

ARE NEW DIAGNOSTIC CRITERIA NEEDED? DIFFUSE IDIOPATHIC SKELETAL HYPEROSTOSIS CONCEALED BY CERVICAL RADICULOPATHY: A CASE REPORT

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Background: Diffuse idiopathic skeletal hyperostosis (DISH) involves widespread ossification of ligaments in the skeleton with associated nonspecific symptoms. Cervical DISH may result in dysphagia and orthopnea but can be missed with accompanying spinal pathology.

Case Report: A 52 year-old man with a history of coronary artery disease presented with worsening neck pain radiating to the bilateral upper extremities. Magnetic resonance imaging revealed ventral osteophytes at C3-C7 and moderate stenosis at C4-C5. The patient underwent cervical epidural steroid injections resulting in partial pain relief. However, axial neck pain persisted and he reported new-onset dysphagia and orthopnea. Subsequent imaging showed C2-C7 anterior osteophytes with tracheal displacement, confirming the DISH diagnosis. The patient underwent resection of the osteophytes and had subsequent complete symptom resolution.

Conclusion: Although radicular symptoms improved with an injection, dysphagia not previously mentioned was concerning. DISH is routinely missed in the setting of spinal pathology, such as radiculopathy. The presence of anterior cervical spine osteophytes should prompt further respiratory or swallow evaluation.

Key words: Pain medicine, spine, dysphagia, orthopnea, DISH, cervical spine

BACKGROUND

Diffuse idiopathic skeletal hyperostosis (DISH) is a systemic musculoskeletal condition first described in 1950 by Dr. Forestier as ossification of ligaments of the axial and appendicular skeleton (1). The incidence of DISH increases as individuals get older. Over 10% of adults above 50 have DISH, which is also more frequently observed in men (1).

Patients often present with stiffness affecting their range of motion and progressively worsening back pain with or without radicular involvement. Additional

symptoms may be present depending on the disease's location, such as in the cervical spine area in close proximity to the trachea and esophagus (1-3). Due to its nonspecific symptoms, DISH is routinely overlooked, which may prove costly.

We present a case of a 52-year-old man whose ongoing neck and radicular pain masked underlying, isolated cervical DISH causing dysphagia and orthopnea. Due to the insidious and potentially lethal nature of this disease, we recommend modifying the current diagnostic criteria to better detect DISH. We also recommend

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establishing novel guidelines to allow for more sensitive monitoring.

CASE PRESENTATION

A 52-year-old man with a history of microvascular coronary artery disease (CAD), renal stones, gastric cancer (in remission), and L3-L4, L4-L5, and L5-S1 fusion presented with worsening neck pain radiating to the bilateral upper extremities. Radicular symptoms radiated along the lateral proximal arms in a roughly C5 distribution. Magnetic resonance imaging (MRI) revealed ventral osteophytes from C3 to C7 and multi-level spondylosis featuring broad-based central disc protrusion at C4-C5, causing mild-to-moderate stenosis.

Electromyography of the upper extremities was unremarkable. The patient had unsuccessful pain relief with acetaminophen, nonsteroidal anti-inflammatory drugs (NSAIDs), duloxetine, gabapentin and opioids. He could not tolerate physical therapy due to ongoing pain. He underwent 2 cervical epidural steroid injections, which resulted in 70% radicular pain relief. His axial neck pain persisted and the patient developed worsening dysphagia and orthopnea.

Computed tomography (CT) of the cervical spine showed prominent anterior osteophytes from the C2 to C7 vertebral levels measuring up to 20 X 25 mm axially with tracheal displacement and stenosis (Figs. 1-3). The patient was diagnosed with DISH and referred to our surgery colleagues. He underwent surgical removal of the osteophytes. The patient reported complete resolution of dysphagia and improved orthopnea. Furthermore, he reported 100% pain relief as well as increased participation in his normal day-to-day activities.

DISCUSSION

DISH is defined per the Resnick criteria (RC), which constitute flowing ossification over at least 4 adjacent vertebrae, sparing of the intervertebral disc, and non-involvement of the apophyseal and sacroiliac joints (2). Growth hormone and insulin-like growth factors are significantly elevated in patients with DISH (2). These factors contribute to abnormal bone formation through inflammatory mediators and induce transformation of mesenchymal cells into fibroblasts and osteoblasts (2,3). Although symptoms primarily develop gradually, serious complications may arise. In addition to orthopnea or dysphagia, as evident in this case, patients are at risk for vertebral fractures, myelopathy, and aspiration pneumonia (4).

