

Case Report

Idiopathic Hypertrophic Cranial Pachymeningitis Misdiagnosed as Acute Subtentorial Hematoma

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A case of idiopathic hypertrophic cranial pachymeningitis (IHCP) misdiagnosed as an acute subdural hematoma is reported. A 37-year-old male patient presented with headache following head trauma 2 weeks earlier. Computerized tomography showed a diffuse high-density lesion along the left tentorium and falx cerebri. Initial chest X-rays revealed a small mass in the right upper lobe with right lower pleural thickening, which suggested lung cancer, such as an adenoma or mediastinal metastasis. During conservative treatment under the diagnosis of a subdural hematoma, left cranial nerve palsies were developed (3rd and 6th), followed by scleritis and uveitis involving both eyes. Magnetic resonance imaging (MRI) revealed an unusual tentorium-falx enhancement on gadolinium-enhanced T1-weighted images. Non-specific chronic inflammation of the pachymeninges was noticed on histopathologic examination following an open biopsy. Systemic steroid treatment was initiated, resulting in dramatic improvement of symptoms. A follow-up brain MRI showed total resolution of the lesion 2 months after steroid treatment. IHCP should be included in the differential diagnosis of subtentorial-enhancing lesions.

KEY WORDS : Pachymeningitis · Subtentorial hematoma · Lower cranial nerve palsy.

INTRODUCTION

Idiopathic hypertrophic cranial pachymeningitis (IHCP) is an extremely rare disorder that predominantly affects male patients¹⁾. IHCP is a chronic, fibrosing, inflammatory process that involves the dura mater of the brain, particularly the falx cerebri and the tentorium^{1,5)}. Several causes of the disorder have been recognized, including infections, autoimmune disorders, and neoplasms¹⁾. However, the exact etiology of IHCP remains unknown. Generally, the presenting symptoms include headaches, cranial nerve palsies, cerebellar dysfunction, and seizures. The laboratory findings in patients with IHCP include mild-to-moderate elevation of C-reactive protein (CRP) and the erythrocyte sedimentation rate (ESR)^{1,8,9)}. Cerebrospinal fluid (CSF) studies in most cases show aseptic inflammatory changes³⁾. Radiographic studies reveal diffuse or localized thickening of the dura mater^{2,6)}.

Magnetic resonance imaging (MRI) studies provide the best evidence for the diagnosis of IHCP^{4,6)}. It is probable that IHCP is an isolated intracranial localization of multifocal fibrosis^{2,4,6)}. Left untreated, the clinical course of patients with IHCP is usually marked by severe headaches, progressive neurologic deterioration, and visual loss^{3,8)}. The clinical course is chronically progressive and is characterized by frequent recurrences^{3,4)}. Combined therapy with steroids and other immunomodulatory agents has been found to be effective^{1,2)}, suggesting that the mechanism underlying pachymeningitis may be an immunological process⁸⁾. The treatment of choice for IHCP is mainly steroid therapy. A rare case of IHCP misdiagnosed as an acute subdural hematoma is reported.

CASE REPORT

A 37-year-old man with headache which developed following head trauma 2 weeks before admission sought evaluation. Brain computed tomography (CT) scans showed a diffuse, high-density lesion along the left tentorium and falx cerebri, suggesting a subdural hemorrhage. A non-contrast enhanced CT was obtained for follow-up, and showed minimal resolution of the hemorrhage (Fig. 1). The patient had no heada-

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Fig. 1. Ten days after head trauma, a computed tomography (CT) scan shows a high-density enhancement along the left tentorium and falx, suggesting a subdural hemorrhage (arrow). A : Non-enhanced image. B : Enhanced image.

