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Oesophageal Manometry and Video-radiology in Patients with Systemic Sclerosis: A Retrospective Study of its Clinical Value

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The aim of this retrospective study was to evaluate the initial video-radiology and manometry in 40 consecutive patients with systemic sclerosis ($n = 21$) or suspected systemic sclerosis ($n = 19$) in relation to oesophageal symptoms; and, furthermore, to evaluate the consequence of radiographic or manometric findings in the oesophagus on diagnosis and treatment. Evaluating oesophageal abnormalities in relation to diagnosis and treatment has, to our knowledge, not been reported before. Video-radiology together with manometry demonstrated oesophageal dysfunction in 80% of patients. Thirteen patients (33%) were asymptomatic in spite of identified motility abnormalities. Following the radiographic and manometric investigation, 9 patients (23%) had a change of diagnosis and, in 20 patients (50%), systemic treatment was instituted or intensified. The study confirms that both manometry and video-radiology are important for the identification of oesophageal motility abnormalities in patients with systemic sclerosis. Oesophageal symptom profiles alone do not predict abnormal findings. ***Key words: oesophagus; motility-disorder; symptoms; treatment.***

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Systemic sclerosis (SS) is a generalized connective tissue disorder of unknown aetiology characterized by fibrosis and degenerative changes of the skin and internal organs including the gastrointestinal tract (1). Atrophy and fibrosis of the smooth muscle result in impaired oesophageal motility, which is common in this disease. Histological examinations of the gastrointestinal tract in patients with SS reveal a non-homogeneous fibrosis and muscular thinning predominantly of the smooth circular muscular layer (2). Functionally, involvement of the digestive tract has been suggested to progress from early neuropathic dysfunction to a later myopathic dysfunction (3, 4). Oesophageal electromyography has shown a disorganized hyperactivity in SS patients with early functional dysphagia, and hypoactivity or no activity in SS patients with functional dysphagia present for several years (5). Diminished lower oesophageal sphincter pressure (LES-P), gastroparesis and associated gastro-oesophageal reflux (GER) occur frequently and may lead to oesophagitis and oesophageal strictures, complications that often are progressive and difficult to treat (6–8). However, patients may have no oesophageal complaints despite abnormal oesophageal motor function (9–11).

Systemic sclerosis is classified according to the preliminary criteria proposed by the American College of Rheumatology (12). Notably absent from this classification of SS is oesophageal dysfunction because of its apparent lack of specificity (10, 12). The significance of hypo-motility as an isolated finding may be unclear, since it occurs in other diseases, in elderly people, and occasionally in otherwise normal persons (10, 13). However, distal oesophageal smooth muscle dysfunction is highly specific for SS, and failure of oesophageal peristalsis occurs at such a frequency that it is a major indication of the disease (14, 15). GER with poor acid clearance results in an increased incidence of complications such as oesophagitis and peptic strictures. A lower oesophageal mucosal ring, when symptomatic called a Schatzki-ring, may be found as a complication to GER, but the aetiology is uncertain. The prevalence of erosive oesophagitis has been found to be 60% in patients with SS (16). Oesophageal dysfunction is an early and frequent complication, and may precede by several years the characteristic skin changes of SS (17). Changes in the oesophagus are unrelated to changes elsewhere in the body, and involvement of the oesophagus has no effect on survival (15, 18, 19). The most common symptoms are heartburn (35–84%) and dysphagia (57–84%) (15, 16, 20). Heartburn is due to the reflux of gastric acid across an incompetent LES, and its dwell time in oesophagus is determined by the efficacy of oesophageal emptying (18, 21). Functional oesophageal dysphagia is secondary to an oesophageal stricture or disturbed oesophageal peristalsis (18).

Video-radiology and manometry are used in the evaluation of oesophageal function in patients with SS or suspected SS (22–27). Oesophageal manometry is stated to be the most sensitive technique, especially in detecting early changes in oesophageal function (9, 28). Using cine-radiography, some authors have found that aperistalsis is specific for SS, but decreased peristalsis is not, leading to a number of false positive findings (13). Video-radiology and manometry are complementary in terms of information. Manometry shows the involvement of the smooth muscle layers in the oesophagus, i.e. changes in pressure; whereas radiology shows the consequences of the pressure changes, i.e. delayed oesophageal emptying, structural abnormalities and, most importantly, GER.

MATERIAL AND METHODS

Records of patients with a diagnosis of SS or suspected SS, who had been referred by the Department of Dermatology to an initial video-radiographic and manometric examination of the oesophagus, were reviewed. The patient group comprised 40 consecutively referred patients, 30 women and 10 men. Twenty-one patients were already diagnosed having SS, while 19 were suspected suffering from SS at the time of the examination. Their mean age was 48 years, range 23–77 years. The median duration of the disease after the diagnosis SS was 4 months, range 0–32 years. The median duration of the disease after the diagnosis suspected SS was 2 months, range 0–11 years. All patients diagnosed as having SS fulfilled the major criteria for SS proposed by the American College of Rheumatology, and were

further divided into 3 subtypes according to cutaneous involvement: (i) limited cutaneous systemic sclerosis with lesions distal to the wrists and ankles; (ii) limited cutaneous systemic extremital sclerosis with lesions above the wrists and ankles; (iii) diffuse cutaneous systemic sclerosis showing lesions of the trunk (12, 29). All patients were interviewed according to a standardized questionnaire concerning oesophageal symptoms such as heartburn, dysphagia and regurgitation (30).

Oesophageal manometry and video-radiology

All patients underwent a combined investigation of manometry and radiology after an overnights fast. In the supine position the patient made 2 wet swallows of 10 ml barium contrast; next a bread barium item (liver paté on rye bread with barium, 6 g) was given twice all with 30-s interval. Finally, the patient drank 180 ml of barium contrast continuously (swilling). Manometry was carried out 5, 10 and 15 cm above the gastro-oesophageal junction. The manometric examination was further complemented with measurement of the LES-P and relaxation during swallowing. After removal of the catheter, radiological examination for GER was performed: the fluoroscope was positioned over the gastro-oesophageal junction and to provoke reflux the patient first received 180 ml barium contrast in the left supine position, then he turned to the right anterior oblique position; secondly the patient was given a bread barium item and fluoroscopy was performed during mastication and swallowing, and during the passage of food through the oesophagus.

Manometric measurement was performed using a triple-lumen catheter with an outer diameter of 1.7 mm and 3 ports spaced 5 cm apart. It was constantly perfused by a hydraulic perfusion pump with a rate of 0.4 ml/min (Arndorfer Medical Specialties, USA). The catheter was connected to an external transducer (Medex Inc., USA), which transmitted the pressure signals to a Polygraph (Synectics PC, Sweden). Tracing could be seen directly on the monitor and was also saved for later analysis.

Manometric diagnoses

The manometric diagnoses were: (i) normal; (ii) achalasia, incomplete or absent relaxation of the lower oesophageal sphincter (LES), and absence of primary peristalsis of the oesophagus; (iii) diffuse oesophageal spasms (DES), defined as repetitive broad-based contractions intermingled with normal peristaltic waves; (iv) hypo-motility, defined as low amplitude contractions occurring peristaltic or non-peristaltic with or without low-resting LES-P; (v) nutcracker oesophagus, defined as peristaltic waves of amplitude >200 mmHg; (vi) non-specific oesophageal motor disorders (NSEMD), determined as abnormal findings devoid of classification according to the definitions above (8).

Radiological diagnoses

The radiological diagnoses were: (i) normal oesophageal emptying; (ii) delayed oesophageal emptying defined as emptying time of more than 15 s for a solid bolus; (iii) oesophageal rings and strictures (continuous barium swallowing in the supine position); and (iv) GER, the radiological test for GER was considered positive, if the barium column reached 5 cm or more proximal to the gastro-oesophageal junction and was reproducible (31, 32).

Statistics

Comparison of symptoms, findings, diagnoses, and treatment were made by the χ^2 test or by Fishers exact test. $p<0.05$ was regarded as the level of statistical significance.

RESULTS

Manometry

Manometry showed at least 1 oesophageal abnormality in 57% of the patients with SS and in 63% of the patients with suspected SS (Table I). The most common abnormal finding in both groups were hypo-motility. All patients with diminished or absent LES-P also had hypomotility.

Video-radiology

Video-radiology showed at least 1 oesophageal abnormality in 81% of patients with SS and in 79% of patients with suspected SS (Table I). Patients with SS had significantly more often reflux (43%) than patients with suspected SS (5%) ($p<0.01$). Among patients with rings/strictures, 3 had reflux.

