Inflammatory pseudotumor (IPT) is an ill-defined entity of unknown origin presenting as a mass-like lesion with inflammation and mesenchymal repair. It is composed of plasma cells, lymphocytes, foreign body giant cells, histiocytes, foam cells, and numerous vascular elements. It is a benign lesion which has been observed in tissues of the respiratory tract, gastrointestinal tract, orbit, soft tissues, lymph nodes, and other organs. However, occurrence of IPT of the spleen is extremely rare and it is frequently misdiagnosed as a malignant neoplasm clinically and radiologically. This report describes a case with IPT of the spleen that was difficult to diagnose before surgery.

CASE REPORT

A 44-year-old female underwent routine health examination at our institution. Physical examination was unremarkable and laboratory data were all within normal ranges. Abdominal ultrasonography showed a hypoechoic splenic hilar lesion about 3 cm in size. Serum levels of CEA, CA153, and AFP were all within normal range. Upper abdominal computed tomography without enhancement demonstrated a rounded mass-like lesion in the hilum of the spleen with low attenuation values, and the lesion exhibited a low density, heterogenous, focal solid mass in post-contrast study (Fig. 1). Therefore, metastatic tumor or primary malignant lesion was highly suspected. Sigmoidoscopy, upper gastrointestinal panendoscopy, and transvaginal ultrasound showed non-specific findings. Double contrast colon series was also negative. Because a malignant splenic tumor could not be ruled out preoperatively, we chose hand-assisted laparoscopic procedure to perform splenectomy for diagnostic purposes, surgical removal of the tumor and to prevent iatrogenic intra-abdominal dissemination.

Intraoperative and postoperative courses were uneventful. The patient was discharged on the 9th postop-
The spleen measured $9 \times 7.5 \times 4$ cm in size and 124 gm in weight. A well-defined, variegated, firm tumor, measuring $3.5 \times 3 \times 3$ cm in size, was found in the hilum. The tumor was just beneath the splenic capsule, which was not invaded by the tumor grossly (Fig. 2). Microscopically, the sections showed a well-circumscribed lesion composed of spindle cells, plasma cells, and vascular channels (Fig. 3). Immunohistochemical

**Pathology**

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staining showed that spindle cells were positive for vimentin and smooth-muscle actin, also presenting numerous diffusely infiltrated CD68-positive cells. Although extravasation of erythrocytes was noted, there were no PAS-positive globules, and the CD34-positive cells were orderly arranged (Fig. 4). Therefore, Kaposi sarcoma could be excluded. Acid-fast stain showed no stainable bacilli, and PCR for DNA of mycobacterial tuberculosis complex was absent. The lesion was thus diagnosed as an IPT of the spleen.

DISCUSSION

Mass-like lesions with histologic features of nonspecific inflammation and mesenchymal repair have been designated as IPTs. Such lesions occasionally have been observed in a variety of tissues, such as the orbit, spinal meninges, digestive system, heart, soft tissues, mesothelial membranes, and respiratory tract. Splenic involvement is extremely rare. To our knowledge, since the first 2 cases of inflammatory pseudotumor of the spleen were reported by Cotelingam and Jaffe in 1984, only 74 cases have been reported in the literature. IPT is a benign well-circumscribed mass, usually solitary, composed of foci of inflammatory cells, mainly plasma cell, and lymphocytes in a fibroblastic stroma. Someren classified the inflammatory pseudotumors into 3 histopathological subtypes: xanthogranuloma type, plasma cell granuloma type, and sclerosing pseudotumor. Various combinations of these 3 subtypes may be seen within a single lesion. Although Cotelingam and Jaffe suggested that the initial pathologic event might be parenchymal necrosis with hemorrhage, Yamaguchi et al. posited that Epstein-Barr virus infection may play a role in the development of splenic inflammatory pseudotumor and the elevation of soluble IL-2 receptor level. Gomez-Roman et al. showed Human herpesvirus-8 present in several pulmonary IPTs, Matsubayashi et al. suggested that rupture of splenic hemangioma may result in IPT, and some authors postulated that the etiology and pathogenesis might be due to ineffective antibiotic therapy, a specific unidentified infectious agent, the granulomatous inflammation process, disturbance of blood supply, autoimmune disorders or an abnormality of lipid metabolism, but these hypotheses have not been confirmed currently. Therefore, the precise etiology of IPT remains unknown.

Clinically, the symptoms of IPT of the spleen are mostly diverse and non-specific, such as left flank or left upper quadrant abdominal pain, or the lesion may be detected as an incidental finding and is usually asymptomatic, as in our case.

Fine-needle aspiration biopsy can be used as the preoperative work-up in other organs, but it is not recommended for a mass in the spleen because of poor specificity, the risk of bleeding, and the fear of spillage of tumor cells if the tumor is malignant.

Sonography can demonstrate IPT as a hypoechoic lesion in the spleen, not typical of a cyst, or shows a large, partially calcified, well-defined echogenic mass. Nonenhanced CT demonstrates a rounded mass with low attenuation values with or without partial calcification. Enhanced CT demonstrates a slightly heterogenous mass in the spleen with minimal enhancement. MRI may show isointense on T1-weighted images and with either increased or decreased signal intensity on T2-weighted images with respect to the surrounding normal spleen. Although sonography, CT scan and MRI aid in the identification of space-occupying lesions of the spleen, these techniques do not permit a preoperative diagnosis.

The differential diagnosis of focal splenic mass with or without calcification is extensive and includes splenic cysts, abscess, hamartoma, organizing hematoma, hemangioma, lymphangioma, plasmacytoma, lipoma and fibroma. Primary benign tumors of the spleen account for only 0.007% of all operations and autopsies. Hemangiomas, cysts, lymphangiomas, and hamartomas are common, while the other diseases are very rare. It is important to distinguish benign tumors of the spleen from malignant tumors, such as malignant lymphoma, malignant fibrous histiocytoma, hemangiosarcoma, fibrosarcoma, and metastatic tumors.

A malignant splenic lesion could not be ruled out preoperatively in our case, and hand-assisted laparoscopic splenectomy was performed for diagnostic purposes and surgical removal of the tumor. Hand-port with endo-bag could isolate the specimen and avoid the specimen having direct contact with the surgical wound when we pulled it out from the abdominal cavity to preventing
iatrogenic intra-abdominal dissemination or skin metastasis. No hemangioma or other tumors accompanied the inflammatory pseudotumor pathologically. Acid-fast stain showed no stainable bacilli and DNA of mycobacterial tuberculosis complex was absent. Therefore, mycobacterial spindle-cell pseudotumor was less likely. In situ hybridization studies for Epstein-Barr virus-encoded RNAs and immunoperoxidase studies for human herpesvirus-8-specific protein were not performed, so the relationship between the IPT and Epstein-Barr virus or human herpesvirus-8 in this case were unknown.

There was no recurrence or development of new lesions in our case after hand-assisted laparoscopic splenectomy for 1 year. Inflammatory pseudotumor is a benign well-circumscribed mass, however, 2 cases with an inflammatory pseudotumor of the liver died as a result of the disease. Therefore, careful postoperative follow-up is necessary.15

In conclusion, inflammatory pseudotumor of the spleen is a rare, benign lesion of controversial etiology. It is very difficult to diagnose this disease by image study preoperatively. A final diagnosis can only be obtained through postoperative histopathological examination.

REFERENCES