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Clinical Image

Spontaneous pneumomediastinum in dermatomyositis☆

Neumomediastino espontáneo en la dermatomiositis

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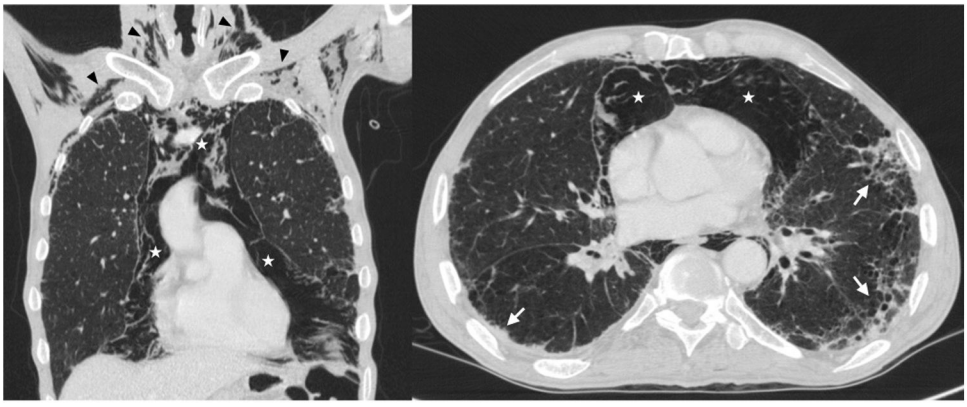

Fig. 1. Pneumomediastinum.

A 63-year-old man consulted for a 1-week history of cough and dyspnea, along with significant weight loss in the previous 8 months. No significant medical history was reported. Examination revealed subcutaneous crepitation in the upper trunk and neck, Gottron papules, and heliotrope rash. He did not present muscle weakness. A CT scan (Fig. 1) showed extensive pneumomediastinum *(asterisks)*, subcutaneous emphysema *(arrowheads)*, and diffuse interstitial lung disease (ILD) *(arrows)*. Clinical laboratory tests were significant for aldolase 13 U/l (normal: 1.2–7.6 U/l) and presence of anti-melanoma differentiation-associated gene 5 (MDA5) antibodies. A muscle biopsy was consistent with dermatomyositis. With a diagnosis of clinically amyopathic dermatomyositis, associated with diffuse ILD and spontaneous pneumomediastinum (PM), treatment began with prednisone and cyclosporine. A CT scan 1 month after diagnosis showed complete resolution of PM. No underlying neoplasia has been detected after 2 years of follow-up.

Fewer than 100 patients with PM associated with dermatomyositis have been described and 80%–90% of cases have demonstrated 3 characteristics: clinically amyopathic disease, ILD, and the presence of anti-MDA5 antibodies.[1,2] Most patients have an established diagnosis of dermatomyositis prior to the development of PM. Prognosis is dependent on the ILD and not the PM, which usually only requires observation until resolution.

References

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