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CASE REPORT

Idiopathic hypertrophic craniocervical pachymeningitis

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Abstract
Purpose Hypertrophic craniocervical pachymeningitis (HCP) is a rare disease causing chronic inflammatory hypertrophy of the cranial and spinal dura mater. To increase awareness of this condition, we report the details of a case here.
Methods We reviewed the case of a 78-year-old man presenting with a rare case of HCP and summarized the clinical features, laboratory evaluations and treatment of the case.
Results In this case, the HCP involved the intracranial dura and high cervical regions, manifesting as lower cranial nerve palsies, headache, and neck pain, developing over 7 months. Magnetic resonance imaging revealed thickening of the dura in the craniocervical region with peripheral enhancement. Steroid therapy was commenced and the symptoms improved rapidly.
Conclusions HCP can be diagnosed by MRI and laboratory investigations. In this case corticosteroid treatment was effective, although care must be taken when slowly reducing the dose. This case highlights HCP as a cause of progressive cerebellomedullar and cervical spinal cord symptoms.

Keywords Hypertrophic craniocervical pachymeningitis · Neuroimage · Treatment

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Introduction

Hypertrophic pachymeningitis (HP) is a rare disorder caused by thickening of the intracranial or spinal dura mater. It occurs in either of these areas alone or as a craniospinal form, which is extremely rare. Chronic headache and multiple cranial neuropathies, radiculopathy, and myelopathy are the most common clinical manifestations. Here, we report an unusual case of HCP involving both the cranial and spinal dura mater.

Case report

In April 2011, a 78-year-old man was admitted to a neurological department with a 3-month history of dysphagia and coughing when attempting to drink. A neurological examination revealed no gag reflex and that the upper and lower limb tendon reflexes were hypoactive. Cerebrospinal fluid (CSF) protein levels were moderately elevated and cell counts were normal. Although a diagnosis was not made, he received 30 mg prednisolone daily for 1 week and improved quickly. In June 2011, the patient was readmitted to a neurological department with a 20-day history of cervicodynia and headache, soon followed by vertigo, nausea, and vomiting. A detailed review of his history revealed that the patient had experienced intermittent neck pain for the previous 2 years. A neurological examination revealed a stiff neck, good motor function, and no sensory deficits or ataxia. However, his C-reactive protein (CRP) level was 32.8 mg/L (normal 0–8) and his erythrocyte sedimentation rate (ESR) was elevated. CSF examination showed an absence of pleocytosis, increased protein levels and normal sugar.

Tests for HIV, syphilis antibodies, thyroid function, rheumatoid factor (RF), antinuclear antibodies, antibodies to Sm, double-stranded DNA, antineutrophil cytoplasmatic antibodies (p-ANCA and c-ANCA), U1RNP, Ro (SS-A), La (SS-B), Jo-1 and tumor markers were all negative. Hemogram, renal and liver function tests, and prostate-specific antigen (PSA) levels were normal. The tuberculin skin test was negative and no tuberculous bacilli were found by CSF culture. A lung CT revealed lung bronchiectasis while abdominal enhanced CT showed multiple liver cysts, a slightly widened bile duct, and kidney cysts. Gd-DTPA enhanced cervical MRI showed a thickening of the dura in the craniocervical region (from C1 to C5) with peripheral enhancement (Fig. 1a–c). A brain MRI showed focal thickening and enhancement of the dura mater over the clivus. A PET study showed extracerebral hypermetabolism at the clivus and the upper cervical spine, but did not find evidence of a tumor. Considering the history and results of other investigations, a diagnosis of possible "idiopathic hypertrophic craniocervical pachymeningitis" was made and the patient began steroid therapy. Following his first admission, steroid treatment was withdrawn too quickly and the patient suffered from a relapse 3 months later. Therefore, on his second admission he was treated with steroid pulse therapy followed by a daily maintenance treatment regime of oral prednisolone (60 mg). The patient continued with an alternate-day oral maintenance dose of prednisolone (10 mg) until the last follow-up and every attempt of slowly withdrawing it had resulted in a return of the cervicodynia and headache.

Discussion

This case can be clinically summarized as an older man with a gradual onset of bulbar paralysis, chronic cervicodynia, and headache. Multiple cranial nerves, including VIII, IX, X, and XI, were involved. Radiological studies revealed thickened dura in the craniocervical region with peripheral enhancement. The patient was treated with prednisone, which resulted in excellent clinical improvement.

