Cranial hypertrophic pachymeningitis is an uncommon disorder in which the intracranial dura mater becomes locally or diffusely thickened, often adhering to the underlying leptomeninges. Intra-axial involvement is extremely rare; only three cases of pachymeningitis involving the cerebral parenchyma have been reported in the literature. We report a rare case of focal idiopathic hypertrophic pachymeningoencephalitis (IHP) focally affecting the right parietal dura mater and adjacent parietal cortex.

Case Report

A 43-year-old male experienced intermittent right parietal headache and numbness of the left lower limb in August 2004. On September 11 of that year, he was sent to our hospital due to rhythmic jerking of the left abdominal muscles that had persisted for more than 4 hours. Neurologic examination revealed mild weakness and impaired sensation in the left lower limb. Electroencephalography disclosed active focal spikes in the right parietal region and brain magnetic resonance imaging showed a well-enhanced lesion involving the right parietal lobe and its overlying dura mater. Surgical removal of the lesion revealed infiltration by abundant chronic inflammatory cells without granuloma formation, caseous necrosis or vasculitis. After surgery, the patient was treated with steroid, which was tapered off 5 months later. Serial magnetic resonance imaging follow-up revealed that the parenchymal mass and perifocal edema had completely disappeared 6 months after the operation.

Key Words: chronic inflammation, meningoencephalitis, pachymeningitis

Cranial hypertrophic pachymeningitis is a diffuse granulomatous disease that involves the dura mater extensively without intra-axial involvement. We report a rare case of focal idiopathic hypertrophic pachymeningoencephalitis (IHP) focally affecting the right parietal dura mater and adjacent parietal lobe. A 43-year-old male suffered from acute onset of rhythmic twitching over the left abdominal muscles that had persisted for more than 4 hours. Neurologic examination revealed mild weakness and impaired sensation in the left lower limb. Electroencephalography disclosed active focal spikes in the right parietal region and brain magnetic resonance imaging showed a well-enhanced lesion involving the right parietal lobe and its overlying dura mater. Surgical removal of the lesion revealed infiltration by abundant chronic inflammatory cells without granuloma formation, caseous necrosis or vasculitis. After surgery, the patient was treated with steroid, which was tapered off 5 months later. Serial magnetic resonance imaging follow-up revealed that the parenchymal mass and perifocal edema had completely disappeared 6 months after the operation. We suggest that early recognition of this rare IHP, together with proper surgical intervention and concomitant steroid therapy, may be beneficial for long-term remission. [J Formos Med Assoc 2008;107(2): 181–184]
sensation of the left lower limb. Electroencephalography disclosed active focal spikes in the right parietal region. Gadolinium-enhanced T1-weighted magnetic resonance imaging (MRI) showed a homogeneously enhanced lesion in the right parietal region involving both the leptomeninges and underlying cortical parenchyma, with marked perifocal edema (Figure 1A). Serologic tests for syphilis and HIV were negative. Autoimmune work-up, including erythrocyte sedimentation rate, antinuclear antibody and extractable nuclear antigen screening, as well as survey of tumor markers such as carcinoembryonic antigen, α-fetoprotein, prostate-specific antigen, CA-125 and CA-153, were all within normal limits. Chest X-ray revealed no indications of hilar lymphadenopathy or parenchymal lesions.

The patient’s seizures were controlled with parenteral phenytoin. A right parietal craniotomy was performed, and the thickened dura and contiguous mass were partially resected.

Tissue and cerebral spinal fluid (CSF) cultures for Mycobacterium tuberculosis, other bacteria and fungi were all negative. Histologic examination of the dura (Figure 2A) and cortex (Figure 2B) revealed infiltration by abundant chronic inflammatory cells. There was no evidence of granuloma formation, caseous necrosis or vasculitis.

Figure 1. (A) Baseline gadolinium-enhanced T1-weighted magnetic resonance imaging (MRI) shows a homogeneously enhanced lesion in the right parietal region (arrow) involving both the leptomeninges and underlying cortical parenchyma, with marked perifocal edema. (B) Gadolinium-enhanced T1-weighted MRI performed 6 months after surgery shows complete resolution of the previous lesion in the leptomeninges and cortical parenchyma.

