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Spontaneous esophageal rupture in adult dermatomyositis

Abstract  Dermatomyositis is a chronic inflammatory myopathy with severe prognosis. A 57-year-old woman suffering from dermatomyositis is presented who, in the course of the disease, developed acute spontaneous esophageal rupture due to dermatomyositis involvement of the esophagus. She was successfully treated with total esophagectomy and stomach interposition. This is the first report of spontaneous rupture of esophagus in dermatomyositis.

Key words  Esophagus • Dermatomyositis • Rupture • Esophagectomy

Case report

At the beginning of 1991 a 57-year-old Caucasian housewife developed a diffuse erythematous skin rash followed by bilateral proximal muscle weakness of the upper and lower limbs and swelling of both hands and feet. The diagnosis of dermatomyositis (DM) was made and she was commenced on methylprednisolone 24 mg daily with good response. She kept fairly well until July 1991, when she was re-admitted to our institution after an episode of pyrexia and a 3-week history of dysphagia with occasional regurgitation of fluids. Hematological and biochemical investigations were within normal limits, apart from a moderately raised creatine phosphokinase (CPK; 452 units), low hematocrit (34%), albuminuria and an erythrocyte sedimentation rate (ESR) of 40 mm in the first hour. The ten-silon pharmacologic test was negative for myasthenia, while electromyography studies as well as a left quadriceps muscle biopsy were typical for polymyositis findings. Computed tomography (CT) of the chest and upper and lower abdomen, as well as bronchoscopy, gastroscopy and skin biopsy, were normal. The diagnosis of DM was confirmed and she was commenced on prednisolone 50 mg daily. During the following 3 months she made fluctuating progress in her clinical state, including a severe arthritis of the left elbow, controlled with increased (80 mg) doses of prednisolone.

Towards the end of November 1991, while still in hospital, she started complaining of a mild epigastric pain with no vomiting. The following day the pain suddenly became severe and was located between the scapulae; she also started complaining of a mild dyspnea. On clinical examination, subcutaneous emphysema was found in the neck and pneumomediastinum was also noted on a chest radiography. Esophagography with Gastrografin swallow was reported as normal, but a CT of the thorax with Gastrografin swallow revealed, apart from air (Fig. 1 A), contrast medium in the mediastinum (Fig. 1 B), thus confirming the clinical assumption of spontaneous esophageal rupture. In view of these findings, surgical intervention was undertaken and no attempt at esophagoscopy was made. At right thoracotomy, free air and bile were found occupying the entire mediastinum, but the esophageal tear could not be detected easily. A subpleural nodule, measuring 2.5 cm at its greatest diameter, was found in the right lower lobe of the lung as well as numerous tiny (1-3 mm) nodules. Total esophagectomy with gastric interposition was decided on and performed via a right thoracotomy and coeliotomy. The large subpleural nodule was removed, as was the gallbladder because it was noted to be enlarged and thick. Her postoperative (PO) course was uneventful, requiring 10 h of mechanical ventilation. By the 3rd PO day a remarkable improvement of her signs and symptoms became obvious and the severe erythematous rash on both arms...
Fig. 1  Computed tomography of the chest after Gastrografin swallow. a The presence of pneumomediastinum is noted. A few nodules are detected in both lung fields. The esophagus is intact at this level. b Contrast medium is present outside the esophageal lumen in the posterior mediastinum (arrow). Normal esophagus is indicated by arrowhead.

Fig. 2  a Muscular layer of the intact esophageal wall adjacent to the tear. Atrophic muscle fibers (arrows) embedded in fibrotic stroma and marked edema separating the muscle bundles (arrowhead). HE, x100. b Muscle fibers showing vacuolar degeneration (arrow), inflammatory cells (arrowheads) and edema in the muscular layer of the esophageal wall. HE, x400

subsided significantly. She was discharged home on the 20th PO day with minimal doses of prednizolone. A year later, however, she developed an anastomotic stricture which was easily treated with dilatation in one treatment. Four years have passed and she remains asymptomatic with no complaints; she requires 10 mg prednizolone daily for maintenance.

Histology showed a 5 mm transmural tear of the middle esophageal section surrounded by necrosis and hemorrhage. The adjacent submucosa was edematous and infiltrated by polymorphs. The muscle layer was thinned and acute inflammatory cells infiltrated the degenerative muscle fibers, which were separated by edema. Atrophic muscle bundles and interstitial fibrosis with relatively little inflammation were also present (Fig. 2). These findings were in keeping with DM involvement of the esophagus. The excised pulmonary nodule consisted of distorted lung parenchyma with unevenly thickened interalveolar septa occupied by dense connective tissue and infiltrated by scattered lymphocytes. These findings were in keeping with interstitial lung fibrosis and nodular scarring associated with DM. The gallbladder showed cholesterolosis and mild chronic cholecystitis.

Discussion

Dermatomyositis is a chronic inflammatory myopathy that may involve all thoracic organs including the esophagus, lung and heart [2, 5, 7]. Urgent life-threatening complications in association with DM very rarely occur. In fact, a few urgent cases of circulatory and respiratory involvement have been reported to date. Cicuttini and Fraser [2] presented a case of recurrent pneumomediastinum in adult
DM and suggested that this may be due to pulmonary vasculitis. Pereira et al. [7] reported a case of pericardial tamponade in juvenile DM. Kobayashi et al. [6] described the case of a 49-year-old woman suffering from DM, who developed acute pneumomediastinum in the presence of interstitial pneumonia. This patient, despite all efforts, died in respiratory failure.

Our patient, who also had lung involvement by DM in the form of nodular scarring, presented with pneumomediastinum and subcutaneous emphysema in the neck. The fact that the patient had previously complained of dysphagia raised the suspicion of severe esophageal involvement and led us eventually to investigate for the possibility of spontaneous perforation of the esophagus. Esophagography was presumably not conclusive because, as was demonstrated histologically, the transmural esophageal tear was small (5 mm) and a leakage could not be detected. However, the diagnosis was easily confirmed by CT of the chest with gastrografin swallow. In our case there was no associated malignancy, although this possibility has been well documented [1].

Spontaneous esophageal rupture in adult DM has never been reported before. However, Cook et al. in 1963 [3] described a type of narcotizing juvenile DM with a 58% fatal outcome. In autopsies, they found arteritis and phlebitis with muscular, neural and fat necrosis involving the wall of the intestinal tract, but with no particular reference to the esophagus. Nevertheless, very recently Disdier et al. [4] reported a case of extensive pharyngeal necrosis, an unusual complication of DM causing dysphagia and dysphonia. Our report is the first case of DM with typical esophageal involvement complicated by necrosis and perforation.

As far as the surgical management is concerned, we believe that total esophagectomy was the treatment of choice, because the tear was not easily detectable at the operative field. Furthermore, the organ was apparently diffusely involved by the disease, thus precluding a safe anastomosis. For these reasons we preferred to approach and mobilize the esophagus through a right thoracotomy and not through a left thoracoabdominal incision, which may be preferable for repairing an esophageal tear in other cases. It is unwise to attempt primary repair of a poorly functioning esophagus, as can be seen in scleroderma [8]; this should also apply to DM. Following surgery, our patient showed remarkable clinical improvement. We believe this was not related to the esophagectomy but was rather due to the continuing treatment with prednizolone.

In summary, esophageal perforation in association with DM is a very rare complication. It may be confused with pneumomediastinum, due to interstitial lung infiltration, but CT of the chest with swallow of contrast material will confirm the former. In our opinion, total esophagectomy is the treatment of choice.

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References