Esophageal dysmotility in scleroderma: A prospective study of 183 cases

Troubles moteurs œsophagiens au cours de la sclérodermie systémique : étude prospective de 183 cas

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Summary

Aims. — The goal of the study was to evaluate the prevalence and risk factors of esophageal motor disorders in systemic sclerosis.

Patients and methods. — In 183 consecutive cases of scleroderma, as diagnosed by American College of Rheumatology criteria (1980). Patients’ mean age was 40.6 ± 13.3 years, the gender ratio was 0.13 and the average duration of disease was 6.8 ± 7.5 years. A localized, cutaneous form was observed in 148 patients (81%) and a diffuse form in 35 (19%). All patients underwent upper gastrointestinal endoscopy and standard esophageal manometry.

Results. — Esophageal symptoms and reflux esophagitis were found in 108 (59%) and 68 (37%) of patients, respectively. Esophageal motor disorders were present in 148 patients (81%), and were associated with a hypotensive lower esophageal sphincter in 114 (62%). The presence of these motor abnormalities was not related to age, gender, skin extension or duration of disease. Esophageal motor disorders were present in almost all patients with esophageal symptoms or esophagitis, and were also found in 48 (64%) of the asymptomatic patients.

Conclusion. — Esophageal motor disorders are frequently seen in scleroderma, especially in cases with clinical symptoms, but are not associated with a specific form of the disease.

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Résumé

Objectif. — Préciser la prévalence et les facteurs de survenue des troubles moteurs œsophagiens de la sclérodermie systémique.


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Introduction

Esophageal disorders are among the most common visceral manifestations of systemic sclerosis [1,2]. These disorders present as a characteristic motor impairment due to a hypotensive lower esophageal sphincter (LES) and/or esophageal motor disorders (EMD). EMDs generally involve hypotonic or atonic contraction waves and/or qualitatively defective peristalsis. The motor disorder is limited to the smooth muscles, which explains why the lower esophagus is involved while the cervical portion and superior esophageal sphincter remain intact [3–5]. Overall, the incidence of manometric anomalies is high—ranging from 70–96%, depending on the report—with a hypotensive LES and EMDs noted in 50 and 60% of cases, respectively [1,2,5]; 52–63% of patients present with both disorders [6,7]. In 48–81% of patients, the EMD is a hypotonic contraction wave [1,2,6], while 23–52% have an atonic wave [2,8–10] and 40–91% present with wave-propagation disorders [1,5,6,10]. These disorders are localized in the distal esophagus in 70% of cases. Involvement of the upper esophagus and upper sphincter are unusual, and generally only seen in advanced-stage disease where massive, extensive gastrointestinal reflex disease (GERD) is the causal mechanism [8,9].

There is still considerable debate over the time of onset and factors associated with EMDs. These factors appear to develop early in the disease course, and their early detection would be useful for identifying early forms of the disease [2,3,10]. Their presence could also be strong predictors of the development of esophageal reflux [8,11]. Yet, despite the recommendations of several authors [8,12], a strategy based on esophageal manometry as part of the standard workup for patients with systemic sclerosis is still not widespread.

Patients and methods

The present study population, recruited prospectively at a single center over a period of more than 19 years (April 1988 to September 2007), comprised 183 consecutive patients with systemic sclerosis, diagnosed according to the 1980 criteria from the American College of Rheumatology [13].

Mixed forms of connective tissue disease and those associated with diabetes were excluded. The patients’ mean age was 40.6 ± 13.3 years (range 11–73 years) and the gender ratio was 0.13. Disease duration was measured from the time of onset of the first clinical manifestation—Raynaud’s phenomenon in 185 patients (86.3%) and an acute skin involvement in the remainder. On average, the disease duration was 6.8 ± 7.5 years (range 6 months–32 years). Using the classification of generalized sclerosis described by LeRoy et al. [14], a localized cutaneous form was noted in 148 of our patients (80.8%) and a diffuse cutaneous form in 35 (19%).

A standardized medical chart was completed for all patients, all of whom underwent upper gastrointestinal endoscopy and esophageal manometry. These explorations were performed during the same consultation as the history-taking and physical examination, and before initiating any antisecretory treatment.

Data collected included the patients’ age, gender, characteristic features of the disease, and presence of symptoms of GERD (pyrosis and/or acid regurgitations) and/or dysphagia. Each symptom was graded on a scale from 0 to 3 by intensity (0 = absent; 1 = mild, could be ignored by the patient; 2 = moderate, could not be ignored, but had no effect on daily life activities; 3 = severe or incapacitating, affecting daily life activities) and by frequency (0 = absent or less than one per month; 1 = less than one per week; 2 = several times per week; and 3 = every day) [15]. The total score was the sum of the two scores for each symptom. Symptoms were then categorized as mild for a total score less than or equal to six, moderate for a total score of seven to 12 and severe for a total score greater than 12, or when one symptom was considered incapacitating every day (score = 9).

Upper gastrointestinal endoscopy was used to identify and stage esophagitis due to GERD, according to the Los Angeles classification. Manometry was performed with a four-channel infusion probe (distilled water infusion rate: 0.5 mL/min). The standard procedure was used, following a 12-hour fast [16]. Normal manometric values were established in our laboratory using a control population of 24 healthy subjects (0.2 gender ratio, mean age 41.7 ± 10.7 years, age range 29–60 years).
Student’s t test, chi² test and a reduced standard deviation (SD) test were used as appropriate to compare means and percentages. The level of significance was P < 0.05.

