Dysphagia in a patient with Esophageal Intramural Pseudo-diverticulosis

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Introduction:

Esophageal intramural pseudo-diverticulosis (EIPD) is a rare disorder that may present with dysphagia. The pathogenesis involves dilation of the excretory ducts of sub-mucosal mucus glands of the esophagus. The etiology is unknown. Most cases of EIPD are associated with esophageal strictures, and many cases are associated with Candida infection. The diagnosis is made either by the observation of numerous esophageal diverticula on upper endoscopy, or by barium esophogram. Treatment consists of testing for and treating underlying causes of chronic inflammation, and dilation of strictures.

Case:

A 57 year old man presented to clinic with dysphagia. He reported intermittent dysphagia to solids for 1 year, with a recent increase in severity and frequency. Now symptoms are present daily predominantly with pills and meats. He localizes symptoms to upper sternal border. He has no difficulty with liquids, no odynophagia, no weight loss, and no acute impactions. He has a 25 pack year history of cigarette use, but quit over ten years ago. He consumes 1-2 alcoholic beverages per week. He has a history of gastroesophageal reflux disease controlled with esomeprazole.

His exam was significant for obesity, but was otherwise unremarkable. A barium esophogram (Figure 1 & 2) was performed which demonstrated innumerable tiny outpouchings extending along the length of the esophagus. Upper endoscopy (Figure 3 & 4) revealed an upper esophageal stricture which was traversed after Savary-Gilliard (Wilson Cook Medical Inc, Winston-Salem NC) dilation. Beyond the stricture numerous small diverticula were seen throughout the esophagus. Biopsies were taken which were diagnostic of Candida esophagitis. The patient was treated with fluconazole and has been asymptomatic for 18 months.
Esophageal intramural pseudo-diverticulosis (EIPD) is a rare disorder first reported in 1960.\textsuperscript{1} EIPD presents predominantly in the sixth and seventh decades and is more common in males. The pathogenesis involves dilation of the excretory ducts of esophageal submucosal mucus glands.\textsuperscript{2} The etiology is unknown but leading theories include: 1) ductal occlusion by mucus, inflammatory debris, or desquamated epithelium leads to ductal dilation. 2) extrinsic compression of ducts due to mucosal fibrosis and/or inflammation leads to ductal dilation.\textsuperscript{2,3}

The most common presenting symptom is dysphagia which may be constant or intermittent. Dysphagia may be present even in the absence of stricture. Esophageal strictures are associated with 90\% of cases. Strictures most commonly occur in the cervical and upper thoracic regions of the esophagus. Strictures are thought to form as a result from localized peridiverticulitis or a secondary consequence of diffuse esophagitis.\textsuperscript{4} Candidiasis is associated with 34-48\% of cases. It is unclear if Candida is causal or found incidentally. Other associations include diabetes mellitus, reflux esophagitis, and chronic alcohol abuse.\textsuperscript{2}

Diagnosis is based on radiographic or endoscopic evaluation. Barium esophogram shows numerous tiny flask or collar button-shaped outpouchings. Computer tomography shows thickening of the esophageal wall and diffuse irregularity of the esophageal lumen. Upper endoscopy reveals small outpouchings in the esophageal lumen usually associated with inflammation and stricture.

EIPD is a benign finding with treatment focused on eradication of the associated infection or
inflammation. In patients found to have strictures as the cause of their dysphasia dilation has been shown to have quick and lasting relief of symptoms in about 96% of cases.

This case demonstrates the correlation of endoscopy and barium esophogram in a patient with EIPD and associated Candida infection. When EIPD is encountered causes of chronic esophagitis such as reflux and Candida should be assessed for and treated.

References:


