A Rare Cause of Left Upper Quadrant Abdominal Pain

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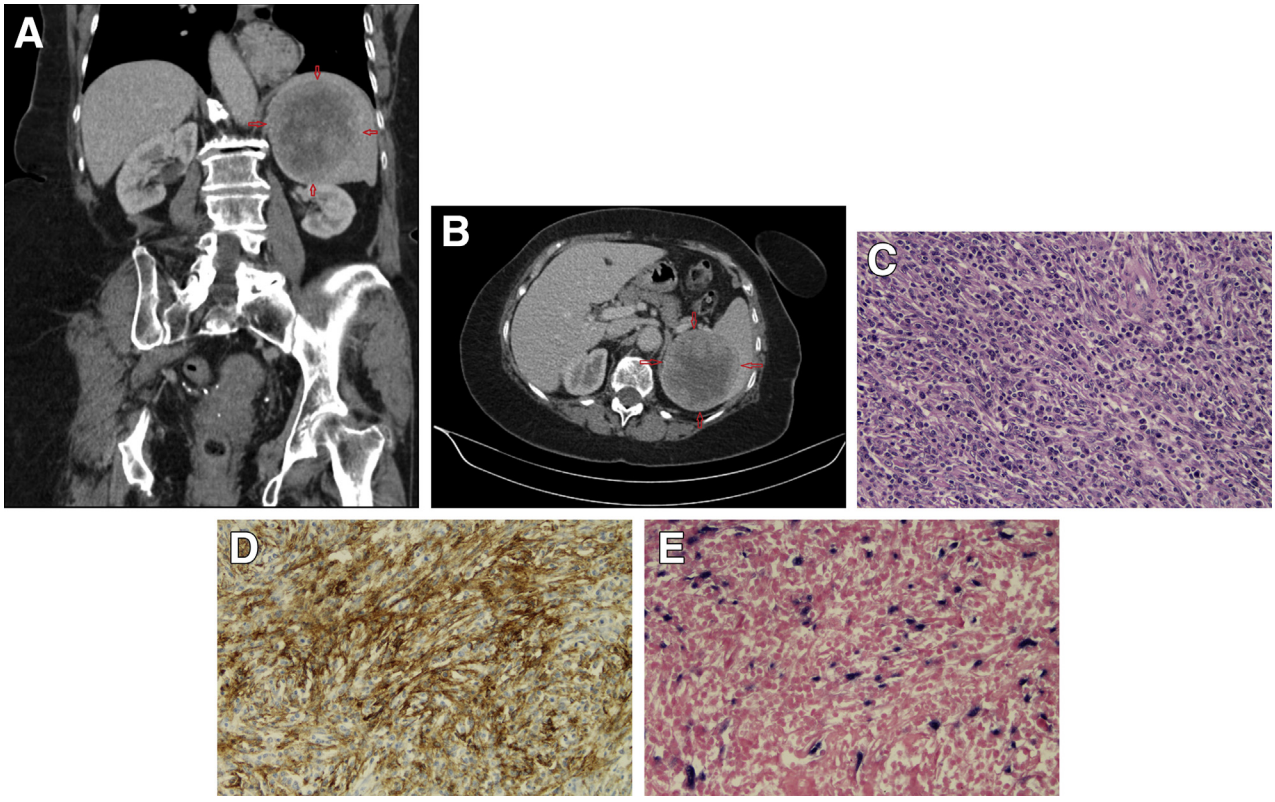

Question: A 77-year-old woman was referred for continuous left upper quadrant abdominal pain and a hypointense splenic mass on T2-weighted magnetic resonance imaging sequences of the spine, ordered for suspicion of degenerative disc disease. Clinical examination and bloodwork were normal. Her history was relevant for abdominal contusion during a car crash 30 years ago, but no splenic lesion was found on surgical exploration at that time. A computed tomography scan showed a well-defined, 12 × 9 × 6-cm lesion with a hypodense necrotic center and isodense peripheral tissue that enhanced after contrast injection, amidst an otherwise normal appearing spleen (Figure *A*, *B*). No distant lesions were found. Because the lesion's size had increased compared with a scan from 2 years earlier and considering a potential malignant process, total splenectomy was performed.

Histology revealed a well-circumscribed encapsulated mass consisting of a chronic inflammatory infiltrate (mostly lymphocytes and plasmacytes accompanied by macrophages and a few giant cells) and spindle cells with mild atypia (Figure *C*, hematoxylin-eosin stain) surrounding a necrotic center. On immunohistochemistry, the atypical spindle cells expressed smooth muscle actin, CD 35 (Figure *D*) and marked positive for Epstein-Barr virus RNA after in situ hybridization (Figure *E*).

What is the diagnosis?

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Conflicts of interest
The authors disclose no conflicts.

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Answer to: Image 4: Epstein-Barr Virus–Associated Inflammatory Pseudotumor-Like Follicular Dendritic Cell Sarcoma

Considering the otherwise normal spleen and liver, as well as the absence of adenopathy or distant lesions, a lymphoma seems implausible. Primary splenic lymphomas and isolated splenic metastases are very rare. The differential diagnosis of an isolated splenic tumor also comprises angiosarcomas, but the clinical presentation is usually more aggressive. Uncommon primary mesenchymal tumors such as other sarcomas, angiolipoma, fibroma, or malignant fibrous histiocytoma have to be considered before surgery because they do not have pathognomonic features on imaging. The differential diagnosis also comprises benign tumors like hemangiomas, lymphangiomas, hamartomas, lipomas, and littoral cell angiomas, but imaging characteristics are different. A history of abdominal trauma could suggest a reactive inflammatory pseudotumor, but definitive diagnosis of this entity requires histology.[1]

In our case the histologic findings are consistent with Epstein-Barr virus–associated inflammatory pseudotumor-like follicular dendritic cell sarcoma, a rare malignant entity, typically occurring in the liver or spleen, and more rarely in lymph nodes. Owing to morphologic similarity, this lesion is often confused with benign lesions like reactive inflammatory pseudotumors or inflammatory myofibroblastic tumors.[2] Patients can be asymptomatic, or complain of abdominal pain or nonspecific symptoms like weight loss or fever. The diagnosis is made by histology and immunohistochemistry. Treatment usually consists of surgical resection. Metastases and recurrence are exceptional in splenic cases.[3] Four months after splenectomy, the patient is asymptomatic.

References

1. Kaza RK, Azar S, Al-Hawary MM, et al. Primary and secondary neoplasms of the spleen. Cancer Imaging 2010; 10:173–182.
2. Hang JF, Wang LC, Lai CR. Cytological features of inflammatory pseudotumor-like follicular dendritic cell sarcoma of spleen: a case report. Diagn Cytopathol 2017;45:230–234.
3. Ge R, Liu C, Yin X, et al. Clinicopathologic characteristics of inflammatory pseudotumor-like follicular dendritic cell sarcoma. Int J Clin Exp Pathol 2014;7:2421–2429.