Treatment for DISH is initially conservative with a trial of physical therapy, medical management, and lifestyle changes, such as modification of diet and losing weight (2). Medications targeted for symptomatic pain relief include NSAIDs, muscle relaxants, and neuropathic medications (2). Surgical intervention becomes necessary for patients when the advancement of the disease hampers their ability to perform daily activities or poses a life-threatening risk. This is particularly evident in cases involving dysphagia, orthopnea, and spinal cord compression (1,2).

Osteophytic growth varies along the axial skeleton. Vasculature overlying the vertebral column serves as a barrier for continued osteophytic expansion owing to the asymmetric osteophyte growth in the thoracic spine, where it is more common than its cervical DISH counterpart (5,6). In stark contrast, growth in the cervical spine arises symmetrically due to the absence of segmental areas which can lead to unmitigated forward bone expansion, hence a higher probability of acquiring dysphagia or orthopnea (7).

After spinal stiffness, dysphagia is the second most common concern for patients with cervical DISH, with an incidence of 0.2% to 0.8% (8-10). The increasing use of CT better delineates structural bony abnormalities and in fact served as the confirmatory test for our patient (2). Given the esophagus is only 1 to 3 mm away from the cervical spine at C5 to C7, dysphagia is most commonly seen with ventral osteophytic growth from C4 to C6 (5,10,11).

Osteophytes cause dysphagia by restricting mobility of the epiglottis, impairing the larynx's anterior and elevating movement, and thereby directly obstruct the transport of the food bolus (10). Seidler et al (10) noted that, on average, the size of the osteophytes causing dysphagia in patients with cervical DISH was 10 or more mm. This was much less than the size measured in our patient, which was greater than 20 mm. Many patients with DISH who have dysphagia will live up to 3 years before having it addressed (12). Although modifying food texture and consistency is preferred initially, surgery via the anterolateral approach has had a high success rate with significant improvement in symptoms and should be considered earlier in the disease process (12).

Patients with cervical DISH may also develop respiratory symptoms due to airway compromise from mechanical osteophyte compression, especially at C3-C4 (13). Respiratory symptoms can also arise from irritation to the laryngeal nerve causing hoarseness,

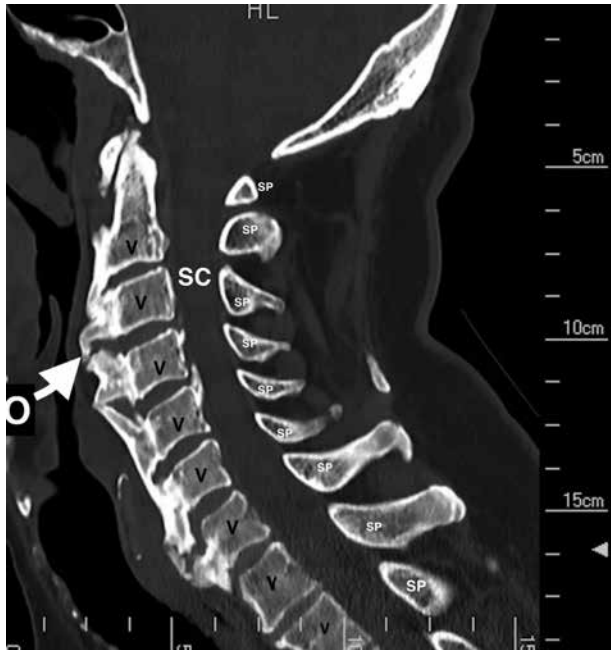


Fig. 1. Computed tomography sagittal view of the cervical spine showcasing contiguous anterior ossification from C2 to C7 vertebra levels. O, contiguous osteophytic growth; SC, spinal canal; V, vertebrae; SP, spinous process.

