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Case Report

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DYSPHAGIA REVEALING A SYSTEMIC SCLEROSIS SINE SCLERODERMA

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ABSTRACT

Background: Systemic sclerosis sine scleroderma (ssSSc) is a rare form of systemic sclerosis without skin involvement. **Case presentation:** We report an interesting case of dysphagia associated with Raynaud phenomenon revealing a ssSSc in a 51-year-old Moroccan woman. Dysphagia is a common symptom in SSc, however it has been rarely reported as the main clinical manifestation of the disease in the absence of cutaneous involvement. **Conclusion:** On the basis of this observation, gastroenterologist should have in mind systemic sclerosis as a differential diagnosis in patients presenting with dysphagia and Raynaud phenomenon even in the absence of any skin involvement.

INTRODUCTION

Systemic sclerosis (SSc) is an autoimmune triggered disease of the connective tissue, arterioles, and microvessels, characterized by the appearance of fibrosis and vascular obliteration.

The clinical features are marked by skin lesions but not only as systemic involvement is frequent. There is also a rare form without cutaneous involvement, called "sine scleroderma".[1] It was first described in 1954 by Abrams et al.[2] Since then, there have been over 200 cases described in the literature.[3 4 5 6]

OBSERVATION

We report the case of a 51-year-old Moroccan woman with a history of diabetes mellitus and adhesive capsulitis. She presented with a 6-month history of persistent dysphagia, initially to solids then to liquids. She also complained of a foreign body sensation in her eyes and a dry mouth and reported a Raynaud phenomenon (RP).

Clinical examination showed erythematous digits corresponding to the reperfusion phase of RP while the patient well described the three phases of the latter. Buccal opening and face mimics were normal. An early sclerodermiform pattern was found on periungueal dermoscopy.

Laboratory analysis including complete blood count, erythrocyte sedimentation rate, C reactive protein didn't reveal any abnormalities. Immunologic test was positive for antinuclear antibodies only.

Shirmer test was positif. Salivary gland biopsy revealed a grade 1 Chisholm and Mason chronic sialadenitis.

An upper gastro-intestinal endoscopy was performed and showed a hypotonic oesophagus. The lower oesophageal sphincter (LES) was passed easily. Oesophageal biopsies a vascular congestion of the chorion with a slight lympho-plasmocytic infiltrate.

High resolution manometry showed the absence of oesophageal contractility and no obstruction of the LES.

Based on the criteria proposed by Poormigham[3], the diagnosis of ssSSc was retained.

To rule out other organ involvement, an exhaustive assessment was performed including dosing of creatinine phosphokinase and aldolase, echocardiography, computed tomography scan, pulmonary function tests and kidney functions evaluation. All of these explorations were normal.

Management was based on hygiene and dietary rules, calcic inhibitors, artificial tears applied regularly and chewing gum to relieve dry mouth. PPI's were delivered in case of gastroesophageal reflux symptoms.

After one-year follow up, our patient didn't show any skin involvement.

DISCUSSION

Systemic sclerosis is a rare disease characterized by anomalies of the microcirculation and by cutaneous and/or visceral fibrotic lesions.[7] There have been 3 main phenotypes of this entity, two that differ only by the extent of the cutaneous sclerosis and the third one is a rare form without cutaneous involvement, called "sine scleroderma".[1] In all these forms, visceral manifestations may occur, in particular peripheral vascular, digestive, cardiopulmonary, and renal.

To diagnose a ssSSC, Poormigham proposed the following criterias: 1) Raynaud's phenomenon or a peripheral vascular equivalent (digital pitting scars, digital-tip ulcers, digital-tip gangrene, abnormal nailfold capillaries), 2) positive ANA, 3) any 1 of the following: distal esophageal hypomotility, small bowel hypomotility, pulmonary interstitial fibrosis, primary pulmonary arterial hypertension (without fibrosis), cardiac involvement typical of scleroderma, or renal failure consistent with scleroderma renal crisis, and 4) no other defined connective tissue or other disease as a cause of 1), 2), or 3). This diagnosis would be more convincing if the ANA specificity was due to an SSc-associated autoantibody.[3]

In the case of our patient, dysphagia was the main symptom leading to the final diagnosis. However, it could have been difficult to retain it if there was no other signs of sclerodermia, such as RP and positive antinuclear antibodies.

Gastrointestinal tract involvement is very common among patients having a SSc, whatever its phenotype. Oesophagus is the most frequently affected part.[8] Two types of linked symptoms are reported : symptoms of gastrooesophageal reflux (GERD) and those related to dysmotility such as dysphagia.[9] The initial assessement of dysphagia is based on upper gastrointestinal endoscopy. It can show a normal aspect of esophagus, lesions related to GERD and/or hypotonic or atonic aspect. Then, evaluation must be completed, when upper gastrointestinal endoscopy does not show any mucosal abnormalities such as esophagitis, strictures or rings, with manometry and if available, high resolution manometry.[8] It confirms the dysmotility and demonstrates diminished or absent peristalsis of the distal two-thirds of the esophagus along with a hypotensive LES.[10]

Other gastrointestinal manifestations such as gastric antral vascular ectasy, small intestine bacterial overgrowth and anorectal dysfunction can often overlap with esophageal manifestations. They should be sought and managed as well.[10]

Management of esophageal dysmotility combines supportive and drug-based care. It is based on hygiene and dietary rules like taking small bites, cut/chew food well, avoid dry or fibrous foods and take plenty of water with solid foods. Many drugs have been used in patients with SSc as prokinetics agents such as metoclopramide, erythromycin and cisapride. However, the experience with the use of prokinetic drugs in SSc patients is bibliographically limited and has had controversial results.[9]

CONCLUSION

Systemic sclerosis sine scleroderma is a rare disease in which different organs can be affected. Among them, the esophagus can be the siege of fibrotic lesions and then lead to dysphagia that can be the only clinical manifestation. Gastroenterologist should be aware of this entity when all other etiologies have been ruled out and, so, other physicians can be invovled in the gestion of this complex pathology.

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