

CASE REPORT

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A case of dermatomyositis complicated with pneumomediastinum

Received: October 6, 2006 / Accepted: December 19, 2006

Abstract We herein describe a 16-year-old boy with pneumomediastinum, pneumothorax, and subcutaneous emphysema as the initial symptoms of dermatomyositis (DM). His pneumomediastinum disappeared after strict bed rest and he was thereafter successfully treated with oral prednisolone and cyclosporine A. His condition was further complicated with mild interstitial lung disease, arrhythmia, and skin ulcers on his fingertips, right elbow, ear, and sacral region. Pneumomediastinum is a rare complication of DM and its pathogenesis remains unclear. We review the literature and discuss the possible mechanism of this disease.

Key words Dermatomyositis · Pneumomediastinum · Skin ulcer · Subcutaneous emphysema · Vasculopathy

Case report

A 16-year-old boy had been treated at another hospital in March 2001 with complaints of a slight fever, muscle weakness, erythema on his fingers, elbows, and knees, and arthralgia of both hands. He was prescribed topical steroids yet his symptoms continued. Several weeks later, he felt acute chest pain and was hospitalized in a respiratory department. His chest X-ray showed pneumomediastinum, pneumothorax, and subcutaneous emphysema. His pneumomediastinum did not improve in spite of bed rest for 3 weeks and he then was transferred to our hospital on the 27 June. He was suspected of having dermatomyositis from the eruption, muscle weakness, arthralgia, and continuous fever.

On initial examination his height was 182.7 cm, weight 68.5 kg, temperature 37.6°C, pulse 70 beats/min, and blood pressure 114/54 mmHg. A physical examination showed

periungual erythema and Gottron's papule on all fingers, and poorly demarcated erythema on both elbows, knees, and shoulders. Subcutaneous emphysema was felt from the supraclavicular fossa to the cervix. Muscle strength tests were performed which demonstrated slight weakness of muscle (4/5) on his proximal extremities. He did not complain of any spontaneous or grasping pain of the muscle. Laboratory data demonstrated slight anemia (red blood cells 412×10^4 , hemoglobin 11.0 g/dl). White blood cell ($3470/\mu\text{l}$) and platelet count (12.3×10^4) were normal. There were slight elevations of aspartate aminotransferase (33 U/l), alanine aminotransferase (35 U/l), lactate dehydrogenase (339 U/l) C-reactive protein (0.6 mg/dl), myoglobin (85 ng/ml), KL-6 (663 U/ml), and soluble interleukin-2 receptor (1580 U/ml) levels. However, the creatine kinase (CK) (43 U/l) and aldolase (3 U/l) levels were normal. The anti-nuclear antibody and anti Jo-1 antibody were negative. Electromyography showed myogenic change in his deltoid muscle and quadriceps muscle. Chest X-ray and computed tomography showed pneumomediastinum around the aorta and in the pericardial space, plus left pneumothorax and subcutaneous emphysema expanding from the supraclavicular fossa to the cervix (Fig. 1A–C). Slight interstitial changes were also apparent in the left lower lobe (Fig. 1D). A biopsy from the erythema of the right elbow showed lymphocytic infiltration around the capillaries, and mucin deposits between the collagen bundles of mid dermis. Muscle biopsy from the right deltoid muscle revealed infiltration of the lymphocytes around interstitial vessels though degeneration, and atrophy of muscle fibers was not observed. From these findings, the patient was diagnosed to have DM.

Although the pneumomediastinum gradually improved and completely disappeared after strict bed rest for 2 months (Fig. 1E), the other complications persisted. He also developed skin ulcers on his fingertips, right elbow, ear, and sacral region (Fig. 2) during the course of the disease. Thus, oral prednisolone (40 mg/day) in combination with cyclosporine A (125 mg/day) was prescribed. Both the interstitial lung disease and skin lesions gradually improved after 2 months and subsequently prednisolone was successfully tapered to

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Fig. 1. A–C Pneumomediastinum around the aorta and in the pericardial space, left pneumothorax, and subcutaneous emphysema expanding from the supraclavicular fossa to the cervix (arrows). **D** Slight

interstitial changes were also apparent in the left lower lobe (arrows). **E** Pneumomediastinum disappeared after strict bed rest for 2 months

10 mg/day. He was discharged from the hospital in December and no recurrence of the pneumomediastinum has since been observed over the following 4 years.

