

An underrecognized cause of dysphagia: Forestier's disease

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INTRODUCTION

Diffuse idiopathic skeletal hyperostosis (DISH), also known as Forestier disease [1], was first described in 1950 by J. Forestier. Flowing calcification along the anterior and lateral sides of the vertebral bodies produces the appearance of candle wax dripping down the spine. Most patients are free of symptoms, so that DISH is usually discovered fortuitously upon plain radiographs of the spine obtained for another reason. A few patients experience spinal pain, spinal stiffness, or dysphagia. We report a case in which the diagnosis was made upon evaluation for dysphagia.

CASE REPORT

A 79-year-old man with an unremarkable medical history was referred to an otorhinolaryngology outpatient clinic for difficulty swallowing solid foods. The dysphagia had worsened gradually over the last few months. He had no history of smoking or alcohol abuse. His general health was good, and he had no

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Abbreviations: ALL, Anterior longitudinal ligament; PLL: Posterior longitudinal ligament.

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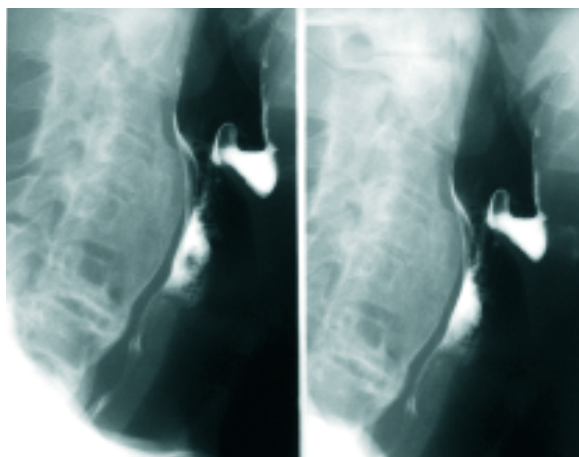
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respiratory symptoms. The oral cavity and oropharynx were normal to physical examination. Endoscopy of the pharynx and larynx showed a posterior bulge consistent with extrinsic compression.

An upper gastrointestinal tract series disclosed flowing calcification along the anterior aspect of five vertebrae, from C3 to C7. The calcification pressed on the posterior wall of the hypopharynx, without causing stasis or blockage of the contrast medium (Figure 1). Computed tomography (CT) (Figure 2a-2b) confirmed the anterior ossification extending from C3 to C7 and impinging on the hypopharynx. A space was clearly visible between the ossification and the spine, establishing that the lesion was due to ossification of the anterior longitudinal ligament (ALL), as opposed to osteophytosis. Also visible on the CT images were ossification of the ALL along the thoracic spine and calcifications on the posterior side of a vertebral body indicating involvement of the posterior longitudinal ligament (PLL). Finally, CT showed fluid-density

Figure 1: Upper gastrointestinal tract series, lateral view.

Flowing ossification along the anterior aspect of the spine impinging on the posterior pharyngeal wall.



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Figure 2A: Computed tomography, axial section through C4 after intravenous injection of an iodinated contrast agent.

Tumor-like mass by pseudotumoral left piriform sinus retention caused.

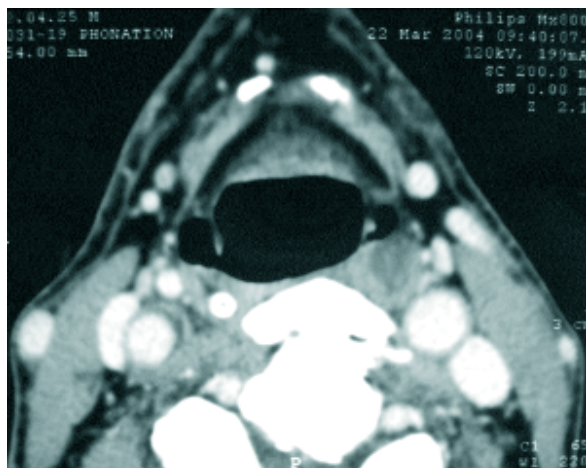
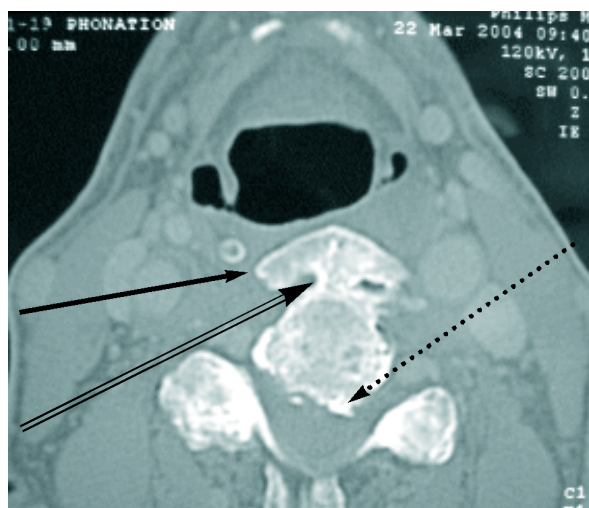


Figure 2B: Computed tomography, axial section through C4 after intravenous injection of an iodinated contrast agent (bony window).

