CASE REPORT

# A Case of Idiopathic Hypertrophic Pachymeningoencephalitis : MR Findings

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Idiopathic hypertrophic pachymeningitis is an idiopathic disorder characterized as a fibrosing inflammatory process involving the dura mater, tentorium and falx cerebri. It sometimes causes cranial nerve paralysis secondary to mass effect or infiltration, but to our knowledge, anosmia with olfactory cranial nerve palsy has never been documented. Recently there have been only a few reports of intraparenchymal involvement (pachymeningoencephalitis). We experienced a case of idiopathic hypertrophic pachymeningoencephalitis presenting with decreased visual acuity and anosmia. Magnetic resonance (MR) imaging showed a dural-arachnoid based en-plaque mass with infiltration to adjacent sulci of the bilateral frontal lobe and optic nerve bilaterally. $T_2$-weighted axial images showed the mass as low signal intensity relative to gray matter, a finding corresponding with dense fibrosis. Marked edema was evident in the frontal lobe bilaterally, which upon microscopic examination was seen to consist of intraparenchymal infiltration with lymphocytes and plasma cells.

## INTRODUCTION

Idiopathic hypertrophic pachymeningitis is an idiopathic disorder characterized as a fibrosing inflammatory process involving the dura mater, tentorium and falx cerebri. Patients commonly present with long standing headaches and sometimes with cranial nerve paralysis secondary to mass effect or infiltration[1]. Involvement of the dura of frontal fossa is rare and to our knowledge, anosmia with olfactory cranial nerve palsy has never been documented[1]. Most of the cases of idiopathic hypertrophic pachymeningitis do not include intraparenchymal involvement. Recently, however, there were a few reports of intraparenchymal involvement (pachymeningoencephalitis)[2)~5)]. We experienced a case of idiopathic hypertrophic pachymeningoencephalitis in a patient presenting with decreased visual acuity of the left eye and anosmia and demonstrable magnetic resonance (MR) findings.

## CASE REPORT

A 65-year-old woman presented 2 weeks af-

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ter noticing decreased visual acuity of the left eye and anosmia. Her past medical history was unremarkable. Neurological examination showed bilateral olfactory nerve palsy, no direct light reflex of the left eye, no indirect light reflex of the right eye and lower half visual field loss of the left eye. The blood cell count and serum chemistries were normal. C-reactive protein was 1.1 mg/dl (normal$<$0.2 mg/dl). Angiotensin converting enzyme (ACE) level and adenosine deaminase (ADA) level were normal. Serum VDRL and STS were negative. Rheumatoid factor and diffuse cytoplasmic anti-neutrophil cytoplasmic antigen (c-ANCA) were negative. A lumbar puncture revealed an elevated protein level of 117 mg/dl (normal, 10–40 mg/dl), a glucose level within normal limits, 84 white blood cells/mm$^3$ (all monocytes), elevated IgG level of 36.6 mg/dl (normal, 1–3 mg/dl). Oligoclonal IgG bands were not seen. Cytologic examination and culture for bacteria, fungi and mycobacteria were negative.

Head MR examination was performed with a Magneton H15 1.5-Tesla machine (Siemens). $T_1$-weighted axial images (TR/TE=600/15 ms) showed an iso-signal intense mass relative to gray matter in the anterior cranial fossa. In $T_2$-weighted axial images (TR/TE=2500/90 ms), the mass showed low signal intensity relative to gray matter (Fig. 1A; $T_2$-weighted imaging of other planes was not done). There was marked edema in the frontal lobes bilaterally (Fig. 1B). Postcontrast $T_1$-weighted coronal images showed intense homogenous enhancement of the dural-arachnoid based en-plaque mass with infiltrating to adjacent sulci of the frontal lobes and optic nerves bilaterally (Fig. 1C, D).

Upon left fronto-temporal craniotomy, the dura mater of the anterior cranial fossa was seen to be thickened and firm. Adhesion with the underlying arachnoid, pia mater and brain was seen. The brain was severely edematous. Biopsy from the dura mater, arachnoid and brain was performed. On microscopic examination, the dura mater and arachnoid was thickened by dense fibrous tissue containing lymphocytes and plasma cells (Fig. 2A). No caseation was seen. Stains and cultures for bacteria, fungi and mycobacteria were negative. The cerebral parenchyma, particularly perivascular areas, was infiltrated with lymphocytes and plasma cells (Fig. 2B). This finding suggested that brain edema was related to inflammatory infiltration as opposed to venous congestion due to compression of the brain parenchyma by the thickened dura.

