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IDIOPATHIC HYPERTROPHIC PACHYMENINGITIS

Pathology

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ABSTRACT

Idiopathic pachymeningitis is a disease with thickened dura and lymphocytic infiltrate. Several causes need to be ruled out followed by immunotherapy.

KEYWORDS

idiopathic, hypertrophic, pachymeningitis

INTRODUCTION:

Hypertrophic pachymeningitis (HP) is an inflammatory process of the dura mater that can occur as a manifestation of sarcoidosis, granulomatosis with polyangiitis, and IgG4-related disease. HP is defined macroscopically by the local or diffuse thickening of the cranial or spinal dura mater, which often becomes adherent to the underlying leptomeninges. [1]

Case Study:

A 32-year-old male presented with seizures and headache for three months. Magnetic resonance imaging showed a lesion varying from 3 mm to 11.2 mm, longer frontal convexity. It was isointense on T1 and hypointense on T2 with homogenous contrast enhancement. (**Figure 1)**

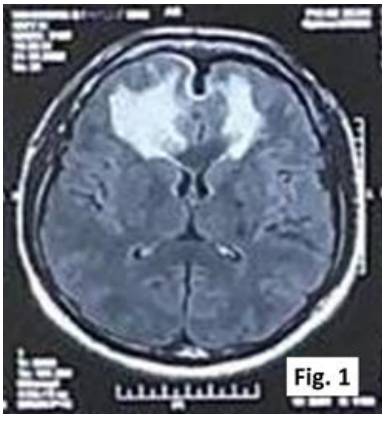

Figure 1: Radioimaging shows a lesion along extra axial basifrontal region bilaterally.

Intraoperative findings showed extra axial flat mass. Microscopic findings were consistent with HP. (**Figure 2)**

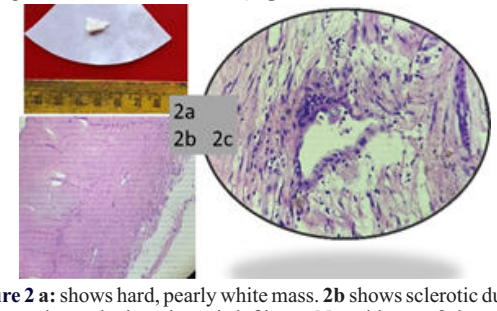

Figure 2 a: shows hard, pearly white mass. **2b** shows sclerotic dura. **2c** shows perivascular lymphocytic infiltrate. No evidence of plasma cells or granuloma.

Laboratory investigations were negative for tuberculosis, HIV, dsDNA, c-ANCA and IgG4. CSF was negative for raised ADA levels. Thus, probable causes such as granulomatous inflammation, systemic lupus erythematosus, Wegener's granulomatosis and IgG4 related disease were ruled out. By exclusion, such HP is termed idiopathic HP (IHP).

The inflammatory process and increased vessel permeability causes a marked contrast enhancement in HP. Contrast enhancement usually decreases with immunotherapy. IHP tend to have homogenous contrast enhancement whereas secondary HP tends to have central hyperintensity on T2. Tissue biopsy is the gold standard for diagnosis. Microscopy should be searched for granulomas, sclerosing thrombophlebitis, obliterative endarteritis etc for tuberculosis, Wegener's granulomatosis, IgG4 disease and neurosyphilis.

Treatment of IHP includes prednisolone, methotrexate and cyclophosphamide [2,3]

CONCLUSIONS:

HP has varied etiologies and varied clinical presentations. Radio imaging and histopathology can be symbiotic to diagnose this condition.

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