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Case Report

Progressive dyspnea and dysphagia due to diffuse idiopathic skeletal hyperostosis: a case report

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Abstract: Objective: The purpose of this study is to describe an uncommon patient with diffuse idiopathic skeletal hyperostosis (DISH) and syringomyelia (SM) resulting in obvious dyspnea and dysphagia, whose symptoms markedly improved after the surgical intervention of anterior cervical approach osteophyte resection from C2-C7. Methods: A 58-year-old Chinese male with 32-years history of SM was admitted to our hospital complaining of mild dyspnea and dysphagia for 5 years. The dysphagia gradually deteriorated in the past 1 year, and he had difficulty in swallowing solid food. DISH, SM, Chiari malformation (CM) were diagnosed by personal history, neurological examination and X-ray, CT and MRI. Hyperintense signal in the cervical and thoracic spinal cord on T2 weighted sequence was observed. Results: Anterior cervical approach osteophyte resection from C2-C7 was performed. The prevertebral fascia was separated and the trachea and esophagus were retracted on the left side and the carotid sheath on the right. The C2-7 level was exposed and identified by lateral scope examination. Large anterior osteophytes were removed with Kerrison rongeurs and high-speed air drill until the anterior spinal surface from C2 to C7 was flat on palpation and on lateral scope control. The symptoms of dyspnea and dysphagia markedly improved after the surgical intervention. During the first postoperative month, the patient was able to continue his daily life. Conclusion: Dyspnea and dysphagia caused by cervical osteophytes may be treated conservatively or surgically. Early surgical treatment provides satisfactory improvement in the patient when conservative treatment fails.

Keywords: Diffuse idiopathic skeletal hyperostosis, syringomyelia, dyspnea, dysphagia, surgery

Introduction

Diffuse idiopathic skeletal hyperostosis (DISH), also known as Forestier's disease [1], is a non-inflammatory degenerative disease of unclear cause, characterized by calcification and ossification of the soft tissues, mainly spinal ligaments and entheses [2-4]. It occurs primarily in the elderly and predominantly in men [5]. It involves the anterolateral aspect of the spine, but also several extra-spinal sites, such as shoulder, elbow, hip, knee and heel [6]. Other characteristic findings are extra-skeletal ossifications or calcifications [7-9] that could cause enthesopathy and respiratory, gastrointestinal, and neurological symptoms due to compression of surrounding tissues.

The underlying pathogenetic mechanism of DISH is poorly understood, but genetic, anatomic, environmental, metabolic, endocrinologic and toxic factors possibly contribute to the development of DISH [10-13]. Particularly, it has been associated with a variety of metabolic disorders, such as obesity, hypertension, Type 2 diabetes [14], ossification of cervical posterior longitudinal ligament (OPLL) [15], non small cell lung cancer (NSCLC) [16], and so on.

Nevertheless, to the best of our knowledge, no association with syringomyelia (SM) has been reported yet so far. In the present study, we report a unique patient with diffuse idiopathic skeletal hyperostosis (DISH) and syringomyelia (SM) resulting in obvious dyspnea and dysphagia, whose symptoms markedly improved after the surgical intervention of anterior cervical approach osteophyte resection from C2-C7, which is rarely reported.

Case report

An appropriate written informed consent was obtained from the patient reported in this study.

