Inflammatory Pseudotumor of the Spleen: Review of clinical presentation and diagnostic methods

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ABSTRACT

We describe a 91-year-old woman with a clinical history of invasive ductal carcinoma of the breast diagnosed in 1991 who was admitted because of dizziness, poor appetite, and some swelling and tenderness over her cheeks. The patient's initial work up revealed a 5-cm well-demarcated hypodense solid lesion in her spleen with abnormally intense uptake on PET/CT scan raising suspicion for malignancy i.e. breast metastasis versus lymphoma. Further review demonstrated the presence of this splenic lesion, though slightly smaller, on a CT scan from ten years earlier (2000). An ultrasonographic guided core needle splenic biopsy was performed and the pathology result revealed histological findings compatible with inflammatory pseudotumor of the spleen. As a result, unnecessary splenectomy was avoided.

CASE REPORT

A 91 year-old female was admitted to our institution with dizziness, poor appetite, as well as swelling and tenderness over her cheeks. The patient had a clinical history of invasive ductal carcinoma of the breast diagnosed in 1991, for which she underwent mastectomy chemotherapy and radiation therapy. Subsequent mammogram tests and surveillance for metastatic disease had been negative, most recently 2 weeks prior to the presentation. Laboratory tests on current admission revealed an elevated ESR (49 mm/h; normal 0-30 mm/h) and a slightly low HGB (9.3 g/dL; normal 12.0-16.0 g/dL). Other laboratory results were within normal limits [normal C-reactive protein, normal white blood cell count (9200/µL; normal 3800 to 9800/ µL), negative Ca 19-9, and negative rheumatologic tests: Anti SM, Anti RNP, Anti SSA, Anti SSB, Anti Scl-70, Anti Jo-1, Anti Chromatin, Anti Centromere, Anti Ribosomal P, Anti DNA (DS), ANA]. Abdominal CT revealed a large, heterogeneous, hypodense lesion in the central spleen, measuring 4.9 x 4.2 cm in axial dimensions in unenhanced study. This mass demonstrated gradual mild enhancement on delayed contrast enhanced images (Figure 1-B). Due to potential concern for metastatic disease, positron-emission tomographic (PET)/CT scan was performed which showed intense abnormal uptake [max standardized uptake value (SUV) of 9.8] within the splenic lesion, suspicious for malignancy. Reviewing the patient's prior studies revealed that she had similar lesion in her spleen on an abdominal CT scan from 9 years prior to this study. The mass was slightly smaller than the current size (Figure 1-A). No interval scans were performed. Differential diagnosis on the PET/CT was lymphoma versus metastatic lesion from the patient's breast cancer (Figure 2). Therefore, core needle splenic biopsy was performed, under ultrasonographic guidance (Figure 3 A and B). Ultrasound revealed a
hypoechoic lesion involving the superior and medial aspect of the spleen measuring approximately 7.0 x 5.0 cm.

On histological examination, the core biopsy revealed a dense infiltrate of uniform, bland spindle cells surrounded by abundant mononuclear cells, including many plasma cells, lymphocytes and histiocytes. Scattered eosinophils and mast cells were also identified. No normal splenic parenchyma was identified within the core biopsy specimen. Given the clinical impression that the core biopsy represented the splenic mass, the histological findings were compatible with a diagnosis of inflammatory pseudotumor of the spleen (IPS) (Figure 4).

**DISCUSSION**

Primary benign splenic tumors are extremely rare. The most common primary benign splenic tumors include hamartomas, hemangiomas, and lymphangiomas [1]. IPS is exceedingly rare. Fewer than 85 cases have been reported in the literature [2]. These tumors have been reported in numerous different locations, including the lymphoid organs, respiratory tract, gastrointestinal tract, meninges, orbit, liver, and soft tissues [3]. The first case of IPS was described by Cotelingam and Jaffe in 1984 [4].

The etiology of IPS remains unknown. Speculated etiologies include: infections i.e. viral infections (most commonly Epstein-Barr Virus), abnormalities leading to infarction, focal parenchymal necrosis with hemorrhage secondary to trauma or coagulopathy, and autoimmune processes [4-6].

IPS does not have a typical presentation. Therefore, clinical diagnosis is difficult. A wide range of clinical presentations have been described for this disease, mainly nonspecific left upper quadrant abdominal pain, fever and splenomegaly, anemia, weight loss, or a discrete splenic mass. It can also be asymptomatic as in our case for approximately 9 years.

Diagnosing IPS is challenging. Most cases of IPS are detected incidentally. There is currently no single diagnostic imaging modality that can distinguish IPS from other splenic neoplasms. Plain radiographs may reveal splenomegaly and in rare cases splenic calcifications [1]. Abdominal ultrasound may demonstrate a well-defined hypoechoic lesion or a partially calcified hyperechoic mass [7]. On color flow Doppler ultrasound these masses are typically hypovascular [8]. Computed tomography (CT) reveals an isodense mass on unenhanced studies. Following IV contrast administration, they appear as a hypodense solid mass with mild delayed enhancement [9]. MRI may show a well-defined isointense mass on T1-weighted images and with either increased or decreased signal intensity on T2-weighted images, with respect to the surrounding normal spleen [9]. The tumor mass shows progressive inhomogeneous enhancement on gadolinium-enhanced images [9]. PET/CT scan usually demonstrates variable uptake, with occasional intense uptake, as was also seen in our case. Intense uptake increases the suspicion for a malignant lesion, especially in patients with a history of malignancy as in our case [6]. Suga et al suggested that a combination of Tc-99m colloid and Ga-67 imaging may be a reliable method to differentiate IPS from hamartoma and lymphoma [10]. IPS shows lack of tracer uptake on both Tc-99m colloid and Ga-67 imaging. This argues against splenic hamartoma which usually show more intense radiocolloid uptake on Tc-99m scan. Additionally, it is different from lymphoma which shows increase uptake on Ga-67 imaging [10]. IPS should also be differentiated from other benign processes (i.e. Hemangiomas that can rupture, causing bleeding in 25% of cases).