Large ossification in the anterior longitudinal ligament (plain arrow) separated from the vertebral body by a cleavage plane (double arrow) and impinging on the posterior pharyngeal wall; moderate ossification in the posterior longitudinal ligament (dashed arrow).



nonenhancing material blunting the left piriform sinus and consistent with retention of secretions.

The sacroiliac joints were normal by plain radiogra-

phy. This finding, together with the appearance of the spinal lesions, led to a diagnosis of DISH. The patient was referred to the surgery department for evaluation of treatment options.

DISCUSSION

DISH is characterized by ossification of tendons and ligaments at their sites of attachment to bone.

Although its origin remains unknown, abnormal vitamin A metabolism or insulin-induced growth hormone stimulation have been suggested [2-3]. Risk factors may include type 2 diabetes, hyperuricemia, and overweight [4].

DISH is common, with a prevalence of 5 to 15% among individuals older than 60 or 70 years [5]. Men are affected predominantly in a ratio of 3/1. Ossification of the ALL is the most common lesion and predominates at the level of thoracic spine. However, concomitant ossification of the PLL and ligamentum flavum is common [6]. Extraspinal lesions may develop, the main sites of involvement being the pelvis, calcaneus, olecranon, and patella [3]. DISH is usually asymptomatic. The main initial symptoms are spinal pain and stiffness, but these do not always prompt a physician visit. The proportion of cases revealed by dysphagia has ranged across studies from 0.1% to 28% [7]. The dysphagia is usually marked, present for solid foods, improved by anterior flexion of the neck, and worsened by extension of the neck. Concomitant symptoms may include a foreign body sensation, odynophagia, reflex otalgia, salivary stasis, dysphonia, dyspnea, sleep apnea, and aspiration. Although DISH causes a mass syndrome in the retropharyngeal space, the epicenter is in the prevertebral space [10]. Thus, the picture should be distinguished from a retropharyngeal mass syndrome. In addition to ossification, inflammation develops, causing mucosal thickening or laryngeal immobility and producing a tumor-like formation. There have been a few reports of neurological complications including spinal cord or nerve root compression, peripheral nerve entrapment, and recurrent laryngeal nerve palsy. Spinal cord or nerve root compression is caused by ossification of the PLL or ligamentum flavum, probably in combination with congenital spinal stenosis.

Because the clinical symptoms lack specificity, the diagnosis rests on imaging studies. The diagnostic cri-

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teria developed by Resnick et al. [9] are still used:

- calcifications and ossifications along the anterolateral aspect of at least four contiguous vertebral bodies,
- normal disk space height in the affected segments, absence of facet joint fusion,
- absence of clinical or radiological evidence of sacroiliitis.

The cleavage plane seen on plain radiographs and clearly delineated on CT sections differentiates DISH from osteophytosis (osteoarthritis). Although plain radiography remains the reference investigation, CT visualizes:

- the cleavage plane,
- ossifications in the PLL,
- ligamentum flavum, moreover, it allows measurement of the anteroposterior diameter of the spinal canal.

Magnetic resonance imaging (MRI) is indicated in patients with neurological manifestations to look for spinal cord compression and concomitant myelopathy. DISH may be discovered on an upper gastrointestinal tract series obtained to investigate dysphagia.

The main differential diagnoses are osteoarthritis when the lesions simulate osteophytes and spondyloarthropathy when they suggest syndesmophytes. Resnick's criteria are invaluable to correct the diagnosis in these situations. Although retropharyngeal calcific tendinitis, discitis, and retropharyngeal abscess can produce a similar retropharyngeal mass, the imaging study findings are very different, without the flowing calcification typical of DISH. Rarely, a flowing paravertebral ossification may require discussion of fluorosis, hypoparathyroidism, or familial hypophosphatemic vitamin D-resistant rickets.

No treatment capable of preventing the progression of vertebral hyperostosis in DISH patients is available to date. The management includes three components, physical, pharmacological, and surgical. Physical therapy may improve range of motion in patients with spinal stiffness. Nonsteroidal antiinflammatory agents or glucocorticoid bolus therapy in combination with a muscle relaxant can be used to improve the symptoms. Dietary precautions aimed at minimizing gastroesophageal reflux are in order in patients with dysphagia. Spinal pain may require analgesic therapy. Surgery is needed to prevent aspiration pneumonia in patients with severe dysphagia related to a markedly prominent ossification [10]. Anterior decompression with stabilization by an interbody cage can be performed. A concomitant space-occupying pharyngeal

lesion should be looked for before initiating the treatment of DISH.

CONCLUSION

DISH, a common after 60 years of age, may be revealed by dysphagia. In an elderly patient with dysphagia and no otorhinolaryngological lesions, an upper gastrointestinal series should be obtained. Plain radiographs ensure the diagnosis, although CT provides additional information. When reading CT sections of the pharynx, the bony structures should be examined carefully. Thorough familiarity with the signs of DISH is essential to ensure the diagnosis of this disease that can cause major morbidity related to swallowing dysfunction and to aspiration. Because DISH is extremely common, it should not be accepted as the cause of dysphagia until other lesions, most notably tumors, have been convincingly ruled out by a thorough otorhinolaryngological examination.

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