

Inflammatory pseudotumor-like follicular dendritic cell tumor of the spleen

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Dear Editor,

A 58-year-old woman was admitted to a hospital with an incidental splenic mass.

Laboratory tests, including those for the tumor marker, were normal. Ultrasonography revealed a well-marginated, round, hypoechoic mass in the spleen (Fig 1A). The mass showed septal echogenic thickening in the internal area. Computed tomography (CT) revealed a well-circumscribed round mass in the spleen; the mass was 5 cm in size. In a dynamic enhancement scan, the mass showed progressive enhancement (Fig 1B-D). In the arterial phase, the mass demonstrated septal-like enhancement at the central portion (Fig 1B). Therefore, the radiological diagnosis was sclerosing angiomatoid nodular transformation (SANT) of the spleen.

The patient underwent resection and splenectomy. The cut surfaces of the tumor revealed a well-circumscribed yellowish soft tissue tumor that was 5×5×4.5 cm. There was no lymph node enlargement in the splenic hilum.

In a gross examination, a well-marginated, thin-walled, yellowish, soft tissue mass was found throughout the spleen (Fig 1E). A microscopic examination revealed that the tumor had an admixture of lymphocytes, plasma cells, and atypical cells on staining with hematoxylin and eosin (400×) (Fig 1F). The use of immunohistochemical staining demonstrated that tumor cells expressed CD31, CD21, and CD23. In situ hybridization for Epstein-barr virus (EBV)-encoded RNA shows positive in almost all tumor cells (Fig 1G) representing inflammatory pseudotumor-like follicular dendritic cell (FDC) tumor.

The patient is disease-free for 2 years after operation without any recurrence.

The term "follicular dendritic cell" tumor was first mentioned by Monde et al. in 1986 (1). Most FDC tumors affect the lymph nodes (2-3). These tumors behave silently, but intra-abdominal cases can behave aggressively like local recurrence (4).

FDC tumors with inflammatory pseudotumor-like pathologies are a separate and unique type of neoplasms. FDC tumors showing inflammatory pseudotumor (IPT) like pathologies are separated from existing FDCs, and the World Health Organization named them as IPT-like variant FDC sarcomas. Until now, few cases have been reported (5-7).

Inflammatory pseudotumor-like FDC sarcomas are regarded as a separate entity mainly owing to the involvement of the liver and spleen with distinct pathological features and a strong association with EBV infection (8). Studies have demonstrated EBER-positive tumor cells by in situ hybridization in almost all cases and the frequent expression of the oncogenic protein EBV-latent membrane positive-1 by immunohistochemical staining and monoclonal episomal EBV by southern blot analysis, suggesting a pathogenic role of this tumor (8).

In contrast to conventional inflammatory pseudotumors, inflammatory pseudotumor-like FDC tumors have a low-grade malignancy (3). There have been limited reports on the imaging features of IPT-like FDC sarcomas (7,9). According to a recent case report (9), these tumors show similar or slightly lower attenuation than splenic parenchyma on performing non-enhanced CT. After contrast enhancement, the fibrotic component of the tumor shows gradual increased enhancement that maximum enhancement is made on the delayed phase. The tumor could have some necrosis and calcification, also.

