

FAYSAL ALTAHAWI

Cleveland Clinic Lerner College of Medicine
of Case Western Reserve University,
Cleveland, OH

M. CHADI ALRAIES, MD, FACP

Department of Hospital Medicine,
Cleveland Clinic

The Clinical Picture

A less common source of dyspnea in scleroderma

A 48-YEAR-OLD MAN reports progressive exercise intolerance, shortness of breath, fatigue, and melena over the past month. He has a long history of Raynaud phenomenon, and 5 months ago he developed severe sclerodactyly in both hands, diagnosed as limited cutaneous systemic sclerosis (scleroderma).

He has no chest pain, swelling of the lower limbs, change in weight, cough, fever, chills, or sick contacts, and he has not traveled recently.

His symptoms began as fatigue and shortness of breath, which worsened until he began having episodes of abdominal pain with melena and dizzy spells, although he never passed out.

He is currently taking long-term low-dose prednisone and mycophenolate mofetil (CellCept) for the systemic sclerosis, and omeprazole (Prilosec) for gastroesophageal reflux. His father had lupus, and his grandmother had colon cancer.

An outpatient workup for sclerosis-related lung and heart involvement is negative. The workup includes computed tomography of the chest, pulmonary function tests, and Doppler echocardiography.

He is afebrile, with a blood pressure of 105/60 mm Hg and a pulse of 98. His cardiopulmonary examination results are normal. He has mild epigastric tenderness without rebound or guarding. His hemoglobin concentration at the time of hospital admission is 7.8 g/dL, down from 14.5 g/dL recorded when limited cutaneous systemic sclerosis was diagnosed. Iron studies reveal iron deficiency.

He receives two units of packed red blood cells and is started on an esomeprazole (Nex-



FIGURE 1. Esophagogastroduodenoscopy of the antrum of the stomach shows the classic “watermelon stomach,” ie, the striped distribution of angioectasia, consistent with gastric antral vascular ectasia. The areas of ectasia were cauterized with argon plasma coagulation.

ium) drip for suspected upper gastrointestinal bleeding. He then undergoes esophagogastroduodenoscopy, which reveals the source of the bleeding: the classic “watermelon” distribution of angioectasia in the antrum of the stomach, consistent with gastric antral vascular ectasia (FIGURE 1).

The antral ectasia is treated with argon plasma coagulation during the endoscopic examination.

Afterward, the patient's hemoglobin stabilizes, and the melena resolves. He is discharged on an oral proton pump inhibitor, with instructions to follow up for another endoscopic session in 1 month.

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GASTROINTESTINAL FEATURES OF SYSTEMIC SCLEROSIS

Sclerodermal disorders have diverse manifestations that always include characteristic cutaneous signs. While there are several well-recognized symptomatic conditions commonly associated with scleroderma, attention must also be paid to the less common causes of these symptoms. Scleroderma has gastrointestinal complications that can easily be missed and may not respond to immunomodulatory or proton pump inhibitor therapy: complications can include esophageal dysmotility, hypomotility, gastric paresis, reflux esophagitis, strictures, drug-related ulcer, malabsorption, bacterial overgrowth, and pseudo-obstruction.¹

This patient had an underrecognized cause of dyspnea in the setting of systemic sclerosis. Vascular symptoms of limited cutaneous systemic sclerosis are typically attributed to Raynaud phenomenon; gastrointestinal symptoms are typically attributed to esophageal dysmotility; and associated dyspnea is often considered to represent pulmonary or cardiac involvement of the sclerosis. However, gastric antral vascular ectasia should be considered in any patient with scleroderma and evidence of anemia.

The prevalence of gastric antral vascular ectasia in patients with systemic sclerosis is estimated to be about 6%.²⁻⁴ It is a relatively rare cause of upper gastrointestinal blood loss that can be clinically silent until the patient develops severe iron deficiency anemia and symptoms of dyspnea, fatigue, or congestive heart failure.

Gastric antral vascular ectasia in scleroderma usually presents as iron deficiency ane-

mia, and only presents overtly as hematemesis or melena 10% to 14% of the time.⁴ Because of the often occult nature of the bleeding, the condition may be clinically silent in the early phase. Symptoms of shortness of breath and fatigue may not develop until the anemia worsens rapidly or becomes severe. Anemia is present in almost all cases of gastric antral vascular ectasia (96% to 100%) and should be a strong clinical clue for early endoscopic evaluation in patients with scleroderma, especially if there is already suspicion of upper gastrointestinal bleeding.²⁻⁵

The distinctive endoscopic streaky pattern of ectasia along the stomach antrum seen in gastric antral vascular ectasia is called “watermelon stomach”^{4,5} because the striped pattern recalls the stripes of a watermelon. The endoscopic appearance can vary, however, from the watermelon pattern to a coalescence of angiodysplastic lesions termed “honeycomb stomach,” which can easily be mistaken for antral gastritis.^{4,5} Therefore, biopsy often serves to confirm the diagnosis, with histologic features including dilated mucosal capillaries with focal fibrin thrombosis and fibromuscular hyperplasia of the lamina propria.

Gastric antral vascular ectasia often requires multiple transfusions of red blood cells, as well as repeated treatments with endoscopic argon plasma coagulation, whereby ionized argon gas is used to conduct an electric current that coagulates the surface of the mucosa to a few millimeters depth.⁴⁻⁶

A knowledge of the association between scleroderma and gastric antral vascular ectasia can lead to earlier recognition and treatment and can avoid unnecessary testing and complications of severe anemia.

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ADDRESS: M. Chadi Alraies, MD, FACP, Department of Hospital Medicine, A13, Cleveland Clinic, 9500 Euclid Avenue, Cleveland, OH 44195; e-mail alraiec@ccf.org.

Pay attention also to the uncommon causes of symptoms of systemic sclerosis