A 31-year-old woman was referred due to a 1-month history of low-grade fever and dry cough. Physical examination showed a bilateral, erythematous, scaly eruption occurring in a symmetric fashion over the metacarpophalangeal and interphalangeal joints, knees, and elbows, suggestive of Gottron’s papules. She also had late inspiratory crackles mainly over the right upper lung field, but no muscle weakness. Laboratory examination revealed negative anti-Jo-1 antibodies and high antinuclear antibody titer (1 : 80), KL-6 (3,050 U/mL). Thoracic computed tomography showed subpleural patchy ground glass attenuation and consolidation in both lobes, with bronchiectasis (Picture 1A). She was diagnosed as having amyopathic dermatomyositis and treated with oral prednisolone plus cyclosporine. Although her condition seemed to improve, one month later, massive pneumomediastinum (Picture 1B, arrowhead) and subcutaneous emphysema (Picture 1B, arrow) suddenly developed, and compressed major vessels and main bronchi. Thereafter, her lung disease showed rapid deterioration, and she died three weeks later. Le Goff et al (1) reported that the prevalence of pneumomediastinum in polymyositis/dermatomyositis (PM/DM) may be estimated at approximately 2.2%, and in most cases, it occurs within one year of the initial onset of signs and symptoms of PM/DM. They also described that the one of the factors associated with poor survival was an absence of muscle weakness and the overall mortality rate was 34.4%, with up to 25% of patients dying during the first month of respiratory distress, as in the present case.

The authors state that they have no Conflict of Interest (COI).

Reference