Image Report

Clinical and radiological features of Forestier’s disease presenting with dysphagia

Giuseppe Roberto Giammalva, Domenico Gerardo Iacopino, Francesca Graziano, Carlo Gulì, Maria Angela Pino, Rosario Maugeri

Unit of Neurosurgery, AOUP "Paolo Giaccone", Postgraduate School in Neurosurgery, Department of Experimental Biomedicine and Clinical Neuroscience, School of Medicine, University of Palermo, Palermo, Italy

E-mail: *Giuseppe Roberto Giammalva ‑ robertogiammalva@live.it; Domenico Gerardo Iacopino ‑ gerardo.iacopino@gmail.com; Francesca Graziano ‑ francesca.graziano03@unipa.it; Carlo Gulì ‑ carlogul@yahoo.it; Maria Angela Pino ‑ mariangelapino@live.it; Rosario Maugeri ‑ rosario.maugeri1977@gmail.com
*Corresponding author

Received: 05 July 2018  Accepted: 02 August 18  Published: 28 November 18

Abstract

Background: Diffuse idiopathic skeletal hyperostosis (DISH), also known as Forestier's disease, is a rheumatologic condition characterized by ossification of the spinal ligaments and tendons. Large anterior osteophytes are typically present in the lower cervical levels, while upper cervical ossification resulting in dysphagia is very rare.

Methods: Here, we presented a patient with Forestier's disease involving massive ossification of the anterior longitudinal ligament extending from C3 to C4 downward contributing to severe dysphagia.

Results: A 65-year-old male presented with cervical pain and dysphagia. The computed tomography of the cervical spine demonstrated massive anterior longitudinal ligament ossification (DISH) extending from C3 to C7. There was an additional large osteophyte at the C3-C4 level, and also a high-grade intracanalicular C6-C7 cervical stenosis due to ossification of the posterior longitudinal ligament. The patient was offered surgical intervention (e.g., resection of the C3-C7 anterior DISH and anterior cervical discectomy/fusion at the C6-C7 level), but he declined.

Conclusions: When conservative management fails to resolve severe dysphagia for cervical DISH/Forestier’s disease, anterior surgical resection is typically performed. In this case, the patient refused surgery and opted for conservative management strategies.

Key Word: Cervical spine, diffuse idiopathic skeletal hyperostosis, Forestier’s disease, non-surgical options

Access this article online

Website: www.surgicalneurologyint.com
DOI: 10.4103/sni.sni_223_18
Quick Response Code:
INTRODUCTION

Diffuse idiopathic skeletal hyperostosis (DISH), also known as Forestier’s disease, is a rheumatologic condition characterized by ossification of the spinal ligaments and tendons. It occurs in up to 30% of the population, but is mostly seen in patients between the ages of 60 and 80.\textsuperscript{[1,2]} Cervical DISH usually appears as large anterior longitudinal ligament osteophytes involving the lower cervical spinal canal; upper cervical DISH resulting in dysphagia is very rare. Here we presented a patient who exhibited C3-C7 anterior cervical DISH with dysphagia, accompanied by C6-C7 intracanalicular cord compression due to ossification of the posterior longitudinal ligament.

METHODS

Clinical history
A 65-year-old male presented with cervical pain and dysphagia (e.g., inability to eat solid food for the past year). The neurological examination revealed no motor or sensory deficits; the patient only exhibited diffuse hyperreflexia to the upper and lower extremities, without Babinski responses.

Radiographic findings
The computed tomography of the cervical spine demonstrated massive anterior longitudinal ligament ossification extending from C3 to C7 (DISH); there was an additional large osteophyte at C3-C4, and high-grade C6-C7 intracanalicular cervical stenosis due to ossification of the posterior longitudinal ligament [Figures 1–4]. The magnetic resonance imaging study confirmed these lesions (e.g., showing displacement of the larynx and trachea anteriorly, and a kinked esophagus) [Figure 5]. Based on these findings, the patient was diagnosed cervical DISH, also known as Forestier’s disease.

