

Idiopathic hypertrophic pachymeningitis as a rare cause of cervical compressive myelopathy

ABSTRACT

The spinal form of idiopathic hypertrophic pachymeningitis (IHP) is a rare condition characterized by a chronic progressive diffuse inflammatory fibrosis of the dura mater, which may evolve to the compression of the spinal cord. We present a case report about IHP focusing on its features in magnetic resonance imaging, which are determined by an intradural extramedullary mass in the cervical spine showing hypointensity on T2-weighted images and peripheral enhancement, causing compression of the spinal cord. Histological analysis showed a nonspecific chronic inflammatory process in dense fibrous tissue. The patient had a good outcome after therapy with steroids.

Keywords: Idiopathic hypertrophic pachymeningitis, cervical spine, magnetic resonance imaging

INTRODUCTION

The spinal form of the idiopathic hypertrophic pachymeningitis (IHP) was first described by Charcot in 1869, being an extremely rare condition.^[1] This disease is characterized by a chronic progressive diffuse inflammatory fibrosis of the dura mater, which may eventually compress the spinal cord. This compressive mechanism determines symptoms related to radiculopathy and/or myelopathy.^[1]

On magnetic resonance imaging (MRI), the characteristic findings of the spinal form of IHP are hypointensity on T2-weighted images, on an intradural extramedullary mass, which extends >1 vertebral level, with 3 distinct patterns of enhancement: (1) homogeneous, (2) linear or nodular enhancement,^[2-6] and (3) peripheral (presumably due to peripheral active inflammation and central fibrosis).^[7,8]

The treatment can be done with corticosteroids; however, some cases require azathioprine/cyclophosphamide therapy or surgical intervention for decompression.^[1]

CASE REPORT

A 51-year-old male patient presented moderate and constant pain in the right scapular region, with irradiation to the back. At that year (2009), the diagnosis was not elucidated.

In September 2014, cervical complaints began, characterized by cervical pain, worsening with movement, with irradiation to his upper limbs. After 3 months, the symptoms progressed to weakness and fine motor impairment of his arms, leading the patient to be admitted to the clinical neurology ward.

Initially, infectious causes were investigated. The following hypotheses were ruled out through clinical, laboratory, and pathological evaluation: tuberculosis, cryptococcosis, Lyme's

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disease, syphilis, AIDS, lupus, vasculitis, bacterial meningitis, neoplasms, and sarcoidosis.

MRI of the cervical and thoracic spine was performed, and the analysis suggested IHP [Figure 1]. The main findings were as follows: a fusiform intradural extramedullary mass in the posterior region of the vertebral canal, extending from C2 to T1 levels, showing marked hypointensity on T2/short tau inversion recovery weighted images; hypointensity on T1-weighted images; and linear peripheral enhancement after contrast administration. The mass was causing compression of the spinal cord, and the space of cerebrospinal fluid was decreased between C2 and T1.

Six days after the MRI, the patient underwent a dura mater biopsy at the C3 level, without any intraoperative complications. Histological analysis showed a nonspecific chronic inflammatory process in dense fibrous tissue, negative for the IgG4 antibody test.

The patient received treatment and all clinical support at the infirmary, with discharge after 3 months of hospitalization, with 60 mg/day of prednisone.

In outpatient follow-up, there were no motor or sensory complaints. Furthermore, a control MRI was performed

6 months after the first examination, demonstrating reduction in the intradural extramedullary mass in the posterior portion of the spinal canal.

DISCUSSION

Whenever there is thickening of the dura mater, neoplasms, infectious diseases, and inflammatory diseases, such as sarcoidosis, granulomatosis with polyangiitis, and IgG4-related hypertrophic pachymeningitis should be considered.^[6-8]

Neurosarcoidosis and IHP are rare causes of hypertrophic pachymeningitis and may be considered as the diagnosis of exclusion.^[6,9] The biopsy of the meninges is necessary if the diagnosis is not yet clear.^[9]

Meningeal sarcoid lesions and IHP do not have specific MRI characteristics to distinguish them from bacterial, fungal, or tuberculous infections of the meninges, leukemic infiltration, or carcinomatous meningitis.^[9] Some clues for neurosarcoidosis are pathological enhancement of brain parenchyma and leptomeninges, with homogeneous nodular or diffuse enhancement, mainly in the basal cisterns and hypothalamic regions.^[9]

Spinal IHP affects mainly the cervical and thoracic regions, causing symptoms of radiculopathy. The histopathological feature is dense fibrosis and inflammatory cell infiltration,^[8] as found in our patient.

IgG4-related disease is an immune-mediated condition that affects multiple organs, mimicking malignant, infectious, and inflammatory disorders. It is a common cause of hypertrophic pachymeningitis^[10] and should be promptly discarded during clinical investigation. Serum IgG4 concentrations may be used for screening but cannot be considered as a single diagnostic marker. The diagnosis is determined by the biopsy, which shows predominant lymphoplasmacytic infiltrate, storiform fibrosis, and obliterative phlebitis.^[10]

After empiric treatment with corticosteroids, most of the diseases investigated would present partial or complete remission, such as vasculitis, lupus, and sarcoidosis. In our case, cerebrospinal fluid examinations and dura mater biopsy were determinant to exclude such diseases.

CONCLUSION

There are many different causes for hypertrophic pachymeningitis, and each one has to be ruled out during investigation. In this paper, we report a rare case of cervical compressive myelopathy

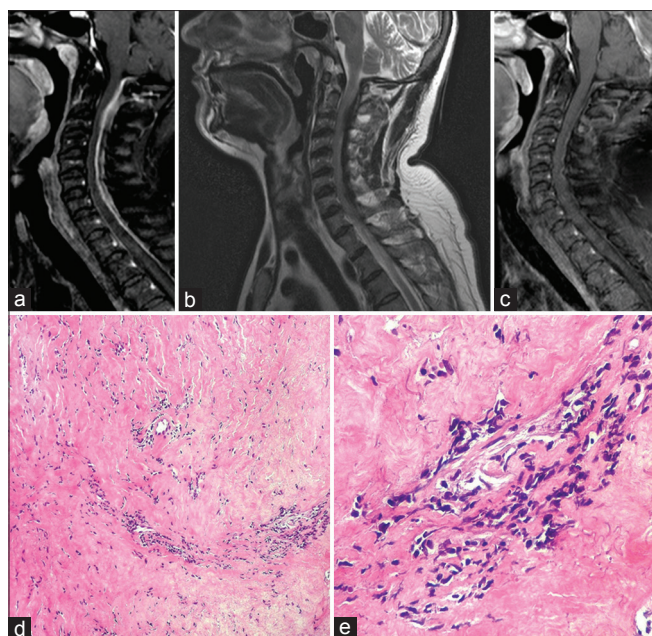


Figure 1: (a) Sagittal T1-weighted images after contrast revealed a fusiform intradural extramedullary mass in the posterior region of the vertebral canal, between C2 and T1 levels. It shows linear peripheral enhancement after contrast administration. (b) Sagittal T2-weighted images showed a marked hypointensity within this mass. (c) Sagittal T1-weighted images after contrast, performed 6 months later, showed important improvement after corticoid treatment. (d) Dural biopsy showing intense fibrosis and nonspecific inflammatory infiltrate (H and E, $\times 100$). (e) Dural biopsy showing intense fibrosis and nonspecific inflammatory infiltrate (H and E, $\times 400$)

caused by IHP, diagnosed according to the exclusion of secondary causes, with a good response to steroids.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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