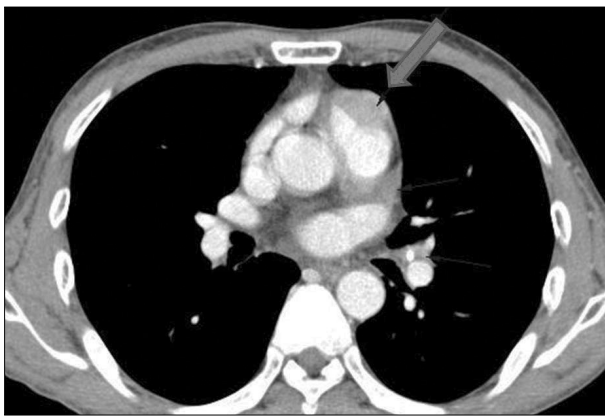


Fig. 2. A chest CT shows an irregular soft tissue density measuring 3.3×1.6 cm in the left upper anterior pericardial region, suggesting a left pericardial metastasis (arrow).

ches, nausea, vomiting, or photophobia prior to the head trauma. Thus, we rendered a diagnosis of acute subdural hemorrhage. However, a small mass in the right upper lobe with right lower pleural thickening was noted on routine chest X-ray. A chest CT suggested lung cancer, such as an adenoma with a mediastinal metastasis (Fig. 2). After admission, further studies focusing on the brain lesion and lung mass were planned, such as a lung biopsy, brain magnetic resonance image (MRI), and open surgery, if needed.

Two days after admission, left oculomotor and abducens (3rd and 6th) nerve paralysis occurred, followed by scleritis and uveitis involving both eyes. Based on the symptoms, we investigated the possible presence of an autoimmune disease, a metastatic brain tumor from lung cancer, or infectious causes. Laboratory tests related to autoimmune diseases (VDRL, rheumatoid arthritis factor, Mantoux test, serum HTLV-1, c and p ANCA, angiotensin converting enzyme, FANA, Euroline, C3, C4, and CH 50) were performed. However, no findings suggesting an autoimmune disease were demonstrated.

Laboratory findings showed an elevated CRP (44.49 mg/ml; normal range, 0-5 mg/mL) and ESR (48 mm/hr;

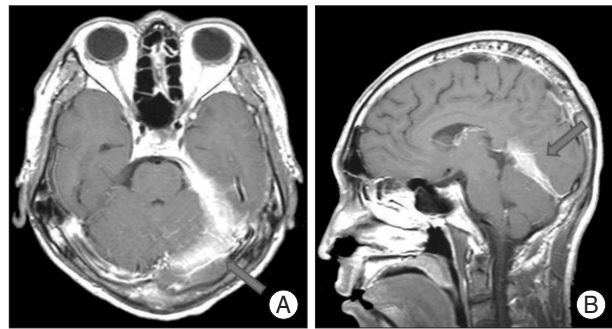


Fig. 3. Enhanced T1-weighted MR images shows thick and homogeneous enhancement along the left tentorium and cerebellar surface suggesting an en plaque meningioma or dural metastasis of lung cancer (arrow). A : Axial view. B : Sagittal view.

normal range, 0-9 mm/hr). The CSF study performed 4 days after admission revealed the following : WBC count, 28/hpf (lymphocyte dominant); total protein level, 99 mg/dL; glucose level, 51mg/dL; color, clear; pressure, 15 cm H₂O. An acid-fast bacilli stain of the CSF was negative. These findings suggested non-infectious inflammatory changes. Enhanced axial and coronal T1-weighted images (WI) of a brain MRI performed 3 days after admission showed dural enhancement on the left tentorium at the surface. The non-enhanced T1WI had a similar signal intensity as the gray matter, and the T2WI had a low signal intensity. There was no definite abnormal signal intensity in the surrounding brain parenchyma. The radiologist diagnosed these findings as an en plaque meningioma or probable dural metastasis from lung cancer (Fig. 3).

A craniotomy was performed for pathologic confirmation and tumor removal. A round craniotomy, 3 cm in size, was performed on the occipital bone above the inion. A thick, rubbery, white accumulation was noted on the surface of the tentorial dura. Some specimens, including dura, were biopsied for pathologic examination.

