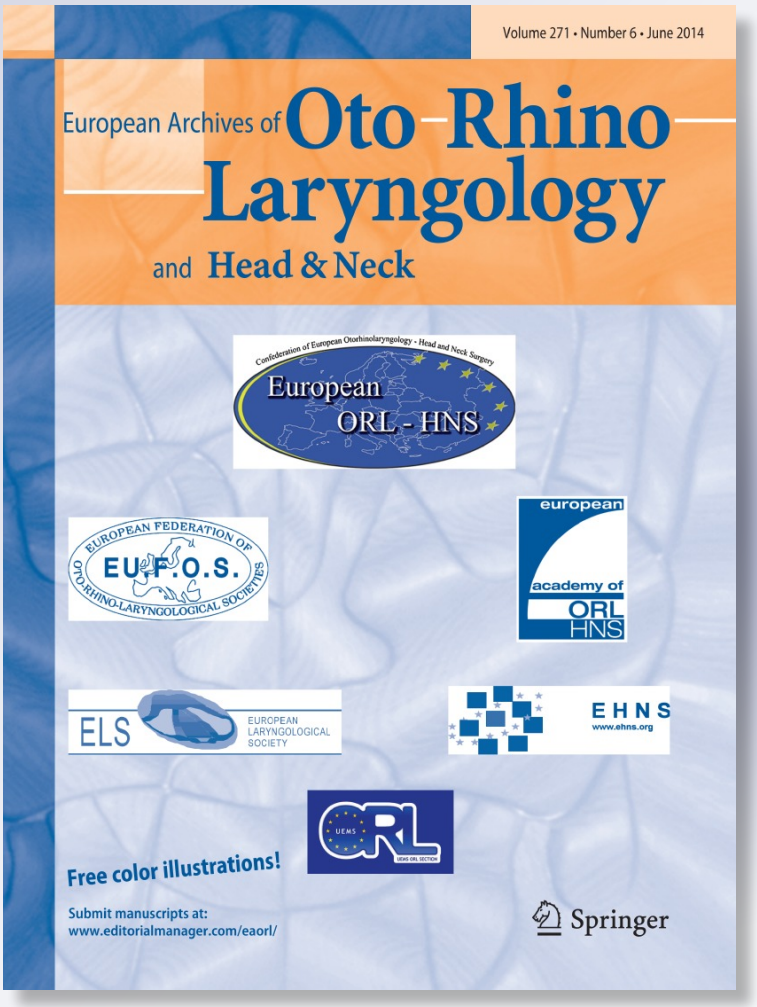
Otolaryngologic manifestations of diffuse idiopathic skeletal hyperostosis

Mosaad Abdel-Aziz, Noha A. Azab, Mohammed Rashed & Ahmed Talaat

European Archives of Oto-Rhino-Laryngology
and Head & Neck

ISSN 0937-4477
Volume 271
Number 6

Eur Arch Otorhinolaryngol (2014)
271:1785-1790
DOI 10.1007/s00405-013-2827-z



Springer

Eur Arch Otorhinolaryngol (2014) 271:1785–1790
DOI 10.1007/s00405-013-2827-z

MISCELLANEOUS

Otolaryngologic manifestations of diffuse idiopathic skeletal hyperostosis

Mosaad Abdel-Aziz · Noha A. Azab · Mohammed Rashed · Ahmed Talaat

Received: 17 October 2013 / Accepted: 13 November 2013 / Published online: 22 November 2013

Abstract Diffuse idiopathic skeletal hyperostosis (DISH) is characterized by formation of large cervical osteophytes that may compress the posterior wall of the aerodigestive tract. It is a rare cause of dysphagia in the elderly. The aim of this study was to investigate the various otolaryngologic manifestations of DISH. Eleven elderly patients with DISH were included in the study. All patients presented with dysphagia that was graded on the swallowing screening tool (EAT-10), and the diagnosis of DISH was based on computed tomographic criteria. The patients were subjected to otolaryngologic examination and flexible laryngoscopy. Polysomnography was used for patients with excessive daytime sleepiness for detection of obstructive sleep apnea (OSA). In addition to dysphagia of varying severity, OSA was found in nine patients, change of voice in six, globus sensation in seven, aspiration in three, and cervical pain in seven. Flexible laryngoscopy showed bulging of the posterior pharyngeal wall in all patients. DISH may be an unrecognized contributory factor to both dysphagia and OSA in the elderly. Change of voice, aspiration, globus sensation, and cervical pain are other otolaryngologic manifestations that may be encountered symptoms of the disease. An otolaryngologist should be aware of the disease that may be overlooked, and computed tomography is a confirmatory diagnostic method.

Keywords Dysphagia · Obstructive sleep apnea · Cervical osteophytes · DISH · Cervical pain

Introduction

Diffuse idiopathic skeletal hyperostosis (DISH) is a skeletal disorder of unknown etiology. Its main characteristic is the ossification of the anterior longitudinal spinal ligament, with large osteophytes that flow down the spine, producing a typical candle wax appearance. It was described in the elderly and may lead to abundant bone formation, ossification, and calcification of connective tissue in spinal and extraspinal sites. The vertebral findings were first described as "senile vertebral ankylosing hyperostosis" by Forestier and Rotes-Querol in 1950 [1, 2]. Any part of the anterior longitudinal ligament may be affected, but the most typical involved sites are the mid-low cervical and the mid-low thoracic vertebral segments where bony ankylosis may develop [3]. Extraspinal manifestations have also been reported. The disease may cause bony hyperostosis at tendon and ligament attachments in different parts of the body [4].

The diagnosis of DISH is primarily radiological [5]. It is probably an asymptomatic condition in many affected individuals, although numerous clinical symptoms have

Electronic supplementary material The online version of this article (doi:10.1007/s00405-013-2827-z) contains supplementary material, which is available to authorized users.

M. Abdel-Aziz (✉)
Department of Otolaryngology, Cairo University, 2 el-salam st., King Faisal, Above El-baraka Bank, Giza, Cairo, Egypt
e-mail: mosabeez@yahoo.com

N. A. Azab
Department of Rheumatology and Rehabilitation, Cairo University, Cairo, Egypt

M. Rashed
Department of Otolaryngology, Beni Suef University, Beni Suef, Egypt

A. Talaat
Department of Otolaryngology, Suez Canal University Hospital, Ismailia, Egypt

Table 1 Clinical characteristics of the patients

Patients	Age (years)	Sex	Level of hyperostosis	EAT-10 score	ESS score	VHI-10 score	Globus sensation	Cervical pain	Aspiration	AHI
1	73	Male	C_2–T_7	34	17	25	Present	Absent	Present	23
2	61	Male	C_3–C_7	10	8	32	Absent	Present	Absent	Not done
3	80	Female	C_4–T_3	26	16	10	Present	Present	Present	26
4	59	Male	C_2–T_7	17	6	8	Absent	Present	Absent	Not done
5	65	Female	C_2–C_7	20	18	16	Present	Present	Absent	21
6	70	Female	C_3–C_6	12	13	22	Present	Absent	Absent	13
7	62	Male	C_2–C_7	15	12	8	Absent	Absent	Absent	11
8	68	Male	C_2–T_7	12	11	9	Absent	Present	Absent	9
9	70	Male	C_3–C_7	32	20	24	Present	Present	Present	35
10	64	Female	C_4–T_3	16	19	30	Present	Present	Absent	18
11	72	Male	C_4–T_3	28	14	6	Present	Absent	Absent	20

C cervical vertebral body, *T* thoracic vertebral body, *EAT-10* swallowing screening tool, *ESS* Epworth Sleepiness Scale, *VHI-10* voice handicap index, *AHI* apnea/hypopnea index

been described including pain, limited range of spinal motion and increased susceptibility to unstable spinal fractures after trivial trauma [6]. In some cases of extensive ossification of the cervical spine, compression of the esophagus and less often the trachea by ossified anterior longitudinal ligament can lead to dysphagia, hoarseness, stridor and dyspnea [7]. Also, large osteophytes may compress the posterior pharyngeal wall resulting in sleep disturbed breathing [8]. The aim of this study was to investigate the otolaryngologic manifestations of DISH.

Materials and methods

The study was conducted on 11 patients who presented with dysphagia and radiologically diagnosed to have DISH. Barium swallow and computed tomography (CT) were done for all patients to confirm the diagnosis and magnetic resonance imaging was requested for patients suspected to have medullary compression. However, medullary compression was not the objective subject of the study. Diagnosis of DISH was based on the radiographic criteria described by Resnick and Niwayama [5]. These criteria include flowing ossification of the anterolateral aspect of at least four contiguous vertebral bodies, relative preservation of the disc height in the involved segments with absence of radiographic changes associated with disc degenerative disorder, and absence of sacroilitis and facet ankylosis. Only patients who fulfilled these criteria were included. Patients who gave history of chronic diseases of the esophagus and/or the respiratory system were excluded. The study was carried out in the period from September 2007 to June 2012. Table 1 summarizes the clinical characteristics of the patients. Informed consent was obtained from the patients, and the principles outlined in the Declaration of Helsinki were followed. In addition, the research protocol was approved by the ethics committee of our institute.

After radiologic confirmation of DISH, patients underwent extensive assessment as follows.

Clinical examination

Medical history was obtained from the patients, with emphasis on throat and airway symptoms. The severity of dysphagia was graded on the swallowing screening tool (EAT-10), in which if the score was 3 or more, the swallowing was considered abnormal [9]. Patients with score up to 20 were subjected to conservative treatment in the form of modification of diet, non-steroidal anti-inflammatory drugs, corticosteroids, muscle relaxants and antireflux medications, and patients with score more than 20 were subjected to surgical reduction of the osteophytes via the anterolateral neck approach [10, 11]. Surgery was performed by the orthopedic surgeons of our institute. Patients' swallows were followed up for at least 6 months. The Epworth Sleepiness Scale (ESS) was used to measure the daytime sleepiness; a score of 10 or more reflects above normal daytime sleepiness [12]. The voice of the patients was assessed using the voice handicap index (VHI-10), and a score more than 11 is considered abnormal [13, 14]. Patients were also questioned about the presence of stridor, aspiration, globus sensation and cervical pain. Full ear, nose and throat, and head and neck examinations were done for the detection of other associated lesions. Special attention was paid to looking at the posterior pharyngeal wall.

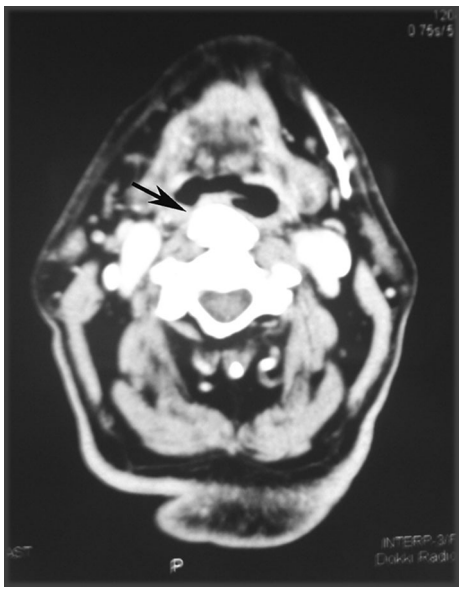


Fig. 1 An axial computed tomography. The *arrow* points to a large osteophyte compressing the posterior pharyngeal wall

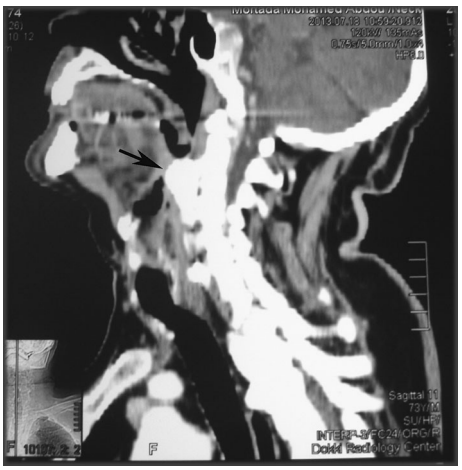


Fig. 2 A sagittal computed tomography. The *arrow* points to the hyperostotic anterior longitudinal spinal ligament with large osteophytes compressing the aerodigestive tract

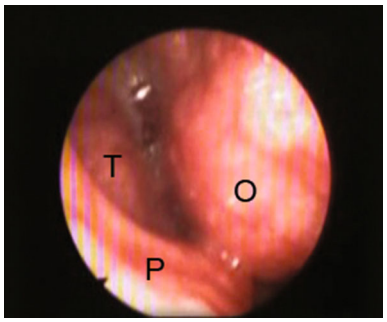


Fig. 3 A nasopharyngoscopic view for a patient with DISH. A bulged posterior pharyngeal wall (O) contacts the soft palate (P) and base of the tongue (T)

Flexible laryngoscopy

Visualization of the upper airway was performed using a fiberoptic flexible laryngoscope. The nose was decongested and anesthetized with a mixture of 4 % lidocaine and 0.05 % oxymetazoline hydrochloride. The endoscope was introduced through the nose down to the larynx and hypopharynx, and any abnormality in the upper airway was recorded.

