



Sigmoid Esophagus in Systemic Sclerosis: A Case series

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Background and Aims

The hallmark feature of esophageal involvement in Systemic sclerosis is absent peristalsis or ineffective esophageal motility [1]. These findings are reported in up to 70-90% of patients with Systemic sclerosis [2,3]. Sigmoid esophagus is a radiological feature commonly seen in advanced achalasia. With the widespread use of barium esophagogram and availability of High Resolution Esophageal Manometry (HREM), a variety of atypical esophageal findings are seen in systemic sclerosis. Our aim was to report these patients with systemic sclerosis with sigmoid esophagus on barium esophagogram and associated demographic characteristics and comorbidities.

Methods

Charts of Systemic sclerosis patients with sigmoid esophagus on barium esophagogram were reviewed and the following variables collected: Age, gender, race, comorbidities and findings on barium esophagogram, HREM, upper endoscopy, esophageal pH monitoring and autoantibody panel.

Results

Patient 1 is a 48 year old male who was undergoing lung transplant evaluation for Interstitial Lung Disease (ILD).

Patient 2 is a 41 year old African American female with systemic sclerosis and pulmonary fibrosis. She was undergoing lung transplant evaluation secondary to her pulmonary fibrosis.

Patient 3 is a 74 year old Caucasian female with a history significant for diabetes mellitus,

Table 1: Summary of Test Results in Systemic sclerosis with sigmoid esophagus on Barium Esophagogram. HH=Hiatal Hernia, CRP=C-reactive protein, ANA=Antinuclear antibody, IRP=integrated relaxation pressure, LES=lower esophageal sphincter, N/A= not available.

Variable	Patient 1	Patient 2	Patient 3	Patient 4	Patient 5	Patient 6
Barium Esophagogram findings:						
Dilated Esophagus	Normal	Present	n/a	Present	Present	Present
Aperistalsis	Present	Present	Present	Present	Present	Present
Delayed esophageal emptying	Absent	Absent	Present	Absent	Present	Present
Impaired LES relaxation	Present	N/A	Present	Present	Absent	Present
Reflux	Absent	Absent	Absent	Absent	Present	Absent
HREM Findings:						
Basal LES pressure (mmHg)	5	0.5	1.2	2.6	-4.9	0.3
Mean IRP of LES	2.7	1.2	3.8	12	0.3	0
Peristalsis	Absent	Absent	Absent	Absent	Absent	Absent
Motility Disorder	Absent contractility	Absent contractility	Absent contractility	Absent contractility	Absent contractility	Absent contractility
pH testing	Normal	Normal	n/a	n/a	Normal	n/a
Upper endoscopy	n/a	n/a	Normal	HH, esophagitis, Schatzki ring	HH	Markedly dilated esophagus
Anti ENA panel	n/a	Elevated CRP and ANA titers	-Elevated CRP and ANA titers -Systemic sclerosis IgG Antibody positive	-Elevated ANA titers -SS-A Antibody positive	-ANA positive -negative for antimitochondrial antibody	- Elevated CRP

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systemic sclerosis and pulmonary hypertension who presented with dysphagia.

Patient 4 is a 58 year old African American male with a history of Systemic sclerosis with skin, joints, lung and gastrointestinal tract involvement, heart failure, chronic kidney disease- stage II, pericardial window secondary to pericardial effusion who presented with dysphagia for solid foods, weight loss and failure to thrive.

Patient 5 is a 51 year old Caucasian male with a history of pulmonary hypertension, systemic sclerosis and interstitial lung disease who presents for a lung transplant evaluation.

Patient 6 is a 74 year old Hispanic female with a long standing history of rheumatoid arthritis who presents with dysphagia to solid food and heartburn.

The findings on HREM, upper endoscopy, pH testing and antibody panel are presented in (Table 1). The timed barium esophagogram showing sigmoid esophagus and delayed esophageal emptying is presented in (Figure 1).

Conclusion

All the 6 patients in this series had sigmoid esophagus on barium esophagogram but had absent contractility with low LES pressures on HREM.

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