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Case Report

Long-term lack of progression after initial treatment of idiopathic hypertrophic pachymeningitis

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ABSTRACT

Idiopathic hypertrophic cranial pachymeningitis (IHCP) is a rare inflammatory disease which is sometimes difficult to diagnose and can lead to misinterpretations of the clinical and imaging findings. The main clinical manifestations are headache, ataxia and cranial nerve palsy. In most of the reported patients continuous medication is needed to avoid disease recurrence. We present a female patient with an 8-year follow-up, no clinical regression and no need for any further medical treatment. Even though most patients with IHCP experience recurrence after diagnosis and initial treatment there were no clinical or imaging signs of relapse in our patient. Our patient is still not under any medical or surgical treatment due to the lack of any significant symptoms.

1. Introduction

Idiopathic hypertrophic cranial pachymeningitis (IHCP) is a rare inflammatory disease characterized by diffuse dural thickening, dural masses, sinus thrombosis and venous congestion.[1] Since the introduction of CT scans and MRI, IHCP has been increasingly reported in the recent literature.[2] The clinical picture can be heterogeneous including headache,[3] ataxia,[1] seizures[4] and cranial nerve palsy[5–7] which are caused by compression of the exit zone of the nerve roots by the hypertrophic basal pachymeningitis. IHCP can even imitate transient ischemic attacks (TIA).[8] The headache is probably related to dural inflammation since there is no evidence of raised intracranial pressure. MRI shows a diffusely thickened dura, usually isointense on T1-weighted MRI, that enhances strongly after paramagnetic contrast injection due to an inflammatory reaction of the pachymeninges. Adequate management is still under debate and depends on the patient's clinical features. Patients with IHCP experience recurrence with clinical or imaging signs of regression and need constant medication[1] (coricosteroids, methotrexate, azathioprine, even minocycline hydrochloride[9]) or surgical treatment. We present a female patient with an 8-year follow-up with no clinical regression after initial treatment and who did not require further medical treatment.

2. Case report

In 2002, a 47-year-old female patient was referred to a department of neurology for diplopia, unspecified headache and drowsiness that had deteriorated over the previous 2 weeks. There was no history of cardiovascular disease, diabetes mellitus or any infectious disease. No regular medication use was reported, apart from the pain killers she occasionally used for her headaches. Over the previous years the patient had suffered from day-long bitemporal headaches, but she did not use more than 2000 mg/week of paracetamol. The neurological examination revealed diplopia due to left abducens nerve palsy. There were also symmetrical enhanced tendon reflexes and bilateral Hoffmann sign. Contrast-enhanced brain CT scans revealed high signal thickening in the cerebral falx and tentorium (Fig. 1a and b). Initially she was treated with high doses of intravenous (IV) dexamethasone for 1 week with complete regression of the symptoms.

Within 1 year the patient was admitted to our department for re-evaluation, even though her clinical status remained stable, and to have an MRI scan. On a T1-weighted MRI scan with gadolinium there was enhancement in the falx and the tentorium, showing a markedly hyperintense thickened right tentorium, consistent with IHCP(Fig. 1e–g), while on a T2-weighted MRI and a T1-weighted MRI without gadolinium enhancement (Fig. 1c) the same area was hypointense. During her 7 years' follow-up, there was always the same enhancement and thickening in the falx and tentorium on brain MRI.

Other causes of pachymeningitis, including intracranial hypotension, sarcoidosis, tuberculosis meningitis, rheumatoid arthritis, syphilis, meningioma and occult malignancy were excluded during the diagnostic work-up (Table 1). Her erythrocyte sedimentation rate was 15 mm/hour and results of the hemogram, biochemistry profile, rheumatoid factor, complement activation, anti-nuclear antibodies, anti-neutrophilic cytoplasmic antibodies, angiotensin converting enzyme, homocysteine and tumour markers were

