The gastrointestinal tract, particularly the oesophagus, is affected in about half of all patients with systemic sclerosis. Only a few studies so far have dealt with the anorectal tract. We studied the anal function using anorectal manometry in 12 patients with limited systemic sclerosis. We also studied the oesophageal function. For the oesophagus, we measured the difference between intragastric and oesophageal pressure, while for the anorectal tract we investigated the maximum resting pressure, the maximum voluntary squeeze effort and the rectoanal inhibitory reflex. Maximum resting pressure and maximum voluntary squeeze effort were found to be decreased in all patients. The rectoanal inhibitory reflex was abnormal in four patients. Statistical analysis showed a significant correlation between maximum resting pressure and maximum voluntary squeeze effort. No correlation was found between oesophageal and anorectal involvement. Anorectal dysfunction is common in patients with limited systemic sclerosis. We suggest that these patients should have an evaluation of their anorectal function including anorectal manometry. Key words: systemic sclerosis; oesophagus; rectum; gut.

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Twelve unselected women with limited SS classified according to Le Roy et al. (3) were recruited in a dermatology clinic and enrolled in the study from 1999 to 2000. Their average age was 59 years. Eleven patients complained of oesophageal symptoms such as dysphagia, regurgitation and vomiting, and heartburn. Constipation defined as three or fewer bowel motions per week was observed in two patients. One had diarrhoea and one had changing stools. Only one patient had faecal incontinence for liquid stools, but she had no rectal prolapse.

Scleroderma was associated with Sjögren syndrome in three patients. The skin exhibited oedematous changes in two patients, sclerodematous in five and sclerotic in five. Telangiectases, calcinosis and ulcerations were present in 12, 7 and 3 patients, respectively. Raynaud’s phenomenon was present in all of them for a mean duration of 18 years. Lungs and heart were involved in three patients. No patients had renal involvement.

None had anti-topoisomerase 1 antibodies by enzyme immunosorbent assay. By indirect immunofluorescence, seven patients had anti-centromere antibodies, one patient had nucleolar-patterned anticentromere antibodies and the remainder had non-specific speckle-patterned anticentromere antibodies.

All patients underwent barium oesophagography, oesophageal manometry and anorectal manometry.

Oesophageal manometry was performed after an overnight fasting period with an 8-hole, water-perfused polyvinylchloride tube (outer diameter: 4.5 mm) introduced through the nose. The four distal side holes were located close to the tip of the tube, 90° apart. The other four openings were positioned more proximally at a distance of 5 cm each and axially oriented at 90°. The lower oesophageal sphincter pressure was recorded by a stationary pull-through method using the four distal openings. The tubular oesophageal function (TEF) was assessed using one of the four distal side holes (placed 5 cm proximal to the lower oesophageal sphincter) and the four proximal openings, thus covering 20 cm of the oesophagus. TEF was analysed for peak contraction pressure, duration of contraction, and co-ordination and propagation velocity after voluntary deglutition of 10 dry and 10 wet swallows (using 5 ml of water). The whole examination lasted from 20 to 40 min. Manometric tracings were interpreted using standard criteria (4). TEF was classified as normal when the amplitude of the peristaltic contractions exceeded the lower range of normal value, and as abnormal when there was no peristalsis or a marked hypo-peristalsis (e.g. weak or multiple peaked contractions with the amplitude below the lower range of normal value) of the lower two-thirds of the oesophageal body. Moreover, we calculated the differences between the oesophageal and intra-gastric pressure (AP).

Anorectal manometry was performed after enemas on the day before and also on the morning of the examination. A water-perfused 8-hole catheter (outer diameter 4.5 mm; perfusion rate 0.8 ml/min) (inner diameter 1.8 mm), located longitudinally 1 cm apart and 90° axially, was passed through the anal canal into the rectum. A rectal distending balloon was fixed at the tip of the probe and filled with air via a separated 1.8 mm opening. The probe was withdrawn until the most distal orifice was just outside the anal sphincter, even...
if the balloon remained inside. The anal pressure was measured. The maximum resting pressure (MRP, normal value > 50 mmHg) represents the resting tone of the anal sphincter.

The patients were then asked to squeeze the anal sphincter as firmly as they could and the maximum pressure recorded under this condition was defined as the maximum squeeze pressure (or maximum voluntary squeeze effort, MVSE, normal value > 70 mmHg). The threshold of rectal sensitivity was determined by slowly inflating the distension balloon with air and was defined as the smallest volume of the balloon that could be perceived by the patient. Finally, the rectoanl inhibitory reflex, RAIR (i.e. relaxation of internal anal sphincter after rectal distension, normal value 20–40 ml of air), was determined by inflating 10–60 ml of air into the balloon; RAIR threshold (i.e. minimum volume at which the reflex could be elicited) was recorded. Anal manometry lasted for 20–30 min (5).

Statistical analysis included the Pearson’s correlation coefficient; a regression line of MSVE on MRP was calculated.

RESULTS

Oesophageal manometry showed oesophageal abnormalities in all patients. In particular, both wave amplitude and duration were decreased in the distal two-thirds of the oesophagus in 11 patients. ΔP values were normal (> 17 mmHg) in 3 patients, mildly reduced (14–17 mmHg) in 3, moderately reduced (10–14 mmHg) in 2 and severely reduced (<10 mmHg) in 4.

Anorectal motor abnormalities were found in all patients (Table I). MRP values were mildly reduced (40–50 mmHg) in 2 patients, moderately reduced (20–40 mmHg) in 9 and severely reduced (<20 mmHg) in one. MVSE values were normal (> 70 mmHg) in one patient, mildly reduced (50–70 mmHg) in 3 and moderately reduced (25–50 mmHg) in 8. RAIR values were normal (insufflation of 20–40 ml of air) in 8 patients and pathological (insufflation of more than 40 ml of air) in 4.

