Cervical myelopathy in diffuse idiopathic skeletal hyperostosis

Case report

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The case of a rapidly progressive cervical myelopathy in a 64-year-old man is presented. Radiological studies revealed a partial extradural block, which at surgery was found to be a focal fibrous, calcified mass associated with the ligamentum flavum. On the basis of the underlying disorder of diffuse idiopathic skeletal hyperostosis (DISH), the etiology of this compression was concluded to be focal fibrous proliferation and dystrophic calcification. The neurological complications of DISH are reviewed. The authors are not aware of any other reports of this cause of myelopathy associated with DISH.

KEY WORDS • hyperostosis • myelopathy • ligamentum flavum • ossification

Diffuse idiopathic skeletal hyperostosis (DISH) is a disorder affecting both the vertebral column and the extra-axial skeleton, and is characterized by ossification of ligaments, with hyperostosis at their points of attachment to bone. The association of neurological symptomatology is unusual.

This report describes a patient with DISH who developed cervical myelopathy. The cause was a unilateral, focal fibrous mass with calcification in the ligamentum flavum and its laminar attachment. This patient had no underlying medical condition other than DISH. The case is discussed, and the neurological aspects of DISH are reviewed.

Case Report

This 64-year-old right-handed man came to the hospital complaining of weakness in both hands for 3 weeks. He had experienced chronic neck pain for approximately 30 years, but noted this to be worse over the 3 to 4 weeks prior to presentation. He described numbness in both hands and weakness in the right hand of sufficient severity to prevent him holding objects. He had no lower-extremity symptoms and no sphincter dysfunction. His medical history included a right-hip arthroplasty with postoperative femoral neuropathy and DISH, initially diagnosed as “arthritis” 30 years before. The patient was being followed by a rheumatologist. His only medications were two antihypertensive agents. There was no history of trauma although during his school days he had participated in boxing.

Examination. Motion of the neck was reduced to 25% of the normal range in all directions, and at rest the neck was held in an abnormally flexed attitude. There was no Lhermitte's sign. The patient exhibited an old Bell's palsy. Wasting and fasciculations were visible in the intrinsic muscles of the hands, more prominent on the right side. Grade 4/5 power was present in the right deltoid, biceps, and triceps muscles, wrist flexor and extensor muscles, and finger flexor and extensor muscles; hand intrinsic muscle power was 3/5. The abnormalities on the left side were wrist flexion and extension power 4/5 and intrinsic muscle power 4/5. Testing of the deep-tendon reflexes disclosed the following power: on the right side, biceps 1/4, triceps 3/4, knee jerk 1/4, and ankle jerk 2/4; and on the left side, biceps 2/4, triceps 3/4, knee jerk 2/4, and ankle jerk 2/4. Plantar responses were flexor bilaterally. Hypalgesia was profound in the right C-5 dermatome and
moderate in the C6–T1 dermatomes. Position and vibration sense was decreased in the right lower extremity.

Laboratory values were all within normal limits, except for the cerebrospinal fluid protein level (1.9 gm/liter). Plain spine x-ray films revealed the typical changes of DISH, which were seen best in the thoracic and lumbar spine (Fig. 1). The cervical spine was in good alignment and showed much less extensive paraspinal calcifications. A myelogram with computerized tomography revealed a partial posterior extradural block at C-4 on the right side (Fig. 2).

**Operation.** In view of the patient’s rapid progression of neurological deficits over a 3-week period, surgery was performed within hours of presentation. Preoperatively, the diagnosis was considered most likely to be extradural compression of either a neoplastic or infectious etiology. A bilateral laminectomy was performed at C-4 and the removed bone was noted to be normal in structure. Beneath it and strongly adherent to the dura was a soft-tissue mass of firm consistency, roughly spherical in shape with a diameter of approximately 1.25 cm. It appeared to be either originating in, or adherent to, the ligamentum flavum, and was attached to the ventral aspect of the C-4 lamina on the left side only. No pus was visible, and the mass was virtually avascular. It was dissected sharply from the dura.

