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Image of the Month
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Image of the Month

Type II Achalasia as the Initial Presentation of Systemic Sclerosis

Keywords: *Achalasia; Esophagus; Functional GI disease; Scleroderma*

Type II Achalasia in a Patient with Scleroderma

A 62-year-old female with a history of coronary artery disease and longstanding Raynaud's syndrome was referred to us for an assessment of noncardiac chest pain, dysphagia and nocturnal reflux. Esophagogastroduodenoscopy was unremarkable with no evidence of stricture or mass. High-resolution esophageal manometry showed Type II achalasia (Figure 1). Ambulatory 24-hour pH study showed no significant acid reflux. A year later, she presented with progression of Raynaud's. Physical examination demonstrated sclerodactyly, digital capillary dropout and chest wall telangiectasias. Bloodwork was significant for positive antinuclear antibody (1:80) and anticentromere antibody (1:320). She was subsequently diagnosed with limited cutaneous systemic sclerosis. Follow-up manometry (3 years later) showed classic 'scleroderma esophagus' (Figure 2).

Discussion

Vasculopathy leading to smooth muscle atrophy and fibrotic replacement is thought to be responsible for the classic finding of 'scleroderma esophagus', characterized by hypotensive lower esophageal sphincter (LES) pressure and aperistalsis in the distal esophagus (1). However, an achalasia-like syndrome with incomplete LES relaxation has been reported in systemic sclerosis (SSc) patients. Park et al. first described aperistalsis and incomplete LES relaxation on conventional manometry in four of seven patients with known SSc (2). The finding of esophageal dilation and delayed esophageal

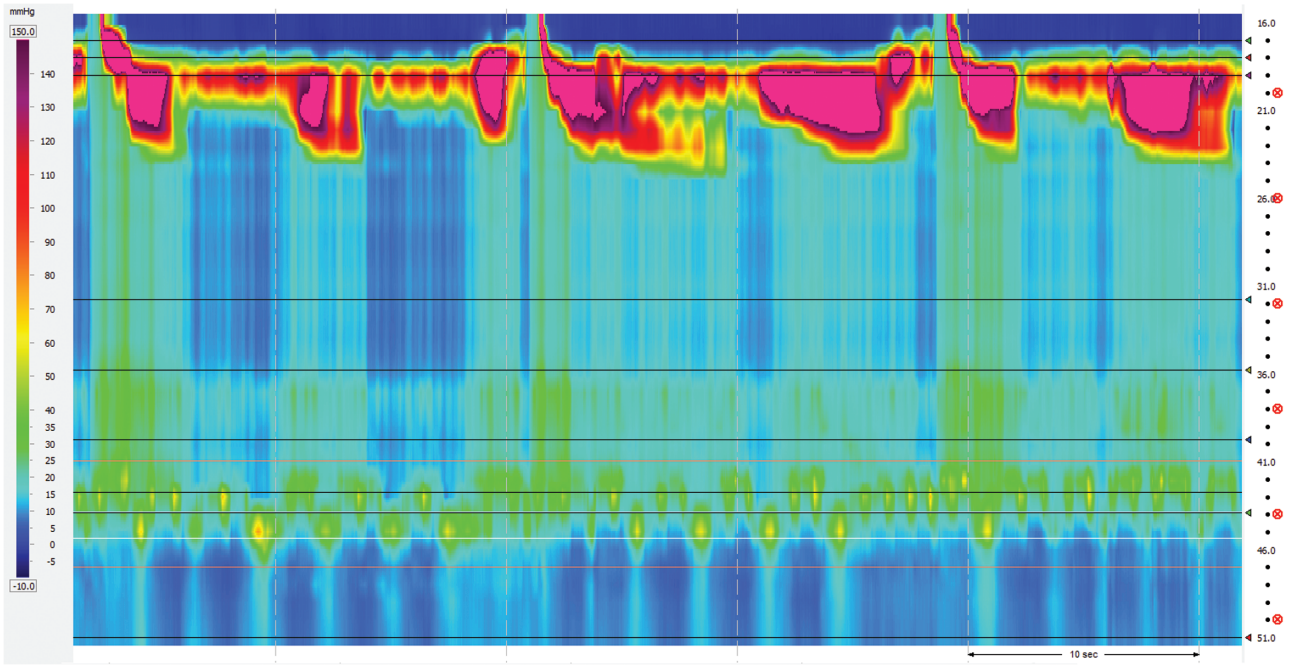

Figure 1. Clouse plot demonstrating Type II achalasia (integrated residual pressure = 19.2, normal < 15 mmHg; panesophageal pressurization with 70% of swallows).

