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Case Report

Cranio cervical tuberculous hypertrophic pachymeningitis

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Abstract

Background: Hypertrophic pachymeningitis is a unique clinical entity characterized by fibrosis and thickening of dura mater resulting in neurological dysfunction. It could be idiopathic or due to variety of inflammatory and infectious conditions. Tuberculous hypertrophic pachymeningitis involving cranio cervical region is rarely reported.

Case Description: A 50-year-old female presented with history of progressive quadriparesis and stiffness of neck for 2 years, dysphagia to liquid for past 3 months. Her condition rapidly deteriorated when another physician prescribed her corticosteroid. Physical examination revealed high cervical compressive myelo-radiculopathy with lower cranial nerve palsy and neck rigidity. Series of serum analysis, cerebrospinal fluid (CSF) study and contrast magnetic resonance imaging (MRI) clinched the diagnosis. She improved on antitubercular treatment.

Conclusion: In case of multilevel cervical compressive myelo-radiculopathy with lower cranial involvement, possibility of hypertrophic pachymeningitis should be kept in mind. Before diagnosing it as idiopathic, infectious causes should be excluded otherwise prescription of corticosteroid will flare up the disease process.

Key Words: Cranio cervical, hypertrophic pachymeningitis, tuberculous

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INTRODUCTION

Hypertrophic thickening of the meninges can be caused by a variety of pathological processes such as inflammatory (tuberculosis, fungal, Lyme's disease, syphillis, Human T-lymphotropic virus (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.[4] It has a nonspecific clinical picture comprising headache, vomiting, cranial nerve palsy, ataxia, raised intracranial pressure, and focal neurological deficit. These symptoms and signs are due to entrapment of cranial nerves, occlusion of cerebrospinal fluid (CSF) flow, venous sinuses, and, rarely, arteries.[2] Involvement of the meninges can be diffuse or focal and both cranial and spinal meninges can be affected. Some diseases selectively involve the dura mater, whereas others affect predominantly the leptomeninge. When limited to the dura mater, it is called pachymeningitis. Tubercular involvement of cranio cervical dura mater is rare.[10] In

this report, we describe the clinical features, radiological findings, and therapeutic options of this unique clinical entity.

CASE REPORT

A 50-year-old female presented with history of progressive quadriparesis and stiffness of neck for 2 years, dysphagia to liquid for past 3 months. She had visited several doctors without any relief for the said complaints. Magnetic resonance imaging (MRI) of cervical spine was done one year back. On the basis of clinical history and supportive MRI finding, she was previously diagnosed as a case of high cervical compressive myelopathy with cord changes due to thickening of ligamentum flavum and posterior longichudinal ligament extending from foramen magnum up to C4 vertebral level. Corticosteroid was prescribed by her previous physician. Following which her condition deteriorated rapidly.Her neck became so stiff that she was unable to move it, numbness extended to all over her body; she developed dysphagia to liquid and became bed ridden. Physical examination revealed high cervical compressive myelo-radiculopathy with lower cranial nerve palsy and rigid neck. Recent contrast MRI was suggestive of circumferentially, grossly thickened, enhancing dura extending from caudal aspect of posterior fossa up to C7 level with variable compression of cord and emerging nerve roots [Figures 1-4]. Radiological features were in favor of hypertrophic pachymeningitis (HPM). An extensive search was carried out to find out the cause of HPM. Her vital signs and other laboratory data were normal except low hemoglobin level (Hb and Hct, 9 g/dL and 32%, respectively), elevated erythrocyte sedimentation rate (ESR) (patient: 110 mm/h, normal: <25 mm/h) and positive tuberculin skin test. Serologic tests for tuberculous antibody were Immunoglobulin M (IgM) positive indicating persisting infection. The serum was negative for rheumatoid factor, antinuclear antibodies, Venereal disease research laboratory (VDRL) test, hepatitis B surface antigen (HBsAg), and antidouble-stranded DNA. Work up for sarcoidosis such as chest radiograph, serum calcium, and angiotensin converting enzyme were negative. Bone marrow examination and ultrasound scan of the abdomen, to exclude a neoplastic and chronic inflammatory process, were normal. CSF analysis after lumbar puncture revealed 80 lymphocytes/mm^3, protein of 90 mg/dl, and sugar of 35 mg/dl. Microbiological evaluation of the CSF for cryptococcal antigen, VDRL, and cultures for acid fast bacilli (AFB) and fungi yielded no positive results. An increase in the adenosine deaminase level in CSF was observed. On basis of radiological, serum analysis, and CSF study, a diagnosis of tuberculous HPM was made. Screening for systemic source of tuberculosis was negative. She was discharged on antituberculosis therapy with regular follow up. At the 3-month follow-up, the patient was able to walk with a cane, neck rigidity vanished, dysphagia and limb spasticity improved. Antitubercular treatment continued for 15 months. At present, she can walk independently with some spasticity.