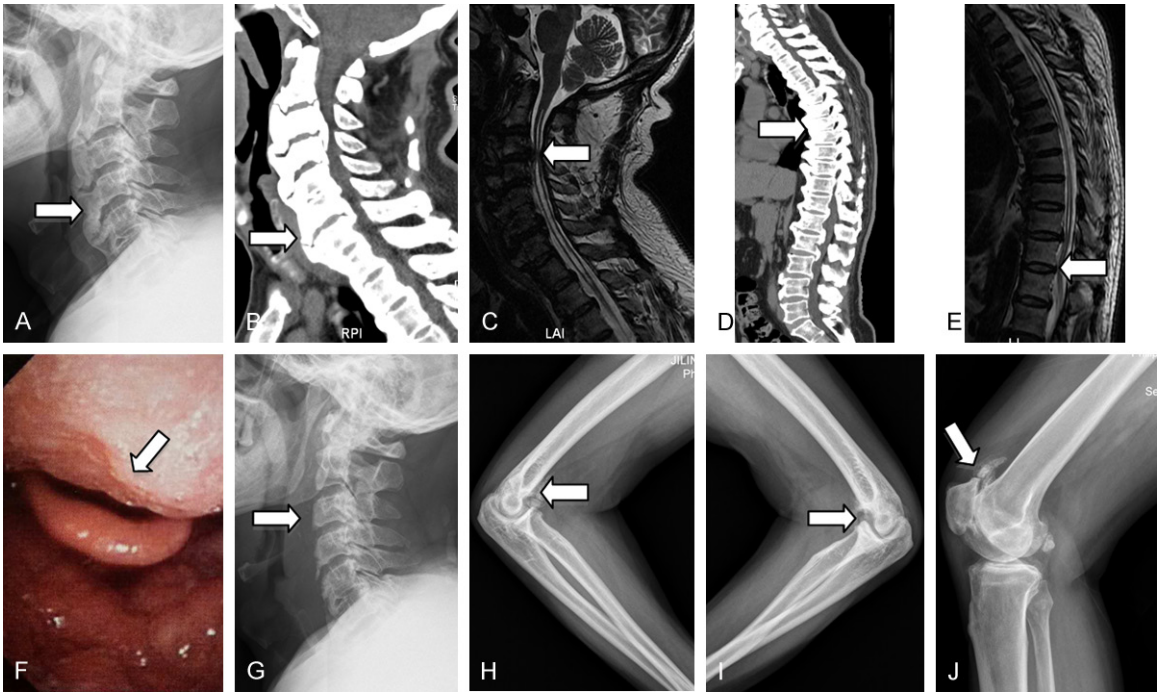

Figure 1. Exuberant bony growth in the water samples along the anterior longitudinal ligament widely (C2-C7) in preoperative X-ray (A), CT (B) and MRI (C), which showed cervical canal stenosis, cerebellar tonsillar herniation and syringomyelia (C2-T2). CT and MRI of thoracic spine indicated massive anterior osteophyte at T1-12 without destruction of the intervertebral disc space (D, E). Pharyngoscopy showed the protrusion of the posterior wall of the pharynx without other abnormalities (F). Postoperative X-ray indicated massive anterior cervical osteophytes (C2-7) had been resected (G). X-ray of other regions showed osteoproliferation of bilateral elbows and right knee (H-J).

A 58-year-old Chinese married non-smoking male was admitted to our hospital complaining of mild dyspnea and dysphagia for 5 years. The dysphagia gradually deteriorated in the past 1 year, and he had difficulty in swallowing solid food. 32 years ago, he was diagnosed with "SM, Chiari malformation (CM)" due to minor hyperpathia with associated severe neck pain. And then he has been using Adenosine Disodiu over the years. The patient had had type 2 diabetes for 10 years and had been admitted to the local hospital on three previous occasions for control of his disease. Physical examination revealed with upper extremity weakness predominating. He complained of being poor in fine movements. Significant sensory deficits were present from his upper torso to the plantar regions of his feet bilaterally, and Morvan sign was positive. Bilateral Hoffman sign and Babinskin sign were present, in addition to brisk lower and upper extremity reflexes, though he could still walk without a cane. Laboratory examination: Fasting plasma glucose (FPG): 8.34 mml/L (3.9-6.1), White blood cell (WBC): 11.9*10^9/L (3.5-9.5), C-reactive protein (CRP): 0.71 mg/L (0.00-7.44), Erythrocyte sedimentation rate (ESR): 4.0 mm/h (<16), Human leukocyte antigen B27 (HLA B27) (-). Initial X-ray and CT of the cervical spine showed the presence of osteophytes and exuberant bony growth along the anterior aspect of the vertebral column widely (C2-7) (**Figure 1A**, **1B**). Preoperative MRI of the cervical spine showed canal stenosis, cerebellar tonsillar herniation and syringomyelia (C2-T2) (**Figure 1C**). CT and MRI of thoracic spine showed massive anterior osteophyte at T1-12 without destruction of the intervertebral disc space and syringomyelia (T4-10) (**Figure 1D**, **1E**). Pharyngoscopy showed the protrusion of the posterior wall of pharynx without other abnormalities, which excluded pharyngeal neoplasm (**Figure 1F**). Electromyography (EMG) indicated cervical and lumbosacral neurogenic damage, and peripheral neuropathy.

