Esophageal involvement in progressive systemic sclerosis

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Geller SA, Campos FPF. Esophageal involvement in progressive systemic sclerosis. Autopsy Case Rep [Internet]. 2013; 3(3): 77-79. http://dx.doi.org/10.4322/acr.2013.031

Progressive systemic sclerosis (PSSc) is a chronic disease of unknown etiology characterized by progressive, abnormal accumulation of fibrous tissue in the skin and many organs. Characteristically, there is induration and thickening of the skin (scleroderma), abnormalities involving muscles, joints, and viscera. The first description was likely by William and Robert Watson in 1754.

In the following century, Robert Graves, in Dublin (1843), and the French physician, Maurice Raynaud (1865), described peripheral vasoconstriction of the hands, relating it to scleroderma, which was present in 90-95\% of the patients with PSSc. It is generally considered that the initiation of PSSc is due to the combined effects of abnormal immune response and vascular damage, leading

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Two different physiopathologic mechanisms are recognized in the scleroderma esophageal dysfunction: a) impairment of neuromuscular electrical transmission; and b) progressive loss of muscle strength in the distal esophagus as well as in the lower esophageal sphincter. It is quite likely that the neurological involvement precedes the muscular atrophy and fibrosis.

Dysphagia and pyrosis are the most common symptoms. Likewise, early satiety, regurgitation of food, progressive weight loss, malnutrition, or impaction of food may also be observed. These symptoms are the result of the disruption of peristalsis, gastroesophageal reflux, peptic stricture, and occasionally the presence of candidiasis. The severity of these abnormalities increases with disease progression, establishing a certain parallelism between the intensity of motor problems and disease severity.

Esophageal manometry detects early dysfunction and is altered in 90% of cases. The most common findings are motor abnormalities, lack or decreased pressure of the distal esophageal high-pressure zone, moderate to severe gastroesophageal reflux, abnormal acid clearance, and decreased frequency of peristaltic contractions. Gastroesophageal reflux rather than impaired motility is the major cause of esophageal symptoms. Endoscopy reveals an atonic esophagus covered by pale mucosa and the presence of ulcers and, often, cicatricial stenosis.

Lateral chest films of patients with scleroderma frequently reveal segmental air or air along the entire esophagus rarely with dilation or air-fluid level; the latter being more suggestive of tumor, stricture, or achalasia.

Keywords: Scleroderma, Systemic; Esophagus.

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