Hypertrophic pachymeningitis
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Introduction

Hypertrophic pachymeningitis is a rare form of diffuse inflammatory disease that causes thickening of the dura mater. It can involve the cranial or the spinal dura or both. The condition 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 relationship to the development of this condition may be debatable. This report describes two cases of hypertrophic cranial pachymeningitis and briefly discusses the clinical and radiographic findings.

Case Reports

Case 1
A 60-year-old lady presented with headache and progressive painless loss of vision in both the eyes over a period of 6 months. On clinical examination, she had no perception of light in both the eyes and ophthalmoscopic examination showed obliteration of cup with slightly hyperemic fundi. There was bilateral involvement of trochlear nerve and partial involvement of oculomotor nerve on the left side. Rest of the neurological examination was normal. Magnetic resonance imaging (MRI) of the brain showed diffuse enhancement with thickening of the meninges. Brain parenchyma was normal [Figures 1 and 2].

Case 2
A 40-year-old male came to ER with headache associated with vomiting since 2 weeks and 1 episode of right focal seizure. Neurological examination showed right hemiparesis, which recovered in 2 days. His past history was not contributory. MR imaging revealed diffuse dural enhancement and thickening, along with left fronto parieto occipital edema. His MRA and MRV were normal [Figure 3].

Both the patient’s routine biochemistry was normal, and they were further evaluated to rule out secondary causes...
The serum was negative for rheumatoid factor, antinuclear antibodies. Enzyme-Linked Immunosorbent Assay for human immunodeficiency virus, Venereal Disease Research Laboratory (VDRL) test, hepatitis B surface antigen (HBsAg) and anti-double stranded DNA were negative. Cerebrospinal fluid analysis revealed normal cell count, protein and sugar levels. Fungal and AFB culture were negative.

**Discussion**

Hypertrophic pachymeningitis is a rare disorder of diverse etiology. It was first described by Charcot and later by Naffziger and Stern.[1] Early reports were in relationship to tuberculosis or syphilis. 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 pathology.[2] Cranial pachymeningitis typically cause progressive cranial nerve palsies, headaches, and cerebellar dysfunction.[1,3,4] Seizure at initial presentation is rare.[1,3,4] Hypertrophic cranial pachymeningitis is best identified by MRI.[7] The diagnosis is established by excluding all other granulomatous and infectious diseases.[8] But in most of the cases the symptomatology and imaging characteristics are enough to come to a reasonable conclusion.

CT or MR imaging studies should be obtained to identify mass lesions in the brain stem or skull base. Non-enhanced CT scans show thickened, hyperdense dura, typically along the tentorium, tentorial ridge, falx, and preoptic brain stem with marked enhancement on contrast administration.[1,2] The T2-weighted MR images typically show relative hypointensity of the thickened meninges, which may be bordered by a thin margin of hyperintensity. Gadolinium-enhanced T1-weighted MR images show marked enhancement of the dural edges.[1,3] Diffuse dural enhancement defined as continuous dural enhancement as seen on MR imaging, encompassing at least 75% of the dural surface was seen in both of our patient. Presence of associated leptomeningeal enhancement or parenchymal abnormalities with the exception of brain edema should suggest an alternate diagnosis.[9,10] Our second patient had edema in left fronto parieto occipital region in addition to dural enhancement, possible following the focal epileptic activity. Hypertrophic pachymeningitis can be diagnosed with contrast MRI of brain when the clinician maintains a high index of suspicion for this condition.

**References**

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