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CASE REPORT

DYSPHAGIA: AN INFREQUENT SYMPTOM OF DIFFUSE IDIOPATHIC SKELETAL HYPEROSTOSIS

ABSTRACT

Diffuse idiopathic skeletal hyperostosis (DISH) is a disorder of unknown aetiology characterised by exuberant hyperostosis of the spinal column, that occasionally leads to bone ankylosis and ossification of extra-spinal entheses. Associations with systemic conditions including obesity, hypertension, diabetes mellitus, hyperinsulinaemia, dyslipidaemia and hyperuricaemia have been reported. The thoracic spine is typically involved in DISH followed by thoracolumbar and cervical involvement. Herein, we present a 63-year-old woman with cervicothoracic spine involvement leading to pharyngeal impingement and dysphagia. Clinicians should be aware of this rare clinical manifestation of DISH of the cervical spine and patients should be informed about and followed up for dysphagia.

Key Words: Dysphagia; Cervical Spine; Diffuse Idiopathic Skeletal Hyperostosis.



OLGU SUNUMU

DISFAJİ: DİFÜZ YAYGIN İDİYOPATİK İSKELET HİPEROSTOZİS SENDROMUNDA GÖRÜLEN BİR SEMPTOM

Öz

Yaygın İdiyopatik İskelet Hiperostozu (DISH) omurganın aşırı hiperostozisi ile karakterize olup; bazen kemiklerde ankiroz ve ekstra-spinal entezis bölgelerinde ossifikasyonlara yol açan, etyolojisi bilinmeyen bir hastalıktır. Obezite, hipertansiyon, diabetes mellitus, hiperinsülinemi, dislipidemi ve hiperürisemi gibi sistemik hastalıklarla ilişkisi bildirilmiştir. DISH sendromunda tipik olarak torakal omurga tutulumu görülür ve bunu torokolomber ve servikal tutulum takip eder. Bu yazıda, farekse bası ve yutma güçlüğüne neden olan servikotorasik omurga tutulumuna sahip 63 yaşında bir kadın hasta sunuldu. Hekimler servikal tutulumla giden DISH sendromunun bu nadir görülen klinik belirtisinin farkında olmalı ve hastalar yutma güçlüğü açısından bilgilendirilip takip edilmelidirler.

Anahtar Sözcükler: Diffüz İdiyopatik İskelet Hiperostozu; Disfaji; Servikal Omurga.



INTRODUCTION

Diffuse idiopathic skeletal hyperostosis (DISH), also known as Forestier's disease (1) is characterized by flowing calcification along the sides of the contiguous vertebrae of the spine. While the aetiology remains unknown, associations with some systemic conditions and comorbidities including obesity, hypertension, diabetes mellitus, hyperinsulinaemia, dyslipidaemia, hyperuricaemia, environmental factors (fluoride), diet and drugs (isotretinoin, acitretin) and metabolic conditions (elevated levels of insulin, insulin-like growth factor-1 and growth hormone, obesity, hyperlipidaemia, hyperuricaemia) have been hypothesised (1–4). Although the exact prevalence and incidence remains unclear, incidence increases with age (5). The thoracic spine is typically and almost always involved followed by thoracolumbar and cervical involvement. DISH is a slowly progressive disorder and might lead to pain and limitation in the affected part of the spine. Some subjects are asymptomatic and are diagnosed incidentally with the observation of ossifications on radiograms obtained for other medical reasons. Due to cervical involvement dysphagia can occur and the incidence varies between 17% and 28% (6). Giant spurs and calcifications in the cervical region may cause dysphagia and airway obstruction. In this report, we discuss a 63-year-old woman with cervicothoracic DISH resulting in dysphagia along with a literature review.

CASE

We report a 63-year-old overweight woman admitted to the outpatient clinic with the complaint of neck pain referring to her shoulders and bilateral hand numbness. Past medical history was unremarkable except for type 2 diabetes mellitus which was diagnosed 20 years ago. She was being administered oral antidiabetic drugs and no other medications. On physical examination, cervical range of motion (ROM) was severely limited and painful in all directions. Neurologic examination revealed hypoactive deep tendon reflexes with grade +4/5 power in the upper extremities. Lower extremity muscle power was 5/5 bilaterally with flexor plantar responses. Sensory examination revealed no abnormalities except bilateral hypoaesthesia in the median nerve territory which was prominent on the right side. Electrophysiological evaluation revealed bilateral carpal tunnel syndrome (CTS) of moderate degree on the right and mild degree on the left side. Laboratory tests including whole blood count, C-reactive protein and erythrocyte sedimentation rate were normal except fasting glucose level (175 mg/dl) and HbA1c=10.9 (normal range=4.0–6.3). She was referred to the endocrinology clinic for the management of blood glucose.

Cervical anteroposterior and lateral spine radiographs revealed the characteristic flowing ossification of DISH along the right anterolateral aspect of the cervical vertebrae from C2–C7 (Figure 1). Similarly, anteroposterior and lateral radi-



Figure 1— **a.** Anteroposterior and **b** lateral radiographs of cervical spine show characteristic flowing ossification along the right anterolateral aspect of the cervical vertebrae from C2–C7 (arrows).