In the pathology report, a dural thickening with no evidence of tumor cells was noticed. The histopathologic examination showed dense fibrosis with an acute and chronic inflammatory cell infiltration and granulomatous inflammation, whereas typical findings of giant cells were absent. In addition, serologic and immunologic tests were normal. The presence of neutrophils, lymphohistiocytes, plasma cell invasion, and fibrosis implied a high probability of meningitis (Fig. 4). These histopathologic findings were consistent with IHCP. The patient underwent systemic steroid treatment 21 days after admission. After administering 20 mg of dexamethasone per day for 6 days, we prescribed prednisolone (20 mg/day) for 1 month.

Cyclosporine (CS) was prescribed by the ophthalmologist at a dose of 50 mg/day for treating scleritis and uveitis 8 days after admission. CS was administered at a dose of 50 mg/day

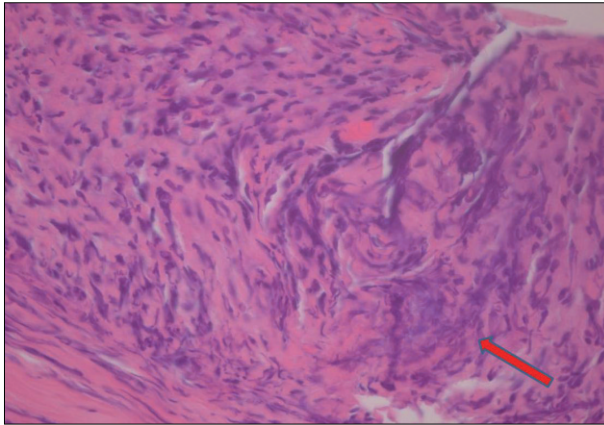


Fig. 4. A histopathologic examination reveals dense fibrosis with an acute and chronic inflammatory cell infiltration, which are consistent with meningitis (arrow; H & E, $\times 400$).

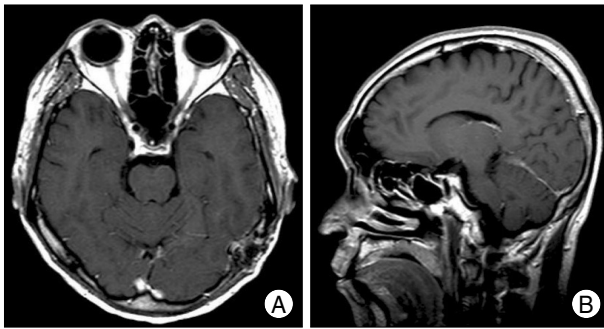


Fig. 5. Follow-up enhanced MRI scans after 2 months of therapy show total resolution of the lesion. A : Axial view. B : Sagittal view.

for 19 days and was then increased to 200 mg/day. Articular symptoms involving both knee joints, such as swelling, tenderness, and local heating, were observed after 5 days of CS (200 mg/day) administration, so CS was stopped. Articular symptoms were improved slowly after discontinuation of CS. The patient showed a marked improvement of headache 2 days after high-dose steroid therapy was started. Improvement of the cranial nerve palsies (3rd and 6th) was observed 15 days after high-dose steroid therapy was administered. The follow-up study performed 2 months after steroid treatment revealed total resolution of the intracranial lesion (Fig. 5). There was no evidence of recurrence after discontinuation of steroids.

DISCUSSION

Headache is the most common symptom of IHCP, reflecting inflammation of the meninges or increased intracranial pressure¹⁰. Cranial nerve palsies caused by fibrous infiltration and ischemic injury around the meninges are the second most common symptom of IHCP². The headaches caused by IHCP are generally severe and treatment-resistant. In our case, the cause of headaches was initially believed to be the

result of an acute or subacute subtentorial hematoma because the headaches developed after head trauma and CT scan findings were very similar to those of acute or subacute subtentorial hematomas. An accurate diagnosis of IHCP, however, was delayed until the development of cranial nerve palsies. With a headache alone, accurate diagnosis is very difficult.