Due that 1-year medications failed and his dysphagia caused weight loss, we considered surgical intervention. Under general endotracheal anesthesia, we firstly performed anterior

approach osteophyte resection from C2-C7. The prevertebral fascia was separated and the trachea and esophagus were retracted on the left side and the carotid sheath on the right. The C2-7 level was exposed and identified by lateral scope examination. Large anterior osteophytes were removed with Kerrison rongeurs and high-speed air drill until the anterior spinal surface from C2 to C7 was flat on palpation and on lateral scope control. Postoperative X-ray showed massive anterior cervical osteophytes (C2-7) had been resected (**Figure 1G**). Moreover, X-ray of other regions in follow-up showed osteoproliferation of bilateral elbows and right knee (**Figure 1H-J**). The patient was mobilized using a semi-rigid collar after surgery. Although neurological improvement was minor, the dysphagia and dyspnea considerably improved without any complications. During the first postoperative month, the patient was able to continue his daily life without any significant problem but reported difficulty in swallowing big bites. His cervical CT showed no re-growth of the osteophytes. He will be performed posterior open-door expansive laminoplasty in a second phase.

Discussion

Diffuse idiopathic skeletal hyperostosis (DISH) is considered an underdiagnosed and mostly asymptomatic nonprimary osteoarthritis [17]. The exact epidemiology of DISH has not yet been described in the medical literature [3], certainly because of the lack of consensus concerning an exact definition of the disease. Likewise, the etiology of DISH remains unknown and the validated diagnostic criteria are absent.

Usually, patients are asymptomatic. Some patients could present local symptoms. Common symptoms of this disease are cervical pain and stiffness; however, it can manifest itself by respiratory compromise due to upper respiratory airway compression and dysphagia due to mechanical obstruction. Rarely, large projecting anterior osteophytes result in esophageal impingement and distortion leading to dyspnea and dysphagia [17-22], which have also been described [23] in the cervical spine, exactly just as this case. The patient in the case was admitted to our hospital mainly complaining of progressive dyspnea and dysphagia for 5 years. The possible mechanisms of dysphagia include mechanical compression to esophagus, pharyngo-esophageal irritation which induces peri-esophageal edema, inflammation, and a local inflammatory reaction resulting in cricopharyngeal spasm and esophageal denervation. However, DISH, as a cause of dysphagia and/or airway obstruction may be an increasing and underappreciated phenomenon, which should be included in the differential diagnosis of dysphagia and airway obstruction.

We present a unique case of a patient with syringomyelia and diffuse idiopathic skeletal hyperostosis. The diagnosis of DISH is based mainly on radiological features [24-27]. The most classical classification criteria were defined by Re snick and Niwayama [28]. These criteria include the involvement of at least four contiguous vertebrae of the thoracic spine, preservation of the intervertebral disc space, and the absence of apophyseal joints and sacroiliac inflammatory changes. Chiari malformations are congenital deformities involving cerebellar tonsillar herniation downward through the foramen magnum [29]. Structurally, greater than 5 mm of tonsillar descent in adults and more than 6 mm in children is consistent with Chiari malformations. Actually, more than 7 mm in the present case. The case we present fulfills all these criteria. Particularly, SM of the patient mainly related to a congenital abnormality - Chiari malformation, while DISH may worsen the severity of SM.