Discussion

Pneumomediastinum is a rare but characteristic complication of DM. To our knowledge, 22 cases (mean age 35, range 10–65 years) including our case have been reported

in the English literature (Table 1). It has become apparent that patients who develop pneumomediastinum tend to have several clinical features, as summarized by Korkmaz et al.¹² These include interstitial lung disease (ILD), cutaneous vasculopathy, a mild or absent creatine kinase elevation, a younger age, and a history of previous systemic glucocorticoid treatment. We observed all of these risk factors except glucocorticoid treatment in our patient.

Although the mechanism of pneumomediastinum in DM is unknown, three possible mechanisms have been proposed. The first is subpleural cyst formation due to intersti-

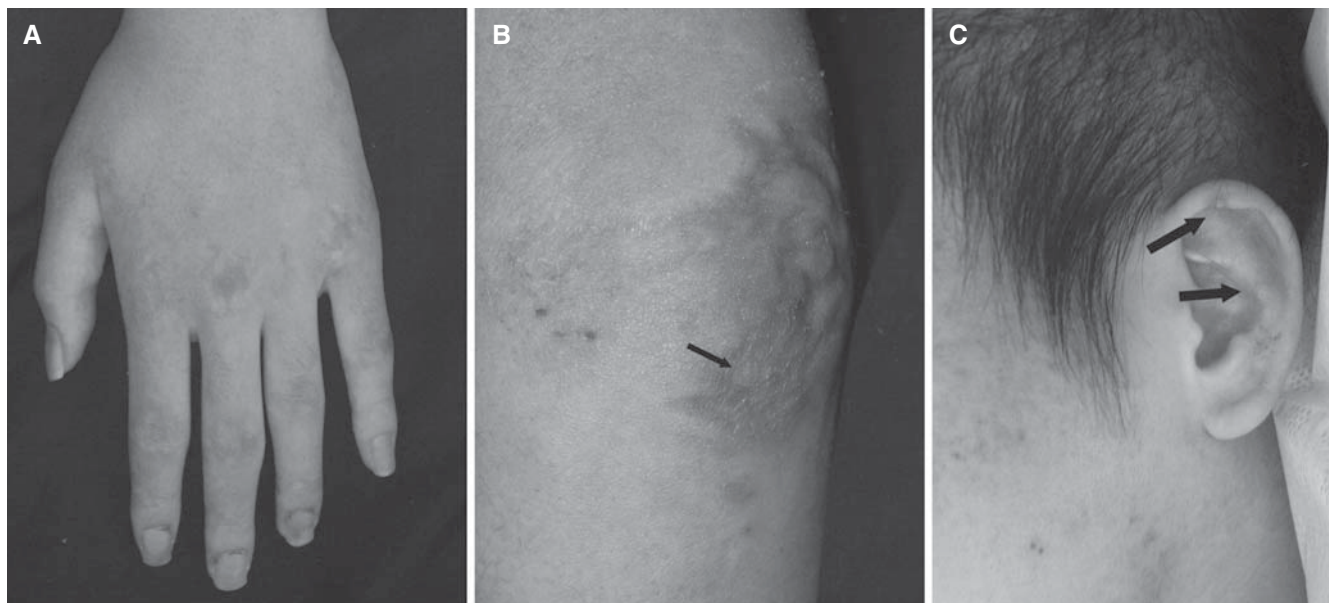


Fig. 2. Skin ulcers were seen on the patient's fingertips (A), right elbow (B), and left ear (C)

