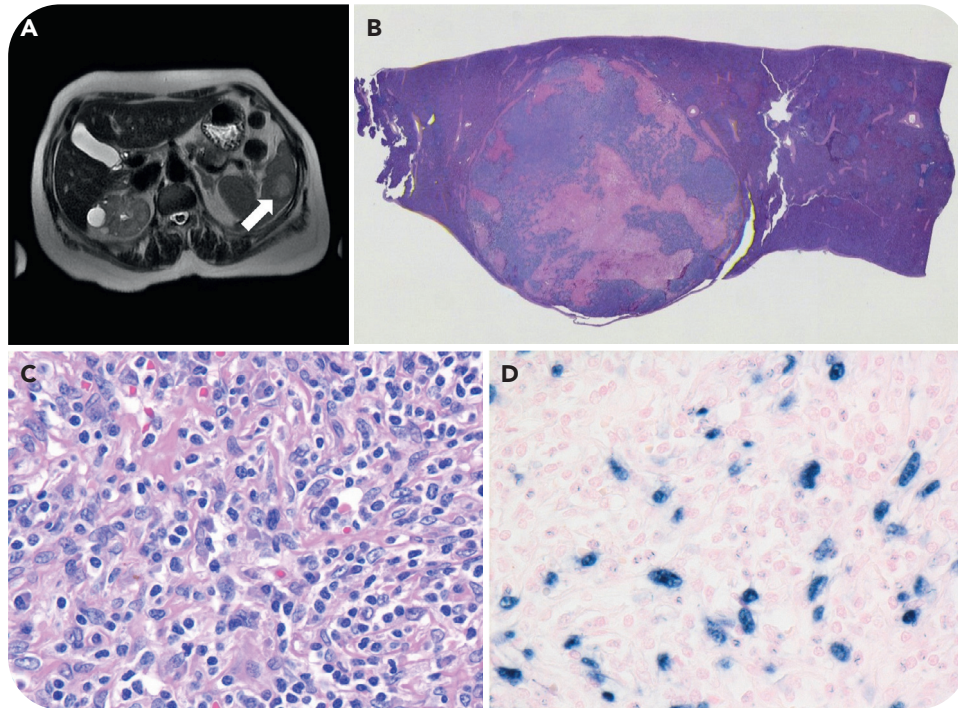


EBV-positive inflammatory FDC/FRC tumor: no longer pseudo or sarcoma!

Ryan W. Hunter and Sandeep Gurbuxani, University of Chicago



A 74-year-old woman, status post partial nephrectomies for bilateral papillary renal cell carcinoma, exhibited a 3.5 cm × 1.4 cm splenic lesion on imaging (panel A, magnetic resonance imaging T2). Hematoxylin and eosin–stained sections of the splenectomy were notable for presence of a well-circumscribed lesion with central area of necrosis (panel B, magnification ×25). Higher magnification showed spindle cell infiltrate mixed with small lymphocytes and plasma cells (panel C, magnification ×500). The spindle cells expressed smooth muscle actin (not shown) and were positive for Epstein-Barr virus (EBV) by in situ hybridization for EBV-encoded small RNA (EBER) (panel D, magnification ×500). CD21, D2-40, and clusterin were not expressed; CD3, CD20, and CD68 highlighted the background inflammatory T-cells, B-cells, and macrophages (not shown).

The findings are consistent with EBV-positive inflammatory follicular dendritic cell (FDC)/fibroblastic reticular cell (FRC) tumor, an indolent proliferation of stromal cells of mesenchymal origin not derived from hematopoietic stem cells. The proposed International Consensus Classification change in nomenclature from the 2016 World Health Organization entity inflammatory pseudotumor-like follicular/fibroblastic dendritic cell sarcoma to EBV-positive inflammatory FDC/FRC tumor acknowledges lineage heterogeneity and the indolent clinical behavior. EBER positivity in the spindle cells present in circumscribed mixed inflammatory splenic lesions is diagnostic for EBV-positive inflammatory FDC/FRC tumor even in the absence of FDC markers.