



Review

Diffuse idiopathic skeletal hyperostosis in pharyngolaryngeal pathology

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ABSTRACT

The objective of this review is to summarize specialty data regarding possible manifestations of diffuse idiopathic skeletal hyperostosis in general otorhinolaryngology practice in order to increase awareness of this entity. Research on the topic was based on consulting specialty books, electronic databases like PubMed, Scopus (Elsevier), Web of Science, Research Gate, Up-to-date and also, on clinical experience. Even though asymptomatic in most patients and many times found during cervical pain investigations or by accident, this overlooked disorder, in rare cases, may cause two of the main complains in practice, dysphagia and dyspnea.

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1. FOREWORD

This paper is a brief review of specialty literature data regarding the possible relation between diffuse idiopathic skeletal hyperostosis, a chronic spine pathology, and a series of complains in general otorhinolaryngology practice. It is meant to draw attention of general otorhinolaryngologists over a usually underestimated pathology that, sometimes, may be responsible for severe dysphagia, weight loss and even airway obstruction.

2. INTRODUCTION

2.1. WHAT IT IS

Osteophytes are bony growths that can be encountered at any spine level either cervical, thoracic or lumbosacral level. Diffuse idiopathic skeletal hyperostosis (DISH), also called Forestier syndrome, is a noninflammatory disease characterized by calcification and ossification of paravertebral ligaments, especially the anterior longitudinal ligament, resulting in exuberant osteophytes formation [1]. Peripheral ethereal ossification and bony spurs can also be found [2, 3].

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2.2. WHY?

Its etiology is yet unknown but several publications correlated osteophytes formation to metabolic disorders such as diabetes mellitus, increased body mass index or higher serum level of uric acid, also genetic factors have been hypothesized [4, 5]. Involvement of growth factors that influence bone formation or remodeling, osteoblasts and osteoclasts, has been considered an important factor in pathophysiology. Insulin-like growth factor, growth hormone, platelet-derived growth factor and insulin have been correlated to DISH [6].

Association with prior treatments with vitamin A, isotretinoin, etretinate, acitretinate or other vitamin A derivatives have been investigated [7, 8].

2.3. HOW OFTEN?

Its incidence was considered by Weinfeld et al., in a study from 1997, between 15%–20% in the elderly people with prevalence greater in males (27.3%) than females (12.8%), based on X-ray findings [1, 6]. Higher data are stated by other studies based on CT-scans of the spine. Hirasawa et al. found a prevalence of DISH based on CT of 27.1%, more than that of x-ray and a higher percentage of men compared to women. [9]. A small study published in 2018 based on spine CT scans of patients over 16 years old, found an overall incidence of 24.4% in population over 40 years of age [10].

3. CLINICAL ASPECTS

DISH is seen most commonly in men over 50 years of age, asymptomatic in most patients but the presence of spinal or extra-spinal ossification can sometimes lead to symptoms like pain, stiffness, a reduced range of articular motion, increased risk of unstable spinal fractures [1]. Dysphagia and even upper airway obstruction have been reported in advanced cases [11].

3.1. SYMPTOMS

Abnormal bone formation that characterizes diffuse idiopathic skeletal hyperostosis may be asymptomatic, may give back pain of various degrees and stiffness, but in some cases, can be the cause of severe dysphagia or dyspnea. Neck pain, usually dull, tolerated, and stiffness are not unusual in older patients presenting in general otorhinolaryngology practice for different complains. These symptoms are caused by DISH too [12].

Patients may complain of impaired swallowing, dysphagia, initially for solid food, many times ignored by patients and doctors both, due to slowly progression of the disease and to other pathologies encountered in elderly patients, more likely to cause dysphagia than bony growths of the spine. As osteophytes presence is asymptomatic in most individuals, inflammatory, humoral, neurological or

neuromuscular pathologies should be excluded first.

Dysphagia in DISH has been explained by several mechanisms, in medical literature on this topic. In time, along with slow growth, anterior large osteophytes exert extrinsic compression on the pharynx or the esophagus depending on the affected level. Besides the mass effect, chronic irritation provoked by osteophytes leads to inflammation and fibrosis of esophageal wall, a cumulative factor in swallowing disorders [12, 13]. Secondary esophageal denervation has been incriminated in dysphagia associated with DISH [14]. Esophagus may be pushed against the hard cricoid cartilage, accompanied by an inflammatory reaction of the soft tissue causing cricopharyngeal spasm [15]. A recent study performed in Spain on 455 patients with dysphagia reported DISH as the cause in 11.2% percent and an incidence of 7:100000 inhabitants per year [16]. Verlaan et al. concluded after a systematic review of 118 articles on this theme, that diffuse idiopathic skeletal hyperostosis as a cause of dysphagia and/or airway obstruction may be an increasing and underappreciated phenomenon and should be included in the differential diagnosis of dysphagia and airway obstruction [17].