A craniospinal form of this condition is extremely rare, with few cases described in the literature [1, 2]. Hypertrophic craniocervical pachymeningitis (HCP) occurs when

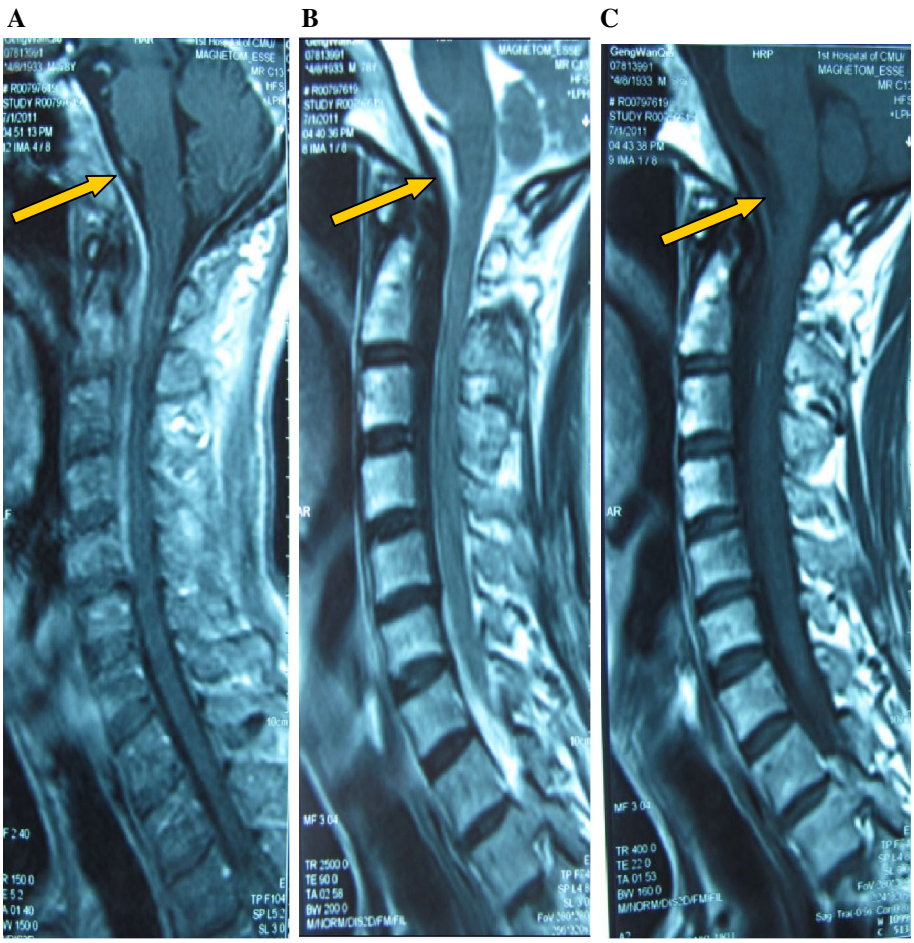

Fig. 1 **a** Sagittal T1-weighted scans after gadolinium-diethylene-triaminepentaacetic acid (Gd-DTPA) administration demonstrates linear enhancement of the thickened dura in the craniocervical region. **b** Sagittal T2-weighted scans MRI shows a dural-based mass of lower T2 signal intensity in the craniocervical region. **c** Sagittal T1-weighted scans MRI shows a dural-based mass of low T1 signal intensity in the craniocervical region

cranial HP extends downwards to the spine [3]. Chief symptoms are chronic headache, cranial nerve palsies, radiculopathy, and myelopathy occurring either alone or in combination [4]. Headache is a universal symptom, which can be focal or diffuse, and at times may be the only symptom for years before other symptoms manifest. Cranial nerve VIII is the most frequently involved cranial nerve, with the next-most frequent being the cranial nerves IX, X and XII [5]. Our patient presented with lower cranial palsies, chronic neck pain, and headache, but no complaint of myelopathy. This may be because the thickened dura matter did not compress the spinal cord. The diagnosis of "idiopathic hypertrophic pachymeningitis" relies on the exclusion of meningioma, lymphoma, tuberculosis, sarcoidosis and other diseases because these may present in a very similar fashion. In our patient, we found no evidence of cancer, infection, or autoimmune disorders based on the tests performed.

HCP has been described as a dural-based mass of low T1 and lower T2 signal intensity extending from the intracranial to spinal area with strong linear enhancement [6]. Friedman and Dumont [7, 8] proposed that MRI revealing an extramedullary mass of low T1 signal intensity and lower T2 signal intensity along with peripheral enhancement is highly indicative of HCP. This may be caused by a peripheral zone of active inflammation along the lesion periphery, rather than attenuated fibrosis throughout the central portions.

The CSF showed increased protein levels and lymphocytic pleocytosis. Elevated CRP levels and ESR were found in approximately 97 % of the patients [9]. A dural biopsy is usually sufficient for pathologic confirmation of the diagnosis; however, our patient refused to undergo this procedure. Corticosteroids have been considered to be the mainstay of conservative treatment and the clinical improvement with these in this case was excellent [9]. In addition, steroid treatment should be tapered off extremely slowly, because recurrence has occurred in many cases when the dose of prednisolone was reduced. If conservative treatment fails, surgery for decompression is required. Surgery serves a dual purpose in achieving the immediate decompression of the spinal cord and obtaining a diagnosis. Recently, the modern surgical strategies of the video-assisted anterior surgical approach and posterior fixation have emerged, being an important update to the classical surgical decompression procedure [10–13]. The patient showed a marked improvement in his clinical symptoms after prednisolone treatment and continues on a low-dose alternate-day steroid therapy, with his disease symptoms being stable at the last follow-up.

We hope that our report will raise awareness of HCP as a cause of progressive cerebellomedullar and cervical spinal cord symptoms.

Conflict of interest None of the authors has any potential conflict of interest.

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