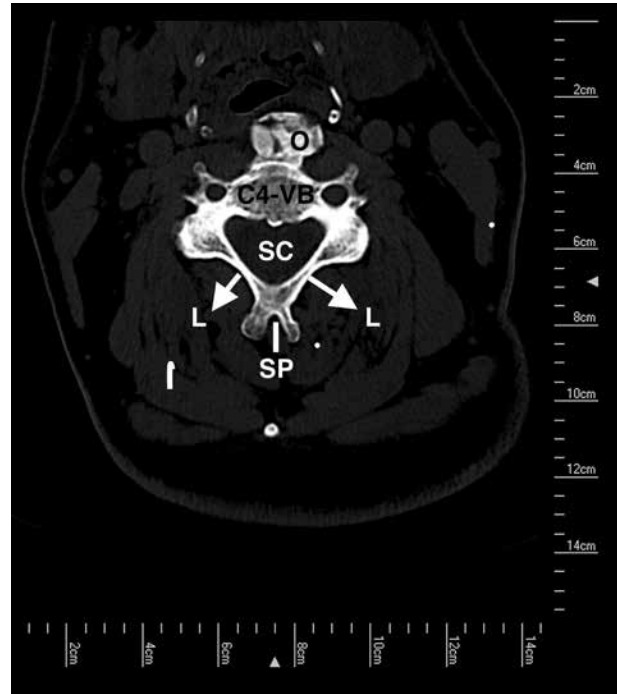


Fig. 2. Computed tomography axial view of the cervical spine at the C4 vertebra level. C4-VB, 4th cervical vertebral body; L, lamina; O, osteophyte; SP, spinous process.

dysphonia, and bilateral vocal cord dysfunction (1,8,9). Moreover, ossification of the sterno-costochondral junctions restricts expansion of the rib cage, further impeding pulmonary function. Routine airway exams commonly miss cervical osteophytes therefore respiratory complaints are attributed to another underlying disease, such as obstructive sleep apnea, especially when the patient may have more befitting risk factors such as obesity (13). Due to DISH progression, orthopnea may evolve to respiratory failure and may render an airway a difficult intubation (1). This highlights the need to collectively initiate a protocol for interval imaging to prevent these complications.

Our patient's MRI revealed a disc protrusion at C4-C5 causing spinal stenosis and confirmed his presentation of bilateral radicular symptoms in the C5 dermatome. Likely, the ventral osteophyte noted growth from C2 to C7 was attributed to spondylosis. Due to the stringent RC needed to diagnose DISH, the disease was not considered. Even when imaging meets the RC, DISH is still missed. For instance, Westerveld et al (11) reviewed approximately 501 chest radiographs referred for non-spine-related conditions and found a 17% prevalence of DISH, although it was not listed on the impression. RC warrants flowing



Fig. 3. Computed tomography axial view of the cervical spine at the C5 vertebra level. C5-VB, 5th cervical vertebral body; L, lamina; O, osteophyte; SP, spinous process.

ossification between 4 contiguous vertebrae, but this alludes to a terminal or late stage form of the disease. DISH therefore is officially diagnosed from the presentation of complications such as dysphagia, dysphonia, hoarseness, orthopnea, or aspiration pneumonia among others (1,4,5). Unfortunately, the clinical course at this junction may point towards a poor prognosis.

As the RC require a tall benchmark to make a DISH diagnosis, understanding associations to this disease may help uncover many hidden DISH diagnoses. Our patient's medical history consisted of CAD. The Framingham score was found to underestimate the risk of cardiovascular disease (CVD) in patients with DISH when compared to their non-DISH counterparts, as 39% developed CVD when only 28.6% were predicted (14). This implies that DISH may serve as an independent risk factor for developing CVD (14). Lantsman et al (15) showed no correlation, however, between CVD and DISH when tracking coronary artery calcification and the stenotic artery classification system through imaging (15). Although evidence for CVD's link to DISH is inconclusive, obesity, diabetes, and metabolic syndrome have been independently linked to DISH (2,3). Given our patient's history of gastric cancer, limited data suggest a correlation. Notably, there has been a published case report of patients with non-small cell lung cell carcinoma diagnosed with DISH (13). Since insulin-like growth factor 1 levels are elevated in various cancers, further studies might also point toward this association.

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There are helpful diagnostic procedures to monitor patients with DISH who are at risk of developing complications. Dynamic video fluoroscopy assesses swallow function alterations by revealing smooth extrinsic indentation in the esophagus (9). Via dynamic video fluoroscopy, peristalsis of the pharyngeal wall is weakened above and below the osteophyte level, causing the obstruction in transporting solids (10). Video laryngoscopy has had success assessing airway and respiratory function, and therefore should be optioned in cases of orthopnea (13).

CONCLUSIONS

DISH is a progressive inflammatory disease that is commonly missed by providers due to its slow growth, nonspecific symptoms, and difficult-to-meet criteria. Spinal pathology is commonplace in the aging population, which further contributes to underdiagnosing DISH. We recommend modifying the current RC in order to diagnose DISH earlier. We also recommend establishing novel guidelines on interval imaging for newly discovered ventral anterior vertebral column osteophytes in order to prevent disastrous complications.

Contributions

Tejas Shah: This author is the corresponding primary author and co-wrote the manuscript.

Hassan Aboumerhi: This author served as the secondary author and co-wrote the manuscript.