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Case Report

Persistent Pneumomediastinum and Dermatomyositis: A Case Report and Review of the Literature

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Abstract: We describe a 42-year-old man with dermatomyositis and interstitial lung disease who presented with anterior neck pain and dyspnoea. Chest radiographs showed subcutaneous emphysema, pneumomediastinum and diffuse reticulonodular infiltration in both lungs. After the administration of high doses of prednisolone, an improvement of pulmonary function and respiratory symptoms was observed but the pneumomediastinum persists 12 months after diagnosis, and without any complication. We review the cases that have been reported thus far of pneumomediastinum associated with dermatomyositis and discuss the possible mechanisms involved. We conclude that pneumomediastinum is not an uncommon complication of dermatomyositis and that its aetiopathogenesis remains very unclear.

Keywords: Dermatomyositis; Interstitial lung disease; Pneumomediastinum

Case report

A 42-year-old man was admitted in February 1999 with a 2-month history of general fatigue and polyarthralgia. He also noticed mild myalgias and moderate proximal muscular weakness. Clinical examination revealed low-grade fever, Gottron's papules on the metacarpophalangeal and proximal joints, and a periorbital heliotrope rash. Routine laboratory tests were normal, including muscle enzymes. Rheumatoid factor, antiplatelet antibodies, cryoglobulin, antinuclear antibodies, ANCA and anticardiolopin antibodies were all negative. Electromyography showed unequivocal signs of myositis and a quadriceps muscle biopsy revealed moderate necrosis of the muscular fibres. Pulmonary function tests showed a restrictive pattern, with a total lung capacity (TLC) of 5.58 l (73 % of the predicted value), a vital capacity (VC) of 3.41 l (59 % of the predicted value) and a forced expiratory lung volume in 1 second (FEV_1) of 3.30 l (75 % of the predicted value). The diffusion coefficient (DL_{CO}/VA') was 60 % of the predicted value. Chest X-ray showed mild interstitial changes in both lower lobes. A diagnosis of dermatomyositis with interstitial lung disease was made. Search for an occult neoplasm was negative. Glucocorticoid therapy was started (three daily pulses of 1 g methylprednisolone, followed by an oral daily prednisolone dose of 0.5 mg/kg/day), together with oral methotrexate (15 mg/week) and hydroxychloroquine (400 mg/day). The arthralgias, myalgias and muscle weakness improved significantly. However, 5 months later the patient experienced dyspnoea, coughing and parietal neck pain. Clinical examination revealed bilateral inspiratory crackles over the lower chest and subcutaneous emphysema. Chest X-ray and chest CT confirmed the presence of subcutaneous and mediastinal emphysema, as well as a worsening of the interstitial changes. Oesophagogastroduodenoscopy and fibreoptic bronchoscopy were normal, as was the analysis of the bronchoalveolar lavage fluid (2 618 000 cells/ml with 83 % macrophages, 14 % lymphocytes and 2.5 % neutrophils). Histology of the lavage was unremarkable. Given the worsening of the dyspnoea, the patient was given five monthly IV pulses of cyclophosphamide (between 0.75 and 1.5 g/m^2), followed by IV gammaglobulins (0.4 g/kg for 5 consecutive days

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Table 1. Pneumomediastinum and dermatomyositis: review of literature

References	Gender	Age	Concomitant pneumothorax	Previous steroid therapy	Concomitant ILD	PM Presentation Duration	Patient Outcome
Bradley *et al.*. [8]	M	42	–	+	+	Once 2 months	+
Cicuttini *et al.* [9]	F	22	–	+	–	Recurrent ?	+
Carmody *et al.* [10]	M	20	–	+	–	Recurrent 5 months	+
Yamanishi *et al.* [11]	M	23	–	+	+	Once 4 days	Fa
	M	57	–	+	+	Once 2 weeks	Fa
Jansen *et al.* [12]	F	33	+	+	+	Once 6 weeks	+
Isfer *et al.* [13]	F	25	–	–	–	Once > 7 weeks	?
Kobayashi *et al.* [14]	F	49	+	+	+	Once ?	Fa
Gayraud *et al.* [15]	F	55	+	–	?	? ?	+
Santiago *et al.* [16]	M	10	–	+	+	Once 3 weeks	Fa
Satomi *et al.* [17]	F	59	–	+	?	Twice < 1 month	+
Bousquet *et al.* [18]	M	39	?	+	+	? ?	+
De Toro *et al.* [19]	F	41	?	+	+	? ?	Fa
Nagai *et al.* [20]	F	65	?	+	+	? ?	Fa
Jang *et al.* [21]	F	32	?	+	+	? ?	+
Matsuda *et al.* [22]	F	41	?	+	+	? ?	Fa
Kono *et al.* [23]	M	30	?	+	+	Once Few days	+
	M	25	?	+	+	? ?	Fa
	M	23	?	+	+	? ?	+
	F	59	?	+	+	? ?	+
This case	M	42	–	+	+	Once > 12 months	+