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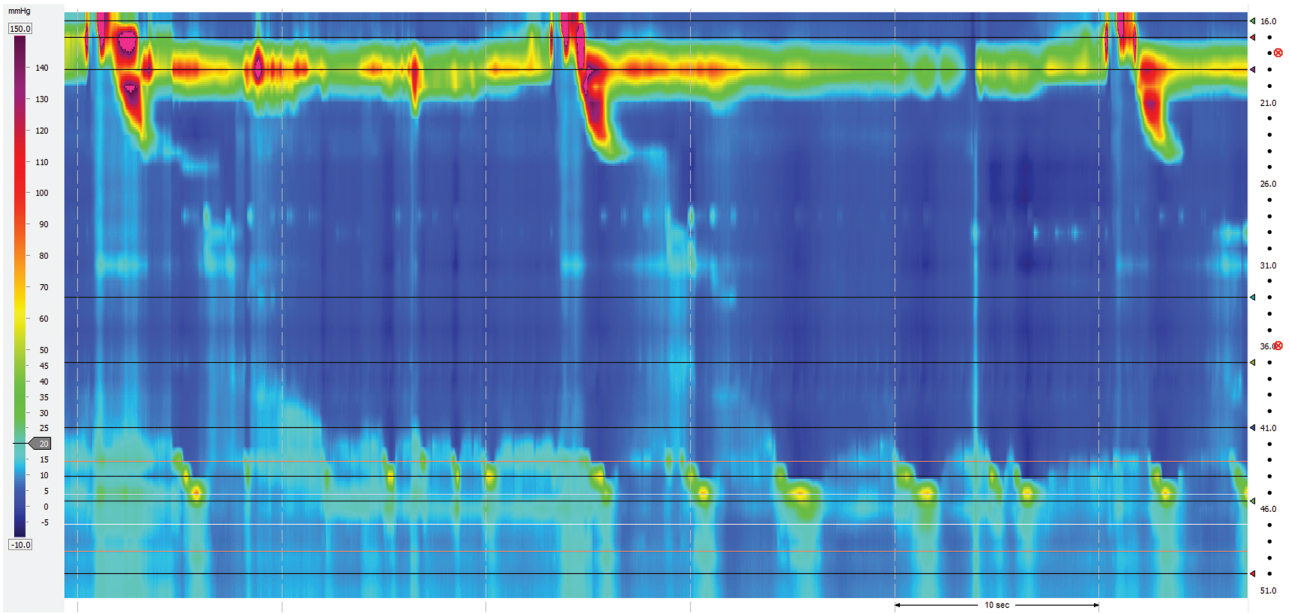

Figure 2. Clouse plot demonstrating hypotensive lower esophageal sphincter, and absent peristalsis, consistent with 'scleroderma esophagus'.

emptying on barium study, associated with manometric evidence of aperistalsis with normal LES relaxation, was documented to precede autoantibody formation and skin changes in a patient who developed SSc; follow-up manometry demonstrated absent motor activity in the smooth muscle esophagus with absent LES tone (3). The pathophysiology of this finding is unclear but may relate to the presence of antimyenteric neuronal antibodies. Antimyenteric neuronal antibodies have been detected in the sera of SSc patients and correlate with the presence of Raynaud's phenomenon (4). Autoantibodies to the myenteric plexus have also been detected in achalasia patients, and may be implicated in the decreased interstitial cells of Cajal and nitric oxide synthase containing neurons seen on histopathologic examination (5). Our case is the first to document on high-resolution manometry the conversion of type II achalasia features to scleroderma esophagus findings. This may yield insight into the pathophysiology of esophageal disease in SSc, which remains poorly understood.

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Matthew Woo, MD, Milli Gupta, MD

Division of Gastroenterology, Department of Medicine, University of Calgary, Calgary, Alberta, Canada

Correspondence: Matthew Woo, MD, FRCPC, Division of Gastroenterology, Department of Medicine, University of Calgary, 3330 Hospital Dr NW, Calgary, AB T2N 4N1, Canada, e-mail: mwoo@qmed.ca

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