

Case Report

Idiopathic hypertrophic pachymeningitis

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ABSTRACT

Idiopathic hypertrophic pachymeningitis is a rare form of granulomatous meningitis. It is mainly a diagnosis by exclusion. Here we are presenting a case of a young female presented with multiple cranial nerve palsies who was diagnosed to have hypertrophic pachymeningitis with the help of MRI brain. All available investigations showed negative results and so possibility of Idiopathic Hypertrophic Pachymeningitis was considered. The patient responded well to steroid therapy.

Keywords: Idiopathic, Pachymeningitis, Steroids

INTRODUCTION

Pachymeningitis is the inflammation of duramater. Idiopathic hypertrophic pachymeningitis is mainly a diagnosis by exclusion as numerous pathological entities produce thickening of the pachymeninges. This condition is extremely rare. It is a fibrosing inflammatory process which involves the duramater, including the tentorium.¹ This condition responds to steroid therapy but recurrences can occur.²

CASE REPORT

A 17 year old female presented with complaints of headache of 6 months duration. She also gives history of double vision and vomiting occasionally for the past 3 months. One month back she developed facial deviation to left followed one week later by incomplete closure of right eye which were persisting. No history of any fever, trauma, altered sensorium or seizures.

Nervous system examination showed a sensory impairment in ophthalmic division of right trigeminal nerve along with LMN type of right facial nerve palsy

and bilateral abducens nerve palsies. Pupils were normal and reacting to light. Fundus was normal. Deep tendon reflexes were brisk bilaterally and plantar was bilaterally flexor. There was no neck stiffness. Other system examinations were within normal limits.

Based on the history and physical findings the differential diagnoses considered were vasculitis, demyelination, chronic meningitis, metastasis and pachymeningitis.

Blood investigations including complete blood count, blood sugar, renal and liver function tests were all normal. Mantoux test, Chest X-ray, Ultrasonogram of abdomen, CT brain and CSF study were also within normal limits. So an MRI brain was taken which showed thickening and enhancement of pachymeninges of tentorium and falx suggestive of hypertrophic pachymeningitis. Vasculitic workup like antinuclear antibody, rheumatoid factor, antineutrophil cytoplasmic antibodies were all negative. Angiotensin converting enzyme level was normal. Blood borne virus screen including HIV, HBsAg, HCV were all negative. CSF study for TB-PCR, VDRL and malignant cells were also negative.



Figure 1: MRI brain showing thickening and enhancement of pachymeninges of falx and tentorium.

With all these clinical and investigation findings a final diagnosis of Idiopathic Hypertrophic Pachymeningitis was made. Patient was given anti oedema measures & steroids with which she had very good symptomatic improvement.

DISCUSSION

Hypertrophic cranial pachymeningitis is a rare, idiopathic form of granulomatous pachymeningitis. These lesions typically cause progressive cranial nerve palsies, headaches, and cerebellar dysfunction. They occur in patients of all age groups but the peak incidence is in the sixth decade. Hypertrophic cranial pachymeningitis is best identified by magnetic resonance imaging. The diagnosis is established by excluding all other granulomatous and infectious diseases.² A dural biopsy is

essential to confirm the diagnosis. Pathological findings consist of thick fibrous dura often associated with chronic inflammatory cell infiltrate comprising lymphocytes and plasma cells. Giant cells, caseation necrosis or epithelioid granuloma or evidence of vasculitis are usually not seen.³ Hypertrophic cranial pachymeningitis is initially responsive to steroid therapy, but in most cases it recurs or progresses despite treatment. Surgical excision of granulomas is occasionally necessary to alleviate a mass effect.² Now there are some evidences suggesting that Idiopathic hypertrophic pachymeningitis is related to the newer disease entity called IgG4 related sclerosing diseases.⁴

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