

Esophageal Intramural Pseudodiverticulosis: An Unusual Cause of Weight Loss in a 70 Year Old Male

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Abstract

Esophageal intramural pseudodiverticulosis is an uncommon condition encountered mostly in elderly males with a variety of symptoms- dysphagia being the most common. The presence of a benign esophageal stricture is usually seen. Association of this condition with gastroesophageal reflux disease and chronic alcohol intake has been reported. Therapy is usually aimed at treatment of the underlying condition. Upper gastrointestinal endoscopy and barium esophageogram remain valuable tools for diagnosis. The authors present the case of a seventy year old male with reflux symptoms and weight loss who was detected to have esophageal intramural pseudodiverticulosis. We opted for a reduced duration of proton pump inhibitor therapy, whilst introducing baclofen- which increases the lower esophageal pressure thereby reducing reflux events. The patient responded well to the administered therapy.

Keywords: Pseudodiverticulosis; Intramural; Stricture; Dysphagia; Benign

Introduction

Esophageal intramural pseudodiverticulosis (EIPD) - an uncommonly encountered condition with an unclear etiology is characterized by the presence of multiple small flask-shaped esophageal outpouchings protruding from the lumen to the wall and pathological abnormal cystic dilatation of the submucosal glands. The incidence of EIPD remains unknown, although an estimated prevalence of 0.15% has been reported [1]. Commonly encountered in elderly males, it is often benign but associations with conditions such as esophageal candidiasis, alcohol consumption, diabetes mellitus, achalasia cardia, gastroesophageal reflux disease, eosinophilic esophagitis and esophageal neoplasms are known. About 230 cases of EIP have been reported world-wide [2]. The predominant symptoms are dysphagia often accompanied by an esophageal stricture and chest pain although a small subset of patients may experience weight loss or remain asymptomatic.

Direct visualization of small diverticular orifices during routine endoscopy may not always be possible³, and hence adjuvant diagnostic tools such as low density barium esophageogram, computerised tomography and endoscopic ultrasound may be useful in special situations.

Therapies are often directed at the likely associated conditions and stricture dilatation is often reserved for patients with progressive severe dysphagia which may necessitate repetition of the procedure.

Case Report

A seventy year old hypertensive south Indian male presented to us with history of chest discomfort, reflux symptoms since six years and weight loss of seventeen kilogrammes over one year. There was no history of dysphagia, nausea, vomiting, gastrointestinal bleeds or odynophagia. He was a smoker with 40 pack years exposure. He had no family history of diseases. He had been treated by multiple physicians on an out-patient basis with proton pump inhibitors (PPIs) over a six year period.

Physical examination revealed a moderately built and nourished, ill-looking individual, with normal sensorium. Oral hygiene was poor and the mucosa and teeth were tobacco stained. Examinations of the cardiovascular, respiratory, gastrointestinal and central nervous system were normal.

Investigations such as Complete blood count, viral serology, liver and renal function test, ultrasonogram of the abdomen, chest X- ray, stool microscopy, electrocardiogram (ECG) and pulmonary function test were normal.

An upper GI endoscopy (OGD) as well as colonoscopy was planned. During OGD, multiple small pseudodiverticular orifices measuring 2 to 3 mm were noted through the entire length of the esophagus (Figure 1). No impaction of food material was observed. An esophageal stricture was not present. Linear mucosal breaks in the lower esophagus compatible with gastroesophageal reflux disease (GERD) Los Angeles class B were seen. Esophageal biopsy performed revealed chronic superficial esophagitis. Barium contrast esophageogram helped us identify small mucosal outpouchings with contrast enhancement parallel to the esophageal wall. Contrast enhanced computed tomographic scan of the thorax demonstrated esophageal wall thickening and small intramural gas collections. Colonoscopy was done and was normal.

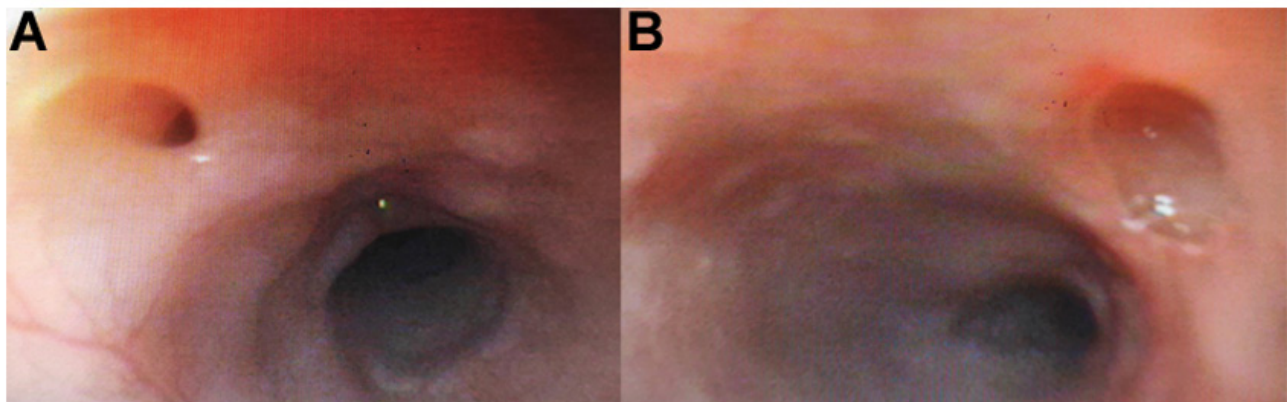


Figure 1A and 1B: Pseudodiverticular orifices seen in mid esophagus.