Polysomnography

Patients who had scores 10 or more on ESS were assessed by overnight polysomnography for at least 6 h (laboratory-based study) in a quiet dark room. Apnea/hypopnea index (AHI) was measured. AHI index was categorized as follows: <5.0, no obstructive sleep apnea (OSA); ≥5.0 to <15.0, mild OSA; ≥15.0 to <30.0, moderate OSA; and ≥30.0, severe OSA [15]. Patients with mild OSA were advised to reduce their body weights, those with moderate OSA were treated with continuous positive airway pressure (CPAP) and those with severe OSA were subjected to surgical reduction of osteophytes [8, 16].

Results

The study included 11 patients (7 males and 4 females) diagnosed to have DISH with radiographic criteria of the disease, with a mean age of 67.6 years at presentation. Barium swallow showed compression of the esophageal wall in the cervico-thoracic region, with no intrinsic lesions of the esophagus. CT showed ossification of the anterior longitudinal ligament with large osteophytes (Figs. 1, 2); the lesion extended from C_2 to T_7 in three patients, from C_4 to T_3 in three patients, from C_2 to C_7 in two patients, from C_3 to C_7 in two patients and from C_3 to C_6 in one patient. Airway compression by osteophytes was detected in three patients: two at the level of the oropharynx and one at the level of the larynx.

Dysphagia was the presenting complaint of all patients with varying severity. The dysphagia score was 20 or less in seven patients (63.5 %) and more than 20 in four patients. Nine patients (82 %) had ESS score more than 10. Six patients (54.5 %) had abnormal VHI-10 score. Seven patients (63.5 %) complained of globus sensation, three had aspiration, seven (63.5 %) had cervical pain and none suffered from stridor. Oropharyngeal examination showed bulging of the posterior pharyngeal wall in all patients with intact healthy mucosa.

Flexible laryngoscopy demonstrated narrowing of the pharyngeal lumen by bulging of the posterior wall in all patients (Fig. 3). The larynx was displaced anteriorly with overhanging of the bulged posterior pharyngeal wall over the arytenoids and posterior part of the laryngeal inlet in nine patients. Fixation of the right vocal cord was detected in one patient.

Apnea/hypopnea index of nine patients was measured; it showed mild OSA in three patients, moderate in five and severe in one.

Seven patients were treated conservatively, while four patients needed surgical intervention. After 6 months, all patients resumed regular diets with decrease in the episodic obstructive attacks during sleep.

Discussion

DISH is a disease characterized by calcification and ossification of soft tissue in the elderly, with predilection for the anterior longitudinal spinal ligament, mostly in the middle and lower thoracic regions, although cervical involvement is found in 76 % of those patients [3, 4]. Its etiology is not yet defined, but there are associations with some metabolic disorders such as diabetes, obesity, hypercholesterolemia and gout [11, 17]. The disease has for a long time thought to be an asymptomatic condition, usually detected incidentally at old age. It has been considered a radiographic entity with little clinical signs and symptoms [17]. However, DISH may lead to spinal rigidity which may pass unnoticed in the elderly, acute or chronic pain caused by dynamic overload of the hyperossified segment and associated root compression, syndromes produced by space occupation and protrusion of hyperostotic bones into the spinal canal producing medullary compression, or outwards, pressing on the esophagus, trachea or larynx. The disease may, also, affect tendon and ligament attachments in different parts of the body [18].

In this study, we examined 11 patients who presented with dysphagia and diagnosed with DISH, aiming to analyze the different otolaryngologic manifestations of those patients. The diagnosis of DISH was based on CT radiographic findings described by Resnick and Niwayama [5]. It is a useful imaging modality in the diagnosis of DISH, as the size and shape of the osteophytes are shown in relation to the esophagus and other important structures [3, 11]. The CT radiographic criteria allow DISH to be differentiated from ankylosing spondylitis and intervertebral osteochondrosis, which are two other diagnoses that can be responsible for vertebral osteophytosis [19].

Dysphagia has been reported to be the main otolaryngologic symptom in DISH [5, 10, 11, 18, 20–22]. Prominent anterior cervical osteophyte is hypothesized to be the result of direct compression of the aerodigestive tract and associated nerves as well as local inflammation that leads to mucosal edema, adhesion formation, fibrosis and cricopharyngeus muscle spasm. The cause of local inflammation is thought to be the result of repetitive mechanical trauma caused by the constant friction of the pharynx and esophagus over the large hypertrophic osteophytes [16, 21]. Dysphagia caused by cervical osteophytes may be treated conservatively or surgically; surgery may be needed when the osteophytes are remarkably large [20]. In our study, we graded the severity of dysphagia on the EAT-10 scoring system [9]. Seven patients had a score of 3–20 and were treated conservatively, while four patients had a score of more than 20 and were treated surgically. All of our patients resumed regular diet within the follow-up period. This strategy of treatment has been used by many authors. Kandogan et al. [23] and Ohki [24] used conservative measures for the treatment of mild dysphagia caused by DISH and achieved successful results. However, they advised surgery for severe cases. Surgical removal of osteophytes has been reported by Carlson et al. [16] and Castellano et al. [19]. Their patients showed significant improvement and most of them resumed regular diet postoperatively.

