sign of hidden inflammatory disease.

Clinical manifestations of IHSP vary according to location and size. In the early stage of the disease, the patient may experience only localized pain. However, as the lesion grows, it may lead to radiculopathy or compressive myelopathy. Typically, the symptoms caused by IHSP have an indolent nature with a chronic

---

**Fig. 1.** Computed tomography scan revealed an osteolytic lesion involving base of the C6 spinous process.

**Fig. 2.** Magnetic resonance imaging revealed an epidural mass involving the dorsal aspect of cervical spinal canal from C5 to C7 level, with low signal intensity on T2 weighted image (A) and non-enhancement on T1 weighted-enhanced image (B).

**Fig. 3.** Photomicrograph of a section of the excised dura mater showed fibrotic pachymeninges with no significant inflammation, which was more apparent at the higher magnification view (Hematoxylin and eosin staining).
Histopathologically, IHSP is characterized by fibrosis and infiltration of inflammatory cells, such as lymphocytes, plasma cells, polymorphonuclear cells, and macrophages, without necrotic change. However, in cases of secondary hypertrophic pachymeningitis, specific histopathologic findings might be different according to the nature of the underlying causative disease. In the present case, there were only scanty inflammatory cells and no evidence of active inflammation. These pathologic findings correspond to MRI findings. Typically, IHSP showed peripheral or nodular enhancement pattern and it represent to zone of inflammation. Nevertheless, in this case, MRI showed no enhanced portion. Less inflammatory reaction, more fibrosis, and no enhanced portion are related to worse response to steroid therapy.

There has been no previous report describing a case of IHSP with osteolytic lesion to our knowledge. In the present case, preoperative CT showed definite osteolytic lesion on the adjacent bone. Without necrotic change. However, in cases of secondary hypertrrophic pachymeningitis, specific histopathologic findings might be different according to the nature of the underlying causative disease. In the present case, there were only scanty inflammatory cells and no evidence of active inflammation. These pathologic findings correspond to MRI findings. Typically, IHSP showed peripheral or nodular enhancement pattern and it represent to zone of inflammation. Nevertheless, in this case, MRI showed no enhanced portion. Less inflammatory reaction, more fibrosis, and no enhanced portion are related to worse response to steroid therapy.

Most reports of IHSP have been limited to short-term follow-up after the initial treatment. According to previous reports, with long term follow-up exceeding 5 years, recurrence was noted in three of five cases. In this regard, repeated follow up are desirable in patient with IHSP even after successful treatment.

CONCLUSION

IHSP is a rare condition, but possibly under-recognized because of its indolent nature and non-specific symptoms. Decompressive surgery should be considered for the patient with definite or progressive neurologic symptoms in order to prevent further deterioration. After the surgical treatment, scheduled follow up is mandatory because IHSP can be recurrent. In addition, IHSP can present as an osteolytic lesion. Differential diagnosis with neoplastic disease, including giant cell tumor, is important.