In the differential diagnosis of DISH: ankylosing spondylitis (AS), osteoarthritis and spondylosis deformans should be considered. Of course, the most meaningful differential diagnosis of DISH requires distinguishing it from AS. An AS is a relatively rare disease with a prevalence of 0.05-1.4%, which is a chronic inflammatory rheumatic disease and tends to affect relatively young white males [30]. Patients with AS are usually symptomatic, but suffer from a myriad of associated conditions such as iritis, uveitis, or ulcerative colitis. Sacroiliac and apophyseal fusion or sclerosis, the earliest symptoms of which are back pain and stiffness, result from inflammation of the sacroiliac joint [31]. This inflammation can gradually spread to the joints between the vertebrae, causing a condition called spondylitis, as well as to other joints including the shoulders, hips, and knees. Whereas the DISH has a prevalence ranging from 2.9% to 25%, which is a systemic condition and not just the result of local mechanical

factors present in each of the involved areas of the skeleton. Moreover, esophageal malignancies should also be considered when dysphagia couldn't be explained by small anterior osteophytes and diagnostic endoscopy should be performed.

The initial treatment of patients with symptomatic DISH should be conservative therapy, which mainly includes anti-inflammatory medication, muscle relaxants, steroids, and so on [32]. Surgical decompression through osteophyte resection is effective for patients who fail conservative treatment, the aim of which is to provide satisfactory decompression of esophagus or airways. Specially, dysphagia or dyspnea, which are severe or resistant to conservative therapy, should be considered adopting surgical treatment. Resection of osteophytes alone by the anterior approach is commonly performed, and many reports have shown that dysphagia resolved well after surgery [18, 33-35], as shown in our case. We treated the patient surgically due to the large size of the cervical osteophytes. Removal of only large cervical osteophytes, rather than multilevel anterior resection of DISH, resulted in rapid and progressive resolution of symptoms within 2 months. The patient in our hospital was mobilized using a semi-rigid collar after surgery. The dysphagia and dyspnea considerably improved without any complications.

However, some reports have described patients with recurrence DISH associated with dysphagia after surgery. Miyamoto et al [34] reported that the postsurgical recurrence of OALL-caused dysphagia in patients with DISH was at an average rate of approximately 1 mm/year and that the incidence of recurrence in segments with mobility was significantly than that in segments without mobility. Re-accumulation of osteophytes a few years after surgical treatment has been reported in the literature and these ossifications tend to grow at mobile vertebral segments rather than immobile vertebral segments as observed during the long-term follow-up of patients [36]. As a result, the fixation of operated vertebral segments via anterior cervical fusion is strongly recommended to prevent recurrence, especially in younger patients [37].

In addition, DISH could cause the ankylosis of spinal column, which can make the spine prone to fracture after even minor trauma. Moreover, such unstable fractures can lead to secondary neurologic deterioration in initially asymptomatic patients [38-40]. In DISH-related spine fractures, delayed diagnosis is reported to be particularly common in nondisplaced fractures after trivial injuries and was associated with neurologic worsening. Due to the difficulty in radiographically visualizing spines affected by DISH, screening of the entire spinal column with an advanced neuroimaging modality (MRI or CT) has been recommended especially for patients with persistent neck or back pain. The prevalence of DISH may be rapidly increasing in modern societies additionally. Thus, awareness of this condition should be increased among neurosurgeons when assessing trauma patients and treating spinal injuries [30, 41]. Further, the prevention of possible complications such as spinal fractures and heterotopic ossification following orthopedic surgical procedures deserves further attention.

In conclusion, DISH is an idiopathic disorder affecting particularly the anterior longitudinal ligament in older age groups. The case illustrates that clinicians including spine surgeons, neurosurgeons, rheumatologists, should be aware of this rare clinical manifestation as the presenting feature of DISH in the cervical spine. Early surgical treatment provides effective and satisfactory improvement in these patients when conservative treatment fails.

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Disclosure of conflict of interest

None.

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