Esophageal motility abnormalities in scleroderma. Report of 17 patients

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ABSTRACT

Objective: Aim of this work was to study esophageal motor abnormalities in patients with systemic sclerosis (SSc).

Patients and methods: We studied 17 consecutive patients with SSc classified according to Le Roy et al. In each patient we performed barium esophagograms and esophageal manometry (EM) and we evaluated in particular lower esophageal sphincter (LES) pressure and relaxation and esophageal peristalsis. We also calculated the differences between the esophageal and intragastric pressure.

Results: Esophageal abnormalities were detected by EM in all patients. Wave amplitude and duration were decreased in 14 and 9 patients respectively. Wave propagation was abnormal in 11 patients and LES pressure was reduced in all but 1. Only two patients with edematous skin changes and 1-year-long Raynaud's phenomenon had a slight esophageal involvement.

Conclusion: Esophageal involvement was detected in all patients even though asymptomatic. A relationship was noted with the duration of the disease, but not with the severity of skin involvement.

KEY WORDS

scleroderma, esophageal abnormalities

Introduction

Esophageal motility abnormalities occur in up to 90% of the patients with systemic sclerosis (SSc), about half of them being asymptomatic. The relationship of such disturbances with the duration of the disease and with SSc specific serology is poorly understood. In this study we compared the esophageal abnormalities with the extent of skin involvement and the disease duration in 17 consecutive SSc patients.

Patients and methods

We studied 14 females and 3 males, mean age 56. According to Le Roy et al (1) they were classified as having limited SSc (lSSc, 14 patients) and diffuse SSc (dSSc, 3 patients). Each patient had a complete history and physical examination. In particular, dysphagia, heartburn, vomiting and substernal pain were recorded. Antinuclear antibodies (ANA) and anti-centromere antibodies (ACA) were detected by indirect immuno-

Table 1. Criteria for scoring symptoms in patients with limited and diffuse systemic sclerosis. Scores ≤3 slight, 4-7 mild, >7 severe involvement.

Medium	Normal wave amplitude (> 25 mmHg)	0
	Mildly reduced wave amplitude (10-25 mmHg)	1
	Severely reduced wave amplitude (< 10 mmHg) or aperistalsis	2
Lower	Normal wave amplitude (> 25 mmHg)	0
	Mildly reduced wave amplitude (10-25 mmHg)	1
	Severely reduced wave amplitude (< 10 mmHg) or aperistalsis	2
Medium	Normal wave duration (3-6 seconds)	0
	Abnormal wave duration (< 3 or > 6 seconds)	1
Lower	Normal wave duration (3-6 seconds)	0
	Abnormal wave duration (< 3 or > 6 seconds)	1
Propagated wave	≥ 75%	0
	50-75%	1
	≤ 50%	2
Lower esophageal sphincter	ΔP normal (18-22mmHg)	0
	ΔP mildly reduced (11-17 mmHg)	1
	ΔP severely reduced (≤ 10mmhg)	2

Total possible score: 10

fluorescence (IIF), anti Scl-70 antibodies were detected by immunodiffusion assay and ELISA.

All patients underwent barium esophagograms and esophageal manometry (EM). The latter was performed by pneumohydraulic capillary infusion system (Arndofer type) (2). We evaluated the pressure of the upper and lower esophageal sphincters (UES-LES) and their relaxation. Esophageal peristalsis was studied by calculating the wave amplitude, duration and propagation.

Moreover, we calculated the differences between the esophageal and intragastric pressure (Δ P) and we measured the abdominal length of LES.

As SSc affects the distal two-thirds of the esophagus, we considered only abnormalities of these segments. To grade the esophageal motility abnormalities, we applied an arbitrary 7-point score scale (Table 1). A score ≤ 3 documented a slight esophageal involvement, 4-7 a mild involvement and >7 a severe one.

Results

Fourteen patients had lSSc and 3 dSSc. The skin exhibited edematous changes in 6 patients, sclero-edematous in 6 and sclerotic in 5. Teleangiectases, calcinosis and ulcerations were present in 15, 11 and 6 patients respectively. Pulmonar, renal and cardiac involvements were present in 12, 7 and 6 patients respectively. One patient had primary biliary cirrhosis, 1 Parkinson's disease and 2 Sjögren's syndrome.

Only 2 patients had anti Scl-70 antibodies, 10 had ACA and the remainders speckled pattern of ANA.

Heartburn was present in 11 patients, dysphagia in 7 and regurgitation in 7. Only 4 patients were completely asymptomatic.

Esophageal abnormalities of some type were detected by EM in all patients. In particular, UES pressure and coordination with pharynx were normal in all patients but 1, and wave amplitude and duration were decreased in the distal two-thirds of the esophagus in 14 and 9 patients respectively. Waves were abnormally propagated in 11 patients and LES pressure was reduced in all but 1. ΔP value was normal in 2 patients, mildly reduced in 2 and severely reduced in 13. LES relaxation was abnormal in all patients. The mean abdominal length of LES was 0,88 centimeters.

The esophageal involvement was scored as slight in 2 patients, mild in 9 and severe in 6. The 2 patients with anti Scl- 70 antibodies scored a mild (4-7) esophageal involvement. Two patients with edematous cutaneous lesions were scored as having mild involvement (≤3). They had Raynaud's phenomenon for only 1 year before (table 2).

All patients with scleroedematous or sclerotic skin involvement had a mild or severe esophageal involvement with Raynaud's phenomenon for many years before the esophageal motor abnormalities were detected (table 2). There was only one patient who scored 6 and had Raynaud's phenomenon for only 2 years before disclosing the manometrical changes. He had also dSSc and anti-Scl70 antibodies.

Table 2. Relationship among score, skin involvement and Raynaud's phenomenon duration

ESOPHAGEAL SCORE	SKIN INVOLVEMENT		
	Edematous	Scleroedematous	Sclerotic
≤ 3	1 year 1 year		
4 - 7	10 years 10 years	10 years 7 years 30 years	2 years 10 years 10 years 20 years 55 years
> 7	16 years 10 years	28 years 5 years	40 years

Years indicate Raynaud's phenomenon duration
* this patient was Scl-70 positive and had dSSc

Discussion

EM proved to be the most sensitive method to detect esophageal hypomotility (3,4) being positive even in asymptomatic patients. Our study confirms that esophageal involvement is very common in SSc. As previously described, incoordination, reduction and abnormal duration of the waves and failure of the LES to relax are its main features. Our study also confirms that there is no relationship between the degree of esophageal dysfunction and the severity of skin changes (5) or the presence of a particular ANA.

In our study, the esophagus involvement seemms to depend on the duration of the disease. The 2 patients with a slight involvement of esophagus (≤3 score) had only edematous skin and experienced Raynaud's phenomenon only since 1 year. The other 4 patients with edematous skin changes and esophageal score >4 had Raynaud's phenomenon for 10 years or more. Moreover, all but one of the patients with scleroedematous and sclerotic skin changes had an esophageal score >4 and Raynaud's phenomenon for 21 years, as an average.

In conclusion, esophageal involvement was detected in all patients even though asymptomatic. Motor abnormalities seem to parallel the duration of the disease, but not the severity of the skin involvement.

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