Idiopathic hypertrophic cranial pachymeningitis (IHCP) is a chronic fibrosing inflammatory condition of dura mater resulting in thickening of dura. It has a non-specific clinical picture comprising headache, vomiting, cranial nerve palsy, ataxia, raised intracranial pressure and focal neurological deficit. These symptoms and signs in IHCP are due to entrapment of cranial nerves, occlusion of CSF flow, venous sinuses and rarely arteries. The diagnosis of IHCP is based on excluding a large number of causes such as inflammatory (tuberculosis, fungal, Lyme’s disease, syphilis, HTLV), collagen vascular disorders (rheumatoid arthritis, Wegner’s granulomatosis, systemic lupus erythematosus, mixed connective tissue disease), multifocal fibrosclerosis, neoplasia (carcinoma, lymphoma, meningioma en plaque) and miscellaneous disorders such as sarcoidosis, hemodialysis, mucopolysaccharidosis and intrathecal drug administration.

The attention to IHCP has been drawn by the availability of CT and MRI scans, following which more and more cases have been described in the recent years. In this issue of Neurology India, 4 patients with IHCP have been reported by Sylaja et al and 3 patients with hypertrophic cranial pachymeningitis (HCP) by Prabhakar et al. The cases reported by Sylaja et al had undergone meningeal biopsy. In HCP, CT scan shows thickened hyperdense dura involving tentorium, falx and basal meninges which enhance intensely on contrast administration. On MRI, thickened dura meter appears isointense or hypointense on T1WI and hyperintense on T2WI sequence which is best seen in coronal or sagittal sections. There is curvilinear enhancement of the thickened meninges following gadolinium administration. In pachymeningitis, curvilinear contrast enhanced segments underneath the inner table of skull do not follow the gyral convolution which is a feature of leptomeningitis. Hypertrophic pachymeningitis may be segmental or diffuse; the former may be confused with tumor metastasis, meningioma en plaque or granuloma. Intracranial hypotension also results in similar MRI picture, therefore, it should be differentiated from IHCP.

The radiological findings although characteristic of HCP, may not reveal the underlying etiology. Presence of associated features such as granuloma, infarction, sinusitis etc may suggest an underlying cause. Exclusion of several underlying causes is an essential feature for the diagnosis of IHCP. Every effort should be made to exclude infections; as infections may flare up following corticosteroid or immuno-suppressive therapy used for the treatment of these patients. The patients with IHCP may be associated with myocarditis due to sharing of common etiological factor, which results in arrhythmias and sudden cardiac death. Electrocardio-gram, therefore, should be carried out in all the patients with IHCP.

Meningeal biopsy is essential for diagnosing IHCP for excluding other causes. There are reports of Pseudomonas aeruginosa, Propionibacterium acnes and otitis media producing HCP. In the case with otitis media, although the organism could not be grown, there were microabscesses and the patient responded to antibiotics. It is, therefore, also important to culture the meningeal biopsy for bacteria. In a study on the role of meningeal biopsy in 25 patients with meningitis; the meningeal biopsy was diagnostic in 5 patients only, revealing tuberculosis in 1, neoplasia in 3 and granulomatous angiitis in 1 patient.
patients, the biopsy, although abnormal, it could neither identify the cause nor alter the management of the patients. The meningeal biopsy should be obtained from the enhancing area to increase the diagnostic yield. In a study on 37 patients with chronic meningitis, definitive diagnosis by meningeal biopsy was possible in 39% which increased to 80%, if the biopsy was obtained from enhancing area; whereas the biopsy from non enhancing area was positive in 9% patients only. In some patients, a sequential biopsy may be necessary if the initial biopsy is inconclusive or patient is deteriorating. In this study, a repeat biopsy was carried out in 4 patients and revealed adenocarcinoma, sarcoidosis, demyelinating disease and chronic inflammation in 1 patient each. The patient diagnosed with IHCP should be closely followed. We have managed a patient with HCP and followed him up without giving corticosteroid. Six months later, he developed tubercular cervical lymphadenopathy.

The diagnosis of IHCP should be made with due precaution especially in our country where infections are common. Attempt should be made to exclude other underlying conditions before prescribing corticosteroid or other immunosuppressant. In view of limitation of radiology and meningeal biopsy, the patient should be closely monitored and any deterioration should lead to review of the patient and even repeat biopsy.

References