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DISEASES OF THE ESOPHAGUS

Images in esophageal diseases

Diffuse esophageal leiomyomatosis

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SUMMARY. **Progressive dysphagia of unknown etiology may still provide a diagnostic challenge despite an increase in the number and quality of investigations available. We describe a 64-year-old man who presented with progressive dysphagia and weight loss. Following a number of investigations, a diagnosis of diffuse esophageal leiomyomatosis was made and the patient was treated appropriately.**

KEY WORDS: **benign tumor of the esophagus, diffuse esophageal leiomyomatosis, dysphagia, esophagectomy, leiomyoma.**

CASE SUMMARY

A 64-year-old male was referred with a 4-month history of intermittent, progressive dysphagia. He described 20 kg of weight loss and his medical history was significant for gastroesophageal reflux disease, smoking, and ischemic heart disease. A barium swallow and upper endoscopy had not identified a cause. A repeat barium swallow demonstrated complete obstruction at the level of the carina (Fig. 1). Subsequent computed tomography (CT) scans of the chest and abdomen (Fig. 2) demonstrated extensive esophageal narrowing with wall thickening extending from the midesophagus to the gastroesophageal junction. No enlarged lymph nodes were noted. Positron emission tomography (PET) demonstrated mildly increased uptake in the lower esophagus, inconsistent with a malignancy. Repeat upper endoscopy did not demonstrate any mucosal obstruction, but by this time, the patient could no longer tolerate oral intake. Nasoenteric feeds were commenced. Endoscopic ultrasonography showed a thickened esophageal wall but biopsies were nondiagnostic. Additional biopsies (obtained via video-assisted thoracoscopy) showed bundles of smooth muscle cells suggestive of an esophageal leiomyoma. At operation, the lower half of the esophagus was thickened and firm to touch (Fig. 3). An Ivor-Lewis esophagectomy was performed without complication. Histopathology confirmed the diagnosis of diffuse esophageal leiomyomatosis. Total tumor length was 150 mm and muscularis propria thickness was 10 mm (normal ~2 mm).

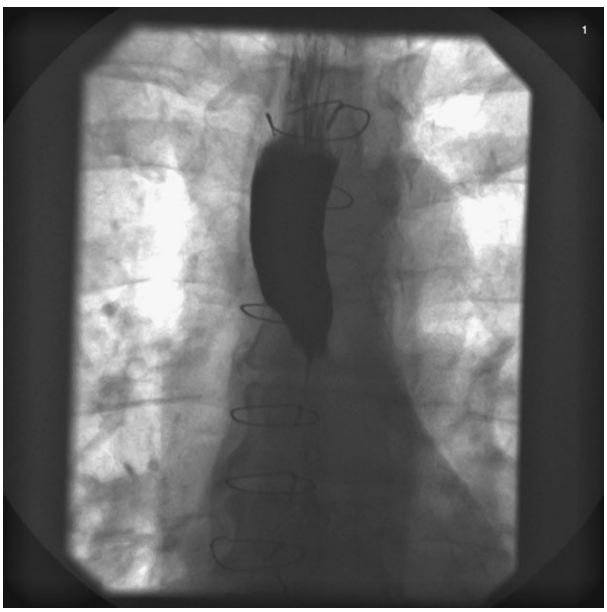

Fig. 1 Barium swallow demonstrating complete obstruction of the esophagus at the level of the carina.

Diffuse esophageal leiomyomatosis (also called corkscrew esophagus, and giant or idiopathic muscular hypertrophy) is a rare, benign tumor of the esophagus that is distinct from the more common esophageal leiomyoma (a solitary, well-circumscribed lesion).[1] In diffuse leiomyomatosis, circumferential proliferation of smooth muscle cells causes marked

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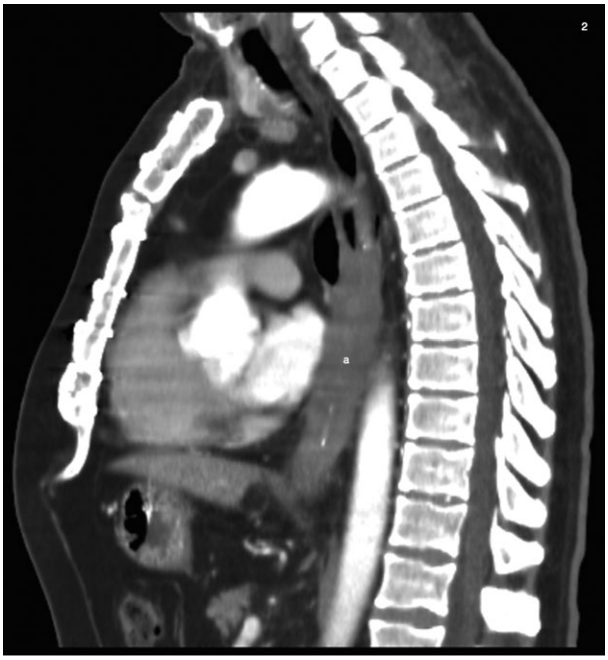

Fig. 2 Contrast enhanced computed tomography scan showing diffuse, smooth thickening of the esophagus, from the carina inferiorly to the gastric fundus.

thickening of the esophagus and subsequent dysmotility. The incidence of diffuse leiomyomatosis is unknown. The esophagus accounts for less than 10% of all gastrointestinal leiomyomas, and leiomyomas account for only 0.4% of all esophageal tumors.[2] Esophagectomy and reconstruction has replaced myotomy as the preferred treatment for diffuse esophageal leiomyomatosis due to its anatomical location in the lower (53%) and middle (35%) thirds of the esophagus.[2–5] At 12-month follow-up, our patient was well with no recurrence of dysphagia.

Acknowledgments

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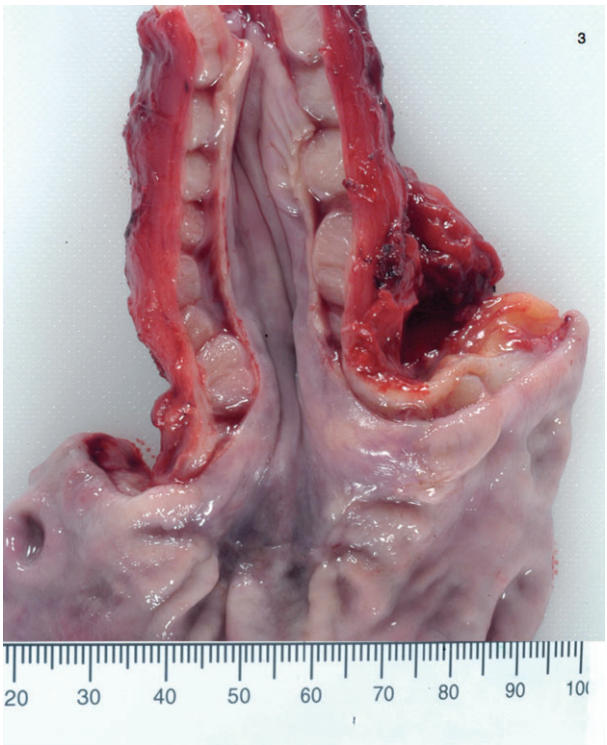

Fig. 3 Macroscopically the esophagus showed concentric rings of hypertrophy. The muscularis propria layer was 10 mm thick (normal ~2 mm).

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