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IDIOPATHIC HYPERTROPHIC PACHYMENINGITIS - A CASE REPORT

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Abstract :
Idiopathic Hypertrophic Pachymeninghits is a rare pathological state, with still unclear aetiopathogenesis.. It presents with headache, cranial Neuropathies and ataxia occurring alone or in combinations. The disease was diagnosed with magnetic resonance imaging (MRI) and histopathological assesment of the pachymeningeal biopsy specimen.The disease may have remitting and relapsing course and usually response to steroids. We report a case of 40 year old man with cranial veriety of this desease. Our patient presented with headache, reccurent episodes of generalised tonic clonic seizures, behavioural disturbance and defective vision in right eye. MRI showed prominent pachymeningeal thickening on imaging. Dura mater biopsy revealed meningeal thickening and non specific chronic inflammation of the dura. Clinical improvement was noted in our patient. Early institution and long term maintenance of steroid theraphy prevents neurologic sequelae.

Keyword : Idiopathic Hypertrophic Pachymeningitis

Introduction:

Hypertrophic pachymeningitis (HP) is a rare disorder of diverse etiology, characterized by fibrosing inflammatory process that thickens the dura mater.[1] Common clinical features include headaches, cranialneuropathies and ataxia.[2] Dural biopsy is essential to exlude secondary causes of pachymeningitis.[3] Until 2008, 60 treated cases of HP have been reported in the English literature.[4] Three more cases were reported from India in 2009.[5,6]There is paucity of data on biopsied cases of HP. From 1997 to 2008, Goyal et al.,[7] Sylaja et al.[3] and Shobaha et al,[8] have documented, respectively, two, four and five cases of biopsy –confirmed “idiopathic” hypertrophic cranial pachymeningitis (IHCPM) from India. We herewith report one biopsy – proven case of IHCPM from our hospital.

Case Report:

A 40 years old man presented in March 2012 with recurrent episodes of generalized tonic clonic seizures of 4 years duration. He had headache, behavioural abnormality in the form of episode of anger outburst. He also had defective vision in right

Investigations	
TC 8200 cells / cmm	CSF Analysis :
DC P62% L36& E2%	sugar 76mgs%
Hb 12gms%	Protein 62mgs%
ESR 44mm/1hr	Cell count - Acelluar
PCV 34%	AFB – staining negative
Platelet count 2.30 lakhs/cumm	Globulin – Negative
Metabolic Parameters Normal	CSF Culture – Negative
Serology for HIV, VDRL, HBSAg – Negative	MRI Brain (Plain and Contrast) – Diffuse
Sr.ACE level : 34 U/L (8-65 u/l)	pachymenigial, thickenic with intese contrast
ANA/C-ANCA/P-ANCA-Negative	enchancement seen.
HRCT Chest Normal	Meningial Biopsy – Non specific lymphocytic
EEG – Normal	infiltration and thickening of dura mater seen. S/o
	chronic ideopathic hypertropic pachymeningitis.

eye for 7 months. There were no constitutional symptoms. He had short burst of irrtablity, lack of insight, MMSE 20/30, visual acuity diminished in right eye in the from of perception of hand movements only. Relative afferent papillary defect (RAPD) present in right eye. His fundi show bilateral primary optic atrophy.

Bilateral finger flexion is brisk; bilateral watenburg sign is positive.Premitive reflexes : Bilateral Palmomental present.Grasp reflex present.Glabellar Tap: Non accommodativeRest of the neurological examination is normal.

