Tuberculous Hypertrophic Pachymeningitis Involving the Posterior Fossa and High Cervical Region
—Case Report—

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Abstract
A 44-year-old female presented with a rare tuberculous hypertrophic pachymeningitis involving the posterior fossa and high cervical region manifesting as progressive multiple cranial nerve pareses and myelopathy developing over 6 months. Magnetic resonance imaging demonstrated the thickened dura mater and associated syrinx. Despite decompressive craniectomy and antituberculous treatment, she died of disseminated intravascular coagulation. Hypertrophic pachymeningitis is probably best treated by the most extensive excision of affected dura mater possible, unless medical treatment can be instituted for an identifiable underlying causative disease.

Key words: hypertrophic pachymeningitis, tuberculosis, posterior fossa, cervical spine, treatment

Introduction
Hypertrophic pachymeningitis is a rare disease causing a chronic inflammatory hypertrophy of dura mater, generally located in the dorsal cervical and thoracic regions in spinal hypertrophic pachymeningitis, and the skull base and posterior fossa in the cranial form. The location may determine the clinical symptoms such as radiculopathy and myelopathy in spinal hypertrophic pachymeningitis and increased intracranial pressure and impairment of cranial nerves in cranial hypertrophic pachymeningitis and increased intracranial pressure and impairment of cranial nerves in cranial hypertrophic pachymeningitis. The etiology is usually obscure, but histological studies may identify a non-specific granulomatous change, commonly consisting of proliferation of collagen fibers with infiltrating inflammatory cells. The disease process is usually progressive. We describe an unusual case of tuberculous hypertrophic pachymeningitis involving both the cranial and spinal dura mater.

Case Report
A 44-year-old female presented with fever of unknown origin, left tinnitus, dysphagia, progressive gait disturbance, numbness of all extremities, and tingling sensation in the left leg. She had been well until August, 1991, when a persistent, intermittent fever started. The cause could not be identified in spite of intensive medical studies, including negative biopsy of the neck lymph node. She was treated with corticosteroids under a presumptive diagnosis of collagen disease. Vertigo and left tinnitus subsequently developed. In December, her neck became so stiff that she found difficulty in rotating her head. In January, 1992, she noticed dysphagia, soon followed by weakness of the left leg and numbness on the left. She then developed double incontinence. The numbness extended to all her body, with a tingling sensation in the left leg. In February, she became unable to walk without assistance.

On admission on February 5, she was pale and
thin. Her blood pressure was 120/80 mmHg and her body temperature was 37.8°C. Neurological examination revealed mild left facial weakness, left deafness, absent left gag reflex, tongue deviation to the left, quadripareisis worse on the left, bilateral hypesthesia below the C3 dermatome, hypalgesia below the right T12 dermatome, bilateral hypopallesthesia worse in the lower dermatomes, hyporeflexia in the upper extremities, hyperreflexia in the lower extremities, and bilateral positive Babinski's sign.

Laboratory examinations revealed leukocytosis with increased serum C-reactive protein, increased serum immunoglobulin G and A, positive antibodies to nuclear antigens, elevated serum hemolytic complement, positive rheumatoid arthritis hemagglutination assay, negative lupus erythematosus cell test, decreased activity of angiotensin-converting enzyme, positive Wassermann’s reaction, and positive Treponema pallidum hemagglutination assay. Urinalysis was normal. The tuberculin skin test was negative.

Lumbar puncture revealed an opening pressure of 160 mmH2O. The cell counts were 12/mm³ with lymphocyte dominance. No atypical cells were observed. The protein, sugar, and chloride contents were 98 mg/dl, 36 mg/dl, and 117 mEq/l, respectively. The tryptophan reaction was positive. No fungi and bacteria were demonstrated by culture.

Roentgenograms of the chest, skull, and cervical spine, and cerebral computed tomographic (CT) scans were normal. Magnetic resonance (MR) imaging demonstrated a lesion, extending from the posterior fossa down to the C3 level and surrounding medulla and the upper cervical cord, as an isointense area on the T1-weighted image and a hyperintense area on the T2-weighted image, and a syrinx around the C5 level (Fig. 1).

On February 7, after the MR study, she suddenly complained of dyspnea and dysphonia. She rapidly lost consciousness and respiratory arrest occurred. Despite immediate resuscitation, she became comatose with flaccid quadriplegia, preserving only the light reflexes.

