Intramural Pseudodiverticulosis of Esophagus

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Abstract
Intramural pseudodiverticulosis of esophagus is a rare disease characterized by intramural sacs communicating with the esophageal lumen. Essentially a radiological diagnosis, the condition is occasionally associated with various esophageal lesions which present as dysphagia. Esophageal stricture is a frequent accompaniment of this entity and dilatation of the stricture gives frequently lasting relief in symptoms. We report here four cases of this entity including a case with corrosive injury to the esophagus, an association not previously reported.

Key words: Intramural pseudodiverticulosis of esophagus.

Introduction
Intramural pseudodiverticulosis of esophagus is an entity associated with dysphagia and intramural sacs communicating with the esophageal lumen. The condition was described in 1960 by Mendel et al. and since then about 60 cases have been reported in the world literature. We report here for the first time from our country four cases with this condition.

Case Reports

Case 1
J, an 8 year old male, had retrosternal burning for 15 years with progressive dysphagia to solids and liquids for 6 months. There was no preceding history of ingestion of corrosive agents. Barium swallow examination showed sliding hiatal hernia, reflux esophagitis with long stricture involving the lower 1/3rd of the esophagus with "pseudodiverticuli" (Fig 1). The patient refused endoscopic dilatation of the stricture. Even after six months of antireflux treatment his symptoms persisted and barium findings remained unchanged.

Case 2
SS, a 57 year old diabetic female, had water-brash, crusting and mild epigastric burning for 2 years. She had no dysphagia. Barium examination revealed multiple pseudodiverticuli in the lower esophagus. Endoscopy showed the entire esophageal mucosa hyperemic, friable and covered with patchy white, thick membranes. Biopsy showed chronic esophagitis with fungal hyaline. Brush smear and culture yielded growth of Candida albicans. Treatment with antireflux measures, ampicillin and metronidazole alleviated her symptoms but follow up barium swallow examination six months later still showed pseudodiverticuli.

Case 3
AK, a 20 year old male, accidentally ingested sulphuric acid in February 1982. He was treated in a hospital with intravenous fluids and antibiotics and 2 weeks later was able to take solids and liquids with some difficulty, which gradually progressed to absolute dysphagia in 3 months. At this stage he presented to our hospital in a grossly undernourished and dehydrated state. Barium swallow examination showed a long stricture of the lower 1/3rd of the esophagus associated with a pseudodiverticulum (Fig 2). There was an intral stricture in the stomach. Dilatation of the esophageal stricture was done upto 45 French size with Erer-Puestow dilators and he was completely relieved of dysphagia. He refused to undergo surgery for the stricture which caused him mild postprandial epigastric fullness and early satiety. Follow up barium examinations four months and two years later revealed continued presence of pseudodiverticulum despite lack of significant symptoms. The patient gained 15 kg of weight in 2 years.

Case 4
VD, a 65 year old female, had eructations and retrosternal burning for 3 years, and intermittent dysphagia for one year. Barium series revealed hiatal hernia with stricture at the lower end of the esophagus and multiple pseudodiverticuli (Fig 3). Endoscopy showed hemorrhagic mucosa with nummular areas where a creamy exudate could be expressed out. They appeared to be openings of pseudodiverticuli. Dilatation of the esophageal stricture with Erer-Puestow dilaters upto 45 French size along with antireflux measures relieved her symptoms, but the pseudodiverticuli persisted even two months later.

Discussion
Originally described as intramural diverticulosis, this condition was later renamed as pseudodiverticulosis because: (i) the whole thickness of the esophageal wall is not involved in the diverticular process, and (ii) the diverticuli develop from dilatation of pre-existing excretory ducts of the esophageal glands and thus are not true outpouchings of the esophageal wall. The condition occurs usually in persons above 60 years of age. Only 5 cases have been described in children between 5 and 15 years of age. Dysphagia, the most common symptom, occurring in 95-100% of cases, was present in 3 of our 4 cases. Dysphagia is usually constant but occasionally intermittent or progressive. Symptoms may run for several months to years (range 3 days to 25 years).