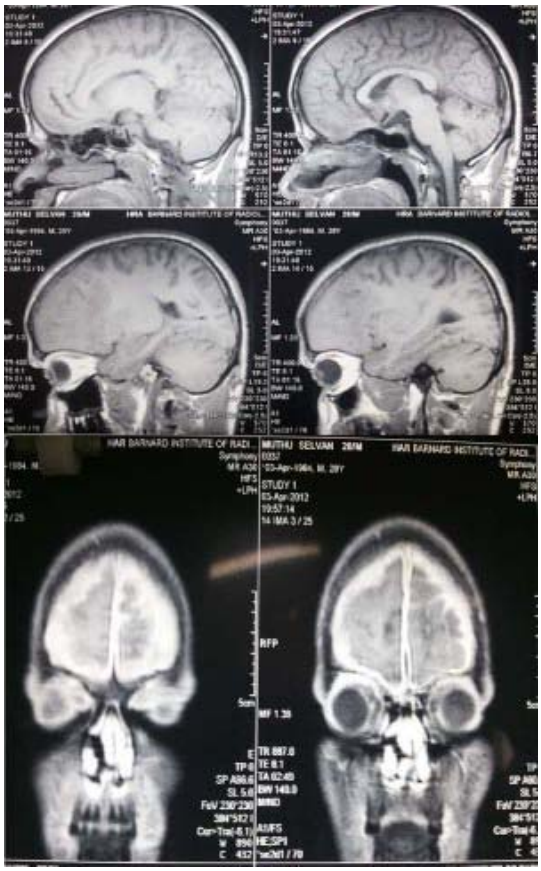
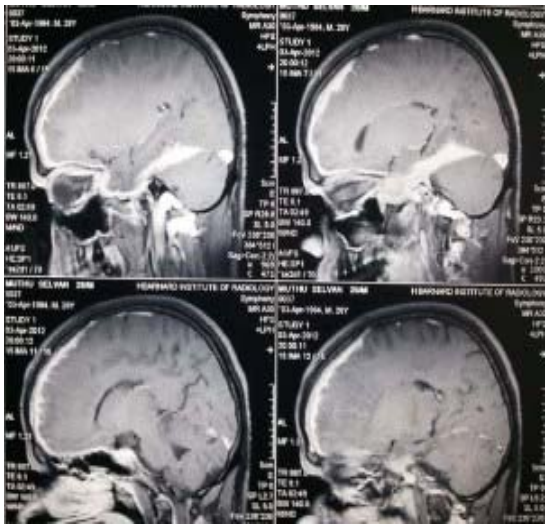


Fig-1: MRI brain T1 Sagital images



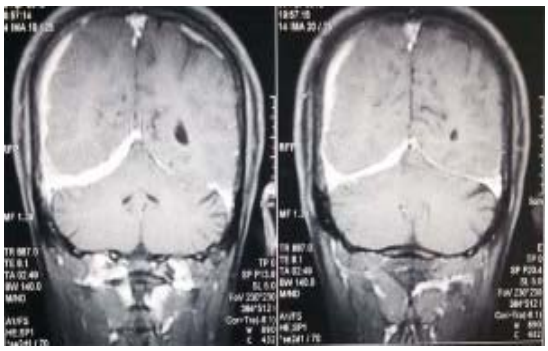
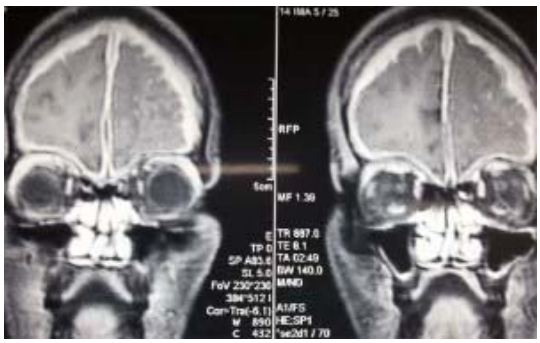


Fig-2: MRI brain T2 Coronal images showing Pachymeningeal Thickening and Contrast enhancement

Fig-3: MRI brain T2 sagittal images showing Pachymeningeal Thickening and Contrast enhancement Fig-4: MRI brain T2 Coronal images showing Pachymeningeal Thickening and Contrast enhancement



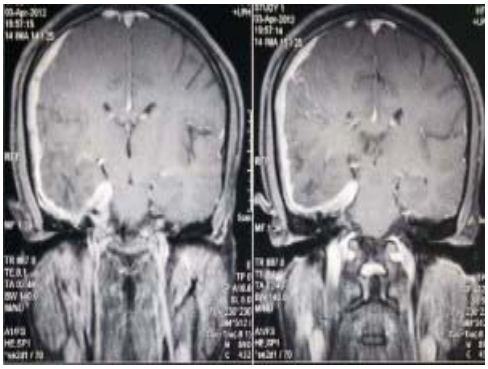


Fig-5: MRI brain T2 Coronal images showing Pachymeningeal Thickening and Contrast enhancement
Fig-6: MRI brain T2 Coronal images showing Pachymeningeal Thickening and Contrast enhancement
Fig-7: MRI brain T2 Axial images showing Pachymeningeal Thickening and Contrast enhancement

Discussion:

IHCPM is a poorly understood inflammatory disease involving the dura mater of the skull base, tentorium and falx cerebrii.(9) HP may be “idiopathic” or “secondary,” where identifiable causes coexist, although their definite relationship may be debatable.[1,10]

Etiopathogenesis:

The exact etiopathogenesis is unknown. It may be an autoimmune disorder or occurring as a direct result of infectious or infiltrative pathology.[7,11] Rossi et al. demonstrated fibrosis and prominent CD4+ T-cell inflammatory infiltrate on dural biopsy in HP, suggesting a probable pathogenetic role for cell-mediated immunity.[9] Riku et al have shown that HP may be a dural lesion of IgG4-related systemic disease.[12]

Clinical Features :

