Pachymeningitis cervicis hypertrophica

Case report


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The authors present the case of a 25-year-old man with idiopathic pachymeningitis hypertrophica that caused cervical radiculopathy. Decompressive surgery produced significant neurological improvement. The etiology and management of the condition are discussed and the literature is reviewed.

KEY WORDS • pachymeningitis cervicis hypertrophica • cervical radiculopathy • nerve root compression

PACHYMENINGITIS hypertrophica is a rare disease causing chronic inflammatory hypertrophy of the dura. The disease has been found throughout the spine, although the cervical and thoracic regions are most commonly affected. The process has also been reported in the base of the skull and the posterior fossa. The natural history is one of progression from local pain to radiculopathy and spinal cord compression, and may be halted by excision of the involved dura. The etiology is usually obscure, although infection, trauma, toxins, metabolic disease, and rheumatoid arthritis have all been implicated.

We are reporting a case of idiopathic pachymeningitis cervicis hypertrophica causing cervical radiculopathy, which responded well to surgery.

Case Report

This 25-year-old male hairdresser presented with a 1-month history of increasing pain in the left side of the neck and left arm, associated with weakness and numbness of the left arm. He had fallen onto the outstretched left hand just prior to the onset of the pain.

First Admission. On examination there was complete paralysis of left shoulder abduction and left elbow flexion. There was also profound weakness of left shoulder adduction and mild weakness of elbow extension with absence of biceps and brachioradialis tendon reflexes, indicating involvement of the left C5-7 nerve roots. A mild decrease in pinprick sensation was present over the entire left arm. There were no other neurological signs. Plain x-ray films of the cervical spine and left shoulder and a cervical myelogram were all normal. Cerebrospinal fluid (CSF) analysis, erythrocyte sedimentation rate, and full blood examination were normal. A provisional diagnosis of brachial neuritis was made. Treatment consisted of physiotherapy.

Second Admission. Four weeks later the patient was readmitted with progressive pain and increased weakness of the left arm, with involvement of the left C-4 to T-1 nerve roots. He had also developed right arm weakness in a distribution indicating C5–6 nerve root involvement, and pinprick sensation was reduced in the right thumb. No long-tract signs were present. Repeat myelography showed lack of filling of some of the lower cervical nerve roots on both sides (Fig. 1). Treponemal serology was negative, and human T cell leukemia virus (HTLV)-III antibody was not detected. Neutrophil function tests and delayed sensitivity skin testing were normal. Computerized tomography (CT) scanning of the head was normal.

Operation. A cervical laminectomy was performed from C-4 to C-7. Marked dural thickening involving the spinal dura and nerve root sheaths was noted. The thickened fibrous tissue was excised from the dura and root sheaths.

Postoperative Course. Postoperative medication in-
FIG. 2. Photomicrograph of the surgical specimen showing dural thickening with nonspecific chronic inflammation and hyaline fibrosis. H & E, x 100.

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ischemia of the spinal cord with lateral and posterior funicular degeneration and cavitation. No microglial or inflammatory reaction was found in the cord of their patient. Chronic compression of the spinal cord may also play a role in the spinal cord pathology.

It has been suggested that pachymeningitis hypertrophica should be suspected when a patient with spinal cord compression has radicular pain in three or more nerve root territories. Charcot and Joffroy described three stages of progression from pain (local and radicular), to signs of nerve root compression, and eventually to spinal cord compression. Our patient experienced the first two stages, but had not progressed to spinal cord compression.

With this disease, plain x-ray films are usually normal. Myelography may show a partial or complete block and dorsal extradural compression with a "beak-like" shape, although there was only evidence of nerve root compression in our case. Computerized tomography myelography can be helpful in showing the thickened dura causing a filling defect dorsally in the spinal canal. The CSF often shows raised protein levels, and the cell count is usually normal. There was no spinal block in our case and the CSF was normal. If the skull or neck is involved, a search for chronic infection in the sinuses should be made, including CT scanning.

The condition may be progressive, as in our case, and surgical decompression by laminectomy and incision or excision of the involved dura with exposure of the spinal cord has been recommended. However, in our case, the excess fibrous tissue was resected, leaving a residual layer of dura. Rapid improvement often follows surgery and may last for years.

The roles of postoperative radiotherapy, steroids, and cyclophosphamide are uncertain. The tissue removed at operation should be cultured.

Pachymeningitis cervicalis hypertrophica is a rare cause of cervical nerve root compression and may cause spinal cord compression. It should be considered in patients who have evidence of nerve root changes at multiple levels but no bone changes on x-ray studies. Early decompressive surgery, including excision of the excess fibrous material will result in neurological improvement.

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References


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