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. (c) Axial CT image showing thorax drainage tubes due to bilateral pneumothoraces (arrows). (d) Note the striking interstitial involvement of both lungs with multiple lung cysts and diffuse thickening of the pulmonary interstitium.

An acute onset of bilateral pneumothoraces is rare in Erdheim-Chester disease (ECD). Previous reports have been described in single cases. ECD is a rare non-Langerhans histiocytosis, with around 50% of cases meeting the criteria of the American Thoracic Society, American College of chest physicians, and European Respiratory Society. There is a strong association between ECD and chronic obstructive pulmonary disease and lung cysts appear in less than 18% of reported cases. 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