IDIOPATHIC HYPERTROPHIC PACHYMENINGITIS

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INTRODUCTION

Idiopathic hypertrophic pachymeningitis (IHP) is a clinical disorder characterized by localized or diffuse thickening and fibrosis of dura matter with no underlying cause demonstrable [1]. IHP is a rare disorder affecting men more than women [2] with peak incidence in 6th decade of life [3]. Common clinical features include headache, multiple cranial nerve palsies and cerebellar dysfunctions occurring alone or in combination. We present a case of a lady, who was diagnosed in our department after suffering from the disease for some time with no diagnosis available.

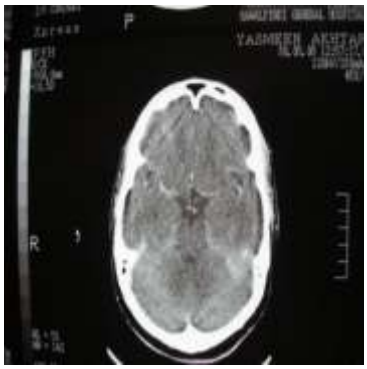
CASE HISTORY

Middle aged lady presented with one month history of drooping of left eyelid along with double vision and pain. She also suffered similar complaints two years back which recovered with treatment from local doctor and homeopath. She also had her computerized tomogram (CT) scan done in last episode with no report available. She is also hypertensive for the same period and complains of palpitation occurring off and on. She had been taking irregular treatment for above complaints. There was no history of surgery, blood transfusion, bronchial asthma, tuberculosis (TB) or Jaundice.

She is non-smoker and non –addict .Married for the last 35 years with 3 daughters and one son. Temperature 98oF BP 120/70 mm of Hg. Cranial nerves III, IV and VI and VII on left affected

Rest of the systemic examination was normal. CSF routine examination was within normal limits.

Previous CT scan was reviewed (Fig 1) and contrast enhanced magnetic resource imaging (MRI) brain done (Fig 2) which showed smooth thickening of dura of tentorium, cerebello-pontine (CP) angle left extending to anterior parts of midbrain. Patient was put on steroids and showed significant improvement after 6 weeks.



Fig, 1: CT Scan shows thickening of dura matter.

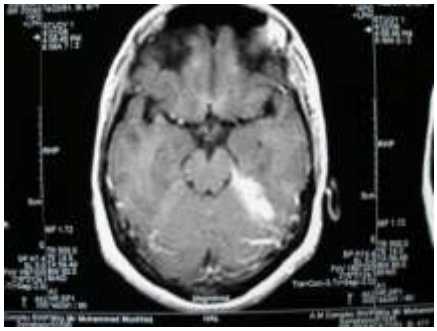


Fig. 2: MRI confirms thickening of dura of tentorium.

DISCUSSION

Hypertrophic pachymeningitis is a unique disorder of diverse etiology. It is characterized by thickening of dura matter, without intra axial involvement, resulting in neurological dysfunction.

The condition may be broadly described as “primary” or “idiopathic” where no cause is found and secondary where identifiable causes co-exist: however exact etiopathogenesis of this entity is still unknown. The underlying disorder may be an autoimmune, infectious or neoplastic process [4, 5]. If the cause of meningeal changes

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remains unidentified after an extensive workup, IHP is diagnosed [6].

IHP is more commonly seen in males with highest incidence in sixties. Patients most commonly present with chronic headache which often resembles chronic migraine with or without other neurological manifestations [7]. Other neurological manifestations include cranial nerve palsies, cerebellar ataxia, seizures and neuro-opthalmic symptoms ranging from visual field defects to complete blindness.

IHP is being increasingly recognized with the advent of CT and MRI. MRI is superior to CT in the diagnosis of hypertrophic pachymeningitis. Unenhanced CT shows thickened hyperdense dura, typically involving the tentorium, falx and prepontine region, which enhances markedly after contrast administration.

On MRI, the thickened dura matter appears isointense or hypointense on T1WS and hypointense on T2WS often associated with a hyperintense edge. Thickening is better appreciated in the coronal and sagittal images. Contrast administration reveals uniform enhancement of thickened meninges. The low MRI signal represents dense fibrosis and enhancement suggests inflammation. Rarely nodular pseudotumoural thickening of dura matter, mimicking multiple meningiomas may occur [8]. Sometime biopsy is usually required to establish the diagnosis of IHP and to exclude other causes of hypertrophic pachymeningitis. Pathological findings include thick fibrous dura often associated with chronic inflammatory cells infiltrate consisting of lymphocytes and plasma cells.

The optimal treatment of IHP is unknown. Spontaneous resolution of clinical symptoms , signs and dural thickening has been reported .Corticosteroid therapy is often effective in arresting progress of disease [10]. Rarely patient have worsened while on steroid therapy.Cyclophosphamide and azathioprine have been found beneficial in steroid non responsive patients. Our patient responded well to steroids with improvement in symptoms and signs over a period of about six weeks. Surgical excision is an option for patients with mass effect. VP shunting is done in symptomatic hydrocephalus. Aim of therapy is to prevent permanent damage to neural structures.

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