Japanese Journal of Gastroenterology and Hepatology

Case Report

Forestier Disease as a Cause of Dysphagia: A Case Report

Fontanella G[1*], Fuggi G[2], Barbieri L[2], Manganiello CAT and Brogna B

[1]Department of Radiology, Ospedale Sacro Cuore di Gesu - Fatebenefratelli, Benevento, Italy

[2]Department of Internal Medicine, Ospedale Sacro Cuore di Gesu - Fatebenefratelli, Benevento, Italy

Received: 22 Mar 2020

Accepted: 01 Apr 2020

Published: 04 Apr 2020

***Corresponding author:**

Giovanni Fontanella, Department of Radiology, Ospedale Sacro Cuore di Gesu - Fatebenefratelli, Benevento, Italy, E-mail: giovanni.fontanella@hotmail.com

1. Abstract

We describe here the clinical history of a 74-year old man presenting with a gradually worsening pharyngeal dysphagia with globus, occasional intra-deglutitory coughing, hoarseness and a 5 kg weight loss in the previous two months. Apart from type II Diabetes Mellitus, the patient's clinical history was unremarkable. The patient was seen by both Gastroenterology and ENT specialists and subsequently referred to our Radiology Unit, where pharyngo-esophageal Barium Swallow and CT were performed and the suspected diagnosis of Forestier Disease was confirmed.

2. Introduction

Forestier disease, also known as Diffuse Idiopathic Skeletal Hyperostosis (DISH) is a rheumatological condition, mainly characterized by calcification and subsequent ossification of ligaments, joint capsules and entheses; this leads, especially in the axial skeleton, to the calcification and ossification of the Anterior Longitudinal Ligament, with the formation of large, confluent osteophytes on the anterior aspect of the spine, especially at cervical and dorsal level [1-4].

DISH much more frequent in males than females and its frequency increases with age. It is only rarely observed before the age of 45, and its prevalence over the age of 65 is ca. 8–10% [5].

This disease, also known as such as spondylosis hyperostotica, spondylitis ossificans ligamentosa, senile ankylosing hyperostosis, physiological vertebral ligamentous calcification, was first described by Forestier and Rotes-Querol in 1950 [7], who established the clinical and radiological criteria for diagnosis, while the disease was named as DISH in 1975, by [5], who outlined the extra-spinal radiological signs and symptoms of the disease.

In fact, while in our case, dysphagia was the main, albeit not only symptom, in all patients with DISH; other symptoms intervene to worsen the patient's clinical condition, such as laryngeal stridor, dyspnea, snoring and hoarseness.

3. Discussion

Our patient is a 74-year-old man with a gradually worsening pharyngeal dysphagia with globus sensation, occasional intra-deglutitory coughing, hoarseness and a 5 kg weight loss in the previous two months, his only declared comorbidity being type II Diabetes Mellitus.

He had previously been seen in a GI private practice, where, after an initial suspect diagnosis of gastroesophageal reflux, an esophageal barium swallow was executed and deemed negative. After two months, a considerable weight loss and a sudden worsening of the dysphagia, he was seen in our institution's GI practice and referred to the Radiology department to have a complete barium swallow with deglutition study.

The differential included all conditions responsible for an oropharyngeal dysphagia, especially neurological/functional diseases; locoregional fistulas, after having had a recent case with similar clinical

findings [9], had to be ruled out, too.

It was immediately clear that, even on the preliminary lateral plain films for functional assessment; there was calcification, ossification and hyperostosis of the Anterior Longitudinal Ligament (ALL), from C2 downwards, especially at the C3-C4 level, where osteophytosis was particularly evident.

We then performed our dynamic barium swallow study which confirmed a diagnosis of DISH, satisfying all radiological criteria for diagnosis: florid, flowing ossification along the anterior aspects of at least four contiguous vertebrae, disc spaces well preserved, with no sacroiliitis or facet joint ankylosis (Figure 1). The anterior osteophyte at the C3-C4 level abutted directly into the pharyngeal mucosal profile, thus considerably restricting the caliber of the pharynx at that level (>1 cm), especially during the pharyngeal phase of deglutition (Figure 2). Other radiographic abnormalities were noted before, during and after deglutition, such as pre-deglutition drooling and oral incontinence, with pooling of contrast into the glossoepiglottic folds and phenomena of both laryngeal penetration and tracheal aspiration of barium (Figure 1), slow epiglottic tilting (Figure 2) and, again, pooling of barium into the glossoepiglottic folds after deglutition (Figure 3), with occasional laryngeal penetration. While the clinical condition of the patient (dysphagia, globus, weight loss) may be attributed mainly to DISH, a few of the additional findings (drooling, contrast pooling, penetration, aspiration) underline the presence of neurological superimposing factors that worsen DISH itself and are to be considered separately from a therapeutic point of view.

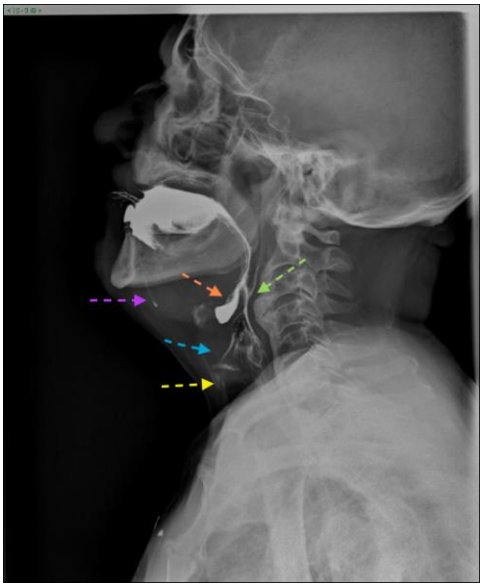

Figure 1: Pre-deglutition phase. Ossification of the Anterior Longitudinal Ligament (green arrow), with normal intervertebral spaces, abutting into the pharyngeal lumen, which is narrowed. Posterior leaking of barium with pooling (orange arrow) into the glossoepiglottic folds, laryngeal penetration (blue arrow) and slight tracheal aspiration (yellow arrow). Drooling (purple arrow) is also observed.

