Case report

Spontaneous pneumomediastinum in adult dermatomyositis

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SUMMARY Spontaneous pneumomediastinum has not been reported in adult polymyositis or dermatomyositis, either in conjunction with spontaneous pneumothorax or in isolation. Spontaneous pneumothorax has been rarely reported as a complication of rheumatoid arthritis, systemic lupus erythematosus, scleroderma, and childhood dermatomyositis. It is associated with active, progressive pulmonary involvement and a poor prognosis. We describe an adult with dermatomyositis and spontaneous pneumomediastinum with a favourable outcome.

Key word: polymyositis.

Spontaneous pneumomediastinum is a rare event which occasionally complicates spontaneous pneumothorax. Several pathogenetic mechanisms have been proposed for spontaneous pneumothorax in rheumatoid arthritis (RA),^{1–3} systemic lupus erythematosus (SLE),^{4–6} scleroderma (PSS),^{7,8} and childhood dermatomyositis.⁹ In these reported cases pulmonary involvement by the connective tissue disease was active and progressive. Many of the patients died from their pulmonary disease. In contrast, we describe a patient with adult onset dermatomyositis who developed subcutaneous emphysema and pneumomediastinum which resolved without any complications or specific interventions.

Case report

This 42 year old white man presented in July 1984 with an erythematous rash, crusted lesions in sun exposed areas, swelling and tenderness of the face and arms, and complaints of weakness in his legs. Pertinent laboratory studies included a creatine phosphokinase of 2260 IU (normal 50–180 IU) and an ethanol level of 278 mg/dl (2.78 g/l). The chest

roentgenogram showed increased interstitial markings, and a room air arterial blood gas showed a P_{O_2} of 77 mmHg and a P_{CO_2} of 41 mmHg. In August 1984 the patient presented with difficulty in swallowing, regurgitation, and inability to sit up or raise his head in bed. He had a classic heliotrope rash on the face and chest with periorbital oedema. Gottron's papules and nailfold capillary dilatation were also present. Profound weakness of the trunk and proximal muscles was noted, and an electromyogram showed typical changes of inflammatory myopathy, with increased insertional activity, positive sharp waves, low amplitude short duration polyphasic motor unit potentials, and fibrillation. Therapy was initiated with prednisone 60 mg/day, with reduction of swelling but little increase in strength. The patient was readmitted in September 1984 because of deterioration in muscle power. Intravenous methylprednisolone 80 mg/day was begun in divided doses, but the patient developed bilateral aspiration pneumonia and required mechanical ventilation. The hospital course was further complicated by cholelithiasis, multiple skin ulcers, and weight loss of 19 kg. Intravenous pyelogram, air contrast barium enema, and flexible sigmoidoscopy did not reveal a malignancy. After resolution of the pneumonia and cholelithiasis, therapy was changed to azathioprine 75 mg/day and prednisone 100 mg/day in divided doses. The patient's strength

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improved but he was unable to stand or walk unassisted at discharge from the hospital. Repeat arterial blood gases and chest roentgenogram were unchanged from his previous baseline. The patient continued regaining strength and weight, but by November 1984 the skin ulcers had deepened and a long sinus tract had developed from the right antecubital fossa extending up the arm, with gross purulent drainage and local cellulitis. The ulcers responded to intravenous antibiotics and an increase in azathioprine to 100 mg/day with a concomitant reduction in prednisone to 80 mg/day.

The patient's skin ulcers improved and he regained the ability to arise unassisted and walk on flat surfaces. In February 1985 he developed asymptomatic enlargement of the neck with palpable subcutaneous emphysema but no local ulcers. The patient noted a morning cough productive of small amounts of whitish sputum, but denied fever, chills, chest pain, and dyspnoea. Examination of the nose, oropharynx, and chest was unremarkable except for subcutaneous emphysema in a mantle distribution. Roentgenograms showed subcutaneous, prever-

tebral, and retropharyngeal air, and extensive pneumomediastinum without concurrent pneumothorax (Fig. 1). The patient refused admission to hospital but returned for outpatient follow up. After one month mediastinal emphysema had nearly resolved radiographically, but Hamman's sign, a precordial 'crunch' with systole, was noted. By two months the subcutaneous emphysema resolved. The patient's clinical condition continued to improve, with healing of the skin ulcers. His strength improved and he gained weight, allowing reduction in the prednisone dose. Pulmonary function tests performed in April 1985 showed normal spirometry and lung volumes, but a diffusion capacity for carbon monoxide was only 40% of predicted.

Discussion

Spontaneous pneumomediastinum is a rare event which occurs most frequently in males in the third decade.¹⁰ The development of spontaneous pneumomediastinum is thought to be due to leakage of air through 'non-partitional' alveolar walls and subsequent dissection medially along pulmonary vessels to the mediastinum.¹¹ Air dissecting peripherally may remain trapped as subpleural blebs or, if the visceral pleura ruptures, may escape into the pleural space, resulting in pneumothorax. Spontaneous pneumomediastinum in isolation is usually benign, particularly if there is a low pressure release of mediastinal air into subcutaneous tissues of the neck and chest. High pressure air in the mediastinum may result in 'air block' with obstruction of pulmonary vascular flow and dampening of respiratory excursion, sometimes called 'malignant pneumomediastinum'.

Several pulmonary diseases are associated with spontaneous pneumothorax, including asthma, chronic bronchitis, emphysema, bronchiectasis, pneumonia, abscess, primary or metastatic cancer, and chronic interstitial lung disease.¹⁰ Since disruption of the alveolus or airway can result in pneumothorax, pneumomediastinum, or both, these associations seem relevant for spontaneous pneumomediastinum and spontaneous pneumothorax.

Causes of spontaneous pneumothorax in RA include rupture of a pleural necrobiotic nodule and rupture of a subpleural bleb.¹⁻³ The latter mechanism is also described in SLE and scleroderma, with bleb formation resulting from interstitial pneumonitis with cystic degeneration of alveoli.⁴ Subpleural pulmonary infarctions from diffuse vasculopathy have been implicated in childhood dermatomyositis.⁹ One case of spontaneous pneumomediastinum with bilateral pneumothoraces has been described in

Fig. 1. Chest roentgenogram showing pneumomediastinum and extensive subcutaneous emphysema in the neck and thorax, but no pneumothorax.

SLE.⁵ No subpleural blebs were found, but diffuse interstitial pneumonitis was present.

One of the risk factors for developing 'malignant pneumomediastinum' is the presence of significant inflammatory pulmonary disease.¹¹ Adult polymyositis and dermatomyositis are frequently complicated by aspiration pneumonitis and interstitial pneumonitis. The frequency of interstitial pneumonitis in polymyositis is usually estimated at 5–10% but may be as high as 30%.¹² Our patient manifested both aspiration pneumonitis and interstitial pneumonitis, but his lung disease was not severe or life threatening at the time he developed spontaneous pneumomediastinum. Likewise, pneumomediastinum did not occur in the setting of intubation and positive pressure ventilation, infection, obstructive airway disease, or malignancy. Spontaneous pneumomediastinum has a reported incidence of two per 10 000 in a college population,¹³ and 88% of cases occur in people under 40 years of age.¹⁴ Polymyositis has a reported incidence of about three per million in white males.¹⁵ The probability of these conditions randomly coexisting in this patient is about seven per hundred billion. This case illustrates that dermatomyositis may be associated with spontaneous pneumomediastinum, probably by the same mechanism as described in SLE, but the spontaneous pneumomediastinum may have a benign course and outcome.

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