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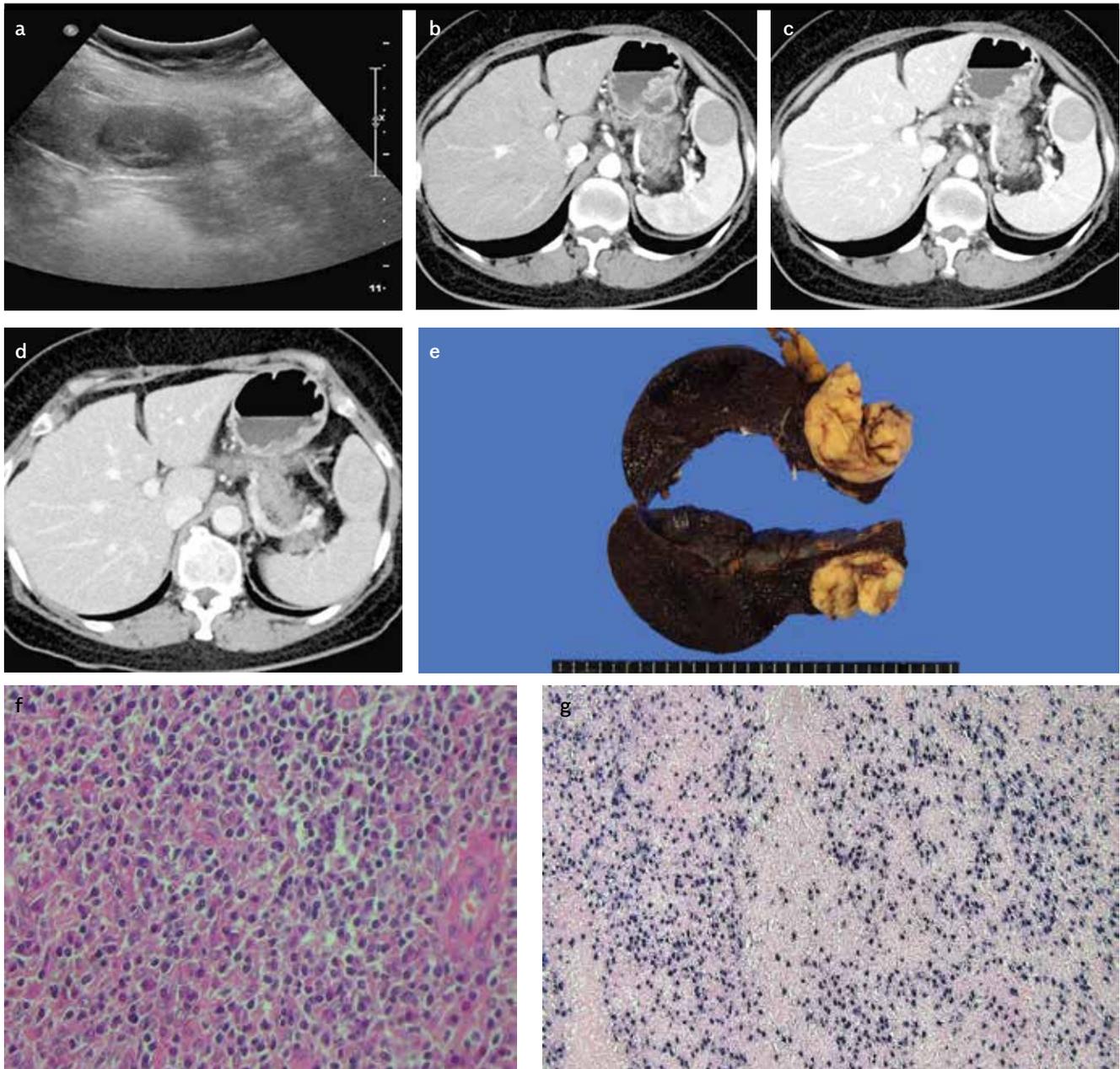


Figure 1. a-g. (a) A 58-year-old woman was admitted to a hospital an incidental splenic mass.

Ultrasonography revealed a well-marginated, round, hypoechoic mass with internal echogenic septal thickening in the spleen.

(b-d) Computed tomography (CT) revealed a well-circumscribed, round mass in the spleen; the mass was 5 cm in size and showed progressive enhancement in a dynamic scan.

(e) The cut surfaces of the tumor revealed a well-circumscribed solid tumor with thin-walled, yellowish, soft tissue mass throughout the spleen.

(f) A microscopic examination revealed that the mass was composed of an admixture of lymphocytes, plasma cells, and atypical tumor cell and (g) positive on EBV-RNA stain.

The central stellate area shows low signal intensities on T1- and T2-weighted magnetic resonance images, which are due to fibrosis and various degrees of necrosis (9). On the sonographic image, IPT-like FDC sarcomas of the

spleen have been reported as hypoechoic masses. In our case, the splenic tumor showed multiple echogenic septal lines in the mass. The differential diagnosis of IPT-like FDC tumors is conventional inflammatory pseudotumor,

because these tumors have similarity by presence of inflammatory cell and fibrotic tissue (9).

Because of the non-specific imaging findings, other common benign splenic tumors such as hemangiomas or hamartomas and malignant splenic tumors such as splenic lymphomas, metastases, and angiosarcomas are not differentiated from this type of tumor (10).

Therefore, it is difficult to differentiate benign tumor such as IPT from a malignant tumor such as lymphoma without pathologic diagnosis.

According to data published thus far, if the tumor is confined to the spleen, total splenectomy is the treatment of choice. There is no requirement of adjuvant therapy (11). However, the effectiveness of adjuvant therapy seems to require more research because the disease itself is rare and the amount of data published thus far is low.

Our patient showed an extremely rare IPT-like FDC tumor that arose from the spleen; the lesion showed progressive enhancement and multifocal septal thickening. Imaging findings suggested a benign splenic tumor such as SANT or hemangioma.

When the splenic tumor shows progressive enhancement, IPT-like FDC tumors should be considered in the differential diagnosis.

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