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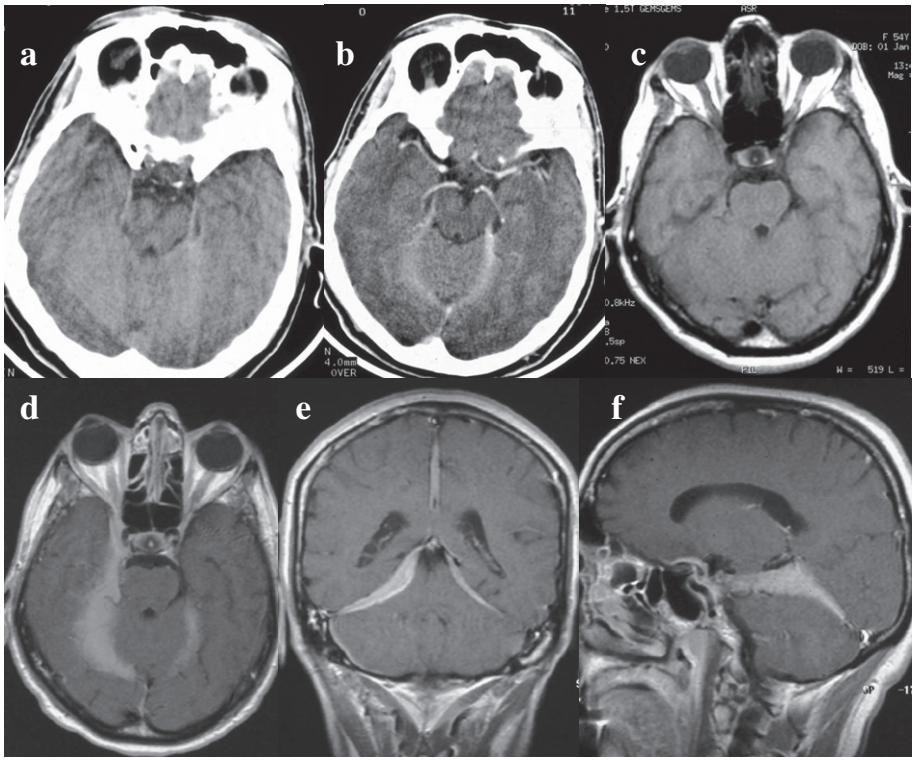

Fig. 1. (a) Axial non-contrast and (b) contrast-enhanced CT scans showing increased enhancement of the tentorium; (c) axial T1-weighted MRI without gadolinium a few days after the CT scan showing hypointense thickening of the tentorium; gadolinium-enhanced (d) axial, (e) coronal, (f) sagittal T1-weighted MRI consistent with idiopathic hypertrophic cranial pachymeningitis showing a markedly hyperintense, thickened right tentorium.

Table 1
Causes of pachymeningitis (abnormal thickening and enhancement of the dura mater on gadolinium-enhanced MRI)

Idiopathic cranial or spinal pachymeningitis
Intracranial hypotension
Spontaneous hypotension
Hypotension occurring after cerebrospinal fluid drainage
Infection
Lyme disease
Mycobacterium tuberculosis
Syphilis
Fungal infection
Cysticercosis
Systemic autoimmune diseases and vasculitides
Wegener granulomatosis
Rheumatoid arthritis
Temporal arteritis
Sarcoidosis
Sjogren syndrome
Malignancy
Meningioma
Dural carcinomatosis
Metastatic disease in adjacent skull

normal. The venereal disease research laboratory (VDRL) test, syphilis *Treponema pallidum* haemagglutination test (TPHA) and human immunodeficiency virus serology were negative. A polymerase chain reaction assay for the detection of *Mycobacterium tuberculosis* DNA was negative. Cerebrospinal fluid (CSF) analyses showed absence of any cells, a normal CSF pressure and negative virologic, parasitologic and bacteriologic analyses. Serum and cerebrospinal fluid immunoelectrophoresis were normal. Chest and abdominal CT scans were also negative.

Although this patient was initially misdiagnosed with a subarachnoid haemorrhage, there was a complete regression of the clinical symptoms from day 2 of her admission. She was treated with steroids.

The patient consented to a meningeal biopsy to confirm the diagnosis. It was decided to undertake a transsphenoidal biopsy as it is a minimally invasive method. In the dural biopsy specimen there were no signs of neoplastic cells, or vasculitis. Granulation tissue infiltrated by lymphocytes and plasma cells was identified, suggesting inflammatory changes consistent with IHCP.

For the subsequent 8 years the patient was followed at least twice per year and had a brain MRI once every 2 years. She complained of her usual mild headache that did not interfere with her everyday activity. Initially we thought of commencing regular medication with corticosteroids, but bearing in mind the possible side effects and the minimal clinical symptoms, the patient consented to remain on oral analgesics, in general, paracetamol.

3. Discussion

Idiopathic hypertrophic cranial pachymeningitis remains a rare inflammatory disease and its cause is still speculative, from infectious agents, mucopolysaccharidosis and intrathecal toxin to fibrosclerotic disease.[2] There are diseases that can imitate the clinical and imaging findings of IHCP including sarcoidosis,[6] primary central nervous system vasculitis,[10] rheumatoid arthritis,[10] Wegener granulomatosis[11] and meningioma.[12]

The role of the dural biopsy is important in the diagnostic process to exclude other causes of IHCP.[13] Considering the high number of relapses reported in the literature, biopsy at an early disease stage must be discussed. Non-specific inflammatory changes in the thickened dura are the most common histopathologic features.

Most patients with IHCP experience recurrence after diagnosis and initial treatment. Our patient showed no clinical signs of relapse and is still not taking any of the recommended medications (corticosteroids, methotrexate or azathioprine), as she is not suffering from any significant symptom.

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4. Conclusion

IHCP is an infrequent disorder that can be identified with CT scans and MRI. In the acute phase, the patient needs early treatment with corticosteroids to achieve remission. However, a patient who is diagnosed with IHCP – if there are no clinical or imaging signs of recurrence – may not need constant medication. Adequate management is still a matter of debate and depends on the patient's clinical features.

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