

Clinical Image

Spontaneous Bilateral Pneumothoraces in Erdheim-Chester Disease

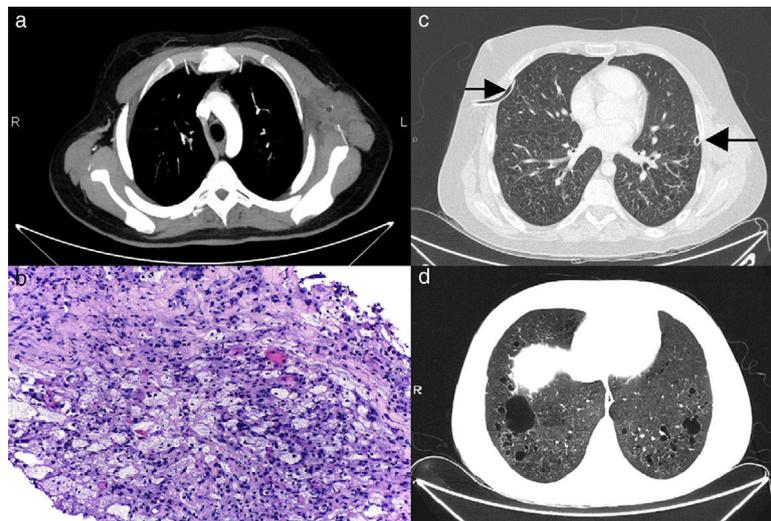


Neumotórax bilateral espontáneo en un caso de enfermedad de Erdheim-Chester

Ruben Eduardo Pacios Blanco<sup>a,\*</sup>, Luis Gorospe Sarasua<sup>a</sup>, María Eugenia Reguero<sup>b</sup>

<sup>a</sup> Department of Clinical Radiology, Ramón y Cajal University Hospital, Madrid, Spain

<sup>b</sup> Department of Pathological Anatomy, Ramón y Cajal University Hospital, Madrid, Spain



**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 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 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 and BRAFV600E 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,<sup>1</sup> which primarily affects male patients between their 5th and 7th decades. More than 50% of ECD patients have BRAFV600E mutations.<sup>2</sup> 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.<sup>3</sup> 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

1. Arnaud L, Pierre I, Beigelman-Aubry C, Capron F, Brun A-L, Rigolet A, et al. Pulmonary involvement in Erdheim-Chester disease: a single-center study of thirty-four patients and a review of the literature. *Arthritis Rheum.* 2010;62:3504–12. <http://dx.doi.org/10.1002/art.27672>.
2. Haroche J, Cohen-Aubart F, Arnaud L, Hervier B, Charlotte F, Drier A, et al. Erdheim-Chester disease. *Rev Med Interne.* 2014;35:715–22.
3. Cives M, Simone V, Rizzo F, Dicuonzo F, Lacalamita M, Ingravallo G, et al. Erdheim-Chester disease: a systematic review. *Crit Rev Oncol Hematol.* 2015;95:1–11.

\* Corresponding author.

E-mail address: [eduardo.pacios@gmail.com](mailto:eduardo.pacios@gmail.com) (R.E. Pacios Blanco).