Idiopathic Hypertrophic Cranial Pachymeningitis with Perifocal Brain Edema

—Case Report—

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Abstract

A 51-year-old female presented with an extremely rare case of idiopathic hypertrophic cranial pachymeningitis manifesting as markedly thickened frontotemporal meninges with expanding perifocal edema. Magnetic resonance imaging with gadolinium revealed enhancement of the thickened dura mater protruding into the brain parenchyma accompanied by focal edema causing a mass effect. Histological examination of a biopsy specimen revealed thickened dura with infiltrating lymphocytes. Serological and immunological tests were normal. No inflammatory response or evidence of malignant tumors was observed. The patient was treated with prednisolone, resulting in marked improvement of the mass effect. High-dose steroid therapy appears to be effective for intracranial pachymeningitis associated with expanding perifocal brain edema.

Key words: pachymeningitis, edema, steroid therapy

Introduction

Chronic hypertrophic cranial pachymeningitis is a rare disorder of diffuse, or sometimes localized, thickening and fibrosis of the dura that affects the spinal cord,^{1,6,28)} tentorium, and posterior part of the falx.^{14,17,33)} Cranial pachymeningitis usually involves the dura mater more extensively without intra-axial involvement. The causes of dural thickening include infectious diseases such as epidural abscess, syphilis,^{8,15)} tuberculosis,^{19,24)} and pseudomonas,⁷⁾ sarcoidosis,^{12,29)} rheumatoid arthritis,^{9,18)} Sjögren's syndrome,¹⁶⁾ fungal infection, Wegener's granulomatosis,⁵⁾ mucopolysaccharidosis,^{25,32)} intracranial fibromatosis,²⁾ and intrathecal administration of corticosteroids.²⁾ However, no clear cause is established in the majority of cases, and thus idiopathic hypertrophic pachymeningitis is a diagnosis of exclusion.

Accurate diagnosis is difficult due to the lack of specific neuroradiological findings. Thickened diffuse dural enhancement with perifocal edema is

uncommon and occurs in patients with various types of inflammation of the meninges, such as bacterial meningitis and sarcoidosis following subdural hemorrhage,³¹⁾ after craniotomy,³⁾ with dural metastases,³⁰⁾ and in intracranial hypotension.^{10,23)} Focal dural enhancement may be seen in patients with meningioma and leptomeningeal metastases. Therefore, histological examination of dura mater specimens is required to make a definitive diagnosis and to determine the nature of adjuvant therapy.

We present a case of idiopathic intracranial hypertrophic pachymeningitis that responded to treatment with prednisolone.

Case Report

A 51-year-old female was admitted to the Department of Neurosurgery at our hospital with severe temporal headache and vomiting. She was afebrile. Neurological examination revealed slight motor weakness of her left upper extremity. She had neither papilledema nor meningeal signs. Laboratory

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Fig. 1 *left*: Computed tomography (CT) scan showing a marked intraaxial edema in the right temporal lobe. *right*: Bone window CT scan showing slight erosion of the right temporal bone.

Fig. 2 *left*: Axial T₁-weighted magnetic resonance (MR) image with gadolinium depicting homogeneously enhanced thick meninges extending to the cortical parenchyma. *right*: Axial T₂-weighted MR image revealing a hypointense mass with marked extension of perifocal edema.

ry tests revealed that the complete blood count, erythrocyte sedimentation rate (ESR), serum C-reactive protein level, and immunoglobulin and complement levels were normal.

Computed tomography (CT) of the head revealed marked perifocal edema in the right temporal lobe (Fig. 1 *left*). A bone window CT scan revealed slight erosion of the inner table of the temporal bone (Fig. 1 *right*). T₁-weighted magnetic resonance (MR) imaging revealed a hypointense area in the thickened right frontotemporal dura mater (mainly temporal). T₁-weighted MR imaging with gadolinium showed homogeneously enhanced meninges with perifocal edema (Fig. 2 *left*). T₂-weighted MR imaging showed the mass of the thickened dura mater as hypointense, and a high intensity area extending to the

Fig. 3 Photomicrograph of the dural biopsy specimen showing mononuclear inflammatory cell infiltration without malignant cells along the fibrous collagenous tissue and perivascular space. HE stain, ×100.

Fig. 4 Axial T₁-weighted magnetic resonance (MR) image with gadolinium (*left*) and T₂-weighted MR image (*right*) following burr hole biopsy and steroid therapy revealing marked reduction of the enhanced mass in the meninges and intraaxial perifocal edema.

white matter corresponding to perifocal edema (Fig. 2 *right*). Angiography revealed no vascular staining or other vascular abnormalities.

