Clinical Image

Spontaneous Bilateral Pneumothoraces in Erdheim-Chester Disease

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An acutely ill 22-year-old man was admitted to hospital with severe chest pain, high-grade dyspnoea and sinus tachycardia. He had a history of low-growth and polyostotic bone alterations since the age of 14. The patient presented 3 months before the current episode with a large left chest wall mass (Fig. 1) that was biopsied, showing xanthomatous CD68-positive, CD1a/S100-negative foamy histiocytes with positive BRAF-V600E mutation, which confirmed Erdheim-Chester disease (ECD) (Fig. 1), as well as bone biopsy results. Emergency chest X-ray showed massive bilateral pneumothoraces, so chest tubes were placed (Fig. 1). Computed tomography (CT) demonstrated multiple lung cysts and diffuse interstitial thickening. Lung biopsy was positive. ECD is a rare non-Langerhans histiocytosis, with around 600 published cases to date,\textsuperscript{1} which primarily affects male patients between their 5th and 7th decades. More than 50% of ECD patients have BRAFV600E mutations.\textsuperscript{2} It encompasses a spectrum of manifestations: osseous, cardiovascular, pulmonary, central nervous system, orbital, neuroendocrine, retroperitoneal and cutaneous. Interstitial pulmonary disease and lung cysts appear in less than 18% of reported cases.\textsuperscript{3} To the best of our knowledge, there have been no cases of bilateral pneumothoraces and fewer than five cases of unilateral pneumothorax complicating ECD have been reported.

References