

Diffuse Idiopathic Skeletal Hyperostosis Causing DYSPHAGIA- A Rare Presentation

S Kaushik, A Karthikeyan, P Omer, R Singh, R Nath

Citation

S Kaushik, A Karthikeyan, P Omer, R Singh, R Nath. *Diffuse Idiopathic Skeletal Hyperostosis Causing DYSPHAGIA- A Rare Presentation*. The Internet Journal of Otorhinolaryngology. 2009 Volume 12 Number 2.

Abstract

An elderly male presenting with complaints of dysphagia was found to have neck movements' restriction and was screened radiographically. He was found to have DISH and the same was excised later to give a complete relief from dysphagia for the patient.

CASE REPORT

A 60 year old male presented to the ENT outpatient department with the complaints of dysphagia to solids for 6 months duration. Dysphagia was progressive and he localized the obstruction to middle neck. He had no history of odynophagia, heartburn and regurgitation of foods.

Clinical oral and oropharyngeal examination was insignificant. Indirect laryngoscopy and fiberoptic examination of laryngopharynx didn't reveal any pathology. Cranial nerve examination was normal. The patient had restricted neck movements.

Figure 1

Fig.1: clinical photograph showing restricted anteflexion and lateral flexion of the neck



A barium swallow radiogram was done and revealed obstruction to the passage of dye opposite to junction of fifth and sixth cervical vertebra. We also found bony bridging anterior to body of fourth, fifth and sixth cervical vertebra.

Figure 2

Fig.2: barium swallow radiogram shows obstruction to the dye passage opposite C5 (white arrow) and DISH (black asterisk) bridging C4-C6 vertebrae.

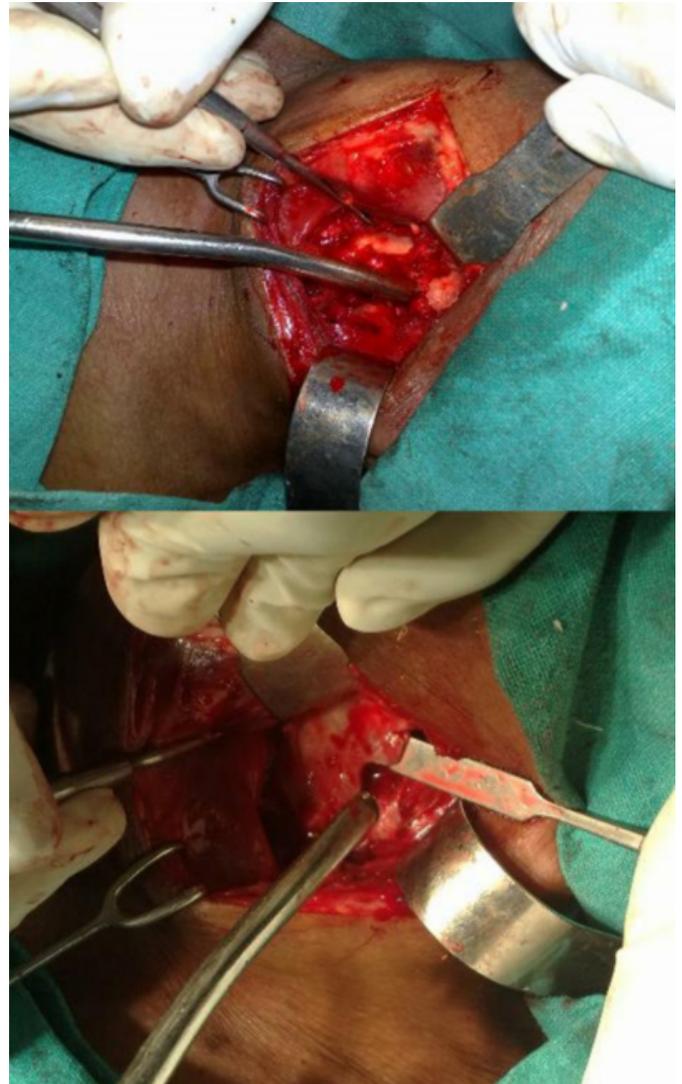


He was diagnosed to have diffuse idiopathic skeletal hyperostosis (syn: Forrester's disease) as a cause of dysphagia and was planned for surgical management.

An anterior lower neck approach was used. Under general anesthesia, the patient was placed supine with sandbag under the shoulders and head turned slightly to the right side. A horizontal neck incision was made starting in the midline at the level of the lower border of the thyroid cartilage and extended laterally upto sternocleidomastoid muscle on the left side. Subplatysmal flaps were elevated. Dissection was carried deeper and medial to sternocleidomastoid muscle and lateral to strap muscles. Internal jugular vein and carotid sheath were identified and retracted laterally. By blunt dissection medial to carotid sheath, the esophagus was identified and retracted anteriorly. Prevertebral fascia was identified and with the longus colli muscle separated and an incision made in the midline with the help of electrocautery. Calcified anterior longitudinal ligament overlying C4, C5, C6 vertebrae was seen and were removed with the assistance of spinal surgeon. Spicules were removed and a gelfoam placed over the raw bone to prevent damage to the esophagus. Ryle's tube was inserted and patient was nil per oral for 1 week postoperative.