A diagnosis of idiopathic hypertrophic pachymeningoencephalitis was made and steroid therapy (betamethasone 12 mg/day) was initiating with tapering for one month. The visual acuity and field restriction of the left eye improved remarkably. Follow up MRI showed the mass and brain edema were decreasing in size. After treatment, the patient became asymptomatic, but 22 months after therapy MRI showed recurrence of thickened dura and falx in the frontal convexity and brain edema (Fig. 3A, B). Radiating linear shadow in the area of brain edema was suspected to be perivascular encephalomalacia due to inflammation. On this basis, the patient was again given steroid therapy (prednisolone 20 mg/day) with tapering for

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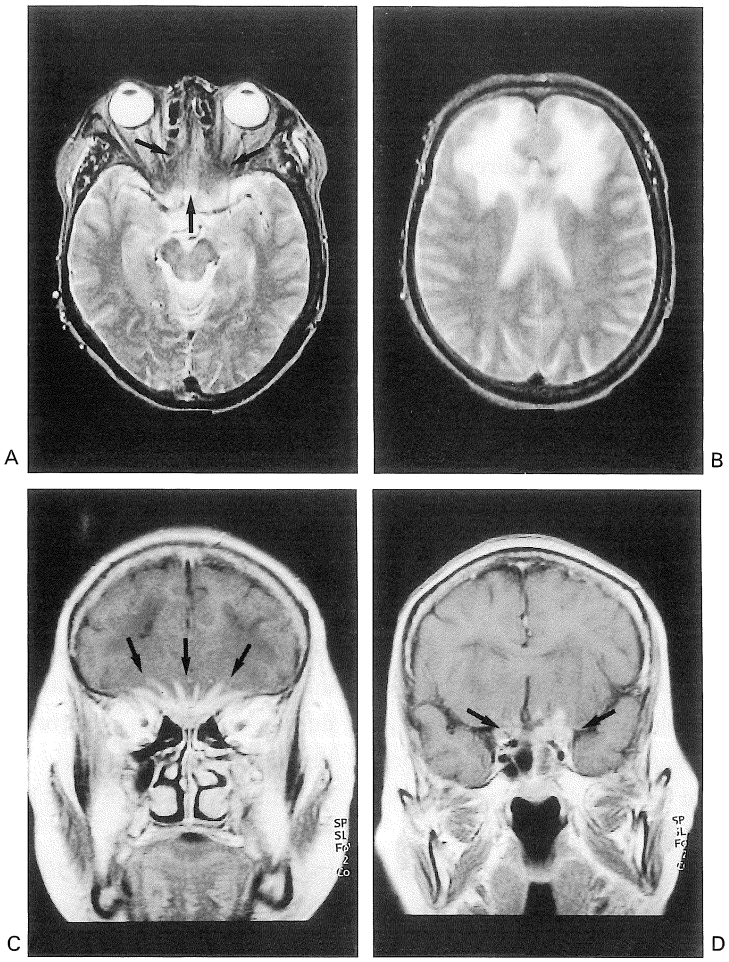


Fig. 1. In $T_2$-weighted axial images (TR/TE=2500/90 ms), the abnormal area shows low signal intensity relative to gray matter (arrow) in the anterior cranial fossa (A). There is marked edema in the frontal lobes bilaterally (B). Postcontrast $T_1$-weighted coronal image (TR/TE=600/15 ms) shows intense homogenous enhancement of the dural-arachnoid based en-plaque mass with infiltrating to adjacent sulci of the frontal lobes bilaterally (arrow) (C). The mass involves the optic nerves bilaterally (arrow) (D).

two months. Follow up MRI showed the recurrent lesion was improving. At present 13 months after the second course of therapy, MRI shows no recurrence.

## DISCUSSION

Idiopathic hypertrophic pachymeningitis is an idiopathic disorder characterized as a fibrosing inflammatory process involving the dura mater, tentorium and falx cerebri. Microscopi-

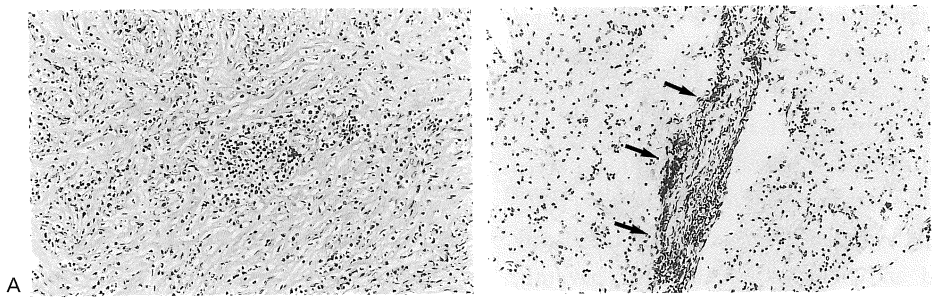


Fig. 2. On microscopic examination, the dura mater and arachnoid was thickened by dense fibrous tissue containing lymphocytes and plasma cells (A), and the cerebral parenchyma, particularly perivascular area (arrow), was infiltrated with lymphocytes and plasma cells (B).

