Idiopathic Hypertrophic Cervical Pachymeningitis: A Case Report with 5 Year Follow Up

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Abstract

Idiopathic hypertrophic pachymeningitis is an extremely rare entity. It usually affects cranial meninges. The spinal form is further uncommon and presents as a chronic progressive disease. We describe a 42 year old female with isolated idiopathic hypertrophic cervical pachymeningitis who had a relapsing remitting course under observation for five years. Laminectomy and immunosuppressive therapy produced temporary and partial relief. The long term course and relevant literature is reviewed.

INTRODUCTION

Hypertrophic spinal pachymeningitis, first described by Charcot and Joffroy in 1869, is characterized by compression of the spinal cord and roots by a marked inflammatory hypertrophy of the spinal cord dura and arachnoid matter. It is an uncommon cause of spinal cord compression. The etiology of idiopathic hypertrophic spinal pachymeningitis is unclear. However some causative factors have been reported. Rheumatoid arthritis, thrombophlebitis, tuberculosis, syphilis, complication of intrathecal prednisolone acetate and metabolic diseases, are some known causes.1 It occurs at all ages and both sexes. Although the disease is found throughout the spine, it is most common in the cervical and thoracic regions. Typically several levels are affected. Involvement of the entire spine has also been described.1 The long term evolution of this form of pachymeningitis is unclear, hence we describe cervical spinal form of pachymeningitis with long term observations of five years.

CASE REPORT

A 42 year old lady presented with neck pain radiating to the outer aspect of both arms, fore arms and thumbs, and deep aches in both arms since November 1996. A month later, tingling and numbness began in both lower limbs and ascended up to the multiple level. This was accompanied by stiffness and slowing of gait. There was no urinary complaint. She had no constitutional symptoms. Past history was not contributory. The examination showed left side C-5,6,7 and right sided C-5,6 muscle weakness. The supinator and biceps jerks were decreased. Triceps, knee and ankle jerks were exaggerated and both planters were extensor. There was a sensory level for pain and touch at T4 level. Based on these findings, a diagnosis of progressive low cervical radiculomyelopathy was made and the patient was investigated. The routine blood tests, X-ray chest, serum ACE levels, ESR, CSF routine examination, IgG, PCR for tuberculosis, VDRL, FTA Abs were normal. MRI scan on post-contrast sagittal T1 weighted image of the cervical spine (Fig. 1a), showed diffusely thickened and densely enhancing pachymeninges anterior and posterior to the cord extending continuously from C1 to C4 compressing the cord. There was a diffuse hyperintense signal within the cord parenchyma extending down to T2 level (Fig. 1b). The thickened pachymeninges were not visualized on T2, being masked by the hyperintense signal of the CSF (Fig. 1b). Thus a diagnosis of idiopathic hypertrophic cervical spinal pachymeningitis producing spinal cord compression was made. She underwent C1-C4 laminectomy, decompression and duroplasty. Histopathology showed chronic inflammation. There were no granulomas or malignancy. She received oral prednisolone 50 mg per day. Three months later she was asymptomatic and at this stage clinical examination was normal. Steroids were tapered off over next six months. Symptoms recurred after six months. Spinal cord symptoms became evident again. Oral prednisolone 30 mg/day was reused for two months and tapered to 10 mg/day over next 6 months. She had partial recovery. Post-contrast MRI at this stage (Fig. 2a) showed that the spinal cord had been decompressed adequately between C2 and C4. There was a residual pachymeningeal thickening seen with the relief of the compression. Sagittal T2 weighted images (Fig. 2b)
performed after surgery show residual intramedullary hyperintense signal which was diminished in extent compared to the preoperative scan (Fig. 1b). At this stage, vascular involvement of cord parenchyma was considered and pentoxifylline was added. Over the next three years, she was on maintenance prednisolone 10 mg/day and azathioprine 100 mg/day with relapsing remitting clinical symptoms and persistent spinal cord signs.

**DISCUSSION**

Our patient had chronic hypertrophic cervical spinal pachymeningitis producing cervical cord compression. The diagnosis of hypertrophic pachymeningitis is usually one of exclusion.\(^2,3,4\) Tuberculosis as an etiology was excluded as ESR, chest X-ray and CSF were normal and CSF PCR for tuberculosis was negative. Syphilitic etiology was excluded as VDRL and FTA Abs were negative. Sarcoidosis of nervous system was also considered as it may present with myelopathy, encephalopathy, facial and peripheral neuropathy. The diagnosis of neurosarcoidosis can be difficult when neurological involvement is the first or only manifestation of the disease. In our patient, ACE levels were normal, serum calcium and chest X-rays were normal and the biopsy ruled out sarcoidosis as there was absence of non-caseating granulomas, histiocytes, Langhans and foreign body giant cells.\(^5\) Thus, the nature of hypertrophic cervical spinal pachymeningitis was considered to be idiopathic.

MRI with gadolinium enhancement proved to be useful and demonstrated the thickened dura. We performed C1-4 laminectomy and duroplasty for the relief of compression.\(^2\) Surgical treatment provided temporary relief of her symptoms, but the natural history of the disease was not modified.

The spinal form of the hypertrophic pachymeningitis is less common, but when it occurs, it presents as a progressive radicular symptoms, the second stage by muscle weakness and atrophy, and the third stage is marked by paraplegia, loss of bladder and bowel control and respiratory distress caused by intercostals and diaphragmatic denervation.\(^5\) The natural history of cranial hypertrophic
Pachymeningitis has been well documented. Among the 33 patients with idiopathic cranial hypertrophic meningitis gathered from literature by Parney et al., 626% experienced full remission without steroid dependence, 15% experienced steroid dependent partial or complete remission, 15% experienced a progressive course inspite of steroid therapy, and 32% died regardless of treatment. In contrast, the course of spinal form is less studied, as it is uncommon, but it is believed to run a progressive course. Our patient however had a relapsing remitting and fluctuating course. She initially made near total recovery with decompression and prednisolone and remained asymptomatic for one year after which the disease recurred and at this stage MRI confirmed the continued presence of hypertrophy of meninges. She required long term low dose prednisolone and azathioprine for five years and still ran a chronic relapsing fluctuating course.

In conclusion, we present a long term follow up demonstrating the uncommon relapsing-remitting course of hypertrophic cervical spinal pachymeningitis with limited response to surgical and medical therapy.

REFERENCES


Announcement

International Conference on Hypertension, CME for Practising Physicians sponsored by Rajiv Gandhi University Health and Science, Bangalore Medical College, Medical Education and Research Trust, Karnataka, Directorate of Medical Education, Govt. of Karnataka will be organised by the International Medical Center, Bangalore from August 1 - 3, 2003 at Bangalore, India.

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