He was prescribed a combination therapy of Ilaprazole and Baclofen (both at 10 mg twice daily) for one month and thereafter continued on Baclofen monotherapy once daily for one three weeks and then alternate day therapy for four weeks. He reported complete amelioration of symptoms and also gained eight kilogrammes in weight. Six months later, an OGD was performed which revealed the continued presence of diverticulae and resolution of GERD. After a period of twelve months from the initial visit, the patient had occasional episodes of reflux which were responsive to intermittent baclofen therapy.

Discussion

Approximately 230 cases of EIPD have been reported since Mendl and co-workers first described the condition in 1960 [2,4]. A male preponderance exists with a sex ratio of 3 males to every 2 females among the affected. Although frequently diagnosed during the sixth and seventh decades of life, the mean age of the patients at the time of diagnosis stands at 53.5 years (range, 0.75 to 86 years) [5].

Although EIPD is characterized histologically by the cystic dilatation of multiple submucosal esophageal mucus glands and distinct excretory ducts thereby producing 1 to 3 mm flask shaped diverticulae with pinpoint mouths distributed linearly along the esophageal wall - more so proximally, the exact pathogenesis and mechanism remains unclear. The intramural cysts extend 3 mm or less beyond the esophageal lumen. The dilated ducts are lined by stratified squamous epithelium, which may appear hyperplastic. The lumen may contain desquamated squamous cells or inflammatory cells. Nonspecific acute or chronic inflammation often surrounds the acini and the ducts and may lead to subsequent submucosal fibrosis or stricture formation. Authors of reports which have documented the association of EIPD with achalasia or esophageal webs have attributed esophageal motility disorders as the causation while others have theorised an obstructive etiology of the glandular ducts due to desquamated epithelium, inflammatory cells, or submucosal fibrosis, or a combination of these leading to myoepithelial dysfunction [6].

A majority of EIPD patients commonly present with complaints of dysphagia, and roughly half of the patients have constant symptoms while 24% and 9% have intermittent and progressive symptoms respectively [7]. The presence of a benign esophageal stricture may occur in 80 - 90% of EIPD cases but dysphagia can occur independent of it as well [8]. Others present with symptoms resembling GERD, chest tightness, vomiting, chest pain which may be mistakenly attributed to cardio-pulmonary disease. A few patients present with hematemesis, malena or weight loss due to anorexia. The association of EIPD with GERD, Eosinophilic esophagitis, Achalasia, Crohn's disease, corrosive injury, tuberculosis and Mallory-Weiss syndrome have been reported [9,10]. A higher frequency is observed in patients with diabetes mellitus and chronic alcoholism [3,9]. In our case, the presence of EIPD may be linked to GERD, as no other motility disorders were present.

Upper GI endoscopy and radiographic evaluation are vital for the diagnosis of EIPD. Barium esophagogram (Figure 2), which remains a highly sensitive tool - will show numerous tiny flask or collar button-shaped outpouchings consistent with pseudodiverticula in longitudinal rows, parallel to the longitudinal esophageal axis [11]. Bridging between adjacent pseudodiverticula may occur, resulting in distinct intramural tracking. CECT thorax might show esophageal wall thickening, diffuse luminal irregularity, and intramural gas collection [12]. An endoscopic ultrasound (EUS) might reveal multiple hyperechoic images in the esophageal wall consistent with intramural gas. Direct visualization of the pseudodiverticular orifices may be challenging for endoscopists and it is reportedly seen in approximately 20% of the cases [8]. The orifices may be better visualised during slow endoscopic withdrawal rather than during insertion.



Figure 2: X-ray Barium swallow showing Pseudodiverticulae of mid esophagus.

A vast majority of EIPD patients are often treated with PPIs, and in many cases- the therapy lasts for decades and is directed towards the assumed etiology rather than the pseudodiverticulosis itself. In our patient, we opted to limit the usage of PPIs and added on baclofen which reduces the frequency of transient lower esophageal sphincter relaxations and increases the lower esophageal sphincter pressure [13]. The resultant resolution of GERD symptoms and confirmation by endoscopy shows us that Baclofen may be successfully used in individuals with EIPD with GERD. Our patient also did not have an esophageal stricture, and his complaints of weight loss could not be attributed to any other condition and he regained some weight after initiation of anti-reflux therapy. Sucralfate has also shown to be effective in some patients who are intolerant to PPI therapy [14].

In those patients with esophageal strictures, necessitating serial dilatations, iatrogenic localised perforations due to endoscopic manipulation can cause anterior mediastinitis and pneumomediastinum and hence dilatations should be performed by highly experienced endoscopists. Balloon dilatation is preferred over rigid dilators. Complications such as mediastinal abscess, malignancy, mediastinitis, esophageal perforation, progressive stenosis, esophageal structuring, bleeding, broncho-esophageal and esophageo-mediastinal fistula, peridiverticulitis, pleural and pericardial effusion are rare and may sometimes necessitate esophagectomy [15]. Although EIPD is often associated with benign strictures of the esophagus, some authors have suggested a higher risk of esophageal carcinoma in EIPD patients and recommend periodic screening [16].

Conclusion

EIPD often presents with symptoms of dysphagia and the presence of benign esophageal strictures occurs in a majority of cases. Diagnosis is usually by an OGD or barium esophageography. Therapy is often directed at the underlying cause and consists of PPIs and anti-reflux therapy. Esophageal stricture dilatation is often done in individuals with progressive dysphagia. Although, complications are rare, routine follow up is recommended. In addition to PPI therapy, treatment with Baclofen and sucralfate may be helpful in patients with GERD associated with EIPD.

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