The exact etiology and pathogenesis of IHCP are unknown. Cranial hypertrophy of the meninges accompanied by headaches and cranial nerve palsies occur not only in IHCP, but also in infectious diseases (e.g., tuberculosis, fungal infection, neurosyphilis, and measles), autoimmune disorders (e.g., sarcoidosis, rheumatoid arthritis, Wegener's granulomatosis, and vasculitis), and neoplastic disorders [primary brain tumors (en plaque meningiomas)]^{1,8,11}. The likely pathogenesis of the cranial nerve deficits in IHCP are fibrous encasement and ischemic damage produced by the hypertrophic dura².

For an accurate diagnosis of IHCP, laboratory testing, neuroradiologic studies, and histopathologic examinations are necessary¹⁰. In our patient, the laboratory findings showed elevated CRP and ESR values. However, the CSF studies for the differential diagnosis of infectious diseases are usually non-diagnostic. However, it is known that CSF protein levels are elevated in 50% of patients with IHCP¹. In this case, the high-density lesion was misdiagnosed as a subdural hematoma due to the patient's recent head trauma. Hyperdense lesions on a plain CT can be caused by various conditions, such as chronic inflammation with lymphocytes, extensive fibrosis with plasma cell invasion, scattered eosinophils and narrowing, occlusion of venous sinuses in relation to thickened sinus walls, or obliteration⁶.

Brain MRI is the diagnostic tool of choice for determining the exact cause of cranial nerve palsies. Most frequently, IHCP affects the tentorium and sagittal falx, followed by the cavernous sinus; however, spinal invasion has rarely been observed⁷. The affected areas can be detected clearly by gadolinium-enhanced MRI studies. Other than these clinical findings, diffuse or focal hypertrophy of the meninges, brain edema, cavernous sinus thrombosis, venous congestion, secondary white matter changes, hydrocephalus, and intracranial hemorrhages may also be seen on a MRI⁹. The "dura tail sign," which is a characteristic MRI finding of meningiomas in T1WI, may also be observed¹⁰. In our case, the mass appeared to be attached to the dura with the dura tail sign; therefore, it appeared to be an en plaque meningioma rather than IHCP. Furthermore, the location of the mass was near the area frequently affected by meningiomas, rather than IHCP. Also, it was necessary to rule out metastatic brain tumors because the chest CT scan showed lung cancer, such as an adenoma with mediastinal metastasis. For the definitive diagnosis of IHCP, the histopathologic examination of the

meninges by biopsy remains crucial. It is only through this process that the confirmation of hypertrophic meninges is possible and the presence of fibrosis and non-specific acute and chronic inflammatory cell infiltration can be confirmed. Furthermore, the absence of granulation tissue and vasculitis can exclude secondary causes of hypertrophic changes in the meninges.

In the treatment of IHCP, the use of corticosteroids is effective in relieving symptoms and preventing further progress. However, during the process of decreasing the steroid dosage, recurrence of symptoms and appearance of other neural deficits can occur. In addition, complications with steroid dependency are often experienced. Combined therapy with steroids and other immunomodulatory agents has been found to be effective¹⁾, suggesting that the mechanism underlying pachymeningitis may be an immunologic process⁸⁾. Recently, successful treatment of IHCP using methotrexate has been reported^{1,3)}. In our case, we used cyclosporine to treat the scleritis. However, complications ensued following this treatment, including joint swelling and tenderness.

CONCLUSION

As in this case, IHCP, including acute SDH and metastatic dural carcinomatosis, should be considered in the differential diagnosis for refractory chronic headaches with or without cranial neuropathy. Enhanced MRI is the key tool for diagnosis. The final diagnosis is confirmed by histopathologic examination. After confirmation of IHCP, clinical improvement is achieved with high-dose steroid treatment. Clinical improvement is monitored by normalization of ESR levels.

The patient was treated successfully with immunomodulatory therapy.

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