Clinical correlation with manometry and radiology

Video-radiology (delayed oesophageal emptying) or manometry (hypo-motility) showed oesophageal dysfunction in 80% of all patients. Among all patients with oesophageal symptoms, 83% had positive findings at video-radiology or manometry (Table I). Thirteen (33%) of all patients were asymptomatic in spite of identified motility abnormalities. Eight patients had normal findings, 4 of these had oesophageal symptoms. The most common symptoms were dysphagia (69%) and heartburn (52%). Patients with oesophageal symptoms compared with asymptomatic patients had a significant higher occurrence of reflux ($p=0.03$), but not of

Table I. *Abnormal findings on oesophageal radiology and manometry in 40 patients with systemic sclerosis (SS) or suspected SS compared with symptoms*

	SS ($n=21$)		Suspected SS ($n=19$)	
	Symptom ($n=14$)	No symptom ($n=7$)	Symptom ($n=9$)	No symptom ($n=10$)
Radiology				
Delayed oesophageal emptying	12	5	7	7
Reflux	8	1	1	0
Rings/strictures	3	1	1	2
Manometry				
Hypo-motility	10	2	6	5
Diminished LES-P	4	1	4	3
NSEMD	0	0	0	1

LES-P = lower oesophageal sphincter pressure; NSEMD = non-specific oesophageal motor disorders.

hypomotility ($p=0.14$) (Table I). In patients with suspected SS and abnormal oesophageal motility, 42% were asymptomatic, whereas only 24% of patients with SS and motility abnormalities were asymptomatic, this difference being non-significant ($p=0.38$). Seven patients had delayed oesophageal emptying shown by radiology, but normal findings at manometry.

Among the 32 patients with abnormal findings at radiological or manometric examination, 25% had a change of diagnosis at the following clinical re-examination.

Systemic immunosuppressive or immunomodulating treatment was instituted in 43% of the patients and intensified in 8% of the patients following this initial video-radiographic and manometric oesophageal examination. Medical antacid treatment was instituted or intensified in 14 patients (35%) and 1 patient was referred for surgical treatment. Oesophageal abnormalities identified by radiology and manometry were significantly related to instituted or intensified immunosuppresive or immunomodulating therapy at the next consultation ($p=0.01$), but not to instituted or intensified antacid treatment ($p=0.39$), whereas, oesophageal symptoms were significantly related to instituted or intensified antacid treatment ($p=0.01$).

DISCUSSION

Manometrically and radiographically proven abnormal peristalsis in the distal oesophagus is present in 80–95% of patients with SS (9, 13, 28). In the present study, video-radiology or manometry identified delayed oesophageal emptying/hypo-motility in 32 patients (80%). Patients with an adynamic oesophagus might be asymptomatic because a swallowed food bolus often descends under the influence of gravity (33). In the present study, dysphagia was the most common symptom. The symptoms of heartburn might have been underestimated because some patients with SS already were in medical therapy for reflux oesophagitis. Many patients with SS are without oesophageal symptoms in spite of motility abnormalities (9, 11, 23, 34). This is confirmed by the present findings, in which 76% of all SS patients without oesophageal symptoms had identifiable oesophageal motility abnormalities. Some authors have stated that symptoms are secondary to reflux (33). The present study demonstrated a significant increased incidence of radiologically proven reflux in patients with oesophageal symptoms. Furthermore, a significant higher incidence of reflux was found in patients with SS compared with patients with suspected SS.

Oesophageal examination in patients with SS or suspected SS is also of importance in the determination of internal organ involvement, as demonstrated presently by a significantly increased change of systemic medical treatment in patients with proven oesophageal abnormalities. At present, no curative therapy exists for SS. However, several treatments, i.e. use of immunosuppressive agents, indicate a disease-moderating effect (35). Therefore, early indication of SS following the demonstration of oesophageal involvement is of major importance.

CONCLUSION

The present study confirmed that many patients with SS or suspected SS have no symptoms from the oesophagus in spite of radiographic or manometric oesophageal abnormalities. We found, that video-radiographic and manometric examination of oesophagus in patients with SS, when used in the evaluation of systemic affection often led to therapeutic changes. A quarter of our patients had a change in diagnosis that was, in part, based on the oesophageal examination.

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