Although swallowing disorders appear more often, dyspnea and other respiratory manifestations have been reported in several cases due to encroachment of anterior protruding osteophytes on the larynx or trachea [18]. As an extremely rare condition, vocal fold paralysis were found in relation to DISH [19]. Other respiratory symptoms, in alterations of cervical spine, can be hoarseness, snoring and laryngeal stridor, and dysphonia. [20] Marks et al. reported a case of severe dyspnea and edema of the laryngeal inlet caused by excessively enlarged cervical osteophytes leading to emergency tracheotomy [20].

Difficult intubation was mentioned and could be expected in advanced DISH. Besides the reduced mobility of the cervical spine, airway management can prove difficult due to anatomic changes like osteophytes protrusion through posterior pharyngeal wall, displacement of the larynx or narrowing of the trachea [18, 21].

In rare cases neurological impairment can be encountered and explained by the ossification of posterior longitudinal ligament and ligamentum flavum that can lead to medullary canal stenosis, spinal cord compression and myelopathy [22]. Upper cervical spine can be affected by retro odontoid masses, atlantoaxial pseudo arthrosis, and atlantoaxial subluxation causing neurological problems [23].

3.2. DIAGNOSIS

Diagnosis is based on clinical findings and imaging. Bulging of the posterior oropharyngeal wall is sometimes seen by general otorhinolaryngologists during routine oropharyngeal examination in elder patients.

A flexible naso-pharyngo-laryngeal endoscopy is very useful to appreciate the hypopharyngeal bulging, laryngeal displacement, superior narrowing of airway and food

passage due to abnormal external compression and also for differential diagnostic with intrinsic pathology. Superior digestive endoscopy may be necessary to exclude other intrinsic causes of inferior dysphagia, even though the procedure carries a higher risk of perforating or damaging the esophagus [24].

X-ray of cervical spine is usually helpful in diagnostic [21, 25]. Typical aspects on lateral X-ray are flowing ossification of the anterior longitudinal ligament while intervertebral disc space is initially preserved and bridging ossification connecting the bodies of adjacent vertebrae [22]. Barium swallow X-rays is useful to diagnose the narrowing of the pharyngo-esophageal tract and the level of impaired food passage [26].

Computed tomography (CT) scans offer a better view of the spine, show ossification of the posterior cervical longitudinal ligament or of the ligamentum flavum, the displacement of the larynx, trachea, esophagus and it has been considered more reliable compared to plane radiography [11, 27]. The tendency of anterior hyperostosis in cervical segments in DISH was hypothesized, in a small, recent study based on CT scans, to be the consequence of the natural barrier formed by the main vessels of the neck [27]. MRI is another useful imaging resource for cervical region pathology.

4. TREATMENT

Asymptomatic patients don't require treatment. Symptomatic patients should seek medical assistance and be treated according to the severity of manifestations.

4.1. CONSERVATIVE TREATMENT

Mild neck pain and stiffness can benefit from physical therapy, analgesic medication and non-steroidal anti-inflammatory drugs such as naproxen or ibuprofen [6].

4.2. SURGICAL TREATMENT

Reserved for severe dysphagia or airway obstruction, osteophyctomy, the surgical removal of large anterior osteophytes, has been reported to have good surgical outcomes by several studies, but the extension of resections is still controversial [29, 30]. Surgical decompression through osteophyctomy was considered by Carlson et al., a good alternative to tracheotomy and feeding tube in thorough selected patients who failed conservative medical management [31] On a long-term follow-up of five patients surgically treated for dysphagia related to diffuse idiopathic skeletal hyperostosis, ranging 24 to 126 months, Scholz et al. reported important improvement, even though recurrence of osteophytes growth was admitted possible [28].

5. CONCLUSIONS

Diffuse idiopathic skeletal hyperostosis is a chronic, overlooked and often asymptomatic disorder that may sometimes cause two of the main complains in otorhinolaryngology practice, dysphagia and dyspnea and should be kept in mind by practitioners for differential diagnostic.