Table 1. Cases of dermatomyositis reported in the literature

First author ^{Ref.}	Age (years)/Sex	Previous steroid therapy	Muscle symptom	ILD	Skin ulcer	Outcome
Matsuda ¹	41/F	+	+	Severe	-	Fatal
Santos ²	41/F	+	+	Severe	NR	Fatal
Nagai ³	65/F	+	+	Severe	-	Fatal
Santiago ⁴	10/M	+	-	Severe	NR	Fatal
Yamanishi ⁵	23/M	+	+	Severe	NR	Fatal
	57/M	+	+	Severe	NR	Fatal
Kono ⁶	25/M	+	+	Severe	+	Fatal
Barvoux ⁷	42/M	+	+	Severe	NR	Alive
Bradley ⁸	42/M	+	+	Mild	+	Alive
Ogawa ⁹	50/F	+	+	Mild	NR	Alive
Bousquet ¹⁰	39/M	+	-	Mild	NR	Alive
Jang ¹¹	32/F	+	-	Mild	+	Alive
Korkmaz ¹²	28/M	+	+	Mild	NR	Alive
Jansen ¹³	33/F	+	+	Not active	NR	Alive
Kono ⁶	30/M	+	+	Stable	+	Alive
	23/M	+	+	+	+	Alive
	59/F	+	+	+	-	Alive
Kuroda ¹⁴	46/M	+	+	+	-	Alive
Carmody ¹⁵	20/M	+	+	-	NR	Alive
Cicuttini ¹⁶	22/F	+	-	-	+	Alive
Isfer ¹⁷	25/F	-	+	-	+	Alive
Our case	16/M	-	+	Mild	+	Alive

ILD, interstitial lung disease; NR, not recorded

tial fibrosis of the lung.¹³ A rupture of the cyst results in pneumomediastinum. The second is vasculopathy.¹⁶ A disruption of the mucosal barrier due to vasculopathy is implicated in the development of pneumomediastinum. The third is a weakening effect of glucocorticoids on the interstitial tissues of the lung.⁵

In our patient, it can be assumed that vasculopathy had played a major role in the development of pneumomediastinum because he developed it before the administration of glucocorticoid and his ILD was mild at the time he developed pneumomediastinum. His skin ulcers on the fingertips,

right elbow, ear, and sacral lesions also suggested the presence of vasculopathy. We waited until his pneumomediastinum disappeared spontaneously before administering glucocorticoid because we thought that glucocorticoid would retard the spontaneous absorption. We started prednisolone in combination with cyclosporine since his fever, muscle weakness, and skin ulcers persisted.

Kono et al.⁶ reviewed 48 patients (mean age 49, range 7–81 years) with DM/polymyositis while focusing mainly on the presence of pneumomediastinum and cutaneous vasculopathy. Between them, four patients had complications

with pneumomediastinum and 3/4 (75%) developed cutaneous vasculopathy while one developed bronchial wall necrosis. They concluded that there was a significant association of pneumomediastinum with cutaneous vasculopathy and a young age but not with the prevalence of lung disease. According to our review of previous reports, 12 patients were described with skin symptoms and eight among them (66.7%) presented with skin ulcers, indicating the presence of vasculopathy. All of the four patients who did not present with skin ulcers had interstitial lung disease and previous steroid therapy. We consider that all of these factors plus an event to increase intra-alveolar pressure such as severe cough and pulmonary function test¹⁰ may act synergically to cause pneumomediastinum.

Pneumomediastinum in DM has been considered to be a symptom predictive of a poor prognosis.¹ In fact, 7/22 cases (37.5%) we reviewed were fatal. We have examined the clinical course of fatal cases and noticed that most of them died of respiratory failure because of uncontrollable or rapidly progressing ILD. All of them had severe ILD and 1/7 (14.3%) presented with skin ulcer. Of the responders, only one (6.7%) had severe ILD and seven (46.7%) presented with skin ulcer. This suggests that the prognosis depends on the progress of ILD. Among the patients we reviewed, eight had complications with severe ILD and 7/8 (87.5%) were fatal. On the other hand, the mortality of patients with mild or absent ILD was 0% (0/14). As mentioned by Nagai et al.,³ we consider that pneumomediastinum in DM can thus be divided into two groups. One group develop complications with severe ILD with or without vasculopathy and high mortality, while the other group shows signs of vasculopathy such as skin ulcer and bronchial wall necrosis with mild or no ILD and a favorable prognosis.

We herein reported a rare case of a patient with DM who developed pneumomediastinum, pneumothorax, and subcutaneous emphysema as the initial symptoms. Although our patient developed several skin ulcers, his ILD responded to glucocorticoid and cyclosporine and he has since shown a favorable response.

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