A right temporal craniotomy was performed and the dura-based mass was partially resected. The dura was thick and yellowish and the mass extended into the brain and adhered strongly to the adjacent brain parenchyma. Histological examination of the dura specimen revealed thickened fibrous tissue with marked lymphocyte infiltration (Fig. 3). Inflammatory cells similar to those infiltrating the dura mater were present in the subarachnoid and Virchow-Robin spaces and part of the brain parenchyma in the resected cortex of the temporal lobe.

No caseous necrosis or multinucleated giant cells were observed. Cerebrospinal fluid (CSF) obtained by postoperative lumbar puncture contained 1 leukocyte/ μ l, 55 mg/dl of sugar, and 27 mg/dl of protein. No bacteria were detected. The CSF cytology was classified as Papanicolou class 2.

No neurological deficit was observed following surgery. The patient was treated with prednisolone at 20 mg/day orally for one week and 10 mg/day for another week, which was tapered off over 2 weeks. Headache, nausea, and vomiting improved following steroid therapy. Two weeks after steroid therapy, MR imaging revealed that the cerebral edema had disappeared and that the mass had spontaneously regressed (Fig. 4). No recurrence was observed one year after discontinuation of steroid therapy.

Discussion

Our case of idiopathic hypertrophic cranial pachymeningitis presented with positive Wassermann reaction and serologic test for syphilis, but a Gram's staining demonstrated no microorganisms, and Ziehl-Neelsen staining and Grocott-Gomori methenamine-silver staining were also negative. Some cases represent an autoimmune process, and consequently we also determined rheumatoid factor, anti-nuclear antibody, and complement levels, all of which were absent or within normal limits. Although elevation of ESR is frequently seen in pachymeningitis, idiopathic cases with normal ESR like ours have been reported. This case showed no evidence of elevation of angiotensin-converting enzyme or adenosine deaminase levels, thereby excluding sarcoidosis and tuberculosis, respectively. Based on these findings, our patient was considered to have a rare idiopathic form of pachymeningitis.

Histological examination found remarkable diffuse thickening of the dura mater with considerable fibrosis and chronic inflammatory cell infiltration, including plasma cells and lymphocytes, in the present case. At surgery, tissue, including brain parenchyma, had adhered tightly to the dura mater, with an unclear border. In addition, the thickened leptomeninges and the temporal cortex had adhered tightly together and CSF was not expelled during resection of tissue for biopsy.

The characteristic MR imaging appearance of pachymeningitis is a hypointense mass lesion surrounded by a thin hyperintense margin on T₂-weighted imaging.^{22,27,34} Normal dura mater (pachymeninges) is composed of dense fibrous connective tissue with relatively little interstitial space.

Thus, although the dura mater lacks a blood-brain barrier, relatively little contrast material accumulates in the interstitial space. Hypertrophic pachymeningitis is characterized by dense fibrosis with scattered inflammatory cells, and the peripheral margin of the lesion contains highly vascularized arachnoid membrane. These features probably cause the enhancement of the dura mater on T₁-weighted imaging with gadolinium.

Interestingly, the lesion in our patient was in the frontotemporal area of the dura mater. T₂-weighted MR imaging showed a large hyperintense area of the white matter indicating marked perifocal edema, similar to that seen in patients with glioblastoma multiforme, meningiomas, and metastatic tumors. We were unable to exclude the possibility of neoplastic disease until histological examination of the biopsy specimen. There was no evidence of malignant tumor, and serum tumor markers and gallium scintigraphy findings were normal. Idiopathic hypertrophic cranial pachymeningitis has been described with an unusual and misleading manifestation mimicking multiple meningiomas.^{4,22,27} Ours and previous observations^{4,22,27} emphasize the necessity for considering idiopathic hypertrophic cranial pachymeningitis as part of the differential diagnosis of meningeal tumors.

Some cases of intracranial lesion with expanding focal edema have been described, but the mechanism of edema remains unknown. Thickening of the dura and inflammation due to chronic cranial pachymeningitis have resulted in occlusion of the bilateral carotid arteries¹¹ or superior sagittal sinus.^{13,15} No abnormalities were found in the superior sagittal sinus and deep veins in our case, but the edema may have resulted from venous congestion^{17,26} due to the mass effect, and vascular occlusion due to thickening of the dura or inflammatory cells infiltrating along the Virchow-Robin space.

Perifocal edema diminished after treatment with corticosteroid in our patient, but the definitive treatment for cranial pachymeningitis has not been established. The cause of pachymeningeal thickening is unknown, and spontaneous resolution may occur. However, postoperative treatment with corticosteroid or immunosuppressive agents, such as azathioprine and cyclophosphamide, may be effective.^{16,17,20-22,33} High-dose corticosteroid therapy should be the treatment of choice, followed by immunosuppressive agents.²² Although steroid therapy does not always present the clinical progression of pachymeningitis, high-dose steroid therapy may be worth trying in patients with pachymeningitis associated with marked perifocal brain edema. Long follow up of more cases is needed to establish the

natural history of the disease and the efficacy of therapy.

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