Refusal of surgery
Despite the dysphagia attributed to C3-C7 DISH and C3-C4 osteophyte, along with C6-C7 ossification of the posterior longitudinal ligament (OPLL), the patient refused surgery. Rather, the patient opted for conservative management, and was discharged on steroids and postural therapy.

DISCUSSION

The diagnosis of Forestier’s disease or DISH is primarily radiological; its etiology is still unknown.\textsuperscript{[3]} Three radiologic criteria are utilized to diagnose DISH: continuous ossification of the anterior longitudinal ligament involving at least four contiguous vertebral
bodies, lack of intervertebral ankyloses or fusion, and preservation of intervertebral disk height.\cite{8} DISH typically involves the lower cervical levels, and only rarely is seen in the upper cervical spine. Often DISH is asymptomatic, and is just incidentally diagnosed on cervical computed tomography and X-ray examinations.\cite{9} However, some cases of massive DISH cause dysphagia (up to 6%), dysphonia, and dyspnea.\cite{1,2,5,9,10} Dysphagia is usually attributed to DISH involving ossification of the anterior longitudinal ligament with ventral osteophytes; this is variously attributed to an inflammatory process, neuropathy, and limitation of pharyngolaryngeal motion. Here, the massive C3-C7 DISH combined with the large cervical osteophyte at C3-C4, near the anatomical anchorage of the esophagus, likely explained this patient’s severe dysphagia.

### Treatment of diffuse idiopathic skeletal hyperostosis

The initial treatment of cervical DISH is typically conservative therapy, for example, the use of anti-inflammatory medication, steroids, muscle relaxants, and postural education to optimize swallowing.\cite{10} Surgery should be considered when conservative management fails and/or in case of severe dysphagia. Common surgical management includes anterior cervical osteophytectomy alone.\cite{1} However, subsequent anterior cervical fusion may be necessary if instability results.\cite{1,4,6,7,9} In the case presented, the patient refused surgery, and was discharged on steroids to pursue postural therapy to facilitate swallowing.

### Financial support and sponsorship

Nil.

### Conflicts of interest

There are no conflicts of interest.

### REFERENCES

1. Carlson ML, Archibald DJ, Graner DE, Kasperbauer JL. Surgical management of dysphagia and airway obstruction in patients with prominent ventral cervical osteophytes. Dysphagia 2011;26:34-40.
2. Egerter AC, Kim ES, Lee DJ, Liu JJ, Cadena G, Panchal RR, et al. Dysphagia secondary to anterior osteophytes of the cervical spine. Global Spine J 2015;5:678-83.
3. Forestier J, Rotes-Querol J. Senile ankylosing hyperostosis of the spine. Ann Rheum Dis 1950;9:321-30.
4. Giammalva GR, Iacopino DG, Maugeri R. Natura abhorret a vacuo. Future perspectives of autologous fibrin glue. Is it time for reappraisal? Surg Neurol Int 2017;8:57.
5. Goh PY, Dobson M, Iseli T, Maartens NF. Forestier’s disease presenting with dysphagia and dysphonia. J Clin Neurosci 2010;17:1336-8.
6. Graziano F, Certo F, Basile L, Maugeri R, Grasso G, Meccio F, et al. Autologous fibrin sealant (Vivostat®) in the neurosurgical practice: Part I: Intracranial surgical procedure. Surg Neurol Int 2015;6:77.
7. Maugeri R, Giammalva GR, Graziano F, Iacopino DG. May autologous fibrin glue alone enhance ossification? An unexpected spinal fusion. World Neurosurg 2016;95:611-2.
8. Resnick D, Niwayama G. Radiographic and pathologic features of spinal involvement in diffuse idiopathic skeletal hyperostosis (DISH). Radiology 1976;119:559-68.
9. Sebaaly A, Boubez G, Sunna T, Wang Z, Alam E, Christopoulos A, et al. Diffuse idiopathic hyperostosis manifesting as dysphagia and bilateral cord paralysis: A case report and literature review. World Neurosurg 2018;111:79-85.
10. 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;11:1058-67.