ILD, interstitial lung disease; PM, pneumomediastinum; ?, data not available;
Patient Outcome: +, favourable outcome; Fa, fatal; ?, unknown.

followed by a single dose given every 6 weeks). One year after diagnosis, his condition remains severe but stable, with the persistence of subcutaneous and mediastinal emphysema. It is noteworthy that the patient did not experience any septic episode despite heavy immunosuppression.

Discussion

Dermatomyositis and polymyositis are inflammatory connective tissue diseases of unknown aetiology, chronic evolution and multisystemic character, involving predominantly the muscles and the skin. Pulmonary manifestations are present in approximately 10 % of cases [1]. Although the association of interstitial lung disease [2] and lung cancer [3] with polymyositis and dermatomyositis is well established, the occurrence of a spontaneous pneumomediastinum in these conditions is relatively rare. Spontaneous pneumomediastinum has been reported in a patient with fibrosing alveolitis [4] and in a few patients with pulmonary fibrosis associated with rheumatoid arthritis [5], systemic lupus erythematosus [6], or of unspecified aetiology [7]. From a review of the English and French literature, it appears that 23 cases (excluding ours) of pneumomediastinum associated with polymyositis and dermatomyositis have been reported [8–26] since it was first described in 1986 [8]. Detailed data are available in 20 cases (Table 1). The rupture of alveoli adjacent to vessels, with subsequent air spreading to the mediastinum, is thought to be the direct cause of the pneumomediastinum [9], although the precise mechanism is unknown. Circuttini et al. [9] hypothesised that vasculitis was the common denominator leading to pneumomediastinum in dermatomyositis patients, as three reported patients suffered from vasculitic cutaneous ulcers [8–10]. Yamanishi et al., [11] mentioned that the possible weakening of the interstitial tissues of the lung, caused by steroid treatment, might favour pneumomediastinum, as in most reported cases of pneumomediastinum in dermatomyositis patients (18 out of 20 in our review), it occurred after steroid therapy. For Jansen et al. [12] pneumomediastinum seems to occur primarily in patients with pre-existing lung disease (15 out of 20 patients in our review had intercurrent interstitial lung disease). However, Isfer et al. [13] reported a 25-year-old woman diagnosed with dermatomyositis who developed pneumomediastinum before steroids were started, and without associated interstitial disease. More recently, Kono et al. [23] reviewed 48 patients with polymyositis/dermatomyositis: Pneumomediastinum was observed in four. A significant association of pneumomediastinum with cutaneous vasculopathy ($p = 0.02$) was shown, and in one patient bronchial necrosis due to vasculopathy was strongly suspected as being responsible for the pneumomediastinum.

Out of the 20 patients reviewed, four died soon after the onset of the pneumomediastinum (4 days to 3 weeks). Four other patients died, but details regarding the pneumomediastinum are not available. The outcome of one patient is unclear. The pneumomediastinum rapidly improved in the others, although recurrence was observed in three cases.

Our case is unusual compared to those previously reported. The patient developed a persistent pneumomediastinum, which is still present 12 months after onset,

without any complication. He was on long-term steroid therapy, including repeated high-dose methylprednisolone IV pulse therapy. He suffered from interstitial lung disease, which improved on steroids, and had no associated pneumothorax.

Conclusion

We report the case of a 42-year-old man with dermatomyositis, who subsequently developed pneumomediastinum which persists 12 months after diagnosis. Various mechanisms have been proposed to explain this association. However, it should be stressed that pneumomediastinum is not an uncommon complication of dermatomyositis and that its aetiopathogenesis remains very unclear.

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