The affected region was decompressed by an emergency suboccipital craniectomy and C1-3 laminectomies. There was no inflammatory exudate or epidural tumor. The dura mater of the posterior fossa was rather whitish, but that of the cervical spine appeared normal. An opening in the dura mater of the posterior fossa was made and extended caudally to the C3 level. The enormously thickened dura mater (as much as 10 mm thick) concealed no tumor, so the excess fibrous tissue was resected, leaving a residual layer of dura mater.
Postoperatively, her condition remained the same. No somatosensory evoked potentials and auditory evoked potentials were recorded, although the visual evoked potentials were normal. On February 12, a spontaneous down-beat nystagmus was observed, but otherwise she remained stable.

Histological examination of the excised specimen showed proliferation of collagen fibers, focally infiltrated by neutrophils intermingled with epithelioid cells and giant cells of Langhans type, and sporadic necrosis (Fig. 2 upper). Tuberculous bacilli were demonstrated by Ziehl-Neelsen staining using a triple thickness section (Fig. 2 lower), confirming the diagnosis of follicular caseous tuberculosis.

Triple antituberculous treatment (streptomycin sulfate 3 g/wk, isoniazid 200 mg/day, rifampicin 450 mg/day) was initiated. On February 19, she opened her eyes and grimaced spontaneously, but communication was impossible. In March, she demonstrated the exaggerated jaw reflex, but was still unresponsive to stimuli, flaccid quadriplegic, and mechanically ventilated. Her neurological condition then remained stable until she died on June 14 from disseminated intravascular coagulation. Autopsy was not permitted.

Discussion

The diagnosis of hypertrophic pachymeningitis is now based on the demonstration of hypertrophic dura mater by CT and MR imaging. CT shows the affected dura as a high-density lesion, enhanced postcontrast. MR imaging demonstrates an isointense lesion with remarkable contrast enhancement on the T1-weighted image, and a hypointense lesion with hyperintense edges on the T2-weighted image, indicating a dense fibrous tissue and inflammatory infiltrates. MR images were helpful in our case, although no study with contrast medium was available because of the rapid deterioration in the patient’s condition. The radiological differential diagnosis includes meningioma en plaque, fibroma, dural carcinomatosis, neurosarcoïdosis, and lymphoma. Patients with advanced spinal hypertrophic pachymeningitis may demonstrate syringomyelic changes, but these have not been radiologically demonstrated. In our case, MR imaging showed a syrinx around the C5 level.

Involvement of both cranial and spinal dura mater has only been reported in two patients. Interestingly, both lesions were continuous as in the present case. Clinical symptoms were chronic headache, nausea, and vomiting in one patient, possibly due to increased intracranial pressure, and chronic headache, bilateral hearing disturbances, right hypoglossal paresis, and cerebellar ataxia in the other. However, only our patient presented with myelopathy. The clinical courses of the present patient and the others suggest that cranial hypertrophic pachymeningitis may extend downwards to the spine.

The obscure etiology of hypertrophic pachymeningitis is usually considered idiopathic, although etiological factors, such as tuberculosis, syphilis, fungal and parasitic infection, sarcoidosis, Wegener’s granulomatosis, rheumatoid arthritis, mucopolysaccharidosis, and multifocal fibrosclerosis, have been verified in rare instances. The etiology of the patients with both cranial and spinal forms were described as rheumatoid arthritis and idiopathic. In our case, the laboratory examinations suggested various etiological factors such as syphilis, rheumatoid arthritis, and tuberculosis. Caseous necrosis with giant cells of Langhans type, especially, was highly suggestive of tuberculosis as the etiology. Tuberculosis was histologically confirmed by Ziehl-Neelsen staining of a triple thickness section, as several examinations of a routine section were negative.

Hypertrophic pachymeningitis is currently treated by surgical decompression of the affected nervous tissue in the early stage of the disease. A rapid clinical improvement is usually obtained and may last for years. However, in addition to ambiguity in the long-term follow-up of clinical conditions and the affected dura mater, the occurrence of reappearance of the disease after surgical excision, and increasing thickness of dura mater after partial removal suggest that surgery may not be curative unless total excision is achieved, which seems unfeasible. Therefore, the optimum surgical treatment may be the widest possible excision of the involved dura mater with exposure of the affected nervous tissue, although only incision of the involved dura mater or resection of the excess fibrous tissue have been effective.

The effectiveness of medical treatment is still uncertain, probably because the etiology and disease process are obscure. However, medical treatment for known etiology is effective to reduce the thickness of the involved dura mater. Corticosteroid therapy in idiopathic cases may also be effective. Studies to identify the etiology and disease process are indispensable for effective use of medical therapy in the pre- and postoperative periods.
References


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