CASE REPORT

Spontaneous pneumomediastinum: an unusual but serious complication of dermatomyositis

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SUMMARY

Spontaneous pneumomediastinum is an unusual but severe and potentially fatal complication of dermatomyositis. The authors reported a case of a 60-year-old woman who was referred for investigation of a persistent cough and dyspnoea. On examination she had features diagnostic of dermatomyositis and she was started on prednisolone. A chest X-ray demonstrated bi-basal reticulonodular changes. A High-resolution CT (HRCT) scan of the chest was arranged and when she had this undertaken 4 weeks after her initial review, gross surgical emphysema, extensive pneumomediastinum and small pneumothoraces were seen. She reported no new symptoms and in particular no deterioration in her dyspnoea. She was treated conservatively with a good outcome.

BACKGROUND

Pneumomediastinum is an unusual but potentially fatal complication of dermatomyositis. We are aware of published reports of only 49 cases. It is almost invariably associated with interstitial lung disease. It usually presents with acute onset of retrosternal chest pain, dyspnoea and swelling of the face and neck from surgical emphysema. Our case is unusual as there was no increase in dyspnoea associated with the development of pneumomediastinum and therefore it is important that physicians need to have a high degree of awareness for the development of this complication.

CASE PRESENTATION

A 60-year-old lady, who had never smoked, was referred with a 3-month history of a persistent unproductive cough associated with fatigue and weight loss. She had developed dyspnoea on exertion over the past month. She had a history of psoriasis since the age of 15 years which she felt had deteriorated and she had developed a rash across her face and upper chest since a holiday in Lanzarote 4 months earlier. When seen in the respiratory department examination revealed bi-basal inspiratory crackles heard to the mid-zones. Her chest radiograph (figure 1) showed reticulonodular shadowing particularly at the bases. A high-resolution CT (HRCT) scan of the chest was arranged as an outpatient and she was referred to the dermatologists for investigation of her rash. She was noted to have Gottron's papules across the knuckles, periungal erythema and a rash on her forehead, eyelids, upper back (shawl sign) and 'V' of her upper chest. She had developed some symmetrical proximal lower limb weakness and muscle

tenderness. A diagnosis of dermatomyositis was made.

INVESTIGATIONS

Creatine kinase was 200 U/L (26–192), alanine transaminase normal, C reactive protein 40 (0–10), antinuclear antibody negative, SS-A (Ro) positive, SS-A (La) negative, RNP, Sm, Scl-70, Jo-1 all negative, c and perinuclear-antineutrophil cytoplasmic autoantibody negative, immunoglobulins and C3/4 complement normal. HRCT scan of the chest (figure 2) showed gross surgical emphysema extending across the anterior thoracic wall into the neck and axilla bilaterally with associated small pneumothoraces and extensive pneumomediastinum with multiple locules. The lungs showed interstitial lung disease with honeycomb pattern in the lung bases.

TREATMENT

She was admitted directly from the radiology department after her HRCT scan of the chest. She did not describe the development of any new symptoms in the 4 weeks between her chest X-ray and HRCT scan of the chest and in particular had had no retrosternal chest pain, face swelling or increase in dyspnoea. She had clinical signs of surgical emphysema of the anterior chest wall and neck and the inspiratory crackles heard on

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Figure 1 Chest X-ray.

Figure 2 High-resolution CT of the chest.

auscultation of the chest were unchanged. Hamman's sign was not appreciated. She was hypoxic in type I respiratory failure with a paO_2 on air of 6.1 kPa, paCO_2 4.7 kPa, pH 7.5. She was treated with high-flow oxygen and initial bed rest for her pneumomediastinum. Prednisolone was increased to 1 mg/kg/day for her dermatomyositis with resolution of the myalgia and significant improvement of her rash within 2 weeks. Her pneumothoraces resolved and surgical emphysema greatly improved by 2 weeks but the pneumomediastinum persisted. She was discharged home on oxygen therapy at 4 L/min.

