

Case report

Pharyngo-oesophageal dysphagia in systemic sclerosis

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SUMMARY A patient with systemic sclerosis developed recurrent dysphagia consistently localised to the pharyngo-oesophageal region. This was due to narrowing of the lumen as a result of gross thickening of the pharyngo-oesophageal muscles. Histologically the changes were typical of systemic sclerosis. The lower oesophagus was less involved. Eventually she died of aspiration pneumonia and respiratory arrest.

Dysphagia is common in systemic sclerosis. It is usually due to dysfunction of the lower oesophageal sphincter which gives rise to gastro-oesophageal reflux, with consequent stricture formation. We report here a previously unrecorded phenomenon, namely, pharyngo-oesophageal dysphagia due to involvement of the upper oesophageal sphincter.

Case report

The patient was a 45-year-old female with a 2-year history of progressive Raynaud's syndrome, progressive limitation of jaw movement, and increasing dysphagia for solids and liquids which was consistently localised to the neck. She had also gradually developed exertional dyspnoea progressing to acute dyspnoea, with frothing at the mouth in the month before admission. These attacks lasted several hours, and occurred every 1-2 days. She was admitted elsewhere and transferred to Hope Hospital. On admission she was very dyspnoeic at rest, with frequent frothing at the mouth. There were typical changes of scleroderma in the face and hands. The chest expansion was restricted and the lungs had coarse crepitations.

Investigations were as follows: erythrocyte sedimentation rate 7 mm/h; haemoglobin 15.1 g/dl; white cell count normal; blood urea 2.2 mmol/l; sodium 139 mmol/l; potassium 5.2 mmol/l; PO_2 7.8 kPa, PCO_2 5.06 kPa; chest x-ray normal; rheumatoid factor test negative; antinuclear factor + ve; IgG 1/10 000, nucleolar staining pattern;

DNA binding normal; extractable nuclear antigen negative. C3, 253 g/l; C4, 122 g/l; IgG, 23.3 g/l; IgA, 2.6 g/l; IgM, 1.4 g/l; CPK, 120 IU/l (normal = 80).

The histological appearance of a skin biopsy was typical of scleroderma. A previous barium swallow examination had shown narrowing of the cervical oesophagus with overspill of barium into the trachea. Aspiration pneumonia secondary to oesophageal obstruction was diagnosed. Though the patient was too ill for endoscopy, Maloney tapered mercury bougies passed into the stomach without resistance.

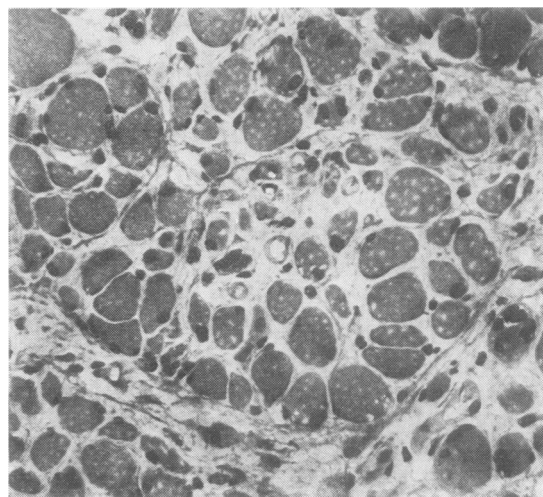


Fig. 1 *Degenerate muscle fibres separated by an oedematous matrix. (Haematoxylin and eosin, $\times 77$).*

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She was treated with physiotherapy and antibiotics but eventually required a tracheotomy and positive pressure ventilation. She improved slowly, and nasogastric feeding was started. She continued to have severe dysphagia for liquids and saliva localised to the pharyngo-oesophageal junction.

Oesophageal manometry showed poor oesophageal peristalsis, a normal lower oesophageal sphincter, but no relaxation of the upper oesophageal sphincter in response to pharyngeal contraction. pH studies

indicated no acid reflux. An endoscope could not be negotiated past the upper oesophageal sphincter.

Operative exploration of the neck revealed diffuse involvement of all neck muscles, with marked thickening of those of the upper oesophagus and of the pharyngeal constrictors. The distal cervical oesophagus was less affected. Removal of a longitudinal strip of muscle from the pharynx and oesophagus increased the lumen of the oesophagus considerably.

