

Spontaneous pneumomediastinum and cutaneous ulcers complicated in a patient with dermatomyositis and interstitial lung disease

Mihye Kwon *Department of Internal Medicine, School of Medicine, Konyang University, Daejeon, South Korea*

In May 2020, a 42-year-old man with a history of dermatomyositis (DM) and interstitial lung disease (ILD) presented with several days of sudden painful swelling in the neck and chest. He was diagnosed as DM eight months ago based on the positive findings on proximal muscle weakness, lower extremity magnetic resonance imaging (MRI) scans, muscle enzyme elevation, electromyography (EMG) and muscle biopsy findings with typical facial rash. He was treated with methotrexate (20 mg/week) and prednisolone 20 mg/day. The anti-Jo-1 was negative, while anti-melanoma differentiation-associated gene-5 (MDA5), and anti-Ro52 antibody were positive. Vital signs were stable and laboratory results were unremarkable. On physical examination, crepitus was palpable around his neck, and ulcers were observed on the Rt. hand and elbow (Figure 1a, b). High-resolution computed tomography showed subcutaneous emphysema and pneumomediastinum without aggravation of previous ILD (Figure 1c). After administering intravenous methylprednisolone 1 mg/kg/day, pneumomediastinum and subcutaneous emphysema improved rapidly, and he was discharged on Day 12. However, a relapse occurred within one week, and he received the

same dose of intravenous steroid with additional cyclosporine. Pneumomediastinum completely resolved (Figure 1d) and digital ulcers considerably subsided. The patient has been followed for 10 months without any relapse.

Pneumomediastinum is a rare complication of DM, and its development has been associated with amyopathic dermatomyositis (ADM), ILD, anti-MDA5 antibody, and particularly with cutaneous ulcers.^{1,2} In a case report, the authors reviewed 15 cases of pneumomediastinum and suggested that it was a complication of DM, but not of pneumomediastinum.³ The authors concluded that vasculitic activity associated with interstitial pneumonitis should be kept in mind in case of spontaneous pneumomediastinum, particularly in young patients with recent disease and cutaneous vasculopathy. Pathogenesis of pneumomediastinum in DM has been considered strongly associated with ILD that one hypothesis is that alveolar rupture secondary to distortion of architecture with an increased intra-alveolar pressure in severely fibrosed lung causes pneumomediastinum. The other one is that pneumomediastinum may develop due to the air leakage secondary to vasculitis.^{4,5} Cutaneous

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Correspondence: Mihye Kwon, MD. School of Medicine, Konyang University, Internal Medicine, 35365 Daejeon, South Korea.
Tel: 821030763029 e-mail: mhkwon@kyuh.ac.kr

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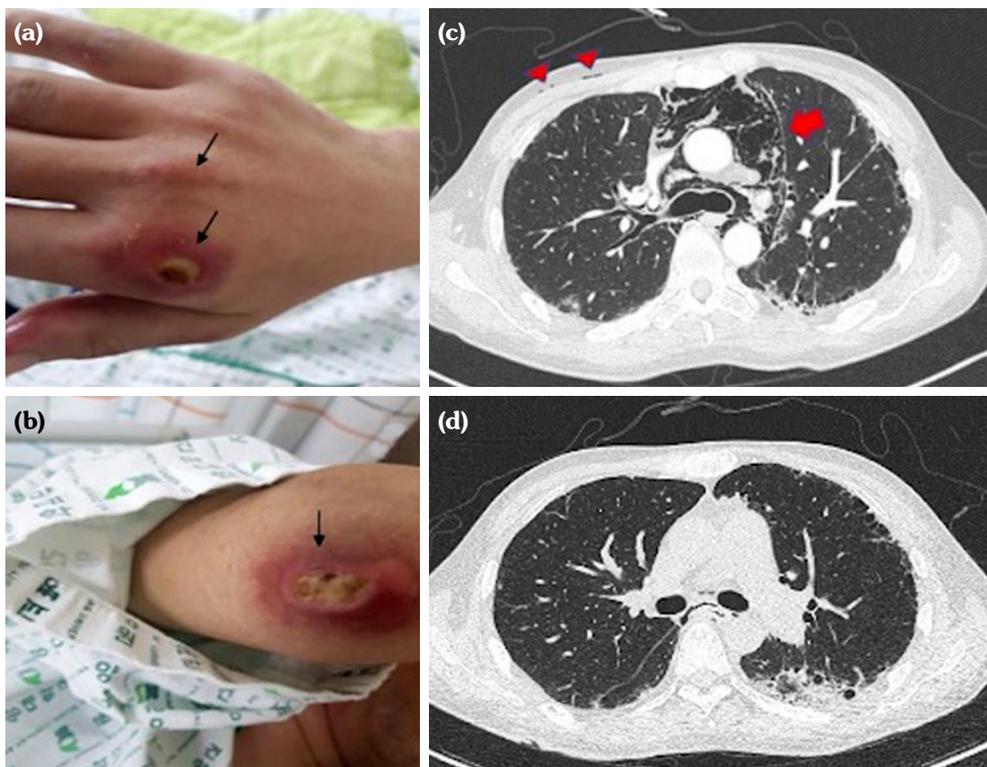


Figure 1. Cutaneous ulcers and HRCT. **(a)** A 7×7 mm-sized deep cutaneous ulcer with plaque was seen and papules were seen on knuckles (thin arrows). **(b)** A 1×1 cm-sized deep ulcer with skin papule on the Rt. elbow. **(c)** Initial HRCT showed air lucency in the mediastinum indicating pneumomediastinum (red arrow) and subcutaneous emphysema (red arrow heads). **(d)** Follow-up HRCT at discharge showed resolution of pneumomediastinum.

HRCT: High-resolution computed tomography.

ulcers are well-known manifestations of vasculopathy and are more frequently associated with pneumomediastinum in DM, regardless of ILD.³⁻⁵ Additionally, anti-MDA-5 antibody, a subset of myositis-specific antibodies known to be related to rapidly progressive ILD, is significantly associated with pneumomediastinum and vasculopathy.^{1,2} Another report has shown its possible association with vasculopathy via type 1 interferon.⁶ Pneumomediastinum in DM attributes to significantly high mortality and aggressive immunosuppressive therapy with steroids, and cyclosporine or cyclophosphamide should be considered.¹⁻³

In conclusion, patients with DM and ILD who develop new-onset chest pain, particularly with accompanying cutaneous ulcers, should be urgently examined for pneumomediastinum and treated aggressively.

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