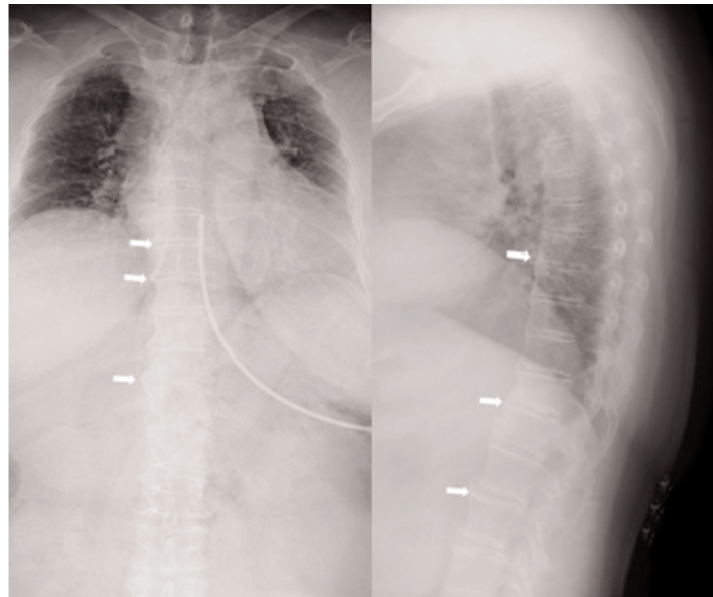


Figure 2— AP and lateral views of thoracic spine show the characteristic flowing osteophytes in the right anterolateral aspect (arrows).

ographs of the thoracic spine also showed characteristic flowing osteophyte formation on the right anterolateral side of thoracic vertebrae (Figure 2).

Upon questioning, the patient reported difficulty swallowing and a foreign body sensation in her throat. She admitted that she had never related the difficulty in swallowing with the neck pain, and therefore, did not bring it up previously. She was informed about the giant osteophytes in the cervical region which might be the cause of her symptoms. Cervical non-enhanced computed tomography (CT) was performed for a detailed assessment of possible pharyngo-oesophageal involvement. Sagittal reconstruction of CT scans demonstrated hyperostosis and anterior cervical fusion from C2–C7 and axial CT imaging demonstrated the pharyngeal compression due to right anterolateral osteophyte formation (Figure 3).

Nonsteroidal anti-inflammatory (NSAID) medication was prescribed together with physiotherapy and an exercise program. Bilateral hand-wrist splints were recommended for the CTS. Neck pain and limitation in cervical ROM improved at the end of a three weeks physical therapy and rehabilitation programme. She was advised to continue with the cervical ROM and strengthening exercises and to adhere to a diabetic diet. She is still on regular follow-ups.

DISCUSSION

Diffuse idiopathic skeletal hyperostosis is shown to be related to systemic conditions and metabolic syndrome (3–5). Our patient was also diabetic and overweight. While decreased range of spinal motion especially a notable loss of thoracic lateral flexion is the most common finding on physical examination, exuberant calcification of ligament as well as spurs could impinge on other structures such as the oesophagus and the larynx, leading to swallowing deficits or dysphonia (7–11). Our patient did not mention swallowing difficulty as a symptom initially. She was questioned about dysphagia after the radiological evaluation which revealed DISH. Dysphagia is a common presenting complaint in otolaryngology practice (12). Hoarseness, stridor, aspiration pneumonia, sleep apnoea, atlantoaxial subluxation or pseudoarthrosis and thoracic outlet syndrome can also be caused by cervical involvement (8,9,13). The risk of vertebral fractures increases with the number of ankylosed vertebra (14).

The most commonly used classification criteria for DISH were defined by Resnick and Niwayama and required flowing anterolateral ossifications of at least four contiguous thoracic vertebral segments, preservation of the intervertebral disk spaces and absence of apophyseal joint degeneration or sacro-



Figure 3— a. Sagittal reconstruction of CT scan demonstrates hyperostosis and anterior cervical fusion from C2–C7 (arrows) **b.** axial CT image shows the pharyngeal compression due to right anterolateral osteophyte (arrow).

iliac inflammatory changes (15). Compared with the thoracic or lumbar spine, alterations in the cervical spine are less frequently encountered (15). Radiographic evaluation of the spine, particularly the thoracic spine, should be obtained to confirm diagnosis among patients with cervical involvement and suspected DISH. Radiographically, the diagnostic criteria of DISH are flowing calcifications and ossifications along the anterolateral aspect of at least four contiguous vertebral bodies. Disk height is usually preserved and excessive disk disease is absent in the involved areas (15). Thoracic and lumbar spine radiography is usually sufficient for the diagnosis of DISH. CT imaging may be performed to evaluate other complications such as pressure symptoms on adjacent organs or fractures.

Conservative treatment of DISH includes physical therapy, exercises, analgesics, NSAIDs and muscle relaxants (5). Occasionally, dysphagia due to esophageal compression may require surgery. When the osteophytes are remarkably large surgery may be needed. (16). For patients who fail conservative treatment, surgical decompression through osteophyctomy is also an effective option (17). Our patient had mild dysphagia which had not yet affected her nutritional status, but she was informed about the possible worsening of symptoms.

This report serves to increase awareness of DISH especially in patients with metabolic syndrome having spinal pain, diminished ROM and stiffness. Besides these well known symptoms of DISH, dysphagia may be the presenting symptom. Clinicians should be aware of dysphagia as a symptom of DISH. They should question and counsel patients about difficulty swallowing and follow them up for possible rare complications such as dyspnoea and undernutrition which require surgical intervention.

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