

INTRODUCTION

In 1950, Jacques Forestier and Jaume Rotes-Querol initially described Forestier Disease; also known as diffuse idiopathic skeletal hyperostosis (DISH). It is a rare, non-inflammatory condition that largely develops in the elderly population and is characterized by calcification and osteophyte formation within the spinal column and peripheral enthesopathies.¹ DISH also leads to fragile bridging osteophytes, which are susceptible to pathological fractures. As many patients go undiagnosed, it becomes of utmost importance for emergency medicine physicians to be aware of this disease. Here, we present the case of a patient with an acutely worsening myriad of symptoms secondary to DISH.

BACKGROUND

- DISH is largely characterized by the formation of bridging vertebral osteophytes and peripheral joint involvement that subsequently lead to a myriad of clinical symptoms. When the vertebral bodies are involved, patients are placed at an increased risk of serious complications including cord compression, pathological fractures, and airway compromise.
- Two sets of criteria have been established: **The Resnick & Niwayama Criteria and the Utsinger Criteria.**²
 - Both state that patients must have 4 contiguous vertebrae involved, the relative joint spaces must be maintained and the SI joints should not be affected.
 - The Utsinger criteria states that patients with only 2 contiguous involved vertebrae or only peripheral and symmetric joint involvement may indicate a probable diagnosis.
- Abnormal osteoblastic differentiation and activity at the enthesis is thought to be the main pathophysiological mechanism. Metabolic factors are thought to contribute to this process, however its role is still ill defined.



Figure 1: Proliferative bony growths at the site of insertion of the Achilles tendon, bilaterally.³

CASE DESCRIPTION

A 78-year-old man, with a Hx of GERD, presented to the ED complaining of a sore throat, difficulty swallowing, a productive cough, and shortness of breath worsening over the past week. He had been experiencing progressively worsening dysphagia and dyspnea for almost a year. Of note, the patient's ability to provide a history was extremely limited by an obvious speech impediment.

- Patient has a PMHx of GERD.**
- Otherwise denied past surgeries, alcohol use, tobacco use, drug use, daily medications and allergies.

Physical:

Vitals – BP: 133/83, HR: 102, RR: 18, T: 37.1 °C, **O2SAT: 100%**

General: Alert, no acute distress. Appears uncomfortable.

HEENT: NT, Atraumatic. Neck supple and trachea midline.

PERRL. EOMI. No pharyngeal erythema or exudate, uvula midline. No signs of peri-tonsillar abscess and no swelling appreciated. Positive hoarse voice and obvious speech impediment.

CV: Regular rate and rhythm. No murmurs. No edema.

Respiratory: Lungs are clear to auscultation. No wheezing, rales, or rhonchi.

**Complete physical exam was performed. Negative, unless stated otherwise.

DIFFERENTIALS

- Epiglottitis.**
- Peritonsillar abscess.**
- Retropharyngeal abscess.**
- GERD related dysphagia.
- Unspecified dysphagia.
- Hernia (paraesophageal vs hiatal).

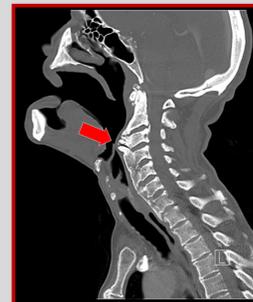


Figure 2.

- Due to the patient's symptoms in combination with his concerning speech impediment, a CT of the neck (**Figure 2**) was performed which revealed large, bulky anterior osteophytes in a contiguous fashion from C2 to C5.

HOSPITAL COURSE

- Neurosurgery was consulted and subsequently they performed an anterior osteophyctomy at C2-3 and C3-4 without complications.
- On postoperative day 1, the patient required suctioning for increased oral secretions as well as respiratory treatments for cough and dyspnea.
- Feeling that he had received all the care he required, the patient left the hospital against medical advice prior to receiving medical clearance.



Figure 3: A 66 y.o. male with DISH; A: Pre-operative sagittal CT scan showing anterior hyperostosis. B: Post-operative sagittal CT scan of same patient.⁴

DISCUSSION

- This case highlights the clinical progression of a patient with DISH as well as some of the associated concerning consequences.
- Patients with cervical DISH will present with worsening symptoms of **dysphagia, dysphonia, globus sensation, cough, and/or dyspnea.** They may also experience sharp pain and restricted range of motion in the affected areas.
- Initial management of patients with DISH is dependent on several factors. Patients with mild disease are typically managed conservatively whereas patients with increasing severity may need to be managed surgically.
- Abdel-Aziz et al. used an Eating Assessment Tool (EAT-10) questionnaire and fiber-optic endoscopy to evaluate for swallowing disorders in 139 patients with DISH. Of the 23 patients identified as having an abnormal swallowing mechanism, 21 were successfully managed with conservative measures (i.e. NSAIDs, corticosteroids, and dietary modifications) while the remaining 2 required an anterior osteophyctomy. **All patients had a drastic reduction in their symptoms.**⁵

FUTURE CONSIDERATIONS

- Patients with DISH are diagnosed after a prolonged disease course and when using the Resnick et al. criteria, it is difficult to make the diagnosis at an early stage. Future studies exploring the benefits of the Utsinger criteria in combination with screening imaging modalities may lead to an earlier diagnosis.⁶ Retrospectively looking at what co-morbidities and demographics may have contributed to the development of DISH will help establish guidelines in deciding which patients warrant screening.