**Pathological Examination.** Histological examination of the surgical specimen revealed fibrous tissue and portions of the ligamentum flavum with degenerative changes and focal dystrophic calcification, with some granulation tissue formation (Fig. 3). There was no evidence of neoplasia, and cultures were negative.

**Postoperative Course.** Within 2 weeks, the patient had recovered normal power in the left upper extremity and had improved to 4+/5 power on the right side. Further improvement was documented at 6 months.

**Discussion**

A case of progressive cervical myelopathy was presented secondary to a focal fibrous, calcified mass involving the ligamentum flavum in an elderly man with DISH. To our knowledge, no such complication has previously been reported in association with this entity. "Diffuse idiopathic skeletal hyperostosis" is a relatively new term used to describe an old disease known by a variety of synonyms including "Forestier's disease," "spondylitis ossificans ligamentosa," and "spondylosis hyperostotica." An "enthesis" is an insertion of muscle or ligament into bone, hence DISH has been referred to as an "enthesopathy" since it is characterized by ossification of paraspinal and extra-axial ligamentous structures, particularly at points of attachment to bone. The three spinal enthesopathy syndromes are ossification of the anterior longitudinal ligament, of the posterior longitudinal ligament, and of the vertebral arch ligaments, with DISH being a diffuse form of these, but with extraspinal involvement as well.

The syndrome of DISH generally presents in middle-aged or elderly patients, and more often in men. Its manifestations are pain, decreasing mobility, and deformity involving (in decreasing order of frequency) the thoracic, lumbar, and cervical spine. Additional complaints may be peripheral skeletal pain or dysphagia. There may be periostitis at ligamentous attachments to bone, and a frequent occurrence of tennis elbow, Achilles tendonitis, and olecranon, patellar, and calcaneal spurs.

In an attempt to refine the terminology and separate the florid DISH syndromes from similar but less extensive diseases, strict radiological diagnostic criteria have been established. These are: 1) flowing calcification and ossification along the anterolateral aspect of at least four contiguous vertebral bodies; 2) relative preservation of disc height; and 3) absence of apophyseal joint bone ankylosis and sacroiliac joint erosion, sclerosis, or fusion. Criterion 1 allows distinction of DISH from simple degenerative spondylitis, Criterion 2 distinguishes DISH from degenerative intervertebral disc disease, and Criterion 3 allows exclusion of ankylosing spondylitis.

The neurosurgical interest of the present case is the relatively rapid progression of a myelopathy in a patient with DISH due to an extradural mass at C-4. The rapid clinical course over 3 weeks and the radiological studies supported a preoperative diagnosis of an epidural neoplastic process or possibly an epidural abscess. It was, therefore, surprising that the compressing lesion was a
Cervical myelopathy in skeletal hyperostosis

FIG. 2. Left: Cervical myelogram, lateral view, showing almost complete extradural obstruction to contrast flow at the C-4 level. The vertebral alignment is good and paravertebral calcification is minimal. Right: Postmyelogram axial computerized tomography scan at the C-4 level showing a posterior extradural mass causing marked spinal cord deformation (arrow).

large focal, globular, fibrous tissue proliferation involving the ligamentum flavum unilaterally at one level. We believe that this unusual lesion which included dystrophic calcification and identifiable ligamentum flavum was a complication of DISH. In this case the involved enthesis was that of ligamentum flavum into the C-4 lamina. While the lesion was adherent to the dura, it did not originate from within it, and therefore did not represent a focal example of pachymeningitis cervicis hypertrophica. The focal nature and the absence of a prominent mononuclear infiltrate distinguishes our case pathologically from the latter disorder. A review of the literature disclosed no similar cases. The only myelopathies related to DISH have specifically involved ossification of the posterior longitudinal ligament or posterior osteophytes or trauma. The only other neurological complications reported in association with DISH have been two cases of multiple peripheral nerve entrapment and one case of thoracic outlet syndrome.

This case was presented in order to add another pathological entity to the list of possible etiologies of compressive myelopathy.

References


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