Inflammatory Pseudotumor of the Spleen

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The case of a 39-year-old man with inflammatory pseudotumor of the spleen is presented. This is an extremely rare benign lesion with histologic features of non-specific inflammatory and reparative changes. The literature is reviewed and this case is compared clinically and radiologically with the previously reported cases. (Internal Medicine 31: 941-945, 1992)

Key words: echography, angiography, CT scan

Introduction

Inflammatory pseudotumors are benign nodular lesions characterized by proliferation of the connective tissue in association with non-specific inflammatory changes. They can occur in a variety of organs. It is very difficult to distinguish primary lesions of the spleen from malignant lymphoma. We recently treated a patient with an inflammatory pseudotumor of the spleen which was diagnosed histopathologically after splenectomy. Here we report this case and discuss it based on a review of relevant studies published in the literature.

Case Report

A 39-year-old man was admitted for medical examination because a tumor-like shadow was found in the spleen by abdominal echography in a routine medical examination. Leukocytosis (leukocyte count, 14,300/μl) had been found during a physical examination 1 year previously, but otherwise his medical history relative to the present illness was uneventful.

On admission, the patient was a well-nourished man of average physique. Neither the liver nor the spleen was palpable. In the laboratory findings on admission, examination of the blood revealed mild leukocytosis with the following values: 5 mm/h erythrocyte sedimentation rate; 531 × 10^4/μl RBC; 15.8 g/dl Hb; 47.3% Ht; 11,500/μl WBC count with 6% Stab., 39% Seg., 5% Eo., 2% Baso., 5% Mono, and 43% Ly., and 30.4 × 10^4/ml Plt. Blood biochemistry revealed normal hepatic and renal functions. Electrolytes were also normal. Results were negative for all tumor markers: 2.0 ng/ml CEA, less than 5.0 ng/ml AFP, 5 U/ml CA19-9 and less than 1.0 ng/ml SCC antigen. Echographic examination of the abdomen disclosed a relatively uniform hypochoeic mass, as large as 26 × 31 × 31 mm, with a somewhat irregular rim near the hilus of the spleen (Fig. 1). Examination by color Doppler technique revealed no abnormal perfusion within the mass. Plain CT abdominal scans revealed a round tumor mass of low density, which partially protruded outside of the capsular surface of the spleen (Fig. 2). On contrast-enhanced scan (Fig. 3), the border of the tumor mass tended to be indistinct and a well-enhanced area was found at the center of the tumor mass. Selective angiography through the splenic artery revealed neither abnormal arteries (Fig. 4) nor contrast medium in the tumor (Fig. 5). For laparotomy, an oblique incision was made in the upper left quadrant of the abdomen. Neither ascitic fluid nor enlarged lymph nodes was noted. Splenectomy was performed because the tumor was not overt. A yellowish-white, firm and elastic well-demarcated tumor mass (30 × 25 × 25 mm) was found near the hilus of the spleen (Fig. 6).

Histopathologically, the lesion was a relatively well demarcated fibrosis-like tissue, showing diffuse proliferation of spindle-shaped fibroblasts with focal infiltration of inflammatory cells, mainly plasma cells, and with partial hyalinization and deposition of hemosiderin. There was neither necrosis nor accumulation of amyloid in the lesion. There was no evidence indicating bacterial or fungal infection. None of the cell populations showed neoplastic changes. Based on these findings, a diagnosis of inflammatory pseudotumor was made. According to Someren's classification (1), this tumor would correspond to a sclerosing pseudotumor due to the marked sclerotic features (Fig. 7).
Inada et al

Fig. 1. Echographic examination of the abdomen disclosed a relatively uniform hypoechoic mass with a somewhat irregular rim near the hilus of the spleen (arrows).

Fig. 2. Plain CT scans revealed a round tumor mass of round low density, which partially protruded out of the capsular surface of the spleen.

Fig. 3. On contrast-enhanced scan, the border of the tumor mass tended to turn indistinct and a well-enhanced area was found at the center of the tumor mass.

Fig. 4. Selective angiography through the splenic artery did not reveal any abnormal artery.

Fig. 5. Selective angiography through the splenic artery did not reveal any tumor stain.

Discussion

Tumors of the spleen are rarely found by routine medical examination. It has been reported that angiomas are predominant among benign tumors of the spleen whereas malignant lymphomas are predominant among malignancies (2, 3). These relatively prevalent two types of lesions have been described in great detail elsewhere (4, 5). Regarding inflammatory pseudotumors which form tumor masses and present non-specific inflammatory changes in association with a variety of changes related to the healing process, the occurrence is rare in the spleen. In a literature search, we could only ascertain 15 cases (including the present case) of this tumor in the spleen (6–14). Consequently, numerous factors remain
Inflammatory Pseudotumor of the Spleen

