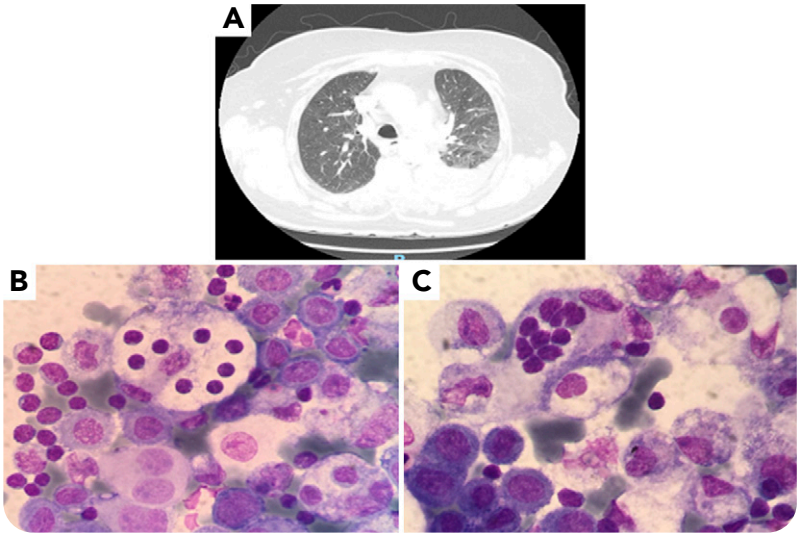
blood work
images in hematology

Unusual histiocytes in a pleural effusion: signature of a rare disease

Christelle El Khoury and Hussein Farhat, Lebanese American University Medical Center



A 79-year-old woman was admitted to the hospital for dyspnea, dry cough, and left-sided chest pain. Her medical history included coronary artery disease, hypertension, and hypothyroidism. No palpable lymphadenopathy or organomegaly was noted on physical examination. Her chest radiograph showed left lower lobe atelectasis and pleural effusion, and her chest computed tomography scan showed significant left-sided pleural effusion with associated volume loss (panel A). Pleural fluid analysis showed proliferating histiocytes that represented 40% of the nucleated cells, with 15% exhibiting emperipolesis by engulfing intact lymphocytes (panels B-C; hematoxylin and eosin stain, original magnification ×1000). They were intermixed with mesothelial cells in a background rich in lymphocytes. Histiocytes were positive for CD68 and S100 and negative for CD1a. No feature indicative of malignant lymphoproliferative disorder was detected by flow cytometry. Findings were consistent with an extranodal variant of Rosai-Dorfman disease. The patient was treated with left pleurodesis and oral prednisone, showing significant improvement.

Rosai-Dorfman disease, also known as sinus histiocytosis with massive lymphadenopathy, is a benign proliferation of histiocytes with no known pathogenesis. It primarily affects cervical lymph nodes with possible involvement of extranodal tissues. Lung involvement occurs in <3% of cases with extranodal disease. Although clonality could not be established in the reported case, somatic mutations affecting *NRAS*, *KRAS*, *MAP2K*, and *ARAF* genes were found.

ASH Image Bank

For additional images, visit the ASH Image Bank, a reference and teaching tool that is continually updated with new atlas and case study images. For more information, visit http://imagebank.hematology.org.

DOI 10.1182/blood.2019001131