IHCPM affects males predominantly. The age ranges from 20 to 78 years (mean, 51 years) The mainlinical features are headache, progressive cranial nerve palsies arid cerebellar dysfunction [9,15]resulting from compression of adjacent structures by hypertrophied pachymeninges.[3,16] Our patient had headache, GTCS, and defective vision. Seizure presentation is a rare feature. Chronic daily headache, often resembling chronic migraine, is the most common manifestation[2].Headache can be the only symptom for years before other symptoms manifest. [17] Riku and Katohave described two patterns of cranial nerve involvement based on site of dural inflammation:Cavernous sinus to superior orbital fissure and falcotentorial to posterior fossa dural involvements. [11,15] Cranial neuropathies were observed in all cases. Presentation with ataxia is less common. Diffuse ischemia, venous sinus congestion and mass effect of thickened tentorium have been incriminated.[11] Symptomatic spinal pachymeningitis either occursalone or as a craniospinal form.[10]

Investigation:

IHCPM is a diagnosis of exclusion.[3,7] A thorough workup includes search for infectious, autoimmune and neoplastic diseases. [10]An overwhelming majority have elevation of ESR.[14] Out patient had ESR of 44 mm/h, CSF in most cases shows variable lymphocytic pleocytosis.[1,11,15,17] Protiein levels are moderately elevented. CSF may be normal in one-fourth of the patients.[17] Our p tient showed lymphocytic pleocytosis and mildly elevated protein level. IHCPM is being increasingly recognized with advent of CT and MRI. CT shows thickened enhancing dura.[3] MRI is the most useful radiological method that reveals diffuse or localized thickening of dura.[15,20] Thickened dura appears isointense to hypotense on both T1 and T2W images, with uniform dense enhancement on contrast study.[7] Dural thickening isappreciated on coronaland sagittal images in the inrterhemispheric fissure, tentoriumand basal dura.[2,3] Out patient showed prominent pachymeningeal changes on imaging. Tentorial and posterior falx involvement was seen. The area of involvement correlated with the clinical picture Dural biopsy is essential to establish the diagnosis of IHCPM and to exclude other causes of pachyrneningitis.[1,8,13,15] Biopsy from an accessible site with CT or MRI documented enhancingand thickened dura mater is more likely to yield a positive etiological diagnosis[3,8,15]. Pathological findings consist of thick fibrous dura often associated with chronic inflammatory cell infiltrate comprising lymphocytes and plasma cells.[8,9,13,15] Giant cells, caseation necrosis or epitheloid granulorna or evidence of vasculitis are usually not seen.[15] Shobha et al in a recent study of 11 cases of HP found specific etiology in only six cases, while the other five cases were of an idiopathic variety. [8] Dural biopsy in our patient was consistent with IHCPM.

Differential Diagnosis:

Differential diagnoses are extensive. Tuberculous meningitis needs careful exclusion.(22) In developing countries, a majority of the patients presenting with features of IHCPM will receive a trial of ATT before alternative diagnoses are considered. Syphilitic pachymeningitis,[2,11] neurosarcoidosis,[2] Wegener's granulomatosis,[23] meningeal carcinomatosis,[2,24] en-plaque meningiomas[2] and intracranial hypotension[2] need exclusion. clinical symptomatology, imaging characteristics, absence of abnormal laboratory and CSF studies,

histopathological basis and long course of disease and responsiveness tostestrongly favor thediagnosis of IHCPM in our case.

Treatment:

The optimal treatment of IHCPM is unknown.[10,25] Untreated, the clinical course is usually marked by severe headache and progressive neurologic deterioration and vision loss.[20] Steroid is themainstay of therapy and is often effective in arresting disease progression.[6,9,10,15,21,25] Serialimaging studies may show reduction in thickness and degree of enhancement of meninges.[2]However, symptoms may become steroid-dependent.[9,21,17,25] Clinical improvementwas noted inour patient. Addition of immunosuppressive agents like azathiprine and cyclophosphamide is requiredin steroid-dependent cases.[2,13,20,21,25]The optimal treatment of IHCPM is unknown.[10,25] Untreated, the clinicalcourse is usually markedby severe headache and progressive neurologic deterioration and vision loss.[20] Steroid is thmainstay of therapy and is often effectivein arresting disease progression.[6,9,10,15,21,25] Serialimaging studies may show reduction in thickness and degree of enhancement of meninges.[2]However, symptoms may become steroid-dependent.[9,21,17,25]Clinical improvement was noted inour patient. Addition of immunosuppressive agents like azathiprine and cyclophosphamide is requiredin steroid-dependent cases.[2,13,20,21,25]

Conclusions:

HP Is an important cause of recurrent cranial neuropathies and headaches. Early institution and long- term maintenance of steroid therapy along with azathioprine may prevent neurologic sequelae.

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