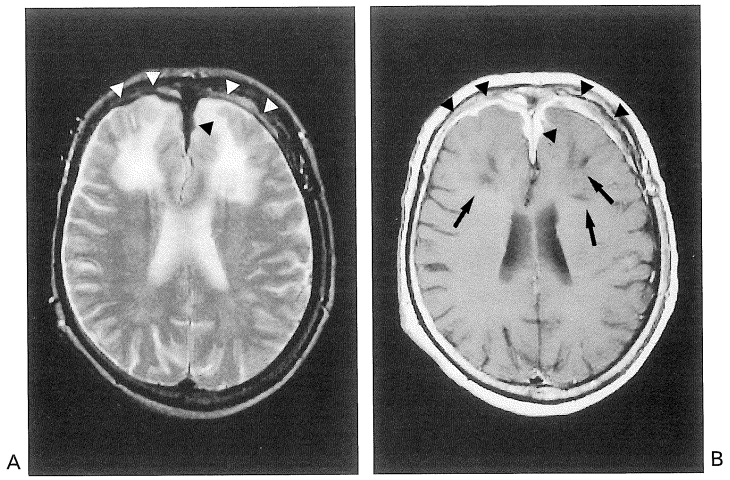


Fig. 3. $T_2$-weighted (A) axial image shows the recurrent lesion (arrowhead) with low signal intensity in the dura and falx of frontal convexity, and brain edema. Postcontrast $T_1$-weighted (B) image shows the lesion (arrowhead) with intense homogenous enhancement. The radiating linear shadow (arrow) in brain edema was suspected to be perivascular enceplalomalacia due to inflammation.

cally, chronic inflammatory cells, including plasma cells and lymphocytes, can be seen. Venous congestion, ischemia, or infarction related to narrowing of dural sinuses or compression of the brain parenchyma from hypertrophic dura may be observed[1]. Most cases of idiopathic hypertrophic pachymeningitis do not include intraparenchymal involvement. Recently, however, there have been a few reports of intraparenchymal involvement (pachymeningoencephalitis) characterized microscopically by infiltration of the cortex and the perivascular spaces with inflammatory cells[2)~5)]. The common site of involvement is around the tentorium cerebelli, posterior third of the falx, clival dura mater, parasellar and cavernous region[6),7)].

The clinical presentation is nonspecific : patients commonly present with long standing headaches, and sometimes with cranial nerve paralysis secondary to mass effect or infiltration and cerebellar ataxia[1]. All cranial nerves except the olfactory nerve may be affected[1]. The eighth cranial nerve is mostly frequently involved and followed by the fifth, seventh, ninth, tenth, and twelfth cranial nerves in equal frequency[1]. To our knowledge, our case is the first to report olfactory nerve palsy. When seen, cerebellar ataxia is thought to be caused by venous ischemia[1]. Among laboratory findings, the erythrocyte sedimentation rate is usually increased as is the C-reactive protein level[2]. The CSF may show elevated levels of cells, protein and IgG[2),3]. For treatment, steroid therapy is known to lead to remission of clinical symptoms and decreased thickness of the dura on neuroimaging. However, patients may become steroid-dependent and some may demonstrate deterioration with tapering of the steroid dose. Recently, the immunosuppressive drug azathioprine has been used[7].

Thickened dura mater is sometimes observed in neoplasmic disorders (dural carcinomatosis, meningioma, plasmacytoma, lymphoma and leukemia), granulomatous disorders (sarcoidosis, Wegener's granulomatosis) and infectious disorders (syphilis, tuberculosis and fungal infections)[8)~17]. The characteristic MR findings of low signal intensity of the thickened dura mater in $T_2$-weighted images corresponds to a dense fibrosis and is useful in the diagnosis of idiopathic hypertrophic pachymeningits[1)~5),7)~9]. However, fungus, sarcoidosis and Wegener's granulomatosis may have fibrous connective tissue, which shows low signal intensity in $T_2$-weighted images[13),14),18),19]. Moreover, these disorders may have intraparenchymal involvement[14),18),19]. Differential diagnosis of idiopathic hypertrophic pachymeningitis from these three disorders is therefore difficult based only on MR findings. Fungal meningitis is secondary to involvement of the lungs or can originate from contiguous sinus cavity, orbit or middle ear[17]. Findings from chest radiographic films and MR studies of the sinus cavities, orbits and middle ears can be useful in diagnosis. In Wegener's granulomatosis, abnormal antibodies against the cytoplasm of certain neutrophil granular proteins (diffuse cytoplasmic anti-neutrophil cytoplasmic antigen; cANCA) in the serum are present in 80–90% of the patients with active systemic disease, so specific assay for these be useful in diagnosis[19]. With involvement secondary to sarcoidosis, any evidence of systemic sarcoidosis, and serum angiotensin-converting enzyme and lysozyme levels can be suggestive for diagnosis.

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