Figure 3

Fig.3: Per-operative photograph showing use of anterior lower neck approach to the cervical spine (above) and the identified calcified anterior longitudinal ligament (below)



The postoperative period was uneventful. There was no cervical spine instability. The patient was followed up at 2 weeks after surgery and reported significant reduction in dysphagia, however there was not much significant improvement in neck movements. He was followed up monthly for the next 6 months and was completely relieved of dysphagia. There was no post-operative hoarseness of voice or oesophageal injury.

Figure 4

Fig.4: shows the resected specimen- calcified anterior longitudinal ligament.



DISCUSSION

Dysphagia can be classified into oropharyngeal and esophageal dysphagia. Esophageal dysphagia can be due to motility disorders, inadequate oropharyngeal bolus transport, inability to pressurize the pharynx, inability to elevate the larynx, discoordination of pharyngeal contraction and cricopharyngeal relaxation; and decreased compliance of the pharyngoesophageal segment secondary to muscle pathology.

Pharyngoesophageal swallowing disorders are usually because of acquired disease involving the central and peripheral nervous system. This includes cerebrovascular accidents, brain stem tumors, poliomyelitis, multiple sclerosis, Parkinson disease, pseudobulbar palsy, peripheral neuropathy, and operative damage to the cranial nerves involved in swallowing. Muscular diseases such as radiation-induced myopathy, dermatomyositis, myotonic dystrophy, and myasthenia gravis are less common causes. Rarely, extrinsic compression by thyromegaly, cervical lymphadenopathy, or hyperostosis of the cervical spine can cause pharyngoesophageal dysphagia^[1].

Forrestier's disease, also known as diffuse idiopathic skeletal hyperostosis (DISH), is an idiopathic rheumatologic abnormality in which exuberant ossification occurs along throughout the body, but most notably the anterior longitudinal ligament of the spine. Diffuse idiopathic skeletal hyperostosis (DISH) is a degenerative musculoskeletal disease that frequently occurs in persons aged 50-75 years^[2]. The prevalence in the adult population is 12 to 28 percent with an increased incidence in patients

with obesity, gout and diabetes^[3]. It is common in Caucasians with a male: female ratio of 2:1. DISH is characterized by the presence of excessive ligamentous calcification and ossification at the spinal and extra spinal locations. The aetiology is uncertain.

Dysphagia due to osteophytic masses was first reported in 1905 by Zahn^[4], and the first surgical removal of the osteophyte was carried out in 1926 by Mosher^[5].

Although most patients with cervical spine involvement are either asymptomatic or have limited cervical spine movement and unspecified pain, possible complications described in the literature are dysphagia, stridor, dyspnea, ossification of the posterior longitudinal ligament and/or myelopathy and hoarseness.

The dysphagia is usually marked, present for solid foods, improved by anterior flexion of the neck, and worsened by extension of the neck. Concomitant symptoms may include a foreign body sensation, odynophagia, reflex otalgia, salivary stasis, dysphonia, dyspnea, sleep apnea, and aspiration^[6].

There are five possible ways that DISH causes dysphagia: (i) giant osteophytes may cause dysphagia by simple mechanical obstruction; (ii) smaller osteophytes may impinge at sites of relative immobility of the oesophagus (anatomically the oesophagus is anchored at the level of the cricoid cartilage and the diaphragm); (iii) inflammation in the immediate vicinity of the osteophyte, provoking oesophageal obstruction; (iv) pain and spasm^[7]; and (v) secondary neurological lesions, e.g. recurrent nerve palsies provoked by the hyperostosis^[8]. The most frequent level of involvement related to dysphagia is C 5-6 followed by C 4-5, C 2-3 being the least common level affected^[9].

Various conservative therapies have been described, including the use of non-steroidal anti-inflammatory drugs^[10], steroid pulses, skeletal relaxants, anti-reflux regimes and bisphosphonates^[11]. Cervical surgery has proven to be useful in the management of severely affected patients^[12] and many surgeons advocate excision combined with fusion of the affected vertebrae to reduce the incidence of relapse.

Radiographic series demonstrate that calcification of the anterior longitudinal ligament in the cervical spine continues until movement is eliminated across adjacent motion segments^[13]. Treatment for DISH is primarily conservative, nonsteroidal anti-inflammatory drugs and myorelaxants are used. But in refractory cases there are two surgical

approaches to remove osteophytes, by lateral cervical or peroral transpharyngeal ways^[14].

