

Cervical Cord Compression In Diffuse Idiopathic Skeletal Hyperostosis

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Diffuse idiopathic skeletal hyperostosis (DISH) is a relatively common disorder predominating in men (65 to 70 percent),¹ with a prevalence of approximately 3 percent in adults older than 40 years of age² and from 12 percent to 15 percent in patients more than 65 years of age.^{3,4} In 1950 Forestier and Rotés-Querol⁵ introduced the term *senile ankylosing hyperostosis of the spine*. DISH has long been regarded as a radiographic entity characterized by calcification and ossification of the anterolateral aspects of the vertebral column whose clinical manifestations (usually thoracolumbar or cervical stiffness) are minor and of little consequence. Some severe neurologic complications, however, resulting from spinal cord compression have been recently reported.⁶⁻⁸ Typically, ossification of the posterior longitudinal ligament of the spine has not been considered a component of this process, and its prevalence is estimated to be approximately 6 percent in individuals more than 65 years of age.⁹ Nevertheless, Resnick, et al.¹⁰ reported that ossification of the posterior longitudinal ligament occurs in 50 percent of patients suffering from DISH. Reported here is a patient who experienced progressive quadriplegia. Diagnostic studies evidenced the existence of both disorders. A brief discussion and recommended strategies follow.

Case Report

A 70-year-old man came to our health center complaining of an inability to walk. He reported a 10-year history of slow progressive weakness in upper and lower extremities and paresthesias in his feet and hands. Cervical and thoracolumbar stiffness and mild neck-to-lower-back pain were also noted, as well as an occasional pain in his shoulders, hips, and knees. There was no bladder

or rectal incontinence. The physical examination showed muscle atrophy, generalized hyperreflexia, and fasciculations in his arms and legs. Sensory loss was observed both in upper and lower extremities. Results of pertinent laboratory tests, including hemogram, proteinogram, sedimentation rate, and glucose, urea, uric acid, calcium, phosphorous, creatine phosphokinase, and aldolase levels, were normal. Serologic tests for syphilis, rheumatoid factor, antinuclear antibody, and HLA-B27 antigen were negative. Standard radiographs of his spine showed flowing calcification of anterolateral aspects extending from C-3 to C-7 (Figure 1), disk spaces were well maintained, and there was no evidence of sacroiliitis. Acetabular hyperostosis was observed in the pelvis radiography. Computed tomographic (CT) scanning of the cervical column (Figure 2) showed ossification of the posterior longitudinal ligament causing severe narrowing of the vertebral canal and compression of the spinal cord. Calcification of the anterolateral ligament was also observed. The electromyogram did not show any generalized lesion of the spinal motor neuron. The patient refused further studies, such as magnetic resonance imaging (MRI). The diagnosis of DISH was made, and the patient was treated with conservative therapy, i.e., continuous skull traction, bed rest, application of a neck brace, and rehabilitation. Despite this treatment, neurologic impairment continued. The patient chose not to have surgery. At the present time, he has total quadriplegia and is confined to a wheelchair.

Discussion

Resnick and coworkers^{1,4,10} have investigated DISH in depth and established three diagnostic criteria: (1) flowing calcification and ossification along the anterolateral aspect of at least four contiguous vertebral bodies with or without associated localized pointed excrescences at the intervening vertebral body-intervertebral disc junctions; (2) relative preservation of intervertebral disk height in the involved vertebral segment

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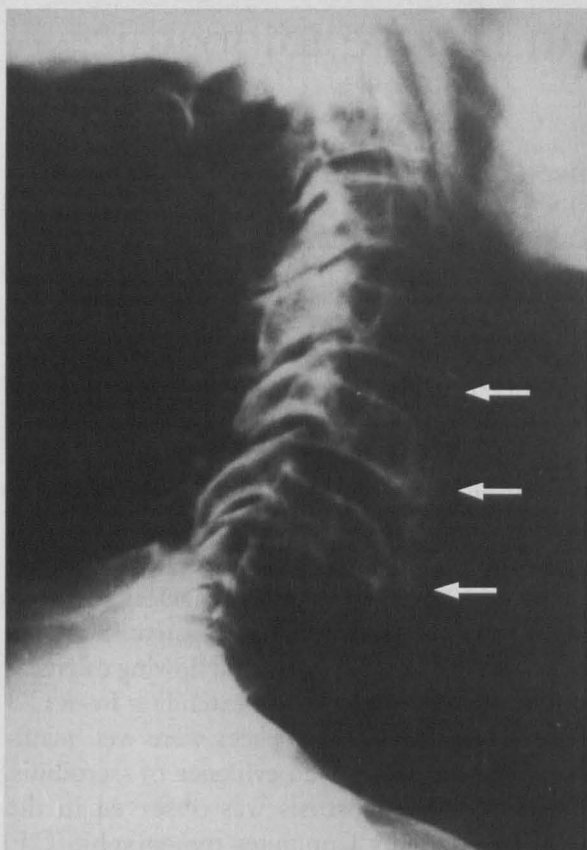


Figure 1. Lateral radiograph of the cervical spine with flowing calcification from C-3 to C-7.

and absence of extensive radiographic changes of degenerative disk disease, including vacuum phenomena and vertebral body marginal sclerosis; and (3) absence of apophyseal joint bony ankylosis and sacroiliac joint erosion, sclerosis, or intra-articular osseous fusion. Our patient met these three diagnostic criteria. Extraspinal radiographic manifestations, such as ligament calcification and ossification and para-articular osteophytes, have also been reported.¹ Although these manifestations can occur at any skeletal site, the hip, knee, heel, shoulder, and elbow locations are the most frequent.