Pseudodiverticuli on a barium swallow study are seen as small 1-2 mm sized outpouchings, either collar button or flask shaped, and these may also show branching. They may be limited to a segment of the esophagus or may involve the entire esophagus. Strictures are associated in 90% of the cases and these may be short or long. Three of our 4 cases had dysphagia and esophageal strictures of varying lengths. Pseudodiverticuli may develop before or after the development of strictures and may be present above or below the stricture. The condition may remain stable and localized for long periods or may progress to involve the entire esophagus.

Barium swallow examination is far more sensitive than endoscopy in diagnosing this entity. It is emphasized that several swallows of barium must be given to diagnose this condition on the assumption that the thick creamy exudate already collected in the pseudodiverticuli is expressed out by multiple swallows
before filling them with barium. Barium swallow study may also detect many associated conditions, viz. hiatal hernia and gastroesophageal reflux (20-30%), motility disturbances (20%), and cervical webs (11%). Two of our cases had hiatal hernia and reflux esophagitis, one had diabetes mellitus and esophageal moniliasis and one had corrosive injury to the esophagus, which has not been described as an associated factor so far.

Endoscopy reveals esophagitis and strictures in a majority of cases, but the openings of pseudodiverticuli are identified in only 20% of cases, as whitish elevations with central openings through which thick creamy white exudate is expressed or as subepithelial cysts or punctate depressions. In some cases the esophageal mucosa looks normal. In one-third of cases esophageal brushing reveals Candida albicans. One of our four cases (case 2) who was a diabetic was found to have Candida albicans.

Autopsy studies in cases of intramural pseudodiverticulosis of esophagus have shown normal external surface of the involved esophagus with thickening of the esophageal wall in the stricturous segment. Histology reveals the pseudodiverticuli to be dilated excretory ducts of mucous glands. They are lined by metaplastic squamous epithelium and are filled with desquamated squamous cells. The mucosa and submucosa are heavily infiltrated by chronic inflammatory cells with no evidence of acute inflammation. All stages of transition from normal mucous gland ducts to ducts showing complete squamous metaplasia are seen in areas adjacent to the pseudodiverticuli. The pseudodiverticuli are situated in the submucosa deeper to the muscularis mucosa and superficial to the muscular coat. The thickening of the esophagus in the region of the stricture is due to submucosal fibrosis and not because of fibrosis of the muscular coat.

Opinions regarding the etiopathogenesis of the condition vary. Based on histological findings the most likely cause appears to be chronic irritation either due to acid reflux or due to chronic infection which leads to squamous metaplasia of the esophageal glands ducts and blockage of these ducts by inflammatory cellular debris and desquamated squamous cells leading to their dilatation. Various known predisposing conditions are esophageal moniliasis, motility disturbances of the esophagus with secondary infections, diabetes mellitus, hiatus hernia with gastroesophageal reflux, esophageal webs, alkaline esophagitis following gastroenterotomy, and esophageal malignancy. Candida may perhaps be a mere secondary contaminant in areas of stasis rather than the cause of the condition.

Treatment by dilatation of associated esophageal stricture is quite effective and nearly always brings quick and lasting relief in dysphagia. Although occasionally pseudodiverticuli disappear completely or undergo reduction in their number following dilatation, in the majority of cases, they persist despite complete relief in symptoms. Associated lesions like fungal infection and hiatal hernia and/or reflex may require appropriate therapy. In 3 of our patients there was marked symptomatic improvement following dilatation of esophageal strictures (cases 3 and 4) and following antifungal and antacid therapy with control of diabetes (case 2). Case 1 did not undergo dilatation of the stricture and continued to be symptomatic despite antireflux treatment. Pseudodiverticuli, however, persist.
sisted in all cases despite relief of symptoms (in cases 2, 3 and 4).

References