



Diffuse Idiopathic Skeletal Hyperostosis: Neurosurgical Cause of Dysphagia

Diffüz İdiopatik İskelet Hiperostozisi: Disfajinin Nöroşirurjikal Sebebi

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ABSTRACT

Diffuse idiopathic skeletal hyperostosis is a chronic enthesopathy which causes excessive formation of osteophytes along the ventral spine in the absence of degenerative, traumatic and inflammatory pathologies of spinal ligaments or paravertebral muscles. Dysphagia is the most common symptom of this disease which may need surgical management. In this article, we report three cases with diffuse idiopathic skeletal hyperostosis treated in our department. The clinical and diagnostic imaging characteristics along with treatments performed and outcomes of these three patients were described. Diffuse idiopathic skeletal hyperostosis is an idiopathic rheumatological disorder which may need surgical treatments in severe forms of the disease.

Key words: Diffuse idiopathic skeletal hyperostosis, Forestier's disease, dysphagia, cervical osteophytes.

ÖZET

Diffüz idiyopatik iskelet hiperostozisi, spinal ligamanlar veya paravertebral kasların dejeneratif, travmatik ve inflamatuvar patolojileri olmaksızın ventral omurga boyunca osteofitlerin aşırı oluşumuna neden olan kronik bir entesopatidir. Disfaji bu hastalığın en sık görülen ve cerrahi tedavi gerektirebilen semptomudur. Bu makalede, kliniğimizde diffüz idiyopatik iskelet hiperostozisi nedeniyle tedavi uygulanan üç vaka sunulmaktadır. Bu üç hastanın klinik ve tanısal görüntüleme özellikleri ile uygulanan tedaviler ve sonuçları değerlendirildi. Diffüz idiyopatik iskelet hiperostozisi, hastalığın ağır formlarında cerrahi tedavi gerektirebilen idiyopatik romatolojik bir hastalıktır.

Anahtar kelimeler: Diffüz idiyopatik iskelet hiperostozisi, Forestier hastalığı, disfaji, servikal osteofit

INTRODUCTION

Diffuse idiopathic skeletal hyperostosis (DISH), also known as Forestier's disease or ankylosing hyperostosis, is characterised by excessive ligamentous calcification and ossification at spinal and extraspinal locations. DISH was first described in 1950 by Forestier and Rotes-Querolas as calcification of anterolateral

perivertebral ligaments¹. In 1975 Resnick named the condition as 'diffuse idiopathic skeletal hyperostosis', defining it as spinal and peripheral enthesopathy². Although most of DISH cases are asymptomatic, dysphagia, dysphonia, cervical subaxial pain, stiffness and decreasing range of motion of the cervical spine are the signs of the symptomatic patients³⁻⁵. Dysphagia is the most

common symptom due to esophageal compression by anterior osteophytes at the C4-5 level⁶.

CASES

Between April 2009 and September 2014, 683 patients who had been hospitalized with cervical spinal pathologies were included in the study. The required data were gathered using medical records of the patients and comprised primary clinical presentations, demographic data, radiological findings, preoperative and postoperative conditions of the patients and treatments applied for their pathologies. The patients who had clinical symptoms such as cervical subaxial pain and dysphagia were diagnosed with the scan techniques including lateral plain cervical radiograph, cervical spinal computed tomography and cervical spinal magnetic resonance imaging techniques. Diffuse idiopathic skeletal hyperostosis was diagnosed in three patients and all three of them had no other diseases accompanying their cervical spinal pathologies. Two of them underwent surgical interventions and one of them was applied conservative treatments.

The first case who was a 47 year-old woman, presented with a 7 month history of gait disorder, dysphagia and progressive weakness of all 4 limbs. Neurological examination revealed quadriparesis and upper motor neuron symptoms such as exaggerated deep tendon reflexes, positive Babinski sign and spasticity. Imaging techniques were performed and co-existence of basilar invagination and diffuse idiopathic skeletal hyperostosis was determined (Figure 1a). Anterior osteophylectomy and posterior cervical decompression and fusion was performed as surgical intervention. Postoperative cervical spinal computed tomography scan (Figure 1b) was performed which showed that the surgery was successful and after surgery in 2 weeks time, her symptoms decreased prominently.

