

Idiopathic Hypertrophic Pachymeningitis: A Case Report from Bangladesh

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Abstract

A 52 years old lady presented with sudden severe headache with the history of similar intense headache twenty years back which ended up with left sided blindness. Her physical examination was unremarkable except optic atrophy of the left eye. Investigation included biochemical work up, imaging studies and CSF study. The MRI of brain with contrast gave the key diagnostic clue with characteristic findings of hypertrophic pachymeningitis. Other investigations helped to rule out possible etiologies and the diagnosis idiopathic hypertrophic pachymeningitis was finally made. The patient has been treated with steroid and enjoyed improvement in her yearlong symptoms. [Journal of National Institute of Neurosciences Bangladesh, 2017;3(2): 110-112]

Keywords: IHP, Meningitis, Headache

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Introduction

Idiopathic hypertrophic pachymeningitis is a rare and unique clinical entity, characterized by thickening and fibrosis of dura mater. Exact etiopathogenesis of this entity is still unknown, but it is speculated to be an autoimmune phenomenon or occur as a direct result of infectious or infiltrative pathology¹. The diagnosis of IHP relies on the exclusion of other possible causes of pachymeningitis, such as neurosarcoidosis, neurosyphilis, tuberculosis, rheumatoid pachymeningitis and Wegener's granulomatosis². The condition may sometimes mimic Tolosa Hunt syndrome or hemicranial continua³. It can now be broadly divided into two forms, 'primary' or 'idiopathic hypertrophic pachymeningitis' where no identifiable cause is found and 'secondary' where identifiable causes co-exist, although their definite role in disease causation is uncertain¹.

Common clinical features include headaches, multiple cranial nerve palsies and cerebellar dysfunction occurring alone or in combination⁴. Headache is a common symptom, which can be focal or diffuse and at times may be the only symptom for years before other symptoms manifest⁴. The headache possibly denotes dural inflammation, whereas dense fibrous encasement and ischemic damage by hypertrophic tissue is the likely explanation for the cranial nerve deficits. Uncommon clinical presentations include encephalitis, hypo-pituitarism, diabetes insipidus, venous sinus thrombosis and hydrocephalus⁶.

Case Presentation

A 52 year female, known diabetic was presented with intense, excruciating headache for last one year. Headache involved the left fronto-parietal region, along

with eye pain, nausea and occasional vomiting. She had history of similar headache 20 years back and became blind in the left side. After becoming blind, her that episode subsided. Thereafter, she did not have any headache or any other complaints in last twenty years. She has no other neurological or systemic complaints and was begging for pain relief. On physical examination she had reduced vision in left eye (no perception of light) and optic atrophy in the same eye and intra-ocular pressure was 10 mmHg in both eyes. Other general and systemic examination was unremarkable. Her investigation report revealed abnormality in brain imaging and subtle changes in some biochemical parameter. Investigations revealed an ESR of 51 mm and negative ANA, ANCA and RA factor. CSF examination showed absence of pleocytosis, a protein level of 70 mg% and normal sugar. Gram's stain, fungal smear and antigen detection and staining for AFB were negative. Serum and CSF VDRL tests were negative. CSF ADA levels were 9.6 U/L. X-ray chest and CT brain were normal and CT of orbit depicted mildly protruded left eye ball. MRI head (plain and contrast) revealed enhancing thick (1 cm) meninges⁴ along left tentorium cerebelli.



Figure I: Axial FLAIR MRI showing iso-hypointense thickening (1 cm) of left side of tentorium cerebelli

The lesion was T1hypo, T2 FLAIR hyper intensity and not partially diffusion restricted. Few tiny T2-FLAIR hyper intensity foci are seen in sub-cortical and paraventricular region of both fronto-parietal regions, which are iso-intense in T1 and are not diffusion restricted (Figure). MRA and MRV of cerebral vessels were normal. Considering the history and results of other investigations, a tentative diagnosis of possible 'idiopathic hypertrophic pachymeningitis' was made and she was started on steroid therapy. Within days she showed a marked improvement in her clinical symptoms.



Figure II: Post contrast T1 axial, sagittal & coronal MRI showing strong enhancement of left side of tentorium cerebelli thickening

Discussion

The case presented as sudden severe headache with the history of similar intense headache twenty years back

which ended up with left sided blindness. The physical examination was unremarkable except optic atrophy of the left eye.

Investigative work up of this condition aims at excluding all possible causes of infectious, noninfectious and malignant etiologies. This includes biochemical work up, imaging studies and CSF study. Elevation of ESR is common. Tests like ACE levels, vasculitic profile, Mantoux test and cultures for bacteria and fungi should be done to rule out specific disease states. CSF is an important investigation, but it showed inconclusive results in this case. Although protein levels were moderately elevated, normal sugar level and absence of pleocytosis failed to point towards any infectious process.

Imaging is an important investigation to identify a meningeal based pathology and exclude mass lesions in the brainstem or skull base. The changes on imaging are probably related to the presence of dense fibrous tissue, with decrease in interstitial space and paucity of imageable free water, accounting for the hypointensity on both T1 and T2W images. Idiopathic hypertrophic cranial pachymeningitis usually involves dura at tentorium cerebelli, cavernous sinus and base of the skull. The extensive involvement at the anterior cranial fossa is extremely rare⁷. Presence of associated leptomeningeal enhancement or parenchymal abnormalities with the exception of brain edema should suggest an alternate diagnosis⁸⁻⁹. The initial non-contrast imaging studies in this patient delayed the diagnosis.

A dural biopsy is considered important for differentiating idiopathic from secondary forms of pachymeningitis⁴. Since many associations and secondary causes for pachymeningeal thickening and enhancement exist, pathological examination of meninges is essential. Pathological features include thickening, fibrosis and presence of inflammatory cells including plasma cells and lymphocytes. Presence of granulomas or vasculitis aids in establishing a specific etiology. In the absence of dural biopsy, clinical symptomatology, imaging characteristics, absence of abnormal laboratory and CSF studies, long course of the disease and responsiveness to steroid therapy point towards an idiopathic variety in this case.

Steroid therapy has been considered the mainstay of therapy in this disease. However, the disease progression might continue despite institution of

steroids and many patients may eventually become steroid dependent⁴. Considering occult tuberculosis, empirical use of antitubercular therapy has been advocated, however, previous studies did not show any improvement⁴. Surgery has both therapeutic and diagnostic benefits⁵. When the radiological or laboratory evaluation is uncertain, but neurological deficits are present, a prompt surgical approach should be considered¹⁰. Postoperative steroid therapy and close observation for recurrence are necessary to ensure a good long-term outcome¹⁰.

Conclusion

Thus, hypertrophic pachymeningitis is an important cause of recurrent cranial neuropathies and headaches. Hypertrophic pachymeningitis can be diagnosed with contrast MRI of brain when the clinician maintains a high index of suspicion for this condition. Nearly accurate diagnosis and treatment may alleviate patient's sufferings, and aim of the therapy is to prevent permanent damage to neural structures.

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