DISCUSSION

HPM is a rare disorder of diverse etiology. Charcot and Joffroy[1] first described it in relation to spinal meninges. Cranio cervical HPM in continuity has been rarely reported.[10] Naffziger and Stern described the first case of idiopathic hypertrophic pachymeningitis (IHPM). The diagnosis of IHPM is based on excluding a large number of causes as described earlier.

In the first stage of spinal HPM, patients experience local and radicular pain. The second stage can present signs of nerve root compression. In addition, in the

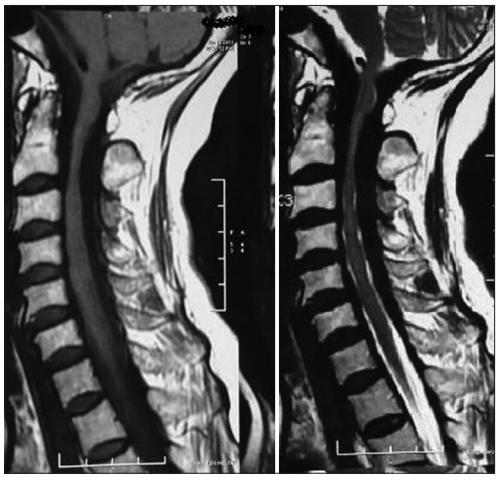

Figure 1: Sagittal T1- and T2-weighted MRI, showing hypointense hypertrophic pachymeninges extending from caudal aspect of posterior fossa up to C7 level with variable cord compression and myelomalacia

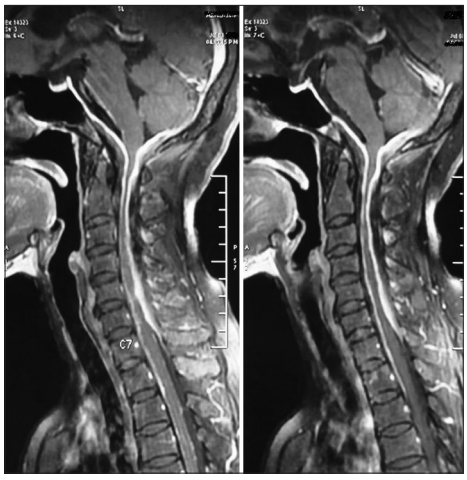

Figure 2: Sagittal T1-weighted image, showing curvilinear enhancement of the inner layer of thickened meninges following gadolinium administration

