

An unusual cause of dysphagia: Isolated IgG 4-related esophageal disease

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A 60-year-male presented with progressively increasing dysphagia accompanied with loss of weight. He denied ingestion of corrosive or any medication. Upper gastrointestinal endoscopy revealed a nonnegotiable stricture at 32 cm from the incisors [Figure 1] and multiple endoscopic biopsies from the mouth of the stricture were noncontributory. Barium swallow confirmed the presence of lower esophageal stricture [Figure 2]. Contrast enhanced computed tomography revealed marked wall thickening of the esophagus at the site of narrowing [Figure 3]. High-frequency endoscopic ultrasound (EUS) at 20 MHz revealed marked thickening of the wall of the esophagus with the loss of wall stratification (maximum thickness 11.2 mm) [Figure 4]. No significant mediastinal lymph nodes were detected on EUS. Endoscopic bougie dilatation of the esophageal stricture was done, and the patient had transient improvement in dysphagia. Positron emission tomography revealed flourodeoxyglucose (FDG) avid esophageal wall thickening (SUV_{max} 2.3) [Figure 5]. No significant

FDG uptake was observed anywhere else in the body. Because of the possibility of hidden malignancy, the patient underwent esophagectomy with gastric pull-up. The resected specimen revealed ulcerated esophageal mucosa and dense submucosal fibro-inflammatory reaction which had storiform pattern and IgG4 rich plasma cell infiltration [Figures 6 and 7]. Serum IgG4 levels done postoperatively were within normal limits. The patient had an anastomotic leak that was successfully managed with endoscopic dilatation and patient is asymptomatic at 8 months of follow-up.

The IgG4 related disease usually involves pancreas with the involvement of biliary tract, salivary glands, lymph nodes, thyroid, kidneys, lung, skin, prostate, and aorta also reported in the literature.^[1,2] Involvement of upper gastrointestinal tract by IgG4-related inflammatory cells is very rare and there are very few case reports

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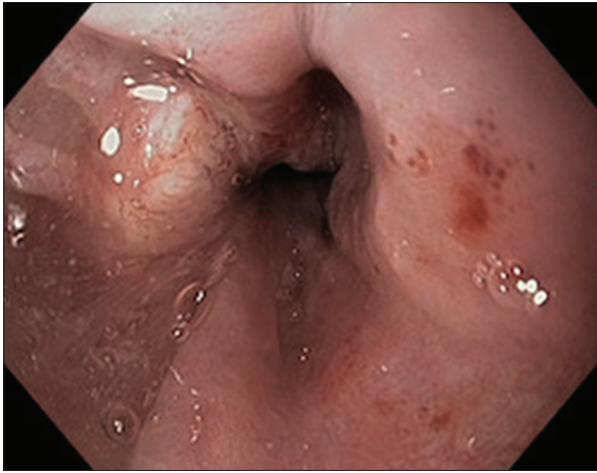


Figure 1. Upper gastrointestinal endoscopy: Nonnegotiable stricture in lower esophagus



Figure 2. Barium swallow: Lower esophageal stricture

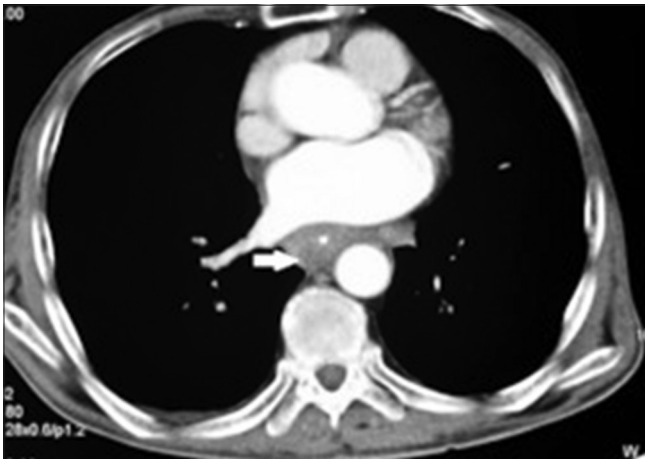


Figure 3. Contrast enhanced computed tomography: Marked wall thickening of the esophagus at the site of narrowing