Results

Esophageal symptoms were found in 108 patients (59%), with GERD symptoms observed in 86 patients (47%) and dysphagia in 84 (45.9%). The clinical complaint was considered mild in 50 patients (27.3%), moderate in 32 (17.5%) and severe in 26 (14.2%). Reflux esophagitis was noted in 68 patients (37%) and hiatus hernia in 43 (23.5%), and both conditions were found in 28 patients.

EMDs were noted in 148 patients (80.8%), comprising decreased amplitude of the post-deglutition contraction-wave (less than 50 mmHg) in 146 (79.8%), including atony in 110 (60%) and isolated increases in the percentage of non-protected wave sequences (greater than 30%) in 30 (16.4%). A hypotensive LES (mean pressure less than 13 mmHg) was observed in 114 patients (62.3%) and was associated with EMDs in 104 (56.8%).

The EMDs were located in the lower two-thirds of the esophagus in 122 patients (82.4%), in the lower third in 13 cases (8.8%) and involved the entire organ in 13 (8.8%). The presence of EMDs was not related to age, gender, disease duration or skin extension (Table 1). However, EMDs were more frequent and more severe in symptomatic patients (Table 2). Thus, the presence of esophageal symptoms had a sensitivity of 67.5% with 77% specificity for the diagnosis of EMDs, and a positive predictive value (PPV) of 92.6% and a negative predictive value of 36%.

EMDs were also more frequent and more marked in patients with erosive esophagitis (Table 3). Distal esophageal contractions alone were significantly weaker in patients with severe or complicated esophagitis (n=24, 0.7 ± 3.4 mmHg) than in patients with moderate esophagitis (Los Angeles A or B, n=44, 10.3 ± 6.1 mmHg; P = 0.005).

Discussion

In this series of patients with scleroderma, seen at a single center in Algeria, EMDs were common and severe, producing esophageal atony in 60% of cases and extending over the lower two-thirds of the esophagus in 83%. EMDs were associated with a hypotensive LES in more than half (57%). These results are in line with earlier reports found in the literature.

In our patients, the presence of EMDs was not related to disease duration. In the literature, EMDs involving propagation disorders have occurred either every year in the disease course [3,17—19] within the year following the diagnosis of systemic sclerosis [10] or even during the so-called “presclerosis phase” of Raynaud’s syndrome, before the development of overt manifestations of scleroderma [2,10,19].

In addition, the type of sclerosis was not related to the presence of EMDs in our patients. Other authors have found no relationship between the severity of EMDs and degree of cutaneous involvement [2,4]. Some have found that EMDs were not more frequent, but were more severe in diffuse forms of the disease [20,21], whereas yet other authors have found EMDs to be more frequent and more severe in diffuse scleroderma [7,22].

However, motor disorders (dysmotility of the esophageal body and hypotensive LES) were more frequent and more severe in patients with peptic esophagitis. In the series reported by Zamost et al., peristalsis was always abnormal in patients with esophagitis, but normal in about half of patients without esophagitis [8].

EMDs were frequently associated with clinical signs of esophageal disease (GERD or dysphagia), but with no strict correlations [4,7,8,23,24]. Our results are similar to those reported by Köhler et al. [25], who demonstrated the better predictive value of clinical symptoms for the diagnosis of EMDs, but also their weak negative predictive value. Weston et al. [26] also observed a higher frequency of manometric disorders in the symptomatic population.

In conclusion, our research shows that esophageal motor disorders are frequent and severe in all forms of systemic sclerosis reported in Algeria, findings that are similar to the results reported in other countries. These motor disorders

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<th>Table 1</th>
<th>Clinical characteristics of the patients.</th>
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<tr>
<td>Esophageal motor disorders</td>
<td>Present (n = 148)</td>
</tr>
<tr>
<td>Mean age (years)</td>
<td>40.9 ± 13.5</td>
</tr>
<tr>
<td>Male/female</td>
<td>16/132</td>
</tr>
<tr>
<td>Disease duration (years)</td>
<td>7 ± 7.4</td>
</tr>
<tr>
<td>Localized form</td>
<td>117 (79%)</td>
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<td>Diffused form</td>
<td>31 (88.5%)</td>
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<th>Table 2</th>
<th>Prevalence and severity of EMDs with or without symptoms.</th>
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<tbody>
<tr>
<td>Esophageal symptoms</td>
<td>Present (n = 108)</td>
</tr>
<tr>
<td>EMDs</td>
<td>100 (92.6%)</td>
</tr>
<tr>
<td>PLES (mmHg)</td>
<td>12.6 ± 6.9</td>
</tr>
<tr>
<td>CWA (mmHg)</td>
<td>8.7 ± 22.6</td>
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<tr>
<th>Table 3</th>
<th>Prevalence and severity of EMDs with or without reflux esophagitis.</th>
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<tr>
<td>Reflux esophagitis</td>
<td>Present (n = 68)</td>
</tr>
<tr>
<td>EMDs</td>
<td>65 (95.6%)</td>
</tr>
<tr>
<td>PLES (mmHg)</td>
<td>11.2 ± 6</td>
</tr>
<tr>
<td>CWA (mmHg)</td>
<td>5.2 ± 16</td>
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</tbody>
</table>

EMDs: esophageal motor disorders; PLES: pressure in lower esophageal sphincter (mean); CWA: contraction-wave amplitude (distal segment).
are frequently seen in patients with erosive esophagitis, but are also present in two-thirds of asymptomatic patients. Manometric screening is highly relevant, at least for asymptomatic patients without esophagitis.

**Conflicts of interest**

None.

**References**