6. REFERENCES

1. Resnick D, Shapiro RF, Wiesner KB, Niwayama G, Utsinger PD, Shaul SR. Diffuse idiopathic skeletal hyperostosis (DISH) [ankylosing hyperostosis of Forestier and Rotes-Querol]. *Semin Arthritis Rheum.* 1978;7(3):153-87. doi: 10.1016/0049-0172(78)90036-7.
2. Maat GJR, Mastwijk RW, van der Velde EA. Skeletal distribution of degenerative changes in vertebral osteophytosis, vertebral osteoarthritis and DISH. *Int J Osteoarchaeol.* 1995; 5:289-98. doi: <https://doi.org/10.1002/oa.1390050308>.
3. Mata S, Fortin PR, Fitzcharles MA, Satarr MR, Joseph L, Watts CS, et al. A controlled study of diffuse idiopathic skeletal hyperostosis. Clinical features and functional status. *Medicine (Baltimore).* 1997;76(2):104-17. doi: 10.1097/00005792-199703000-00003.
4. Denko CW, Malemud CJ. Body mass index and blood glucose: correlations with serum insulin, growth hormone, and insulin-like growth factor-1 levels in patients with diffuse idiopathic skeletal hyperostosis (DISH). *Rheumatol Int.* 2006;26(4):292-7. doi: 10.1007/s00296-005-0588-8.
5. Sarzi-Puttini P, Atzeni F. New developments in our understanding of DISH (diffuse idiopathic skeletal hyperostosis). *Curr Opin Rheumatol.* 2004;16(3):287-92. doi: 10.1097/00002281-200405000-00021.
6. Mader R, Verlaan JJ, Buskila D. Diffuse idiopathic skeletal hyperostosis: clinical features and pathogenic mechanisms. *Nat Rev Rheumatol.* 2013;9(12):741-50. doi: 10.1038/nrrheum.2013.165.
7. DiGiovanna JJ, Helfgott RK, Gerber LH, Peck GL. Extraspinal tendon and ligament calcification associated with long-term therapy with etretinate. *N Engl J Med.* 1986;315(19):1177-82. doi: 10.1056/NEJM198611063151901.
8. Troillet N, Gerster JC. [Forestier disease and metabolism disorders. A prospective controlled study of 25 cases]. *Rev Rhum Ed Fr.* 1993;60(4):274-9.
9. Hirasawa A, Wakao N, Kamiya M, Takeuchi M, Kawanami K, Murotani K, et al. The prevalence of diffuse idiopathic skeletal hyperostosis in Japan - the first report of measurement by CT and review of the literature. *J Orthop Sci.* 2016;21(3):287-90. doi: 10.1016/j.jos.2016.02.001.
10. Weinfeld RM, Olson PN, Maki DD, Griffiths HJ. The prevalence of diffuse idiopathic skeletal hyperostosis (DISH) in two large American Midwest metropolitan hospital populations. *Skeletal Radiol.* 1997;26(4):222-5. doi: 10.1007/s002560050225.
11. Kim BS, Moon MS, Yoon MG, Kim ST, Kim SJ, Kim MS, et al. Prevalence of Diffuse Idiopathic Skeletal Hyperostosis Diagnosed by Whole Spine Computed Tomography: A Preliminary Study. *Clin Orthop Surg.* 2018;10(1):41-6. doi: 10.4055/cios.2018.10.1.41.
12. Vaishya R, Vijay V, Nwagbara IC, Agarwal AK. Diffuse idiopathic skeletal hyperostosis (DISH) - A common but less known cause of back pain. *J Clin Orthop Trauma.* 2017;8(2):191-6. doi: 10.1016/j.jcot.2016.11.006.
13. Masiero S, Padoan E, Bazzi M, Ponzoni A. Dysphagia due to diffuse idiopathic skeletal hyperostosis: an analysis of five cases. *Rheumatol Int.* 2010;30(5):681-5. doi: 10.1007/s00296-009-0967-7.
14. McCafferty RR, Harrison MJ, Tamas LB, Larkins MV. Ossification of the anterior longitudinal ligament and Forestier's disease: an analysis of seven cases. *J Neurosurg.* 1995;83(1):13-7. doi: 10.3171/jns.1995.83.1.0013.
15. Papadopoulos SM, Chen JC, Feldenzer JA, Bucci MN, McGillicuddy JE. Anterior cervical osteophytes as a cause of progressive dysphagia. *Acta Neurochir (Wien).* 1989;101(1-2):63-5. doi: 10.1007/BF01410071.
16. García Callejo FJ, Oishi N, López Sánchez I, Pallarés Martí B, Rubio Fernández A, Gómez Gómez MJ. Incidence of diffuse idiopathic skeletal

