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INFLAMMATORY PSEUDOTUMOR-LIKE FOLLICULAR DENDRITIC CELL SARCOMA OF THE LIVER

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CASE REPORT
A 53-year-old woman with a history of HLA-B27 positive polyarthritis underwent splenectomy because of an incidental splenic lesion, which was confirmed as an inflammatory pseudotumor (IPT). Afterwards, two liver lesions were found and their histopathological examination revealed inflammatory pseudotumor-like follicular dendritic cell sarcoma (IPT-like FDCS). The patient received NSAIDs, corticosteroids, antibiotics and azathioprine, without response. Within the next few months, there was an abrupt clinical worsening due to rapid progression of the hepatic lesions and massive hepatomegaly (Figs. 1 and 2). New biopsies were obtained, showing undifferentiated sarcoma (Fig. 3). The patient started chemotherapy with doxorubicin, but eventually died.

DISCUSSION
FDSC is a rare, low-grade, malignant tumor originating from follicular dendritic cells in germinal centers. The IPT-like variant predominates among women and is even less frequent. IPT-like FDCS mainly involves the spleen and liver and is associated with Epstein-Barr virus (EBV). Histopathological features comprise spindle cells within a prominent lymphoplasmacytic infiltrate (1, 2), similar to IPT, but with an immunophenotype positive for follicular dendritic cell markers (including CD21, CD23, and CD35), and positive for in situ hybridization of EBV encoded RNA. While IPT usually responds to anti-inflammatory drugs, IPT-like FDCS requires surgery as a first-choice
therapy. Chemotherapy and/or radiotherapy are applied for advanced or incompletely resected tumors. The recurrence rate is low, with favorable long-term outcomes (3). A torpid clinical course may indicate malignization. This was the case for our patient. The final biopsy showed marked cellular atypia and vanishing of dendritic cell markers, thus confirming undifferentiated sarcoma.

REFERENCES


Figure 1. On the left, venous phase of axial abdominal contrast-enhanced computed tomography (CT) cuts. Hepatomegaly secondary to multiple ill-defined, confluent hepatic lesions, with heterogeneous density and enhanced peripheral vascularization, consistent with cystic/necrotic changes. A repletion defect in an enlarged left portal vein due to partial thrombosis (*). On the right, axial cuts from a positron emission tomography (PET)/CT scan showing increased fluorodeoxyglucose uptake in the hepatic lesions.

Figure 2. Arterial phase of coronal abdominal contrast-enhanced computed tomography (CT) cuts (left) and positron emission tomography (PET)/CT scan (right). The abdominal structures are displaced by the massive hepatomegaly towards the left hemiabdomen. This condition was causing abdominal distension, constipation and marked urinary frequency. An undifferentiated transformation was suspected due to the rapid tumor growth and the torpid clinical course.
Figure 3. Histological images from the last biopsy. The tumor was composed of oval and spindle cells in an inflammatory background. Immunohistochemical analyses were performed to evaluate the expression of dendritic cell markers (including CD21, CD23 and CD35) but they were not present. In situ hybridization of Epstein-Barr virus encoded RNA was also negative. This information was consistent with an undifferentiated sarcoma.