

PICTORIAL CME

Esophageal Scleroderma- Changes in Esophageal Manometry

Mayank Jain



Fig. 1: Manometry

Systemic sclerosis (scleroderma) commonly involves the esophagus. It mainly causes esophageal motility disturbances which lead to gastroesophageal reflux and its complications.^{1,2}

A forty year old lady presented with history of gradually progressive dysphagia for solids and liquids for last one month. Upper gastrointestinal endoscopy done at an outside centre revealed reflux esophagitis (Los Angeles Classification-C). She was given proton

pump inhibitors for 4 weeks with which she had excellent improvement. She returned three months later with worsening dysphagia. She had lost around 2 kgs and noted skin thickening. Her appetite remained good. On high resolution manometry (Figure 1), basal lower esophageal sphincter pressures were normal and mean integrated relaxation pressures was <15. There was absent contractility in the distal 2/3 of the esophagus (smooth muscle portion). The possibility of scleroderma was considered. Anti nuclear antibodies were positive. She was started on high dose PPI with good relief in symptoms. Over a six month follow up, she noted Raynaud's phenomenon and progressive skin tightening suggesting disease progression.

Progressive systemic sclerosis (PSS) causes smooth muscle atrophy and fibrosis of the distal two-thirds of the esophagus. Motility studies show reduced-amplitude or absent

peristaltic contractions in this region and normal or decreased lower esophageal sphincter pressure. Patients complain of dysphagia, heartburn, and regurgitation due to reflux and dysmotility. Complications include strictures (17% - 29%) and Barrett esophagus (up to 37%). Symptoms correlate poorly with evidence of esophagitis or abnormal 24-hour pH recordings.³

References

1. Hostein J, Fourné J. Gastrointestinal manifestations of collagen diseases. *Digestive Diseases and Sciences* 1986; 4:240-252.
2. Lock G, Holstege A, Lang B, Schölmerich J. Gastrointestinal manifestations of progressive systemic sclerosis. *Am J Gastroenterol* 1997; 92:763-71.
3. Ebert EC. Esophageal disease in scleroderma. *J Clin Gastroenterol* 2006; 40:769-75.

Choithram Hospital and Research Centre, Indore, Madhya Pradesh
Received: 27.09.2016; Accepted: 22.08.2018