CONCLUSION

- Patients suffering from DISH can present a unique challenge to physicians, especially in the emergency setting. Due to the weakening bony architecture, DISH has been associated with undiagnosed and unstable vertebral body fractures following minor trauma.⁷ Emergency physicians must keep this in mind as these patients may suffer irreversible neurological damage.
- DISH can also present as an unusual cause of airway obstruction. Undiagnosed patients may require a surgical cricothyrotomy as orotracheal intubation may not be possible secondary to mechanical obstruction.⁸ Prompt recognition of potentially debilitating symptoms and proper airway management are of the utmost importance in the acute management patients with DISH.

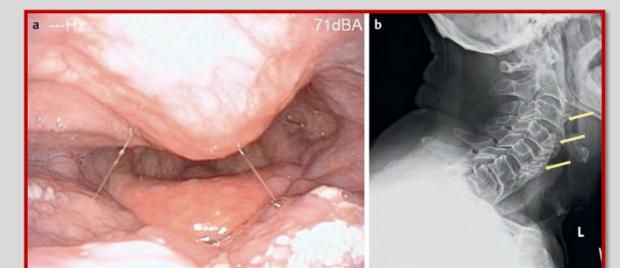


Figure 4: A 75 y.o. female with DISH involving the cervical spine; A: Endoscopic image depicting mechanical obstruction. B: Lateral radiograph of the same patient.⁹

REFERENCES

- Nascimento FA, Gatto LAM, Lages RO, Neto JM, Demarini Z, Koppe GL. Diffuse Idiopathic Skeletal Hyperostosis: A review.
- Resnick D, Niwayama G. Radiographic and pathologic features of spinal involvement in diffuse idiopathic skeletal hyperostosis (DISH). *Radiology* 1976;119:559, and Utsinger PD. Diffuse idiopathic skeletal hyperostosis. *Clin Rheum Dis* 1985; 11:325.
- Oliveri L, et al. "Heel Enthesopathy of Diffuse Idiopathic Skeletal Hyperostosis Resembling Entesitis of Spondyloarthritis." *The Journal of Rheumatology*, vol. 37, no. 1, 2009, pp. 382-383. doi:10.3899/jrheum.090214.
- Alsalimi S, Bagdadi A, Alkhatry A, et al. (March 31, 2018) Urgent Anterior Cervical Osteophyctomy for an Asymptomatic Cervical Hyperostosis to Overcome Failed Intubation. *Cureus* 10(3): e2400. doi:10.7759/cureus.2400
- Abdel-Aziz M, Arab N, Lashen H, Naguib N, Reda R. Swallowing disorders among patients with diffuse idiopathic skeletal hyperostosis. *Acta Oto-Laryngologica*. 2017;137(6):623-626. doi:10.1080/00016489.2016.1272136.
- Mader R, Verlaan J-J, Eshed I, et al. Diffuse idiopathic skeletal hyperostosis (DISH): where we are now and where to go next. *RAM Open*. 2017;3(1):e000472. doi:10.1136/ramopen-2017-000472.
- Callahan, Edward P, et al. Complications following minor trauma in a patient with diffuse idiopathic skeletal hyperostosis. *Annals of Emergency Medicine*, Volume 22, Issue 6, 1067-1070. Published June, 1993.
- Bind JH, Biggs TC, Karkos PD, Repanos C. Diffuse idiopathic skeletal hyperostosis as an acute airway presentation requiring urgent tracheostomy. *The American Journal of Emergency Medicine*. 2015;33(5). doi:10.1016/j.ajem.2014.10.050.
- Arens, Christoph & Herrmann, Ingo F. & Rohrbach, Saskia & Schwemmel, Cornelia & Nawka, Tadeus. (2015). Position paper of the German Society of Oto-Rhino-Laryngology, Head and Neck Surgery and the German Society of Phoniatrics and Pediatric Audiology – Current state of clinical and endoscopic diagnostics, evaluation, and therapy of swallowing disorders in children. *GMS Current Topics in Otorhinolaryngology - Head and Neck Surgery*. 14. 10.3205/ct000117.