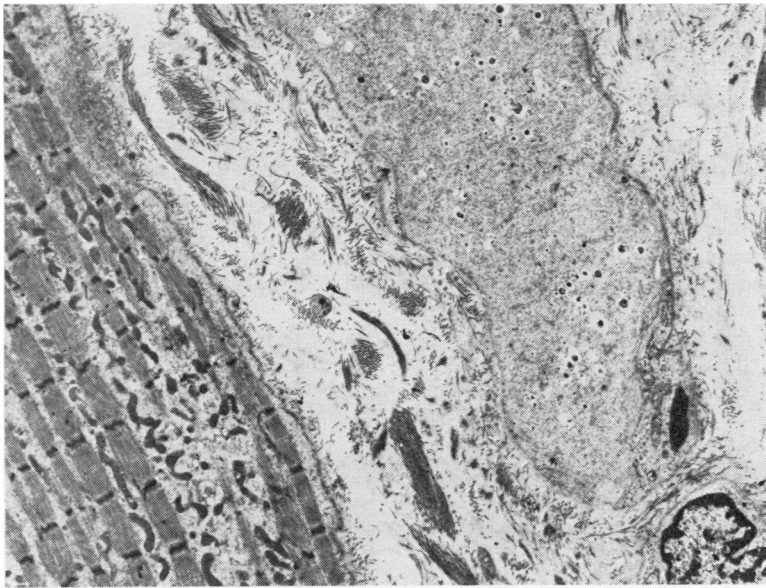


Fig. 2 Collagen between a degenerate muscle fibre (left) and a necrotic fibre (right). (Electron micrograph, $\times 3290$).

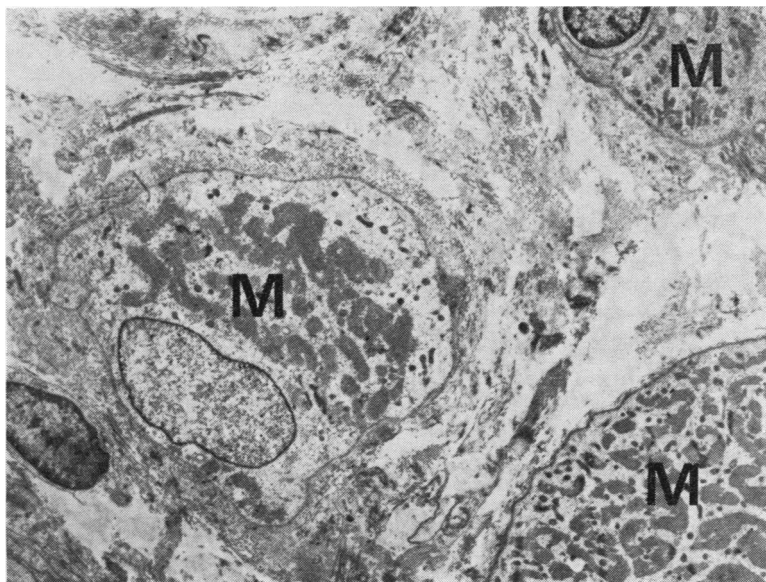


Fig. 3 Muscle fibres (M) separated by a collagenous matrix and showing widened intermyofibrillar spaces. (Electron micrograph, $\times 4444$).

Histological and ultrastructural examination of the specimen revealed muscle fibres separated to a variable extent by an oedematous collagenous matrix (Fig. 1). The muscle fibres showed varying degrees of atrophy and degeneration (Fig. 2) with conspicuous widening of the intermyofibrillar spaces (Fig. 3). Vasculitis and inflammatory cell infiltration were absent.

After this operation the swallowing improved, with no aspiration into the trachea. It was impossible, however, to wean her off the ventilator, as the vital capacity was only 300 ml. Eventually she died after a prolonged respiratory arrest. Permission for a necropsy was refused.

Discussion

Systemic sclerosis is known to affect the full thickness of the oesophagus. Involvement of the mucosa appears to be of no importance, but infiltration of the muscle and destruction of Auerbach's plexus leads to poor propulsive activity and laxity of the lower oesophageal sphincter.^{1 2} Gastro-oesophageal reflux occurs and peptic strictures develop rapidly, possibly hastened by the reduced clearance of refluxed acid. The upper third of the oesophagus is usually spared.²

Pharyngo-oesophageal dysphagia may occur as a reflex mechanism in patients with gastro-oesophageal reflux.^{3 4} In the present patient there was no evidence of reflux either on barium examination or during monitoring with a pH probe in the lower oesophagus. Although propulsive activity as demonstrated by oesophageal manometry was poor, the lower oesophageal sphincter was of normal tone, and the principal abnormality appeared to be the

complete inability of the upper oesophageal sphincter to relax in response to pharyngeal contraction. The dysphagia resulting from this was aggravated by the involvement of the mouth and pharynx by the disease. At operation gross thickening of the muscles of the lower pharynx and upper oesophagus was obvious, with the pathological changes of systemic sclerosis. The distal cervical oesophagus was less severely affected.

So far as we are aware only 1 patient with scleroderma has previously been reported as having dysphagia localised to the neck.⁵ Cruse and his colleagues, however, did not think that this single case was significant. We have now seen 3 further patients with dysphagia localised to the neck, but have not yet had the opportunity to investigate them adequately. It is possible that this type of dysphagia is more common in systemic sclerosis than has been previously realised.

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