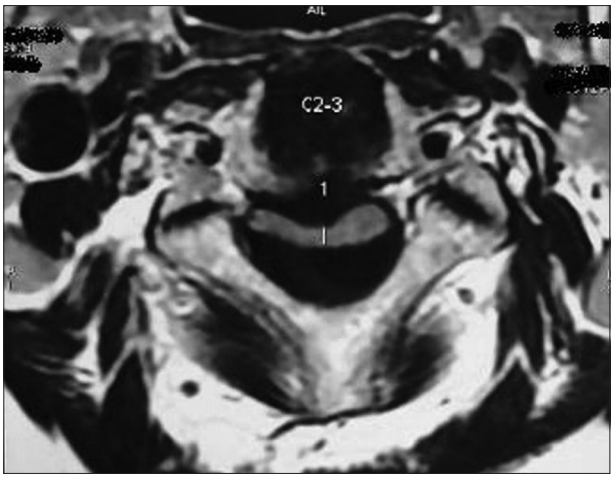

Figure 3: Axial T1-weighted MRI, showing circumferentially compressed cord along with nerve roots due to grossly hypertrophic pachymeninges

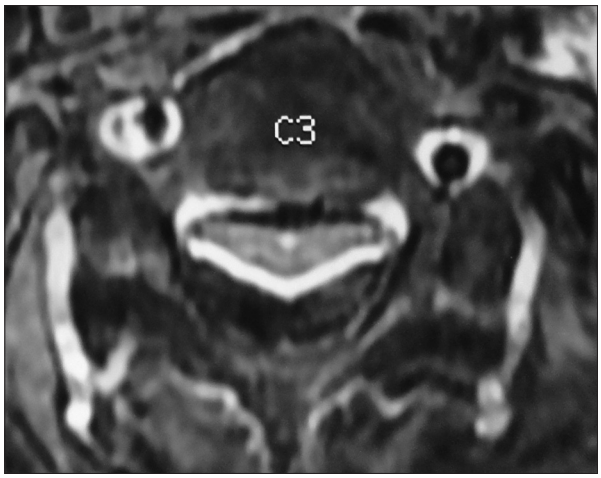

Figure 4: AxialT1-weighted MRI, showing enhancement of the inner layer of thickened meninges following gadolinium administration

third stage, patients eventually suffer from spinal cord compression.[5,7] Parney *et al.*[6] reported headache, cranial nerve palsy, and ataxia in 88%, 62%, and 32% of the cranial HPM cases, respectively. Our patients have experienced features of both cranial and spinal HPM.

HPM is being increasingly recognized with the advent of computed tomography (CT) and MRI. On MRI, thickened dura mater appears isointense or hypointense on T1WI and hypointense on T2WI sequence with a hyperintense edge, which is best seen in coronal or sagittal sections.[8] There is curvilinear enhancement of the thickened meninges following gadolinium administration. In our case, initial diagnosis was wrong as contrast MRI was not done. The radiological findings, although characteristic of HPM, may not reveal the underlying etiology. Exclusion of several underlying causes is an essential feature for the diagnosis of IHPM. Meningeal biopsy is essential in this regard.[2] In our case, meningeal biopsy was not required as there were sufficient serological and CSF study supporting tuberculous nature.

The natural course of IHPM is poorly understood. Spontaneous resolution, response to steroids, steroid dependency, remitting, and relapsing course have been documented.[3] Every effort should be made to exclude infectious causes before prescribing steroid, as infections may flare up following corticosteroid therapy, which was observed in our case. Early surgical excision of spinal IHPM has been documented, as rapid clinical improvement is usually noted. However, it is difficult to excise long segment lesion from the ventral portion of the spinal cord.[9] Therefore, the optimum surgical treatment may be the widest possible excision of the involved dura mater with exposure of the affected nervous tissue. In our case, though early surgical decompression would have rapid clinical improvement, we preferred conservative approach as pathology was an infective cause, which was likely to respond to therapy.

CONCLUSION

In case of multilevel cervical compressive myelo-radiculopathy with lower cranial involvement, possibility of HPM should be kept in mind. Before diagnosing it as idiopathic, infectious causes should be excluded; otherwise prescription of corticosteroid will flare up the disease process. Though early surgical excision has been documented for IHPM, a course of antitubercular treatment seems appropriate for tubercular causes.

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