Multifactorial Dysphagia: DISH and Eosinophilic Esophagitis


* Dept of Otolaryngology, University of Iowa, ^ Dept of Internal Medicine, University of Iowa, ° Dept of Orthopedic Surgery, University of Iowa

Introduction

Swallowing is a complex, multi-stage event with oral, pharyngeal, and esophageal phases. A thorough clinical examination for swallowing complaints begins to differentiate whether the problem is due to anatomic, mechanical, or neurologic etiologies. Based on the clinical suspicion, additional tests may be beneficial, including fluoroscopic and direct imaging methods, and electrophysiologic measurements. A multi-disciplinary approach may also be advantageous, as in this case of dysphagia from diffuse idiopathic skeletal hyperostosis (DISH) and eosinophilic esophagitis (EE) which incorporated care from otolaryngology, speech pathology, orthopedic surgery, and gastroenterology.

Case Report

A 60 year old man with 5 years of progressive dysphagia and recent weight loss was referred to a tertiary care center to address prominent cervical osteophytes (Figure 1) associated with DISH (Forestier’s disease) as the presumed cause of his dysphagia. Transnasal esophagoscopy identified a mass effect by the cervical osteophyte deforming the posterior pharyngeal wall (Figure 3), as well as trachealization of the esophagus with luminal narrowing and small white lesions (Figure 4). A large thyroid mass was also incidentally identified on exam with needle biopsy identifying a follicular neoplasm. A modified barium swallow study identified vallecular and pyriform retention with overlying laryngeal penetration. Transoral endoscopy was then performed by gastroenterology with biopsy of the white lesions confirming EE; esophageal dilation was also performed, which improved but did not eliminate the dysphagia. Medical therapy with oral steroids and proton pump inhibitors failed to further improve the dysphagia. Resection of the of C3-C6 osteophytes by orthopedic surgery (Figure 2) was performed with concurrent thyroid lobectomy and eventual diagnosis of Hashimoto’s thyroiditis and benign nodule. The degree of dysphagia worsened immediately and required nasogastric feedings, but gradually improved to the point that four weeks later all intake was tolerated orally. On one year follow-up, the patient reported improvement in swallowing.

Figure 1. Pre-operative lateral cervical radiograph shows multi-level degenerative change of the cervical spine, with large anterior flowing osteophytes seen extending from C3 – C7 levels.

Figure 2. Post-operative lateral cervical radiograph shows removal of anterior paravertebral ossifications. Drains are in place.

Figure 2. Cervical osteophytes from DISH causing deformation of the posterior pharyngeal wall.

Diffuse Idiopathic Skeletal Hyperostosis

Diffuse idiopathic skeletal hyperostosis is a condition characterized by the calcification and ossification of soft tissue, with predilection for the spinal anterior longitudinal ligament in older men. The production of flowing osteophytes was first described by Forestier and Rotes–Querol in 1950. Diagnosis is based on radiographic abnormalities defined using the criteria of Resnick and Niwayama1. The role of endoscopy is controversial as it may identify an associated ulcer or filling defect, but is associated with procedural risks. The cause of DISH remains unknown, but several risk factors are implicated on the basis of its frequent association with various metabolic conditions such as dyslipidemia, hyperinsulinemia, hyperuricemia, and hypertension. The dysphagia is due to mechanical obstruction in the initial stages and later due to inflammation and fibrosis2. DISH is extremely common, affecting 12% to 28% of the adult population, but dysphagia from cervical spine osteophytosis affects only 0.1% to 6% of adults in their lifetime3. Treatment is usually conservative with physical therapy, stretching and range of motion exercises, and NSAIDs. If this is ineffective, a steroid pulse, muscle relaxant, and trial of anti-reflux medication may be prescribed. If conservative management fails or if cervical myelopathy is present, surgical decompression may be necessary.

Figure 4. Transnasal esophagoscopy demonstrates trachealization of the esophagus, luminal narrowing, and small white lesions.

Figure 5. Microscopy of the esophageal biopsy demonstrates greater than 15 eosinophils per high powered field (40X).

Eosinophilic Esophagitis

Eosinophilic esophagitis is a clinicopathologic disorder characterized by a dense esophageal eosinophilia and upper GI symptoms in the absence of gastroesophageal reflux disease (GERD), as evidenced by a normal pH monitoring study or lack of response to proton-pump inhibitor treatment4. It has received a high level of research and clinical interest in the gastroenterology literature since it was first described in 1978 by Landres et al. In contrast, there has been a paucity of reports in otolaryngology even though EE may cause airway complaints as well as dysphagia5. In one report, failure of laryngotracheal reconstruction was attributed to untreated EE6.

EE is predominantly seen in Caucasian males, with a 3:1 male to female ratio, but is found in all age and ethnic groups7. Whether there is increasing prevalence8 or increasing recognition9 is controversial. Adults often present with solid food dysphagia, heartburn, and food impaction, while children present with heartburn, abdominal pain, nausea, and vomiting7,9. The pathogenesis is unknown, but candidate etiologies include food and aeroallergen hypersensitivity, and an unusual presentation of GERD. Features of EE seen on upper endoscopy include transient or fixed rings, luminal narrowing, small white lesions (eosinophil microabscesses), longitudinal furrowing and shearing, and friable “crépe paper mucosa”5,9. However, there are no pathognomonic endoscopic findings, so biopsy specimens should be obtained regardless of the gross appearance of the mucosa. The degree of mucosal hyper eosinophilia to define EE is controversial, but greater than 15 eosinophils per high-powered field has been the minimal criteria.

The treatment for EE is controversial with no randomized, placebo-controlled studies in adults, and only one study in a pediatric population. In a recent review of therapeutic trials for EE, Bohm and Richter9 suggested starting with allergy testing and a restrictive diet where indicated. Restrictive and elemental diets have been particularly effective in children10. Next are two options: swallowed or systemic steroid therapy for 4–8 wk, or dilation with PPIs to address possible co-existing GERD9. Because adults likely have both an inflammatory and a fibrotic component to their disease, esophageal dilation may be necessary to prevent recurrent food impaction. The drawback of steroids are the side effects and high relapse rate (45%10) after the steroids are discontinued, whereas dilation has a risk of esophageal perforation and does not address the underlying eosinophilic infiltration. Other therapies on the horizon include IL-5 monoclonal antibodies and leukotriene receptor antagonists.

Conclusions

Dysphagia due to DISH is rare, whereas EE is more common and can also cause airway symptoms. DISH is medically radiographically. EE should be considered when patients fail to respond to conventional reflux therapy, and requires an endoscopic esophageal biopsy. Management begins with conservative medical therapy. Surgical decompression of cervical osteophytes may be helpful for refractory DISH. New therapies are being developed for EE as more is learned about the underlying etiology. Given the myriad treatment modalities, a multidisciplinary approach may provide the most appropriate management.

References