The management of dysphagia resulting from Diffuse Idiopathic Skeletal Hyperostosis is purely surgical. Cervical osteophylectomy may be done via an anterolateral extrapharyngeal approach (with lateral retraction of the carotid sheath and its contents and medial retraction of the laryngopharynx and the esophagus) or via the posterolateral extrapharyngeal approach (with anteromedial retraction of the of the neurovascular and aerodigestive structures). Transpharyngeal approach is also described. Concomitant cervical fusion or discectomy are not felt to be necessary. Large osteophytes also present a risk of esophageal injury during the operative exposure. The esophagus may be difficult to mobilize and somewhat adherent to other cervical fascia due to local inflammatory reaction. Re-ossification with new osteophyte formation may rarely occur and repeat operation may be indicated^[15] if dysphagia symptoms return. In a long term study done by Akhtar^[16], all the 7 patients operated by the left anterior neck approach, showed improvement in dysphagia and only 2 out of 7 showed recurrent dysphagia.

CONCLUSION

Diffuse Idiopathic Skeletal Hyperostosis, also known as Forrester's disease, is a rheumatologic abnormality with exuberant ossification, are asymptomatic except for restricted neck movements. They may present with dysphagia, mild cases of which can be managed with NSAIDS and bisphosphonates, severe cases requiring surgery. Anterior neck approach has been evaluated largely in such patients and had a good outcome. Esophageal and recurrent laryngeal nerve injury are the complications of the surgery and stabilization of the spine is not advocated. Recurrence of dysphagia after surgery was minimal and

needs revision surgery.

References

1. Charles Brunnicardi F, Dana K. Andersen, Timothy R. Billiar, David L. Dunn, John G. Hunter, Raphael E. Pollock (eds): Schwartz's manual of surgery, 8th ed. McGRAW-HILL, 2006, pp 597-598.
2. Matan AJ, Hsu J, Fredrickson BA: Management of respiratory compromise caused by cervical osteophytes: a case report and review of the literature. *Spine J* 2002; 2: 456-459.
3. Kiss C, Szilagyi M, Paksy A, et al: Risk factors for diffuse idiopathic skeletal hyperostosis. *Rheumatology (Oxford)* 2002; 41(1):27-30.
4. Zahn H. Ein Fall von Abknicking der Speiserohre durch vertebrale Ekchondrosen. *Munch Med Wochenschr* 1905; 52:1680-2.
5. Mosher HP: Exostoses of the cervical vertebrae as a cause for difficulty in swallowing. *Laryngoscope*. 1926; 36:181-182.
6. Kmucha ST, Cravens RB: DISH syndrome and its role in dysphagia. *Otolaryngol Head Neck Surg*. 1994; 110(4):431-436.
7. Masayoshi O: Dysphagia complications in ankylosing spinal hyperostosis and ossification of the posterior longitudinal ligament. *Spine*. 1993; 18:391-14.
8. Coelto C: DISH with dysphagia: a neurological mechanism? *Clin Exp Rheumatol*. 1995; 13:268.
9. Hilding DA, Tachdjian MO: Dysphagia and hypertrophic spurring of the cervical spine. *N Engl J Med*. 1960; 263:11-14.
10. Eviatar E, Harel M: Diffuse idiopathic skeletal hyperostosis with dysphagia. *J Laryngol Otol*. 1987; 101(6):627-632.
11. Fulton JD: Analgesic use of etidronate in Forestier's disease. *Lancet*. 1992; 340:1287.
12. Krause P: Cervical hyperostosis: a rare cause of dysphagia. *Eur Spine J*. 1994; 13:56-8.
13. Suzuki K, Ishida Y, Ohmori K: Long term follow-up of diffuse idiopathic skeletal hyperostosis in the cervical spine. Analysis of progression of ossification. *Neuroradiology*. 1991; 33:427-431.
14. Uppal S, Wheatley AH: Transpharyngeal approach for the treatment of dysphagia due to Forestier's disease. *J Laryngol Otol*. 1999; 113:366-368.
15. Hirano H, Suzuki H, Sakakibara T, et al: Dysphagia due to hypertrophic cervical osteophytes. *Clin Orthop*. 1982; 167:168-72.
16. Akhtar S: The management of dysphagia in skeletal hyperostosis. *J Laryngol Otol*. 2000;114:154-7.

Author Information

Sandeep Kaushik, MS

Head of the Department, Department of ENT, GSVM Medical College

Anand Karthikeyan, MS

Junior Resident-final year, Department of ENT, GSVM Medical College

Poonam Omer, MS

Junior Resident-final year, Department of ENT, GSVM Medical College

R K Singh, MS

Department of ENT, GSVM Medical College

Rajendra Nath, MS

Professor, Department of Orthopedics, GSVM Medical College