Despite evidence that DISH is an extremely common entity, numerous reports fail to delineate the broad clinical spectrum of this disorder. Nevertheless, clinical data from several reported series^{1,5,10} show that such symptoms as thoracolumbar or cervical stiffness, restricted motion, dysphagia, or bone and joint pain are not uncommon. Because these clinical manifestations can be noted in the context of several rheumatoid diseases, the differential diagnosis of DISH should be made using radiographic studies. Degenerative

disk disease osteoarthritis, spondylosis deformans, and ankylosing spondylitis have different radiographic features that allow us to differentiate these diseases from DISH.¹ Laboratory results for patients with DISH are nonspecific and therefore should be used in conjunction with radiographic evaluation to exclude other conditions, such as inflammatory, infectious, rheumatoid, or collagen vascular diseases.

In the case reported here, the diagnosis of DISH can be made. The question then would be, is DISH responsible for all the symptoms? Certainly, it seems that the patient's quadriparesis was not a consequence of this fairly common radiographic diagnosis. In fact, according to our literature review, the case in which symptomatic spinal cord compression resulted exclusively from DISH was unique.⁸

In our case, the patient was suffering from symptoms caused by DISH, as well as a slow progressive spastic quadriparesis and some sensory loss. This clinical feature corresponds to transversal injury of the spinal cord at the cervical level.

Differential diagnosis of several diseases should be considered at this point. Initially, the slow progression of symptoms allows us to leave out of



Figure 2. Cervical computed tomographic scanning: 1. Calcification of anterolateral aspects of vertebral column. 2. Calcification of posterior longitudinal ligament of spine.

consideration such acute disorders as traumatism, hemorrhage, or ischemia of the spinal cord. Acute or subacute infectious diseases, such as viral myelitis, can also be ruled out. Chronic infections, such as tuberculosis and brucellosis, can also cause spinal cord damage, but it is difficult to recognize these diseases in the clinical history of our patient. Neurosyphilis also can be excluded, as anamnesis did not reveal previous infections and serologic tests were negative. Multiple sclerosis that is characterized by recurrent crises of local or multifocal neurologic dysfunction of the central nervous system¹¹ should also be considered. Our patient, however, did not have any remission throughout the course of the illness, and only a unique neurologic focus was detected. Finally, amyotrophic lateral sclerosis could be considered in the differential diagnosis, because the physical examination revealed muscular atrophy and fasciculations. The patient, however, described a clear sensorial dysfunction, and the electromyogram did not detect any generalized lesion at the spinal motor neuron; therefore, we can confidently dismiss this diagnosis, too.

At this point it can be suggested that spinal cord injury is caused by intramedullar or extramedullar compression. The main causes of spinal cord compression are as follows:

1. Skeletal conditions: disk hernia, osteophytosis, and any other spinal canal stenosis
2. Primary tumors: neurofibroma, meningioma, ependymoma, astrocytoma, or angioma
3. Metastasis
4. Infections: abscess, Pott disease, or granuloma
5. Syringomyelia

CT scanning (Figure 2) showed an osseous density zone that corresponds anatomically to the ossified posterior longitudinal ligament, causing a marked stenosis at the spinal canal. Because no evidence of expansive processes in the spinal cord or vertebral bodies was observed in the serial tomographic images, the diagnosis of DISH associated with ossification of the posterior longitudinal ligament can be made, with the latter being responsible for both the cervical cord compression and the neurologic symptoms. Association between DISH and ossification of the posterior longitudinal ligament has been described previously.¹⁰ It is important to emphasize that this association occurs in most of the

patients with DISH and progressive or acute myelopathy,^{6,7,9} as we observed in our patient.

Because it is difficult to recognize ossification of the posterior longitudinal ligament on a standard lateral view of the cervical spine, CT scanning, MRI, and contrast myelography have been the recommended techniques to evidence this condition and its complications.⁹ Contrast myelography, however, should be avoided because sudden neurologic deterioration can occur as Griffiths and Fitzjohn have described.⁷

Management of DISH in patients with or without moderate symptoms should be conservative, i.e., bed rest, cervical traction, and rehabilitation. Severe symptoms can alert us to the coexistence of ossification of the posterior longitudinal ligament. In these cases a trial of conservative therapy, together with brief courses of high-dose dexamethasone, is indicated to reduce associated edema of the spinal cord. Surgery should be considered both in patients who have persistent or progressive myelopathy despite conservative treatment and in patients who are experiencing rapidly progressive myelopathy.

Conclusion

Although the case reported described a relatively common disorder in general practice, this condition might not be noticed because its clinical manifestations are minor and of little importance, and radiographic studies are necessary for diagnosis. Nevertheless, the combination of DISH and ossification of the posterior longitudinal ligament can cause severe neurologic complications, as occurred in our patient. We think it is important to diagnose this disorder accurately in patients with thoracolumbar or cervical pain or stiffness. These patients should also receive follow-up care, specially if neurologic symptoms, such as weakness, fasciculations, or muscle atrophy, are present.

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