Figure 2. Histologic examination of: (A) the dura reveals marked lymphocyte infiltration; (B) the parietal cortex shows marked lymphocyte infiltration.
Immunohistochemical studies for subtypes of the infiltrating lymphocytes revealed that they were polyclonal in nature.

After surgery, the patient was treated with intravenous methylprednisolone (1 g/day) for 1 week, followed by oral prednisolone (60 mg/day). The steroid was tapered off 5 months later. Concomitantly, he received antiepileptic drugs (phenytoin and topiramate) for 16 months. Serial MRI follow-up revealed that the parenchymal mass and perifocal edema had completely disappeared 6 months after the operation (Figure 1B). Over a 2-year follow-up period, the patient has remained seizure-free after discontinuation of steroid and anticonvulsant treatment.

Discussion

Hypertrophic pachymeningitis refers to a chronic, circumscribed, inflammatory thickening of the dura. This type of meningeal reaction, which is now exceedingly uncommon, was first described by Charcot and Joffroy. Its etiology includes infections, autoimmune and vasculitic disease. IHP is considered immunological in origin. The differential diagnosis includes meningioma, dural carcinomatosis, lymphoma, histiocytosis, syphilis, tuberculosis, fungus infection, Wegener’s granulomatosis, sarcoidosis, Sjögren’s syndrome, and giant cell arteritis. In the presented case, a serologic test for syphilis was negative and the absence of necrotizing vasculitis ruled out a diagnosis of Wegener’s granulomatosis. Tissue and CSF cultures for Mycobacterium tuberculosis were negative. There was no evidence of lymphoma from immunohistochemical staining. Neoplasm was ruled out by histologic and tumor marker studies. Thus, we concluded that the correct diagnosis was idiopathic pachymeningitis with intracerebral extension, i.e. pachymeningoencephalitis.

The characteristic MRI appearance of hypertrophic pachymeningitis shows gadolinium enhancement that may be linear or nodular, involving especially the falx cerebi, tentorium and cavernous sinus. In our patient, gadolinium-enhanced T1-weighted MRI showed a homogeneously enhanced lesion in the right parietal region involving the leptomeninges and extending to the underlying cortical parenchyma with marked perifocal edema. Histopathologically, there are usually lymphocyte, plasma cell and epithelioid cell infiltration with or without granulomatous reaction. In our patient, the dura and cortex revealed similar infiltration by abundant chronic inflammatory cells.

The most common symptoms of hypertrophic pachymeningitis are chronic headache and cranial neuropathies. Depending on the site of thickening, there may be compression of the optic nerve, cranial nerve pass through the cavernous sinus or, less often, lower cranial nerves. Cerebral symptoms include cerebellar ataxia, hemiparesis and altered mentation. Cranial pachymeningitis with cerebral parenchymal involvement is extremely rare. Of the four reported instances, including the presented case, two that involved the temporal cortex and adjacent cerebral sinus presented with increased intracranial pressure and cranial nerve palsies. The remaining two individuals with parietal cortex involvement presented with seizure. Convulsion or status epilepticus, as observed in the case presented herein, is extremely rare.

IHP is a progressive disorder, but in most cases, headache and neurologic dysfunction improve with high-dose oral corticosteroid therapy. However, steroid dependence may occur. Dural thickening may decrease with treatment, but MRI enhancement usually persists. In some patients, surgical excision of the thickened dura may be necessary to prevent irreversible neurologic damage. Based on the experience garnered in the two reported cases of IHP and from our own patient, it appears reasonable to suggest that early surgical excision and concomitant steroid treatment may lead to good clinical recovery. In contrast, Lee et al reported a case where hypertrophic pachymeningitis was treated only with immunosuppressive agents; progressive intracranial involvement developed, which could not be halted by later decompressive surgery.
We suggest that early recognition of this rare pachymeningoencephalitis, with proper surgical intervention for tissue diagnosis and concomitant steroid therapy, may be beneficial for long-term remission.

References