Statistical analysis showed a significant correlation only between MRP and MVSE ($r = 0.595$, $p = 0.041$) (Fig. 1).

DISCUSSION

The anorectal tract consists of a posterior-anterior fissure surrounded by two overlapping muscles. The internal anal sphincter is a smooth muscle, while the external anal sphincter is a striated muscle consisting

<table>
<thead>
<tr>
<th>Patient no.</th>
<th>Systemic involvement</th>
<th>Raynaud’s duration (years)</th>
<th>Esophageal involvement $\Delta P$ (mmHg)*</th>
<th>MRP (mmHg)**</th>
<th>MSVE (mmHg)***</th>
<th>RAIR (ml of air)****</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Lungs, heart</td>
<td>31</td>
<td>8.9</td>
<td>27</td>
<td>45.8</td>
<td>40</td>
</tr>
<tr>
<td>2</td>
<td>15</td>
<td>20.3</td>
<td>22.8</td>
<td>57.8</td>
<td>40</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>5</td>
<td>12.5</td>
<td>45.3</td>
<td>68.7</td>
<td>30</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>15</td>
<td>11</td>
<td>27.9</td>
<td>41.3</td>
<td>30</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>Heart</td>
<td>21</td>
<td>15.6</td>
<td>41.8</td>
<td>NE</td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>Heart</td>
<td>1</td>
<td>16.4</td>
<td>63.7</td>
<td>80</td>
<td></td>
</tr>
<tr>
<td>7</td>
<td>30</td>
<td>18.9</td>
<td>22.1</td>
<td>28.2</td>
<td>50</td>
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</tr>
<tr>
<td>8</td>
<td>Lungs</td>
<td>11</td>
<td>15.3</td>
<td>30.5</td>
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<td></td>
</tr>
<tr>
<td>9</td>
<td>33</td>
<td>20.5</td>
<td>40</td>
<td>80</td>
<td>40</td>
<td></td>
</tr>
<tr>
<td>10</td>
<td>Lungs</td>
<td>10</td>
<td>3.5</td>
<td>44.9</td>
<td>20</td>
<td></td>
</tr>
<tr>
<td>11</td>
<td>Heart</td>
<td>31</td>
<td>8.4</td>
<td>42.5</td>
<td>NE</td>
<td></td>
</tr>
<tr>
<td>12</td>
<td>Heart</td>
<td>15</td>
<td>9.2</td>
<td>29.1</td>
<td>40</td>
<td></td>
</tr>
<tr>
<td>Mean ± SD</td>
<td>–</td>
<td>13.4 ± 5.3</td>
<td>28.1 ± 8.8</td>
<td>47.9 ± 16.5</td>
<td>49.2 ± 23.9</td>
<td></td>
</tr>
</tbody>
</table>

$\Delta P$: difference between the oesophageal and intra-gastric pressure. MRP: maximum resting pressure. MSVE: maximum voluntary squeeze effort. RAIR: rectoanal inhibitory reflex.

Normal values: * > 17 mmHg; ** > 50 mmHg; *** > 70 mmHg; **** 20–40 ml of air.

NE: not elicited. For calculation, NE has been considered = 100.
of three parts. Internal and external anal sphincters represent the involuntary and voluntary components of defecation and its control.

None of the muscles ever relax completely. Their activity increases as the abdominal pressure augments, peaks during voluntary contraction and decreases during defecation. The anal canal is rich in sensory structures for receiving tactile, pressure or thermal stimuli.

In SS, involvement of the area is initially characterized by an alteration of the autonomic nervous system and to a lesser extent of the interstitium of the smooth muscle, later, by its prominent atrophy and by the collagenous replacement of the rectal muscularis propria. All these alterations cause the anal sphincter to lose its function.

Published reports on the dysfunction of the terminal segment of the gastrointestinal tract have yielded inconsistent results: MRP has been found to be lower than in controls; MVSE normal (6, 7) or decreased (8, 9); and RAIR reduced in amplitude (6, 8), absent (6, 7, 9, 10) or even paradoxical in a considerable number of patients (7). However, few of the reported cases distinguish between limited and diffuse SS. Hamel-Roy et al. (6) reported even on cases having morphea. In addition to such heterogeneity, the duration of the disease is often omitted.

In contrast to previous studies, our patients were homogeneous in that they all had limited SS, were all antitopoisomerase I antibody-negative and were mostly asymptomatic. According to the literature, therefore, they were not expected to have an internal organ involvement, except the impairment of oesophageal motility that affects about 70% (11). Yet, all their anorectal parameters proved altered. In particular, both the internal (MRP decreased in 100% of our patients) and the external anal sphincters (RAIR abnormal in 4 patients, MSVE decreased in 12 patients) appeared to be involved. In our patients, such abnormalities started so early that one of them happened to have her anorectal function impaired only 1 year after developing Raynaud’s phenomenon (Table I). Our findings, therefore, contrast with those of Lock et al., who, in a group of unselected patients, rarely found an abnormal anorectal function (12).

Although anorectal alterations were constantly associated with oesophageal involvement, no direct correlation between RAIR and oesophageal motor abnormalities was found, in contrast to the data of Hamel-Roy (6) but in agreement with those of Jaffin et al. (13). We did not investigate the sexual bearing of our patients. It could be, therefore, that some of the alterations are due to relaxation of the anal sphincter rather than to sclerosis. In conclusion, we found that anorectal dysfunctions, common in patients with limited SS, can lead to faecal incontinence or rectal prolapse. Anorectal manometry should therefore be added to the routine evaluation of patients with SS also in view of the new therapeutic options, such as octreotide (14), which has been reported, in one case, to obtain spectacular benefit on skin induration and gut function.

REFERENCES