Clinical Image

Spontaneous Bilateral Pneumothoraces in Erdheim-Chester Disease

Neumotórax bilateral espontáneo en un caso de enfermedad de Erdheim-Chester

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An acutely ill 22-year-old man was admitted to hospital with severe chest pain, high-grade dyspnea 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 BRAFV600E. (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.

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.

Fig. 1. (a) Axial CT image demonstrating a large soft tissue mass in the left chest wall (asterisk); (b) Chest wall mass histological section confirming xanthomatous CD68-positive, CD1a/S100-negative foamy histiocytes with positive BRAFV600E. (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.

References


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