OUTCOME AND FOLLOW-UP

A follow HRCT scan at 6 months showed no pneumothorax, no surgical emphysema and minimal residual pneumomediastinum. There was persistent interstitial lung disease with marked improvement in the ground glass opacification but residual interlobular septal thickening and associated traction bronchiectasis. The patient remains on long-term oxygen therapy with significant restriction of daily activities as a result of her breathlessness.

DISCUSSION

DM is an idiopathic inflammatory myopathy resulting in proximal muscle weakness characterised by a variety of skin manifestations. It is a multisystem disorder associated with interstitial lung disease (ILD), oesophageal weakness leading to dysphagia, cardiac involvement with myocarditis and increased risk of myocardial infarction, as well as an increased risk of malignancy.

ILD is present in 10–43% of cases and is associated with a poor outcome.¹ In the majority of cases ILD either precedes or occurs simultaneously with the development of DM. Non-specific interstitial pneumonia (NSIP) pattern of ILD is most frequently encountered in DM and follows a similar course to patients with idiopathic NSIP with a survival rate better than that of idiopathic pulmonary fibrosis. When a usual interstitial pneumonia pattern does occur it is associated with rapidly progressive disease.^{2,3}

Pneumomediastinum is a rare and potentially fatal complication of DM. In a review of 31 patients with pneumomediastinum and DM mortality was 34% with 25% dying within the first month and cumulative estimated Kaplan-Meier survival rate

of 55% at 2 years.⁴ Poor survival was associated with the absence of muscle weakness or an initial decrease in vital capacity or carbon monoxide diffusion capacity before the onset of pneumomediastinum. Although DM is more common in women, pneumomediastinum complicating DM was slightly more frequent in men and occurred in younger patients (mean age 34 years).⁵

Spontaneous pneumomediastinum is usually a self-limiting condition. Attempt to identify precipitating factors, such as the Valsalva manoeuvre, illicit drug ingestion, vigorous vomiting or cough or activities that may lead to barotrauma (eg, scuba diving, flying). It results from rupture of alveoli adjacent to the mediastinum as a result of marked increases in intra-alveolar pressure. The mechanism underlying alveolar rupture in patients with DM is not clear but is thought to involve either distortion of lung tissue architecture due to ILD or vasculitic lesions within the respiratory tract resulting in bronchial or alveolar wall necrosis.⁶

Treatment for pneumomediastinum is usually conservative with high-flow oxygen, to support hypoxia and wash out nitrogen from the blood increasing the partial pressure gradient for gas absorption. There is some debate over the role of steroids and immunosuppressive agents. In our case and the majority of cases in the literature patients had already been started on corticosteroids. It has also been suggested that weakening of the interstitial tissue of the lungs by corticosteroids might favour the development of pneumomediastinum.⁷ Patients started on corticosteroids have been found to have a worse prognosis,⁸ however it is unclear whether this is associated with the corticosteroids or whether the use of steroids reflects more severe or progressive ILD. There are few reports of the use of immunosuppressive therapy but in one series seven out of eight patients had resolution of the spontaneous pneumomediastinum with no mortality and in a case of refractory DM complicated by spontaneous pneumomediastinum without ILD rituximab was used successfully.⁹

Learning points

- ▶ Pneumomediastinum is an unusual complication of dermatomyositis and is associated with a high mortality.
- ▶ It is usually associated with interstitial lung disease but the best treatment regimen is unclear. It has been postulated that corticosteroids may precipitate pneumomediastinum in patients with dermatomyositis-interstitial lung disease.
- ▶ It usually presents with chest pain and breathlessness but can be asymptomatic and as such physicians treating patients with dermatomyositis need to have a high index of suspicion.

Contributors JA was involved in the initial diagnosis, treatment and follow-up of the patient and also obtained the consent and drafted the initial manuscript and also was involved in all stages of editing this case report. SP was involved in the review and editing of the initial draft and was the consultant involved in the treatment of the patient. GW was also involved in contributing to the initial draft. LMD was involved in editing and also in the literature search of the above case.

Competing interests None.

Patient consent Obtained.

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