Table 1. A summary of the Splenic Inflammatory Pseudotumor

<table>
<thead>
<tr>
<th>No of reference cases</th>
<th>Age/sex</th>
<th>Constitutional symptoms</th>
<th>Laboratory Data</th>
<th>Tumor size</th>
<th>CT. US Angiography findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Cotelingam et al (1984)</td>
<td>60/M</td>
<td>—</td>
<td>WBC: 14,000/μl</td>
<td>3.0cm in diameter</td>
<td>—</td>
</tr>
<tr>
<td>2. Cotelingam et al (1984)</td>
<td>66/M</td>
<td>dyspepsia, weight loss</td>
<td>WBC: 4,500/μl</td>
<td>5.5 x 4.5 x 4.5 cm</td>
<td>CT: low density enhanced mass (contrast study) US: hypoechoic AG: hypervascular</td>
</tr>
<tr>
<td>3. Misawa et al (1985)</td>
<td>50/M</td>
<td>—</td>
<td>WBC: 18,000 ~ 20,000/μl</td>
<td>6.0 cm in diameter</td>
<td>CT: mixed density</td>
</tr>
<tr>
<td>4. Alpern et al (1986)</td>
<td>67/F</td>
<td>nausea</td>
<td>WBC: 10,000 ~ 20,000/μl</td>
<td>6.0 cm in diameter</td>
<td>—</td>
</tr>
<tr>
<td>5. Sheahan et al (1988)</td>
<td>34/M</td>
<td>left upper abdominal pain</td>
<td>—</td>
<td>8.5 x 7.0 x 5.5 cm</td>
<td>—</td>
</tr>
<tr>
<td>9. Chang et al (1989)</td>
<td>64/M</td>
<td>fever, nocturia</td>
<td>WBC: 12,800/μl</td>
<td>7 x 7.5 cm</td>
<td>US: hypoechoic</td>
</tr>
<tr>
<td>10. FU et al (1989)</td>
<td>19/M</td>
<td>fever, local tenderness</td>
<td>WBC: 12,900/μl</td>
<td>2 cm in diameter</td>
<td>—</td>
</tr>
<tr>
<td>11. FU et al (1989)</td>
<td>56/M</td>
<td>—</td>
<td>WBC: 6,800/μl, ESR 75/h, hypergammapathy (polyclonal)</td>
<td>10 cm in diameter</td>
<td>CT: AG relatively hypovascular mass relatively</td>
</tr>
<tr>
<td>13. Wiernik et al (1990)</td>
<td>66/M</td>
<td>fever, anemia</td>
<td>WBC: 11,500/μl</td>
<td>11.5 x 7.5 x 6.5 cm</td>
<td>CT: mixed enhanced</td>
</tr>
<tr>
<td>14. Tomita et al (1991)</td>
<td>50/M</td>
<td>—</td>
<td>WBC: 12,900/μl</td>
<td>4 x 5 cm</td>
<td>—</td>
</tr>
<tr>
<td>15. our case</td>
<td>39/M</td>
<td>—</td>
<td>WBC: 11,500/μl</td>
<td>2.6 x 3.1 x 3.1 cm</td>
<td>CT: hypodensity well-enhanced (contrast study) US: hypoechoic</td>
</tr>
</tbody>
</table>
A yellowish-white, firm and elastic well demarcated tumor mass was found near the hilus of the spleen.

The lesion was a relatively well demarcated, pale staining tissue compressing the spleen (arrows). (H&E. ×25) b) The lesion showed diffuse proliferation of spindle-shaped fibroblasts with focal infiltration of inflammatory cells. (H&E. ×250.)

to be clarified.

In this review we found a 11:4 male preponderance, and the patients ranged in age from 19 to 75 years. Most lesions were detected accidentally. Inflammatory pseudotumor was associated with fever in only 3 cases (11, 12, 14). Blood examination showed polyclonal gammopathy in 1 (12) and leukocytosis in 6 (6, 8, 11, 12, 14). Based on these findings, bacterial infection and immunological derangement have been proposed to be factors in the pathogenesis of this pseudotumor. In the present case, however, since the presence of leukocytosis (leukocyte count, 11,000/μl) persisted even three months after splenectomy, bacterial infection was unlikely.

A differential diagnosis prior to the operation could not be established by imaging techniques such as echography, CT scan and angiography in any of the 15 patients. Echography, performed in 4 (7, 11, 13) of the 15 cases, revealed a hypoechoic mass in 3 (7, 11). Plain CT scan showed a low density mass in 5 (7, 12, 13, 14) of 6 patients studied. Enhanced CT was performed in 5 cases. The border of the tumor mass tended to be indistinct but enhancement of the inner part ranged from relatively uniform to heterogeneous. The findings of the present case, in which high density was increasingly seen near the central area, did not coincide with those of other cases, and thus this was not considered to be disease specific. Due to the characteristic functions of the reticuloendothelium, when an inflammatory pseudotumor in the spleen is found, first it must be differentiated from lymphatic neoplasma, in particular malignant lymphomas. In fact, the diagnosis of malignant lymphoma was made in some of the inflammatory pseudotumor cases listed in Table 1. The diagnosis of malignant lymphoma in the present case was also difficult to exclude. In recent echographic studies on malignant lesions of the spleen, malignant lymphomas and inflammatory pseudotumors are revealed as a hypoechoic mass. Therefore the differential diagnosis of these two lesions can not be made by echography. In plain CT scans, malignant lymphomas appear as masses of low density, but become distinctive by contrast study because the surrounding normal tissue is enhanced to a greater extent than the tumor tissue (15). This property is characteristic of malignant lymphomas and may be used to distinguish them from inflammatory pseudotumors.

Inflammatory pseudotumors have been histopathologically regarded as plasma cell granulomas, xanthogranulomas, mast cell tumors and histiocytomas (1). Due to the diversity of types of lesions, as reflected by the varied names they have been given, some of the reported lesions should be distinguished from angioma or angiosarcoma. Moreover, a lesion undergoing calcification is liable to be confounded with hamartomas and multiple lesions must be distinguished from sarcoidosis. Consequently, to establish the diagnosis of inflammatory pseudotumor, cytologic examination of specimens obtained by invasive needle aspiration biopsy under echographic guidance (16) is the necessary cytologic examination in some instances.
References