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Pneumomediastinum and Subcutaneous Facial Emphysema in Antimelanoma Differentiation-Associated Gene 5 Antibody–Positive Dermatomyositis

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Pneumomediastinum is not a rare complication of antimelanoma differentiation-associated gene 5 (anti-MDA5)–positive dermatomyositis (DM)-associated interstitial lung disease (ILD). Here, we reported a rare case of subcutaneous facial and posterior pharyngeal emphysema due to pneumomediastinum in anti-MDA5-positive DM.

A 48-year-old woman presented with progressively increasing right cheek swelling and dysphagia (Figure 1A). Computed tomography (CT) of the head and chest revealed subcutaneous facial and posterior pharyngeal emphysema and pneumomediastinum, as well as signs of ILD (Figures 1B,C). Fourteen months ago, the patient initially manifested with heliotrope rash and Gottron papules/signs over the metacarpal, interphalangeal, elbow, and knee joints; these were accompanied by dyspnea on exertion, myalgia, and proximal muscle weakness. Chest CT showed signs of ILD. Screening of myositis-specific antibodies

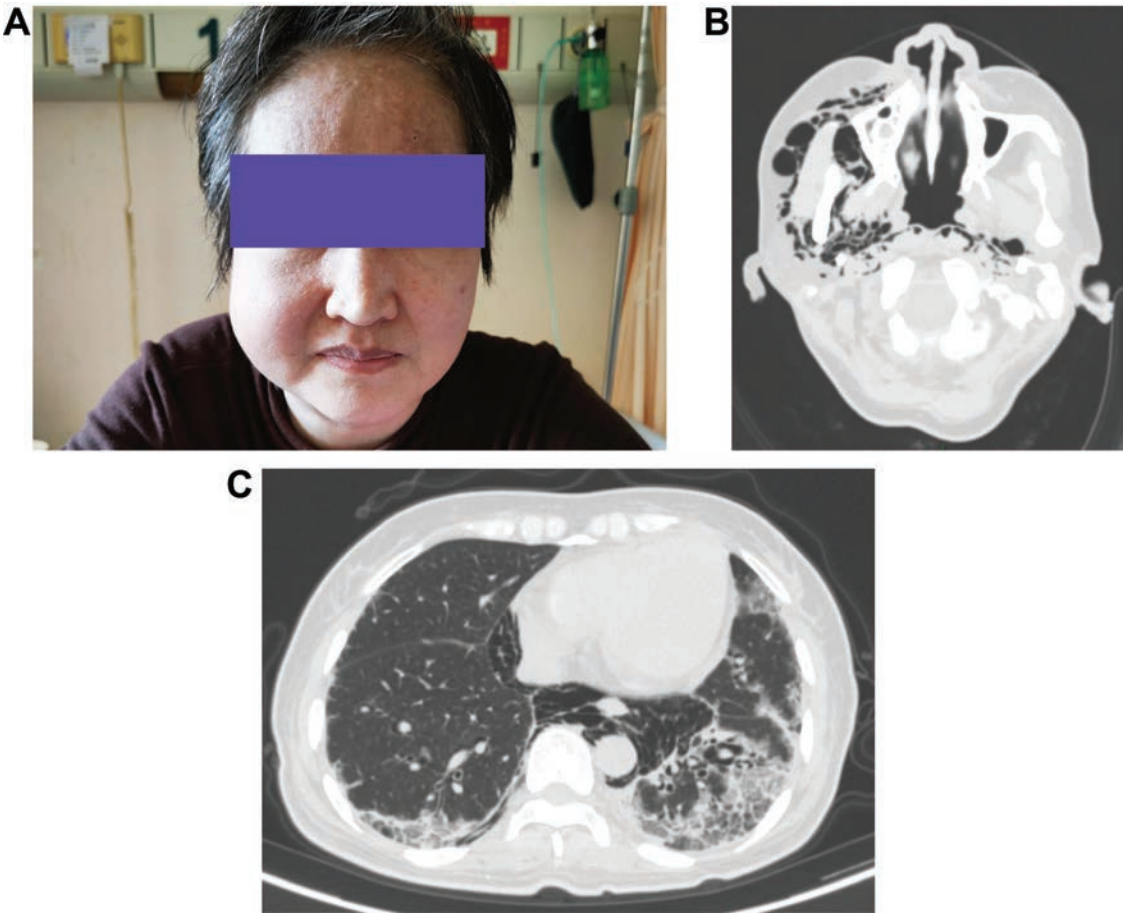

Figure 1. (A-C) Subcutaneous facial and posterior pharyngeal emphysema, pneumomediastinum, as well as signs of interstitial lung disease in anti-MDA5–positive dermatomyositis.

revealed strong positivity of anti-MDA5 antibody, and the diagnosis of anti-MDA5–positive DM was confirmed.[1] The patient initially received combined intensive immunosuppressive therapy (systemic glucocorticoids, tofacitinib [TOF], tacrolimus, and cyclophosphamide [CYC]).[2,3] Under this therapeutic regimen, her dyspnea abated and the severity of ILD on CT remained stable. TOF and CYC were withdrawn due to risk of infection 5 months ago. However, she began to suffer recurrence of Gottron papules/signs and new-onset right cheek swelling and dysphagia 2 months ago.

In summary, we report a rare case of subcutaneous facial and posterior pharyngeal emphysema due to pneumomediastinum in anti-MDA5-positive DM. This serious complication occurred 1 year after disease onset, accompanied with the recurrence of typical cutaneous manifestations, following the discontinuation of intensive immunosuppressive treatment. Anti-MDA5–positive DM associated ILD often presents with progressive disease course within the first 6 months. However, chronic course and relapse of disease might happen as in our case, and deserves long-term close follow-up.

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