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Erdheim-Chester disease: A case with unusual pulmonary manifestations

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Erdheim-Chester Disease (ECD), a rare multisystemic non-Langerhans' cell histocytosis of unknown etiology, primarily affects middle-aged adults. Pulmonary manifestations occur in 15-33% of patients and significantly increase morbidity and mortality. We report a 60-year-old female smoker who presented with dyspnea, weakness and recurring reticulonodular lung infiltrates on CT imaging. Past medical history includes lower extremity pain, bilateral hip replacement and shoulder arthroplasty for degenerative joint disease. Chest radiographic findings were suggestive of possible malignancy or sarcoidosis with progressive centrilobular nodules in a tree-in-bud pattern. Bronchoscopic examination was negative for obstructive endobronchial lesions. The histologic findings on right lung wedge resection revealed interstitial infiltration by finely to coarsely vacuolated macrophages, occasional lymphoid aggregates, and collagenous fibrosis in a perivascular, interlobular septal, and peribronchial pattern. Few macrophages were present in alveolar spaces possibly suggesting chronic exogenous lipoid pneumonia (Fig. 1A and B). An isolated necrotizing granuloma of presumed infectious etiology was also observed. A six-year interval progression of her interstitial lung disease prompted repeat surgical biopsy, of the left lung, with similar histological findings, excluding necrotizing granulomas. Immunophenotypically, the histiocytes were CD68 positive, focally positive for Factor XIIIa and negative for S100 and CD1a (Fig. 1C). Review of the slides of left shoulder bone and synovium showed dense infiltration of histologically and immunophenotypically similar histiocytic cells (Fig. 1D), supporting the diagnosis of ECD. This case raises the question of possible predisposition to infectious granulomatous disease in ECD and emphasizes the importance of review of prior non-pulmonary biopsies to establish the diagnosis, especially in radiographically or histologically atypical cases.

Biography

Karam Han has completed her medical degree from Boston University School of Medicine. She is currently completing her residency at MetroHealth Medical Center, Case Western Reserve University in Cleveland, Ohio.

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