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Systemic Sclerosis with Gastrointestinal manifestations: a unique presentation

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Abstract

Introduction: Systemic sclerosis is a chronic autoimmune disease of unknown etiology. Disease course usually begins with Raynaud's phenomenon followed by skin sclerosis and internal organ involvement. Diagnosis is made based on the clinical symptoms, presence of antibodies and endoscopy with biopsy.

Aims: We present a case of a 60-year female who was followed by GI clinic for treatment resistant dyspepsia, bloating and nausea.

Methods: Stomach biopsy revealed gastric mucosa showing focal vascular ectasia. There was significantly increased fibrosis involving the muscularis mucosae and propria. Following biopsy results, Scl-70 Ab test was performed, which was positive thus supporting the diagnosis of systemic sclerosis.

Conclusion: Full thickness gastrointestinal tract biopsies of systemic sclerosis cases are rarely seen in routine surgical pathology practice. This case is unique because of the complexity of clinical presentation requiring open gastric wall biopsy but demonstrates the value of pathologic evaluation for diagnosis of rare autoimmune disorders such as systemic sclerosis.

Background

- Systemic sclerosis (SSc) is a rare autoimmune chronic autoimmune disease of unknown etiology.
- It is characterized by fibroproliferative alterations of the microvasculature leading to fibrosis and loss of function of the skin and internal organs.
- Disease course usually begins with Raynaud's phenomenon followed by skin sclerosis and internal organ involvement.
- Gastrointestinal manifestations of SSc are the most commonly encountered complications of the disease affecting nearly 90% of the SSc population. Among these complications, the esophagus and the anorectum are the most commonly affected¹.
- The two most common manifestations of SSc in the stomach are gastroparesis and gastric antral vascular ectasia (GAVE) resulting from neuropathic damage and vasculopathy respectively¹.
- Diagnosis is made based on the clinical symptoms, presence of antibodies and endoscopy with biopsy.

Methods

- Sixty-year-old female presented at the gastroenterology clinic for treatment resistant dyspepsia, bloating and nausea.
- Past medical history was significant for fibromyalgia, Raynaud's disease and transient ischemic attack.
- For evaluation of dyspepsia, she underwent endoscopy with gastric and esophageal biopsies, which were superficial and showed only non-specific chronic inflammation.
- 24-hour pH monitoring results were unhelpful in making a definitive diagnosis.
- Esophageal manometry showed a hypertensive lower esophageal sphincter for which she underwent Heller's myotomy.
- Her symptoms persisted and she was scheduled to undergo laparoscopy and open gastric wall biopsy.

Photos

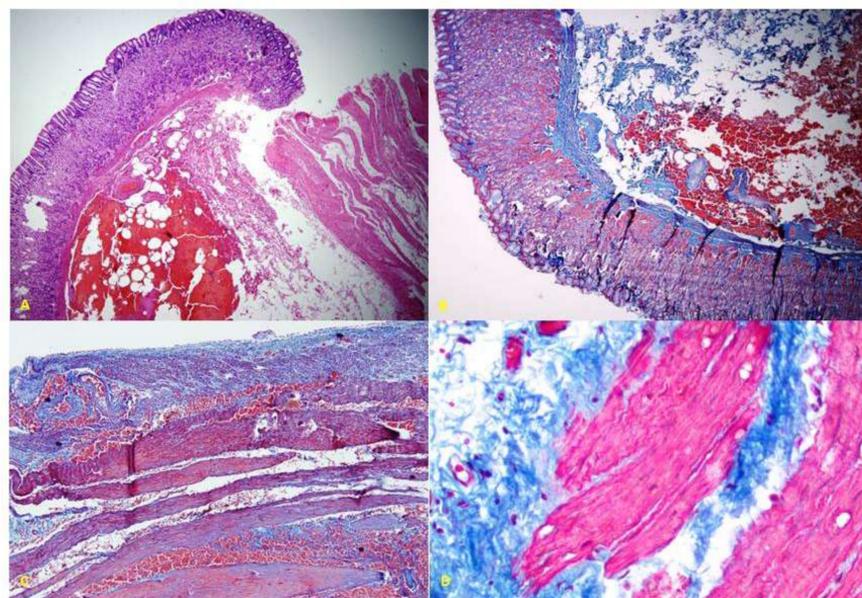


Figure A. Gastric mucosal demonstrating increased fibrosis involving muscularis mucosae and propria.

Figure B-D Trichrome stain highlighting fibrosis in muscularis mucosae and propria

Results

- Open gastric wall biopsy revealed gastric mucosa showing focal vascular ectasia.
- There was significantly increased fibrosis involving the muscularis mucosae and propria (Figure A), highlighted by trichrome stain (Figure B-D).
- Following biopsy results, Scl-70 Ab test was performed, and it turned out to be positive (42, N<20 units). Thus, supporting the diagnosis of systemic sclerosis.

Conclusions

- Full thickness gastrointestinal tract biopsies of systemic sclerosis cases are rarely seen in routine surgical pathology practice.
- While classification criteria for systemic sclerosis do not incorporate the gastrointestinal tract manifestations that are often present in this disease, it is the most common internal organ involved².
- Proper diagnostics and therapeutics for gastrointestinal involvement require the treating physician to have an understanding of an integrated approach and potential medication adverse effects.
- This case highlights the complexity of clinical presentation requiring open gastric wall biopsy and demonstrates the value of pathologic evaluation for diagnosis of rare autoimmune disorders such as systemic sclerosis.

Sample Bibliography

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