

Esophageal Intramural Pseudo-Diverticulosis - A Case Report

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Introduction

Esophageal intramural pseudo diverticulosis (EIPD) is a rare benign pathology which is characterized by multiple small flask shaped evaginations of the esophageal wall, usually with the prominent symptoms of dysphagia in 80% of the patients [1]. The exact pathophysiology is uncertain but in one study, 26 postmortem studies revealed that these lesions represent cystic dilation of sub mucosal gland ducts [2]. Based on the literature review this entity is more common in men. Most cases are associated with esophageal strictures, gastro esophageal reflux disease and less commonly with esophageal candidiasis, achalasia, Crohn's disease, Diabetes and Alcoholism. The common finding in endoscopy is the distinctive small outpouchings on direct visualization. However, Barium esophagogram is valuable in diagnosis as literature has suggested that endoscopy might be able to detect orifices in only 20% of the patients [3].

Case Report

Patient is a 52-year-old African-American male with past medical history of esophageal candidiasis, hypertension, hyperlipidemia, benign prostatic hyperplasia who presented to the Emergency room after having 4-5 episodes of severe hematemesis. The symptoms started 2 days earlier when he started having nausea that gradually worsened. It was associated with moderate epigastric pain which was non-radiating and increased with the oral intake. Patient also reported 1 episode of dark tarry stool, although he denied any hematochezia or bright red blood per rectum. Patient also denied any headache, dizziness, chest pain, alcohol or NSAIDs use. An endoscopy performed in 2009 showed severe candida esophagitis with extensive white clumpy exudate throughout the esophagus which was treated with antifungals.

In Emergency room, patient required fluid resuscitation however the physical examination was completely unremarkable. Lab workup showed anemia which required blood transfusions. CT abdomen and pelvis reported diffuse wall thickening involving the distal esophagus. The Gastroenterology team was consulted immediately and patient was referred for endoscopy. The endoscopic examination revealed many cascading deep diverticula with the visible signs of active bleeding, at the level of distal esophagus measuring from 2-4 mm with different depths. As seen in the Figure 1, upper esophagogastroduodenoscopy reveals several small orifices indicating the presence of esophageal diverticulas. The gastro esophageal junction was normal. The diagnosis of esophageal intramural pseudo diverticulosis was established and patient was managed conservatively.

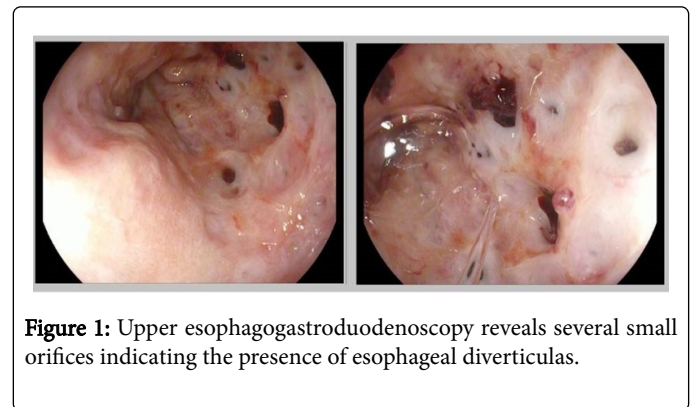


Figure 1: Upper esophagogastroduodenoscopy reveals several small orifices indicating the presence of esophageal diverticulas.

Discussion

Esophageal intramural pseudo diverticulosis, first reported in 1960 by Mendl et al. [4] is a very rare benign pathology caused by the impaired excretion of submucosal glands which ultimately leads to duct dilation giving the impression of diverticula. The exact pathogenesis is unknown, however, theories by Wightman, Lupovitch and Murney include duct occlusion by metaplasia, inflammation or elevated intraluminal pressure respectively [5]. It can be segmental in 40% (upper 14%, middle 14%, and lower 12%) or diffuse in 60% [6]. The role of Candida has been reported controversial whether it is a cause or found incidentally. However in our patient it was not found at the time of diagnosis of Esophageal Intramural Pseudo-diverticulosis (EIPD) but was treated 4 years ago which gives us a clue if Candida has been one of the causes as well.

Also, esophageal narrowing is reported in one study where the EIPD was found in 0.15% of the 14,350 patients who underwent esophagogram [7]. This is one of the reasons nearly 80% of the patients present with dysphagia for which dilation is done to relieve the symptoms. Although esophageal narrowing was also reported in our patient on CT studies, but the clinical presentation was upper gastrointestinal bleed, which is very rare, with no sign or symptoms reported for dysphagia. The EIPD has also been linked to esophageal malignancies in studies, with a statistical significant difference found between patients with an esophageal malignancy vs benign cohort ($p < 0.0002$) [8]. It also implies that esophageal cancer should be ruled out if stricture is found during an endoscopy. Since the lesion is intramural the biopsies are nonspecific and only possible from the surgical specimens.

Diagnosis is based on radiological and endoscopic examinations. Our patient presented with Upper gastrointestinal bleed which made endoscopic examination as first diagnostic and therapeutic tool.

Otherwise as reported double contrast studies are considered more sensitive in literature as compared to endoscopy which might miss the direct visualization of the small orifices. The treatment is mostly for the amelioration of the symptoms. When dysphagia due to the stricture develops, dilation is the treatment modality of choice which provides highly effective results in some cases. However pseudodiverticula persisted in majority of the cases despite symptomatic relief. For patient who present with chest pain or intermittent contractions are benefitted from esophagomyotomy or calcium channel blockers [9]. Peridiverticulitis is a rare complication associated with EPID in literature.

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