Tuberculous meningitis presenting as an obstructive myelopathy

Case report

BRIEN VLCEK, M.D., KIM J. BURCHIEL, M.D., AND THOMAS GORDON, M.D.

Department of Neurology, The Children's Orthopedic Hospital and Medical Center, Seattle, Washington

Subacute paraplegia progressing over 3 months due to spinal cord compression was the presenting symptom of tuberculous meningitis in this patient with a normal chest x-ray film and no radiological or autopsy evidence of Pott's vertebral tuberculosis. The obstructive myelopathy was the result of proliferative granulomatous meningitis. A review of the literature indicates that this is a very unusual presentation of tuberculous meningitis.

KEY WORDS - paraplegia - tuberculous meningitis - myelopathy - granulomatous arachnoiditis - tuberculosis

MYELOPATHY with spinal subarachnoid obstruction secondary to granulomatous arachnoiditis was first described by Sir Victor Horsley in 1909.11 Syphilis, fungal infections such as cryptococcosis or coccidioidomycosis, chemical meningitis due to the subarachnoid injection of foreign materials such as Pantopaque, antibiotics, talc, and surgical sponges, and even chronic reactions to herniated disc material or trauma have all been implicated as causative agents.2,6,8,9,18 There is also a large collection of cases in which the cause has been undetermined. Most reports of this condition antedate the 1960's.

In 1947, Ransome and Montiero15 first called attention to the possibility of tuberculous myelopathy occurring in the absence of Pott's disease in a report on four patients from Singapore. These patients presented with relatively rapidly developing paraplegia without other neurological manifestations of tuberculous meningitis. Yet, because of the rarity of this form of tuberculosis, this etiology is usually not considered in patients with progressive paraplegia. We present a case that exemplifies this disorder, and discuss the clinical, historical, laboratory, and radiological features that may aid in diagnosis.

Case Report

This 73-year-old native American woman had had pneumococcal meningitis 3 years previously, which left her with mild dementia, hearing loss, and a left-sided focal motor seizure disorder requiring phenytoin treatment. Ventricular dilatation thought to be secondary to atrophy was noted on computerized tomography (CT) scanning 2 years prior to her present admission. She remained stable, living alone and independently. She experienced the gradual onset of lower-extremity weakness and paresthesias over the 3 months preceding admission. Three days before admission to another hospital there was marked increase in lower-extremity weakness with frequent falling episodes and the onset of low-back pain.

Examination. The general physical examination on admission was recorded as unremarkable. The patient had a temperature of 98.8°F. There was no evidence of meningismus, and the straight-leg raising test was negative bilaterally. A post-void residual urinary volume of 1500 cc was documented by catheterization. Neurological examination showed her to be oriented only to person and place, with poor short-term memory. Cranial nerve function was normal. Diffuse weakness of the lower extremities, more on the right than on the left, was found, and proprioceptive and vibratory sensation was decreased below the knees bilaterally. Deep-tendon reflexes were normally active in the upper extremities, patellar tendon reflexes were hyperactive, and a right Babinski response was present. Laboratory tests were unremarkable except for a serum sodium level of...
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119 mEq/liter and an erythrocyte sedimentation rate of 60 mm/hr. A chest x-ray film was unremarkable.

On the following morning, the patient had a prolonged left-sided focal motor seizure involving both the arm and the leg. The serum phenytoin level was not detectable and the serum sodium concentration was 128 mEq/liter. Post-ictally, she had a left hemiplegia and was obtunded. A CT scan with contrast enhancement showed only ventricular dilatation and cortical atrophy which appeared to be unchanged. Her hemiplegia and obtundation cleared by the following morning, but she continued to have lower-extremity weakness, greater on the right than on the left, hyperactive knee-jerk reflexes, bilateral Babinski signs, and decreased sensation to all modalities in both legs without any clear sensory demarcation on the trunk. Stiffness of the neck was not noted. Metrizamide myelography was performed and interpreted as showing “no gross abnormality to T-11.” Cerebrospinal fluid (CSF) was xanthochromic with only 78 mm H2O opening pressure, and coagulated at room temperature. The CSF analysis revealed 487 white blood cells (87% polymorphonuclear cells, 13% lymphocytes), 45 red blood cells, 4120 mg/dl protein, and 22 mg/dl glucose. Her temperature was now 103°F. Antibiotics were instituted and myelography with Pantopaque was repeated. This showed a complete block from T-12 to T-4 with irregular flow of the contrast material above the upper myelographic block (Fig. 1).

Operation. An epidural abscess of the spinal cord was thought to be a strong possibility, and the patient was taken immediately to the operating room where an extensive decompressive thoracic laminectomy was undertaken. However, extradural exploration showed no evidence of abscess or other compressing mass lesion. Intradural inspection at T11-12 revealed a thick whitish purulent exudate over the leptomeninges and spinal cord with venous and arterial thromboses and obvious infarction of the underlying spinal cord. The involved arachnoid and thrombosed vein were biopsied.

Postoperative Course. Tuberculous or fungal meningitis was considered to be most likely, so in addition to broad-spectrum antibiotic coverage, the patient was given isoniazid, ethambutol, rifampin, 5-fluorocytosine, and amphotericin B. Despite this treatment she remained febrile and continued a steadily deteriorating course, first losing upper-extremity function, then developing hypoventilation with periods of apnea necessitating intubation and mechanical ventilation. A CT scan of the head showed no change. Nevertheless, an external ventriculostomy was inserted with the finding of normal ventricular pressure. Tuberculin skin tests with appropriate controls showed anergy and subsequently acid-fast bacilli were demonstrated both in the biopsy specimen and in the patient’s sputum. No bacteria were isolated from the CSF.

There was progressive neurological deterioration, with loss of gag reflex and swallowing function, loss of pupillary light reflexes, and anisocoria, followed by intractable loss of all brain-stem function. The patient died on the 7th postoperative day.

Pathological Examination. Biopsy specimens of the meninges revealed thrombosis with focal necrosis of the walls of several veins and arterioles with infiltration by polymorphonuclear leukocytes, histocytes, and plasma cells. Many acid-fast bacilli were demonstrated. No giant cells or frank granulomas were seen.

The significant general autopsy findings included tuberculosis of both lungs, the adrenal glands, and abdominal lymph nodes, but no vertebral caries were present. The cerebral meninges were cloudy and thickened, particularly over the frontal poles, but the basal meninges were only slightly thickened. There was one 3- to 4-mm granuloma in the right orbital frontal cortex, which microscopically had ruptured into the adjacent subarachnoid space. There was a second, very small, fibrotic granuloma in the left parieto-occipital white matter. The spinal cord was encased in a thick fibrous exudate, with fibrous strands connecting the leptomeninges to the inner surface of the dura (Fig. 2). This exudate was most extensive around the thoracic cord. Microscopically, the spinal leptomeninges were thickened and fibrotic with granulomatous infiltration consisting of lymphocytes, plasma cells, and macrophages. At the midthoracic level the underlying spinal cord was compressed and necrotic. There was a circumferential rim of necrosis of the cord approximately 3 to 4 mm deep throughout the entire length of the thoracic cord.

![Fig. 1. Myelograms showing complete block with an irregular contour extending from T-3 on the left to T-4 on the right (left), and a second block at T-12 (right).](image-url)
Spinal meningeal blood vessels showed varying degrees of infiltration, thrombosis, fibrosis, and recanalization (Fig. 3). Rare acid-fast bacilli were identified only in the basal meninges, and not in the spinal cord sections.