Figure 2: Pharyngeal phase of deglutition. Important narrowing of the pharyngeal lumen (green arrow) with difficult and slow passage of contrast into the esophagus. Laryngeal penetration is still evident, due to a slow and uncoordinated epiglottic tilting.

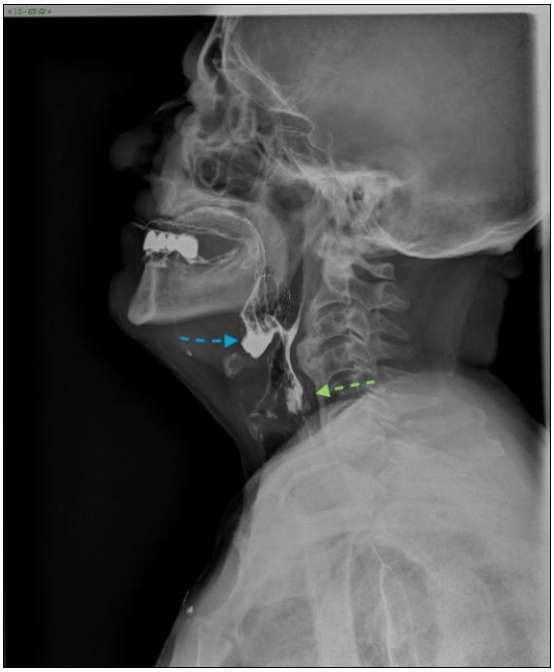

Figure 3: Post-deglutition phase. Pooling of contrast into the glossoepiglottic folds (blue arrow) with occasional downwards leaking and laryngeal penetration (green arrow). These findings, along with the slight tracheal aspiration are responsible for the hoarseness and coughing of the patient.

A dedicated CT study of the spine was performed two days later and confirmed complete ossification of the ALL, from C2 to the sacrum and calcification/ossification of the interspinous ligament, especially at the dorsal spine, a finding peculiar to this case (Figure 4).

Figure 4: Sagittal CT scan of the spine shows the calcification of the ALL (thin arrow) and of the interspinous ligament (thick arrow), a finding peculiar to this patient.

4. Conclusion

There is no clear etiology for the disease. Many associations have been made between DISH and acromegaly, obesity, genetic factors such as hypervitaminosis A, HLA-B27, HLA-B5, HLA-A11, ankylosing spondylitis, infectious diseases and type II mellitus diabetes, the only comorbidity found in the case we discussed in this paper [4].

The spine is not the exclusive location of the disease because Achilles tendon insertion, patellar tendon insertion, plantar fascia, shoulders, olecranon and metacarpophalangeal joints may all be affected [6]. Being this disease a rheumatological pathology, mainly affecting the spine, other symptoms more related to the latter may be present, such as stiffness, back pain, tendinitis, and compression myelopathy due to ossification of both Anterior and Posterior Longitudinal Ligaments, pain due to vertebral fracture or subluxation [8].

Our aim in this case report is not only to show Forestier disease as a potential cause of dysphagia, but to underline the importance of careful execution and report of barium swallow, which must always include the oro-pharyngeal phase of deglutition, as pointed out in Ekberg's seminal work on dysphagia [10], in which the radiologic examination is proposed as a direct extension of the clinical and neurological examinations and the importance of a dedicated radiologist is crucial to avoid imprecise or delayed diagnosis.

References

1. Mader R. Current therapeutic options in the management of diffuse idiopathic skeletal hyperostosis. Expert Opinion Pharmacother. 2005; 6: 1313-8.
2. Sarzi-Puttini P. New developments in our understanding of DISH. Curr Opin Rheumatol. 2004; 16: 287-92.
3. Mader R, Sarzi-Puttini P, Atzeni F, Olivieri I, Pappone N, Verlaan JJ et al. Extraspinal manifestations of diffuse idiopathic skeletal hyperostosis. Rheumatology. 2009; 48: 1478-81.
4. Mader R, Verlaan J, Buskila D. Diffuse idiopathic skeletal hyperostosis: clinical features and pathogenic mechanisms., Nat Rev Rheumatol. 2013; 9: 741-50.
5. Resnick D, Shaul SR, Robins JM. Diffuse idiopathic skeletal hyperostosis (DISH): Forestier's disease with extraspinal manifestations. Radiology. 1975; 115: 513-24.
6. Mader R. Clinical manifestations of diffuse idiopathic skeletal hyperostosis of the cervical spine. Semin Arthritis Rheum. 2002; 32: 130-5.
7. Forestier J and Rotes-Querol J. "Senile ankylosing hyperostosis of the spine." Annals of the rheumatic diseases 1950; 9:321-30.
8. Aydin E, Akdogan V, Akkuzu B, Kirbaş I, Ozgirgin ON. Six cases of Forestier syndrome, a rare cause of dysphagia,Acta Oto-Laryngologica. 2006; 126: 775-8.
9. Fontanella G. Tracheoesophageal Fistula in Chemo-Radio Treated Mediastinal Bulky Non-Hodgkin Lymphoma. Japanese Journal of Gastroenterology and Hepatology. 2020; 3: 1-3.
10. Ekberg O. Radiology of the Pharynx and the Esophagus, Spring- er-Verlag Berlin Heidelberg. 2004; 23.