Histological findings in the spleen affected by adult-onset Still’s disease: a report of three cases

Sirs,

Diagnosis of adult-onset Still’s disease (AOSD) is based on sets of several clinical and laboratory criteria because no specific test is available. The differential diagnosis includes malignant lymphoma and infectious diseases. A recent case series using positron emission tomography/computed tomography (PET/CT) showed high frequency accumulation of injected 18F-fluorodeoxyglucose (FDG) in the spleen and bone marrow of AOSD patients (1). Cell types contributing to the splenic 18F-FDG accumulation in AOSD have not been identified, while granulocytic hyperplasia in bone marrow reported as a common feature of AOSD may be responsible for the FDG uptake by bone marrow (2). Here we describe three female cases presenting fever of unknown origin, in which accumulation of 18F-FDG was found in both the spleen and bone marrow on PET/CT, and on whom spleen and bone marrow biopsies were performed to distinguish AOSD and malignant lymphoma. The histological analysis ruled out the possibility of malignant lymphoma, and the diagnosis of AOSD was made according to the Yamaguchi criteria in each case.

Case 1
A 48-year-old Japanese woman was admitted to our hospital with a high spiking fever and polyarthralgia. Laboratory examination revealed neutrophilia and elevated lactate dehydrogenase (LDH), C-reactive protein (CRP), and ferritin levels. PET/CT demonstrated a profound 18F-FDG accumulation in the spleen, bone marrow, and cervical, axillary, mediastinal and para-aortic lymph nodes (Fig. 1a). Biopsy specimen from leftinguinal lump did not include lymph node tissue, and the superficial lymph node in other areas seemed inadequate for biopsy because of their small size. Thus the percutaneous CT-guided spleen biopsy and bone marrow biopsy were conducted to distinguish between AOSD and malignant lymphoma. The spleen biopsy showed extensive infiltration of neutrophils and mild infiltration of plasma cells, lymphocytes, and eosinophils in the red pulp (Fig. 1d). In the bone marrow, neutrophil hyperplasia and histiocytic hemophagocytosis were observed.

Case 2
A 37-year-old Japanese woman was admitted to our hospital with a high spiking fever, rash on her hands and left arm, and swollen lymph nodes in the bilateral submandibular areas. Laboratory tests showed mild elevation of LDH and CRP levels, PET/CT revealed mild hepatomegaly, mild splenomegaly and swollen lymph nodes in the bilateral submandibular space. 18F-FDG uptake was observed in the spleen and bone marrow (Fig. 1b). Spleen biopsy showed micro-abscesses characterised by focal dense infiltration of neutrophils and sparse infiltration of eosinophils, lymphocytes and plasma cells in the red pulp (Fig. 1e). Bone marrow biopsy revealed normocellular bone marrow with normal myeloid/erythroid ratio.

18F-FDG PET/CT is considered to be useful for monitoring disease activity in AOSD, but unable to exclude malignant lymphoma (1, 3). Exclusion of malignant lymphoma in spleen is required in some cases suspicious of AOSD presenting splenomegaly. Further, percutaneous spleen biopsy is recently considered to be a safe and effective procedure for the evaluation of neoplasm in the spleen (4). However, limited data are available regarding the histopathology of spleen biopsy specimens in AOSD. Denzut et al. performed minilaparoscopy-guided spleen biopsy in four patients with splenomegaly of unknown origin (5). Notably, one of these patients had a history of AOSD and the spleen biopsy showed granulocytic infiltration. Another case of AOSD with peritonitis and granulocytic infiltration in the spleen has also been reported (6). These two reports are consistent with the observations in our cases. In summary, our cases suggest that infiltration of neutrophils into the red pulp may contribute to 18F-FDG uptake by the spleen in AOSD patients.

References:

Fig. 1. 18F-FDG PET/CT images and histological findings of the spleen in AOSD.

a-c: 18F-FDG PET/CT images of cases 1 (a), 2 (b), and 3 (c). Uptake of 18F-FDG was observed in the bone marrow (arrows) and enlarged spleen (asterisks) in all three cases. 18F-FDG also accumulated in the lymph nodes in case 1 (not shown) and liver in case 3 (an arrowhead).

d-f: Histological findings of the spleen in cases 1 (d), 2 (e), and 3 (f). Severe infiltration of neutrophils was identified in the red pulp in each case. Haematoxylin-eosin staining: ×400.
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References


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