Narrowing of the airway by large osteophytes may lead to OSA and even stridor. The osteophytes compress the posterior pharyngeal wall and may even impinge on the larynx and trachea [8, 19, 22]. In our study, we detected OSA in nine patients: mild in three who were advised to reduce their body weight, moderate in five who were managed with CPAP and severe in one who underwent surgical reduction of osteophytes. All patients showed satisfactory improvement with decreased episodic obstructive attacks. Fuerderer et al. [8] found OSA due to DISH in three patients who were treated with surgical reduction of osteophytes. They achieved a decrease of apneic attacks postoperatively. Stridor due to hyperostosis of the cervical spine associated with DISH has been reported by Carlson et al. [16], Castellano et al. [19] and Vengust et al. [22]. None of our patients demonstrated stridor.

Other otolaryngologic symptoms that may be present in DISH are dysphonia, aspiration, globus sensation and cervical pain [10, 21, 22, 25]. In our study, we found voice changes in six patients; we used the self-report questionnaire VHI-10 that is internationally accepted for self-perception measurement of voice problems [13, 14]. Also, we detected aspiration in three patients, globus sensation in seven patients and cervical pain in seven patients. Flexible laryngoscopy showed fixed right vocal cord in one patient, and cricoarytenoid joint fixation was found in DISH by

Anand et al. [25] and Verstraete et al. [26]. The ankylosis of the cricoarytenoid joint may be due to mechanical compression of the osteophytes on the cricoid and arytenoid cartilages, which subsequently causes chondritis [25]. As the posterior pharyngeal wall projects anteriorly, it may lead to stagnation of food and fluid in the pyriform sinuses and impaired laryngeal elevation with subsequent aspiration and globus sensation [24]. Cervical pain is explained by dynamic overload of the hyperossified segment and associated root compression. Also, it may be caused by anterior soft tissue compression by the osteophytes [8, 18].

It is worth mentioning that our study is based on a somewhat small sample of patients, due to the rarity of the disease. Also, dysphagia was the leading symptom for the diagnosis of DISH, and so we could not determine if the disease would present first by another otolaryngologic symptom or not. Follow-up polysomnography and validated questionnaires were not used as a confirmatory method for symptom relief.

Conclusion

Diffuse idiopathic skeletal hyperostosis (DISH) may be an unrecognized contributory factor to both dysphagia and OSA in the elderly. Change of voice, aspiration, globus sensation and cervical pain are other otolaryngologic manifestations that may be encountered in the disease. An otolaryngologist should be aware of the disease that may be overlooked, and computed tomography is a confirmatory diagnostic method.

Acknowledgments We are grateful to the staff of the departments of radiology and orthopedic surgery in our institute for their assistance and contribution.

Conflict of interest The authors declare that they have no conflict of interest.