References
1. Adler JR, Sheridan W, Kosek J, Linder S: Pachymeningitis associated with a pulmonary nodule. Neurosurgery 29: 283-287, 1991
2. Agdal N, Hagdrup HK, Wanzini GL: Pachymeningitis cervicalis hypertrophica. Acta Derm Venerol 60: 184-186, 1980
3. Astrom KE, Lidholm SO: Extensive intracranial lesions in a case of orbital non-specific granuloma combined with polyarteritis nodosa. J Clin Pathol 16: 137-143, 1963
4. Bucy PC, Freeman LW: Hypertrophic spinal pachymeningitis with special reference to appropriate surgical treatment. J Neurosurg 9: 564-578, 1952
5. Dumont AS, Clark AW, Sevick RJ, Myles ST: Idiopathic hypertrophic pachymeningitis: a report of two patients and review of the literature. Can J Neurol Sci 27: 333-340, 2000
6. Friedman D, Flanders A, Tartaglino L: Contrast-enhanced MR imaging of idiopathic hypertrophic craniospinal pachymeningitis. AJR Am J Roentgenol 160: 908-911, 1993
7. Guidetti B, La Torre E: Hypertrophic spinal pachymeningitis. J Neurosurg 26: 496-503, 1967
8. Ito Z, Osawa Y, Matsuyama Y, Aoki T, Harada A, Ishiiuro N: Recurrence of hypertrophic spinal pachymeningitis. Report of two cases and review of the literature. J Neurosurg Spine 4: 509-513, 2006
9. Kao KP, Huang CI, Shen DE, Ho JT, Chang T, Chu FL: Non-obstructive idiopathic pachymeningitis cervicalis hypertrophica. J Neurol Neurosurg Psychiatry 49: 1441-1444, 1986
10. Kawano Y, Kira J: Chronic hypertrophic cranial pachymeningitis associated with HTLV-1 infection. J Neurol Neurosurg Psychiatry 59: 435-437, 1995
11. Khadidkar SV, Menezes K, Parekh HN, Ureikar M, Bhagwati SN: Idiopathic hypertrophic cervical pachymeningitis: a case report with 5 year follow up. J Assoc Physicians India 51: 391-393, 2003
12. Kim JH Park YM, Chin DK: Idiopathic hypertrophic spinal pachymeningitis: report of two cases and review of the literature. J Korean Neurosurg Soc 50: 392-395, 2011
13. Kupersmith MJ, Martin V, Heller G, Shah A, Mitnick HJ: Idiopathic hypertrophic pachymeningitis. Neurology 62: 686-694, 2004
14. Lee YC, Chueng YC, Hsu SW, Lui CC: Idiopathic hypertrophic cranial pachymeningitis: case report with 7 years of imaging follow-up. AJNR Am J Neuroradiol 24: 119-123, 2003
15. Levine MR, Kaye L, Mair S, Bates J: Multifocal fibrosclerosis. Report of a case of bilateral idiopathic sclerosing pseudotumor and retroperitoneal fibrosis. Arch Ophthalmol 111: 841-843, 1993
16. Martin N, Masson C, Henin D, Mompont D, Marsault C, Nahum H: Hypertrophic cranial pachymeningitis: assessment with CT and MR imaging. AJNR Am J Neuroradiol 10: 477-484, 1989
17. Mikawa Y, Watanabe R, Hino Y, Hirano K: Hypertrophic spinal pachymeningitis. Spine (Phila Pa) 1976: 19: 620-625, 1994
18. Moore AP, Rolfe ER, Jones EL: Pachymeningitis cranialis hypertrophica. J Neurol Neurosurg Psychiatry 48: 942-944, 1985
19. Naffziger HC, Stern WE: Chronic pachymeningitic report of a case and review of the literature. Arch Neurol Psychiatry 62: 383-411, 1949
20. Nishino H, Rubino FA, Parisi JE: The spectrum of neurologic involvement in Wegener's granulomatosis. Neurology 43: 1334-1337, 1993
21. Nishizaki T, Iwamoto F, Uesugi S, Akimura T, Yamashita K, Ito H: Idiopathic cranial pachymeningoencephalitis focally affecting the parietal dura mater and adjacent brain parenchyma: case report. Neurosurgery 40: 840-843, discussion 843, 1997
22. Noble SC, Chandler WF, Lloyd RV: Intracranial extension of orbital pseu-
Idiopathic Hypertrophic Spinal Pachymeningitis with an Osteolytic Lesion | TK Jee, et al.

dotumor: a case report. *Neurosurgery* 18: 798-801, 1986
23. Pai S, Welsh CT, Patel S, Rumboldt Z: Idiopathic hypertrophic spinal pachymeningitis: report of two cases with typical MR imaging findings. *AJNR Am J Neuroradiol* 28: 590-592, 2007
24. Park SH, Whang CJ, Sohn M, Oh YC, Lee CH, Whang YJ: Idiopathic hypertrophic spinal pachymeningitis: a case report. *J Korean Med Sci* 16: 683-688, 2001
25. Riku S, Kato S: Idiopathic hypertrophic pachymeningitis. *Neuropathology* 23: 335-344, 2003
26. Rosenfeld JV, Kaye AH, Davis S, Gonzales M: Pachymeningitis cervicallis hypertrophica. Case report. *J Neurosurg* 66: 137-139, 1987
27. Schiess RJ, Coscia ME, McClellan GA: *Petriellidium boydii* pachymeningitis treated with miconazole and ketoconazole. *Neurosurgery* 14: 220-224, 1984
28. Tsutsui M, Yasuda T, Kanamori M, Hori T, Kimura T: Long-term outcome of idiopathic hypertrophic thoracic pachymeningitis. *Eur Spine J* 21 Suppl 4: S404-S407, 2012
29. Yamashita K, Suzuki Y, Yoshizumi H, Takahashi JB, Nogawa T: Tuberculous hypertrophic pachymeningitis involving the posterior fossa and high cervical region—case report. *Neurrol Med Chir (Tokyo)* 34: 100-103, 1994