The second patient was a 60 year-old man who was admitted to our department with a 4

month history of dysphagia and neck pain. Before these, he had no similar complaints or any medical history. On his neurological examination, he did not display any neurological deficits. Lateral plain X-ray, cervical spine computed tomography and cervical spinal magnetic resonance imaging (Figure 2) techniques were performed. These scans showed large bridging osteophytes on the anterior aspect of all cervical and upper thoracic vertebrae and his diagnosis was diffuse idiopathic skeletal hyperostosis. After thorough discussion with the patient, he did not approve the surgical intervention and conservative treatments were performed.

The third patient was a 39 year-old woman who presented with a 1 year history of dysphagia. Her neurological examination was intact and her only symptom was progressive dysphagia. Lateral plain cervical X-ray and cervical spinal computed tomography techniques revealed large symptomatic osteophytes at the C1-C2 levels (Figure 3a and 3b) and contiguous bridging osteophytes on the anterolateral aspect of the lower cervical vertebrae (Figure 4). Peri-oral-transpharyngeal approach and anterior osteophylectomy was performed to the C1-C2 levels. Postoperatively in 6 weeks, full recovery was achieved and the patient had no similar symptoms.



Figure 1a. Sagittal cervical computed tomography scan of case 1 showing co-existence of basilar invagination and diffuse idiopathic skeletal hyperostosis



Figure 1b. Postoperative sagittal cervical computed tomography scan of case 1 demonstrating complete removal of the osteophytes



Figure 2b. Sagittal cervical computed tomography scan of case 2



Figure 2a. Lateral plain cervical radiograph of case 2



Figure 2c. Sagittal cervical magnetic resonance imaging scan of case 2 demonstrating large bridging osteophytes on the anterior aspect of all cervical and upper thoracic vertebrae



Figure 3a. Lateral plain cervical radiograph of case 3



Figure 3b. Sagittal cervical computed tomography scan of case 3 revealing large symptomatic osteophytes at the C1-C2 levels



Figure 4. Sagittal cervical computed tomography scan of case 3 showing contiguous bridging osteophytes on the anterolateral aspect of the lower cervical vertebrae

DISCUSSION

Osteophytic formations at the vertebral column are seen commonly, however dysphagia due to these formations is considered as an uncommon entity. In the literature, DISH is mentioned as one of the most common cause of anterior cervical hyperosteozytosis-induced dysphagia requiring surgical management^{5,7-9}. This disease is characterized by ossification of the anterior ligament and the production of flowing osteophytes involving especially the right side of the spine. The osteophytes are generally located in the thoracic, lumbar and cervical vertebrae (97%, 90%, 78%, respectively). The entire vertebral column is affected in 70% of all cases⁵. DISH can also spread to extraspinal enthesal regions in the peripheral joints such as the peripatellar ligaments, the Achilles tendon insertion, the plantar fascia and the olecranon⁵. DISH occurs mostly after the fifth decade of life and male to female ratio was found 2/1^{1,10}. It has been found in 12% to 28% of autopsy specimens^{11,12}.

The aetiology of this disease has not been defined but there are associations with the diseases like diabetes mellitus, hypercholesterolaemia, hyperuricaemia, hypoparathyroidism, acromegaly and ankylosing spondylitis¹³. In fact, the oldest described factor for ankylosing hyperostosis is obesity^{1,2,12}. In a case control study involving 131 subjects, the body mass index was significantly higher in patients¹⁴. Treatment of acne with vitamin derived retinoids was also noticed to be associated with similar manifestations in some patients especially with Etretinate which is no longer available for clinical use¹⁵. Other retinoids, like Acitretin may also cause extraspinal hyperostosis¹⁶. Increased levels of insulin and insulin-like growth factor 1 and HLA-B8 positiveness were also determined in the patients with ankylosing hyperostosis^{17,18}.

The most common symptoms of DISH are pain, stiffness, dysphagia and decreased range of motion. Dysphagia related to ankylosing hyperostosis affecting the cervical spine has a

reported prevalence of 28% and only 10% of them required surgical resection¹⁹. Dysphagia related to DISH may be due to several mechanisms: 1) mechanical obstruction by an enlarged osteophyte compressing or deviating the esophageal lumen; 2) growth of a relatively small osteophyte at sites where the esophagus is anatomically anchored (level of the cricoid cartilage and diaphragm); 3) inflammation of the soft tissue due to repetitive motion across the osteophyte promoting esophageal obstruction; 4) pain and muscle spasm from an enlarged osteophyte causing esophageal narrowing; 5) osteophyte-induced restriction in mobility of the epiglottis as well as limitation in movement of the larynx; 6) indentation of pharynx wall by an osteophyte resulting in retention of food in piriform sinus^{3,7,9,20}.

Ankylosing hyperostosis can also cause neurological deficits by the following ways especially affecting the cervical spine: 1) reduced flexibility of the spine; 2) spinal canal narrowing secondary to ossification of anterior and posterior longitudinal ligaments; 3) atlantoaxial subluxation of the cervical spine²¹. In the autopsy series performed by Boache et al, the incidence of the involvement of the cervical spine was found 75% of the patients diabetes mellitus and the increasing age may be the mainly risk factors for traumatic spinal cord injuries in the patients with DISH^{11,22}.

The diagnosis of DISH is based mainly on the following radiologic features: 1) presence of flowing new bone formation along the anterolateral aspect of at least four contiguous vertebral bodies; 2) absence of degenerative disc disease and relative preservation of intervertebral disc heights; 3) absence of inflammatory changes in facet or sacroiliac joints^{4,12,22}.

Lateral plain X-ray imaging is the first step in the diagnosis of DISH. After the demonstration of osteophytes in the lateral plain X-rays, computed tomography of the spine should be performed. These imaging techniques may be useful to better delineate the bony anatomy. After these steps, esophagram and diagnostic endoscopy should be

performed for the differential diagnosis of esophageal pathologies such as strictures, infections or malignancies. Magnetic resonance imaging of the spine is also a useful technique to show the involvement of the spine in the patients who have neurological deficits related to DISH.

Treatment managements of DISH are divided between conservative and surgical modalities. Patients with milder symptoms should be treated with conservative managements including modification of diet, swallow therapies, prescription of non-steroidal anti-inflammatory drugs, corticosteroids, muscle relaxants and anti-reflux drugs. When conservative treatments fail, surgical modalities should be performed. Treatment resistant pain and excessive loss of weight due to progressive dysphagia are the main indications for surgical resection of the osteophytes. Peri-oral-transpharyngeal route for C1 and C2 vertebrae or anterolateral servical approach for C3 to C7 vertebrae is preferred^{3,9,23}. Although the complication rate appears low, specific risks associated with these approaches include vocal cord paralysis, Horner syndrome, cervical instability and esophageal or tracheal perforation caused by thinning of the walls from chronic osteophytic compression^{3,9,24,25}. Re-accumulation of osteophytes after the surgery is another problem. In the literature; it has been reported that recurrence of these ossifications tend to grow at mobile vertebral segments rather than immobile segments. As a result; resection of osteophytes combined with cervical fusion is recommended to prevent recurrence, especially in younger patients^{26,27}.

Diffuse idiopathic skeletal hyperostosis (DISH) is an idiopathic rheumatological disorder which may need surgical treatments in severe forms of the disease. Co-existence of dysphagia with the pain at the cervical and thoracic regions should remind us this pathology, especially in older age groups. Patients with milder symptoms should be treated with conservative modalities including diet modifications and medications. When

conservative treatments fail, surgical modalities should be performed.

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