References

1. Forestier J, Rotes-Querol J (1950) Senile ankylosing hyperostosis of the spine. Ann Rheum Dis 9:321–330
2. Giuffra V, Giusiani S, Fornaciari A, Villari N, Vitiello A, Fornaciari G (2010) Diffuse idiopathic skeletal hyperostosis in the Medici, Grand Dukes of Florence (XVI century). Eur Spine J 19:S103–S107
3. Paja L, Molnár E, Ôsz B, Tiszlavicz L, Palkó A, Coqueugniot H, Dutour O, Pálfi G (2010) Diffuse idiopathic skeletal hyperostosis—appearance and diagnostics in Hungarian osteoarcheological materials. Acta Biol Szeged 54:75–81
4. Resnick D, Shaul SR, Robins JM (1975) Diffuse idiopathic skeletal hyperostosis (DISH): Forestier's disease with extraspinal manifestations. Radiology 115:513–524
5. Resnick D, Niwayama G (1976) Radiographic and pathologic features of spinal involvement in diffuse idiopathic skeletal hyperostosis (DISH). Radiology 119:559–568
6. Mader R, Sarzi-Puttini P, Atzeni F, Olivieri I, Pappone N, Verlaan J, Buskila D (2009) Extraspinal manifestations of diffuse idiopathic skeletal hyperostosis. Rheumatology 48:1478–1481
7. McCafferty RR, Harrison MJ, Tamas LB et al (1996) Ossification of the anterior longitudinal ligament and Forestier's disease: an analysis of seven cases. J Neurosurg 83:13–17
8. Fuerderer S, Eysel-gosepath K, Schröder U, Delank K-S, Eysel P (2004) Retro-pharyngeal obstruction in association with osteophytes of the cervical spine. J Bone Joint Surg (Br) 86-B:837–840
9. Belafsky PC, Mouadeb DA, Rees CJ, Pryor JC, Postma GN, Allen J, Leonard RJ (2008) Validity and reliability of the eating assessment tool (EAT-10). Ann Otol Rhinol Laryngol 117:919–924
10. Aydin E, Akdogan V, Akkuzu B, Kirbas I (2006) Six cases of Forestier syndrome, a rare cause of dysphagia. Acta Otolaryngol 126:775–778
11. Srivastava S, Ciapryna N, Bovill I (2008) Diffuse idiopathic skeletal hyperostosis as an overlooked cause of dysphagia: a case report. J Med Case Rep 2:287. doi:10.1186/1752-1947-2-287
12. Johns MW (1992) Reliability and factor analysis of the Epworth Sleepiness Scale. Sleep 15:376–381
13. Rosen CA, Lee AS, Osborne J, Zullo T, Murry T (2004) Development and validation of the voice handicap index-10. Laryngoscope 114:1549–1556
14. Arffa RE, Krishna P, Gartner-Schmidt J, Rosen CA (2012) Normative values for the voice handicap index-10. J Voice 26:462–465
15. Banerjee D (2008) Obstructive sleep apnea: medical management. In: Gleeson M, Browning GG, Burton MJ, Clarke R, Hibbert J, Jones NS et al (eds) Scott–Brown's otorhinolaryngology, head and neck surgery, 7th edn. Oxford University Press, London, pp 2313–2324
16. Carlson ML, Archibald DJ, Graner DE, Kasperbauer JL (2011) Surgical management of dysphagia and airway obstruction in patients with prominent ventral cervical osteophytes. Dysphagia 26:34–40
17. Anand SS, Das G, Chakraborty DP, Saha SP, Tripathi P (2012) Diffuse idiopathic skeletal hyperostosis: a case report. Neurology Asia 17:365–368
18. Rotes-Querol J (1996) Clinical manifestations of diffuse idiopathic skeletal hyperostosis (DISH). Br J Rheumatol 35:1193–1196
19. Castellano DM, Sinacori JT, Karakla DW (2006) Stridor and dysphagia in diffuse idiopathic skeletal hyperostosis (DISH). Laryngoscope 116:341–344
20. Solaroglu I, Okutan O, Karakus M, Saygili B, Beskonakli E (2008) Dysphagia due to diffuse idiopathic skeletal hyperostosis of the cervical spine. Turk Neurosurg 18:409–411
21. Nimmons G, Van Daele DJ, Hoffman HT, Rao SS, Clark CR (2010) Multifactorial dysphagia: diffuse idiopathic skeletal hyperostosis and eosinophilic esophagitis. Laryngoscope 120:23–25
22. Vengust R, Mihalic R, Turel M (2010) Two different causes of acute respiratory failure in a patient with diffuse idiopathic skeletal hyperostosis and ankylosed cervical spine. Eur Spine J 19:S130–S134
23. Kandogan T, Sezgin Ö, Dalgiç A (2012) Diffuse idiopathic skeletal hyperostosis (DISH): a rare cause of dysphagia. Ege J Med 51:259–261
24. Ohki M (2012) Dysphagia due to diffuse idiopathic skeletal hyperostosis. Case Rep Otolaryngol. doi:10.1155/2012/123825

25. Anand V, Vikram Vel VR, Purushothaman PK, Rajesh Kumar MS (2011) Cricoarytenoid joint fixation in diffuse idiopathic skeletal hyperostosis (DISH): a case report. Indian J Otolaryngol Head Neck Surg 63:S55–S57

26. Verstraete WL, De Cauwer HG, Verhulst D, Jacobs F (1998) Vocal cord immobilisation in diffuse idiopathic skeletal hyperostosis (DISH). Acta Otorhinolaryngol Belg 52:79–84