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Pulmonary puzzle

ANSWER

From the question on page 492.

Macroscopic findings favoured a diagnosis of mesothelioma. Microscopy revealed instead that the pleural thickening comprised fibrosis within which abundant CD68-positive histiocytes and Touton-type giant cells were present, these staining with S-100 but not CD1a. There was also microscopic involvement of the lung along the distribution of the lymphatic vessels and areas of yellow firm thickening in the epicardial fat adjacent to the coronary arteries demonstrating identical pathology. A diagnosis of Erdheim-Chester disease (ECD) was made.

ECD is a non-Langerhans histiocytic disorder primarily affecting bone. Over 50% of cases show involvement of other tissues including the retroperitoneum, retro-orbital tissues, heart, pituitary-hypothalamic axis and lung.¹ Diagnosis is usually based on long bone radiographic changes.^{1–4} Bone scanning reveals symmetrical and abnormally increased distal lower limb long bone uptake. Pulmonary CT findings include distinctive interlobular and visceral pleural thickening associated with patchy reticular and centrilobular opacities and areas of ground-glass attenuation.⁴ Histopathologically, lung involvement in ECD is characterised by the accumulation of foamy or clear histiocytes with variable amounts of fibrosis and lymphoplasmacytic inflammatory infiltrate arranged in a lymphangitic pattern.¹ Immunohistochemistry shows consistent staining for CD68 and negative staining for CD1a. Ultrastructural studies reveal an absence of Birbeck granules.²

Pulmonary ECD is typically diagnosed by a constellation of radiographic evidence and other key characteristic visceral signs with a virtually pathognomic bone profile. There is only one other case report of ECD mimicking mesothelioma.⁵ Diagnosis could only be made from microscopy at necropsy, highlighting the importance of tissue diagnosis rather than relying on macroscopic features alone.

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