- hyperostosis from a model of dysphagia. *Acta Otorrinolaringol Esp.* 2020;71(2):78-82. doi: 10.1016/j.otorri.2019.02.003.
17. Verlaan JJ, Boswijk PF, de Ru JA, Dhert WJ, Oner FC. Diffuse idiopathic skeletal hyperostosis of the cervical spine: an underestimated cause of dysphagia and airway obstruction. *Spine J.* 2011;11(11):1058-67. doi: 10.1016/j.spinee.2011.09.014.
18. Baxi V, Gaiwal S. Diffuse idiopathic skeletal hyperostosis of cervical spine - An unusual cause of difficult flexible fiber optic intubation. *Saudi J Anaesth.* 2010;4(1):17-19. doi: 10.4103/1658-354X.62609.
19. Virk JS, Majithia A, Lingam RK, Singh A. Cervical osteophytes causing vocal fold paralysis: case report and literature review. *J Laryngol Otol.* 2012;126(9):963-5. doi: 10.1017/S0022215112001259.
20. Marks B, Schober E, Swoboda H. Diffuse idiopathic skeletal hyperostosis causing obstructing laryngeal edema. *Eur Arch Otorhinolaryngol.* 1998;255(5):256-8. doi: 10.1007/s004050050053.
21. Gosavi K, Dey P, Swami S. Airway Management in Case of Diffuse Idiopathic Skeletal Hyperostosis. *Asian J Neurosurg.* 2018;13(4):1260-3. doi: 10.4103/ajns.AJNS_235_17.
22. Colina M, Govoni M, De Leonardi F, Trotta F. [Diffuse idiopathic skeletal hyperostosis (D.I.S.H.)]. *Reumatismo.* 2006;58(2):104-11. doi: 10.4081/reumatismo.2006.104.
23. Zhang C, Ruan D, He Q, Wen T, Yang P. Progressive dysphagia and neck pain due to diffuse idiopathic skeletal hyperostosis of the cervical spine: a case report and literature review. *Clin Interv Aging.* 2014;9:553-7. doi: 10.2147/CIA.S60146.
24. Solaroğlu I, Okutan O, Karakuş M, Saygili B, Beşkonaklı E. Dysphagia due to diffuse idiopathic skeletal hyperostosis of the cervical spine. *Turk Neurosurg.* 2008;18(4):409-11.
25. Aydın E, Akdoğan V, Akkuzu B, Kirbaş I, Özgirgin ON. Six cases of Forestier syndrome, a rare cause of dysphagia. *Acta Otolaryngol.* 2006;126(7):775-8. doi: 10.1080/00016480500504192.
26. Dutta S, Biswas KD, Mukherjee A, Basu A, Das SW, Sen I, et al. Dysphagia due to forestier disease: three cases and systematic literature review. *Indian J Otolaryngol Head Neck Surg.* 2014;66(Suppl 1):379-84. doi: 10.1007/s12070-011-0334-3.
27. Bakker JT, Kuperus JS, Kuiff HJ, Oner FC, de Jong PA, Verlaan JJ. Morphological characteristics of diffuse idiopathic skeletal hyperostosis in the cervical spine. *PLoS One.* 2017;12(11):e0188414. doi: 10.1371/journal.pone.0188414.
28. Scholz C, Naseri Y, Hohenhaus M, Hubbe U, Klingler JH. Long-term results after surgical treatment of diffuse idiopathic skeletal hyperostosis (DISH) causing dysphagia. *J Clin Neurosci.* 2019;67:151-5. doi: 10.1016/j.jocn.2019.05.057.
29. Kos MP, van Royen BJ, David EF, Mahieu HF. Anterior cervical osteophytes resulting in severe dysphagia and aspiration: two case reports and literature review. *J Laryngol Otol.* 2009;123(10):1169-73. doi: 10.1017/S0022215109005246.
30. Yoshioka K, Murakami H, Demura S, Kato S, Yonezawa N, Takahashi N, et al. Surgical Treatment for Cervical Diffuse Idiopathic Skeletal Hyperostosis as a Cause of Dysphagia. *Spine Surg Relat Res.* 2018;2(3):197-201. doi: 10.22603/ssrr.2017-0045.
31. Carlson ML, Archibald DJ, Graner DE, Kasperbauer JL. Surgical management of dysphagia and airway obstruction in patients with prominent ventral cervical osteophytes. *Dysphagia.* 2011;26(1):34-40. doi: 10.1007/s00455-009-9264-6.