DISHphagia – A RIDDLE UNWRAPPED A CLINICAL CASE WITH LITERATURE REVIEW

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Abstract. *Diffuse idiopathic skeletal hyperostosis (DISH) is a common but underdiagnosed systemic skeletal disease. It is characterized by calcifications affecting mainly the spinal anterior longitudinal ligament. In the majority of cases, the patients are asymptomatic, but cervical osteophytes can sometimes cause hoarseness, dysphagia (DISHphagia) and even dyspnea. Case description:* A 61-year-old man was admitted to our department with complaints of difficulty in swallowing and weight loss. Dysphagia had been increasing gradually for nine months. Barium swallow esophagram revealed asymmetric swallowing with expansion above the upper esophageal sphincter without other abnormalities. The extension was confirmed by esophago-gastro-duodenoscopy (EGD). Furthermore, CT scan of the thorax clearly demonstrated degenerative changes of the cervical and thoracic region, extensive ossification of the anterior longitudinal ligament, and osteophytes from C2-C7 with a forward displacement of the esophagus by 14 mm. The so-called "wax dripping down the candle" phenomenon was as well observed. **Conclusion:** DISH is a systematic, musculo-skeletal disease of older adults with unknown etiology. Dysphagia is the most common symptom of the disease and might be caused by osteophytes of the cervical region. We presented a case of DISH with a rare localization of the osteophytes in the cervical region C2-C7. Due to the increasing incidence of the Forestier's syndrome and its associated "DISHphagia", the gastroenterologist should increase the awareness of this underestimated disease and improve the diagnostic approach.

Key words: Diffuse idiopathic skeletal hyperostosis, DISH, dysphagia, DISHphagia, osteophytes

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RECEIVED: 21 September 2020, **ACCEPTED:** 20 November 2020

INTRODUCTION

Dysphagia is a subjective feeling of impairment in the passage of solid foods or liquids from the oropharynx to the stomach [1, 2]. A

variety of benign and malignant diseases, as well as medications and rheumatological conditions, can result in dysphagia [1]. The difficulty in swallowing is classified as acute or chronic [1]. However, without treatment, it can lead to significantly impaired

quality of life [3, 4]. A rare cause of dysphagia is the Forestier's syndrome, known as Diffuse idiopathic skeletal hyperostosis (DISH) [5-9].

This disorder was first described in 1950 by Forestier and his student Rotés-Querol and was renamed as DISH by Resnick in 1975 [10, 11]. This condition is a systemic non-inflammatory disease, characterized by calcification and ossification of the spinal ligaments and it may affect all extraspinal locations, in which tendons and ligaments are attached to the bone [12, 13].

The exact prevalence of DISH remains unknown, but studies suggest that males are more affected than females and that the incidence correlates positively with the age of the patients [11, 14, 15]. The etiology of the Forestier's syndrome is still unraveled as well. However, Forestier's syndrome is linked to different endocrine and metabolic factors [6, 16, 17]. As dysphagia is the most common symptom in DISH, gastroenterologists should be familiar with this syndrome [18].

Herein we report a rare clinical case of a patient with DISH-associated dysphagia. In an attempt to better understand this underestimated syndrome, we review and discuss the available literature.

CLINICAL CASE PRESENTATION

A 61-year-old man was admitted to our department with complaints of difficulty in swallowing, a feeling of foreign body in the esophagus and weight loss. Dysphagia had been gradually increasing for nine months. The patient was diagnosed in another hospital with GERD grade A, cicatrized ulcer of the stomach and gastritis following esophagogastroduodenoscopy (EGD). Although he was treated with PPI and prokinetics, his symptoms remained. Due to type 2 diabetes mellitus, the patient received treatment with Metformin. The detailed physical examination revealed no pathological findings. The laboratory parameters, including HLA-B27 antigen, were within the reference range. Fatty liver disease was found by abdominal ultrasound. Barium swallow esophagram revealed asymmetric swallowing with expansion above the upper esophageal sphincter as shown in (Fig. 1), with no other abnormalities. During EGD, we confirmed the extension and observed a hiatal hernia with gastroesophageal reflux (GERD) grade IV. Furthermore, computed tomography of the thorax clearly demonstrated degenerative changes in the cervical and thoracic region, with extensive ossification of the anterior longitudinal ligament, and osteophytes from C2-C7 with a forward displacement of the esophagus by 14 mm (Fig. 2). The so-called "wax dripping down the candle" phenomenon was observed as well (Fig. 3). These findings confirmed the presence diffuse id-

opathic skeletal hyperostosis or so-called Forestier's disease. The patient was thereafter consulted with a neurosurgeon and an otorhinolaryngologist and referred to a rheumatologist for further treatment.