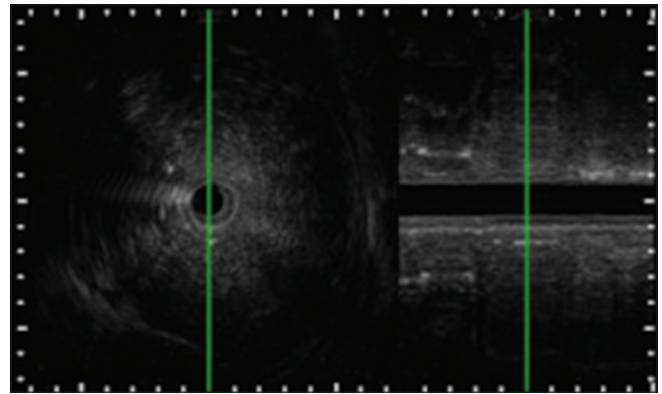


Figure 4. High-frequency endoscopic ultrasound: Marked thickening of the wall of the esophagus with loss of wall stratification

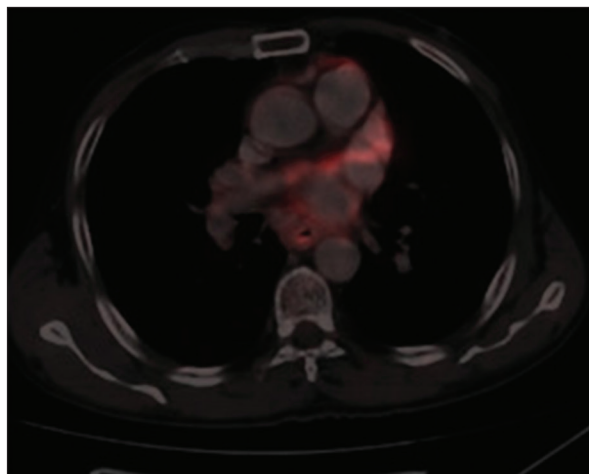


Figure 5. Positron emission tomography-computed tomography: Flourodeoxyglucose avid esophageal wall thickening (SUV_{max} 2.3)

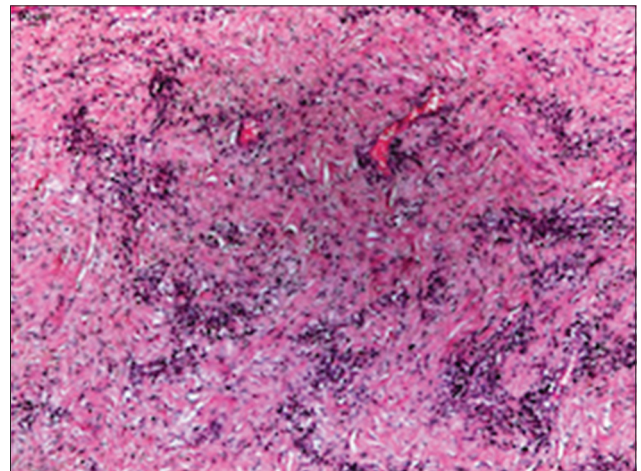


Figure 6. Microphotograph: Fibro-inflammatory reaction with storiform pattern and plasma rich infiltrate (H and E, $\times 40$)

describing IgG4-related esophageal disease with the majority of patients being treated with surgery.^[2-4]

In spite of rarity, IgG4 related esophageal disease should be considered in the differential diagnosis of unexplained esophageal strictures.

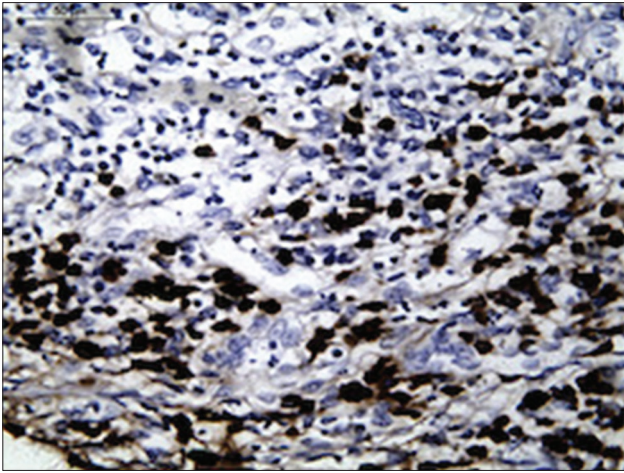


Figure 7. Immunohistochemistry: IgG4 rich plasma cells (×40)

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Conflicts of interest

There are no conflicts of interest.

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