Discussion

Pott's paraplegia, caused by spinal cord compression secondary to tuberculous vertebral caries, is a fairly common cause of myelopathy in developing countries where tuberculosis is prevalent. Plain vertebral x-ray films almost always reveal vertebral caries, and certainly vertebral bone involvement is a frequent finding at autopsy. The pathology in these cases is typically one of extradural compression without meningitis. There are two other forms of myelopathy secondary to tuberculosis that are quite rare and not part of Pott's paraplegia: 1) circumscribed solitary granulomas of a clear tuberculous etiology found within the spinal cord as well as in intra- or extradural locations; and 2) proliferative encasing granulomatous arachnoiditis with compression and vasculitic thromboses of spinal cord vessels. Our case represents this second form of myelopathy with diffuse granulomatous arachnoiditis over many spinal cord segments reminiscent of the four cases described by Ransome and Montiero. Brooks, et al., reported similar findings in 15 of 80 "gravely ill" patients suffering from classic tuberculous meningitis between 1947 and 1953 in England. These 15 patients all had complete myelographic block, myelopathy, and similar exudative proliferative cord-encasing pathology. However, all of these patients had a relatively long-standing severe classical type of tuberculous meningitis prior to the onset of their myelopathy. Only two cases similar to ours have been reported in the United States. The Indian literature and experience concerning tuberculous myelopathy is the most extensive. Wadia and Dastur described 70 Indian patients treated between 1958 and 1967 who had tuberculous granulomatous arachnoiditis and myelopathy in the absence of Pott's disease. This represents the largest series of patients with this problem in the literature, and comes from a country in which tuberculosis is the most common cause of paraplegia secondary to Pott's disease. Between 1% and 5% of the population of India suffers from active tuberculosis.

The following clinical, laboratory, and pathological characteristics are typically seen in cases of tuberculous meningitis with myelopathy: 1) Myelopathy with early signs usually beginning 2 months before the more rapidly progressing stage that develops over 2 to 5 days. These latter acute signs include radicular pain, paresthesias, and loss of sensation (most commonly loss of position sense, weakness, and bladder dysfunction). 2) Absence of clinical or radiological evidence of acute or chronic extraneural tuberculosis or classical tuberculous meningitis. 3) An absence of Pott's disease. 4) Myelographic findings are varied but may show classic signs of arachnoiditis in addition to spinal block. 5) Spinal block and CSF protein levels greater than 1 gm/dl. 6) Cerebrospinal fluid cellular reaction is usually lymphocytic. 7) There may be a superficial circumferential rim of necrosis of the spinal cord only a few millimeters in thickness.

The pathogenesis of this condition probably involves a variation of Rich's hypothesis, with an initial hematogenous spread of tubercle bacilli to the central nervous system giving rise to a small focal tuberculosis adjacent to the subarachnoid space which eventually ruptures resulting in tuberculous meningitis. It may be that, with previous exposure to tuberculosis and the development of partial immunity, the resulting inflammatory reaction is more intense and thus more localized to the site of granuloma rupture in these patients, as has been demonstrated experimentally by Tandon, et al. The larger area of intracranial subarachnoid space

Fig. 2. Autopsy specimen showing the spinal cord encased in a thick fibrous exudate, and severe arachnoiditis with fibrous strands connecting the leptomeninges to the inner surface of the dura.

Fig. 3. Photomicrograph of the spinal leptomeningeal artery showing thrombosis, fibrosis, and some recanalization. H & E, × 40.
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and greater vascular supply of the brain compared to the spinal cord may account for the preponderance of cases of tuberculous meningitis primarily giving rise to intracranial symptoms and intracranial pathology.

The mortality rate from exudative tuberculous meningitis with myelopathy remains high, in the range of 30%. Early diagnosis, which may require biopsy with histological and/or microbiological evidence of acid-fast bacilli, and prompt antituberculous chemotherapy still hold the most promise of cure for this disease. Corticosteroids may also be helpful, but the value of decompressive laminectomy remains uncertain. However, as our patient exemplifies, this diagnosis is usually not suspected because of the absence of clinical or radiological evidence of acute or chronic extraneural tuberculosis and of classical tuberculous meningitis.

Conclusions

Myelopathy due to tuberculous proliferative granulomatous arachnoiditis is extremely rare in developed countries. However, given the proper historical and clinical setting, this entity should be considered in patients who may have emigrated from regions where tuberculosis is common or in those indigenous populations in which tuberculosis is more prevalent. Prompt diagnosis, which may require surgical intervention, and rapid initiation of antituberculous therapy may lead to a satisfactory outcome.

References


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Address reprint requests to: Brien Vlcek, M.D., Department of Neurology, The Children's Orthopedic Hospital and Medical Center, P.O. Box C5371, Seattle, Washington 98105.