Fig. 1. Asymmetric swallowing (mainly at the left side)

Fig. 2. Osteophytes and extensive ossification of the anterior longitudinal ligament. Extensive flowing ossification along the anterior aspects of the cervical vertebrae merging with anterior bridging osteophytes that lead to oropharynx impingement and displacement in the anterior plane

Fig. 3. The "Wax dripping down the candle" phenomenon is shown. Extended ossification along the anterior aspects of the thoracic vertebrae

DISCUSSION

Diffuse idiopathic skeletal hyperostosis (DISH) is a common but underdiagnosed systemic musculo-skeletal disease, which is characterized by calcification and ossification of the spinal ligaments and affecting generally the peripheral skeleton [8, 12, 13, 19]. The most affected part of the spine is the thoracic region, followed by the lumbar and cervical ones [20]. In the majority of cases, the patients are asymptomatic and usually the diagnosis is made based on abnormal findings from radiologic imaging performed for other reasons [5, 19, 21-23]. To make the diagnosis of DISH, the patient has to fulfill three radiologic criteria, originally proposed by Resnick and Niwayama [24]. A very suggestive image of DISH is the so-called "wax dripping down the candle" phenomenon, which was also observed in our patient [20, 25]. In the cervical region, the most frequently affected vertebrae are in the C4-C7 range, which makes the upper involvement of the cervical vertebrae very rare [26, 27].

In our clinical case, the involved segment of the skeleton was C2-7, which is very rarely reported in the literature. The cervical osteophytes in DISH resulted in dysphagia, dysphonia, hoarseness and even dyspnea [5, 20]. Most of the symptomatic patients have osteophytes larger than 10 mm [19, 28, 29].

Forestier's syndrome is observed more frequently in males [11, 14]. The etiology of DISH is still unclear [16]. The disease is often associated with obesity, diabetes mellitus, long-term use of retinoids and other vitamin A derivatives [17, 30, 31]. Higher insulin and growth hormone levels have been registered in patients with DISH, which might explain the osteoblast growth [32, 33]. In our case, the patient had both type 2 diabetes and metabolic syndrome.

H.P. Mosher was the first who in 1926 described dysphagia secondary to cervical osteophytes [34]. Interestingly, about 10% of the patients with difficulty in swallowing have cervical osteophytes [35]. Around 6% of the patients with DISH will possibly develop difficulty in swallowing during their lifetime [36]. The anterior osteophytes in the cervical spine in DISH can compress the esophagus, which may result in dysphagia [5, 8, 9, 37]. Some authors use the colloquial term DISHphagia [38, 39]. Other possible causes of dysphagia due to the large osteophytes are the compression of the esophageal's lumen from the osteophytes, cricopharyngeal muscle spasms and neuropathy from recurrent laryngeal nerve injury [28]. Patients with DISH more often experience difficulty in swallowing solid food than liquids [18, 36].

Due to its asymptomatic clinical course and difficult diagnosis, the disease is underestimated and underdiagnosed [5, 21]. The diagnosis may be delayed for approximately 10 years [40]. Computed tomography is the "gold standard" for "the diagnosis of Forestier's syndrome, while X-ray imaging with Barium swallowing is a useful method to determine the localization and the degree of esophageal compression [5, 21, 41]. On the other hand, endoscopy is indicated in each patient with dysphagia [2]. The EGD ought to be performed minimally invasive due to the risk of trauma and associated esophageal carcinoma [42].

The treatment of DISH depends on the symptoms and their severity [16]. In the early stages of the DISH, the treatment is nonsurgical and the patients are prescribed dietary restrictions, antireflux medication, anti-inflammatory drugs, steroids and muscle relaxants [7, 9, 21]. Most of the patients with DISH respond well to conservative treatment. However, in those with no improvement, a surgical procedure is required [6, 7]. The weight loss is a well-known indication for surgery in Forestier's syndrome [21]. The resection of the osteophyte is an approved reliable and safe method for the treatment of patients with DISHphagia. Nevertheless dysphagia is a well-established post-operative complication [5, 43-46]. The relief of the complaints starts immediately in the postoperative period or within the first 3 to 6 months [6, 7, 9]. A postoperative reappearance of the osteophytes has already been described with an approximate growth rate of 1 mm/year [28].

CONCLUSION

DISH is a systematic, non-inflammatory, musculo-skeletal disease in older adults and of unknown etiology. Dysphagia is the most common symptom of the disease and might be caused by osteophytes in the cervical region. We presented a rare case of DISH with localization of the osteophytes in the cervical region (C2-C7). Due to the increasing incidence of the Forestier's syndrome and its associated "DISHphagia" the gastroenterologist should always be aware of this underestimated disease and correctly guide the diagnostic approach.

Disclosure summary: The authors have nothing to disclose.

Acknowledgements: This work is supported by the Bulgarian Ministry of Education and Science under the National Program for Research "Young Scientists and Post-doctoral Students"

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