Currently, no imaging technique allows definitive preoperative diagnosis. Therefore, the definitive diagnosis can only be made after surgical removal of the spleen and histological evaluation [2]. Microscopic findings are characterized by a spectrum of nonspecific inflammatory changes and are composed mainly of plasma cells, mature lymphocytes, histiocytes and rarely eosinophils in a fibroblastic stroma [10]. Multiple studies have reported splenectomy as the best method of obtaining histopathologic specimen [1,2]. This method has been used both for diagnosis and treatment of this entity. Additionally, fine needle aspiration, performed in previous cases has not been 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 [1]. In our case, we safely performed core needle biopsy. Since conservative therapy, and not splenectomy, was the treatment of choice for our patient, we avoided unnecessary splenectomy. Ultrasound guided core splenic biopsy provided a fast, and safe approach to performing splenic biopsy. Utilizing a coaxial system, caudocranial approach, and along with subsequent closing of the biopsy tract after completing the biopsy we were able to provide a safe and effective approach to splenic biopsy. This spared the patient the need for surgical intervention to obtain a definitive diagnosis. Few reports have also shown that percutaneous image guided biopsy of the spleen is both safe and effective. Keogan et al, performed image-guided percutaneous biopsy on 20 patients. They reported that the complication rates of splenic biopsies was less than 2 percent, which is well within the limits of those associated with biopsies of other abdominal organs [11]. In addition, needle-tract seeding is an exceedingly rare complication, with an estimated occurrence rate of less than 0.01% following percutaneous abdominal fine-needle biopsy [12].

**TEACHING POINT**

Inflammatory pseudotumor of the spleen is a benign lesion with unclear etiology that should be differentiated from a malignant process. Currently no imaging modality can distinguish these lesions. However diagnosis can be safely done by percutaneous guided biopsy.
REFERENCES


FIGURES

Figure 1. A: Axial contrast enhanced computed tomography in a 91-year-old female with IPS. The scan was performed 10 years prior to the current presentation, revealing a hypodense mass within the spleen with minimal peripheral enhancement (white arrows). B: Axial computed tomography of the spleen performed at the patient's current admission rederemonstrating a hypodense mass within the spleen, with central low attenuation and minimal peripheral enhancement (white arrows). The size and appearance seems similar to the prior study [Scan from 2000: 120 kVp and 225 mAs; 10 mm thickness. For Scan from 2009: Siemens Sensation 16; 120 kVp and 320 mAs; 110 ml of intravenous contrast, Optiray (Ioversol)-320, injected 2 ml/sec.]
Figure 2. Unenhanced CT scan (upper row) with PET scan images (middle row) and fused PET/CT scan (lower row images) of a splenic lesion, in a 91-year-old female with IPS. Images demonstrate focal area of increased radiotracer uptake in the spleen (black arrows on the PET scan and yellow arrows on the fusion images). This lesion corresponds with the area of low attenuation in the spleen on unenhanced CT scan (white arrows). The max SUV value of the lesion within the spleen is 9.8. (Philips GEMINI's PET/CT scanner; Performed 60 minutes after injection of 17.1 mCi FDG tracer, with 5 mm slice thickness, CT-based attenuation correction algorithm using two iterations and 8 subsets, 120 kVp, 250 mAs).
Figure 3. A. Grayscale ultrasound image in a 91-year-old female with IPS demonstrates a hypoechoic lesion within the spleen (long white arrow). Image A demonstrates the tract of biopsy needle going through the hypoechoic lesion (short white arrows). B. Post procedure color Doppler evaluation of the site of biopsy demonstrates no evidence of hemorrhage. (Toshiba Aplio XG; curved 3.5-5 MHz)

Figure 4 (left). Representative microscopic image of the core biopsy specimen demonstrating a bland spindle cell population with an associated mixed cellular infiltrate composed of plasma cells, lymphocytes, histiocytes, eosinophils and mast cells (hematoxylin and eosin stain, original magnification, ×200)
<table>
<thead>
<tr>
<th>Disease</th>
<th>Ultrasound</th>
<th>CT scan</th>
<th>MRI</th>
<th>PET/CT scan</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Pseudotumor of the spleen</strong></td>
<td>Well-defined hypoechoic lesion or a partially calcified hyperechoic mass, hypovascular on color flow Doppler ultrasound.</td>
<td>Hypodense mass with occasional calcification with heterogeneous slow enhancement.</td>
<td>Low to iso-intense mass on T1-weighted images and high intense mass with surrounding low intensity on T2-weighted images.</td>
<td>Occasional intense uptake.</td>
</tr>
<tr>
<td><strong>Splenic abscess</strong></td>
<td>Short duration of presentation with a well-defined hypoechoic lesion with fluid component and no calcification. Color flow surrounding the lesion on Doppler ultrasound.</td>
<td>Hypodense mass with fluid attenuation and no calcification. Peripheral ring enhancement.</td>
<td>Low intensity on T1WI, high on T2WI. Peripheral enhancement.</td>
<td>Intense uptake, mainly peripheral.</td>
</tr>
<tr>
<td><strong>Primary Splenic Lymphoma, and Hamartoma</strong></td>
<td>Well-defined hypoechoic lesion with or without calcifications.</td>
<td>Hypodense mass with occasional calcification with heterogeneous enhancement.</td>
<td>Low to iso-intense mass on T1WI and high intense mass on T2WI.</td>
<td>Intense uptake.</td>
</tr>
<tr>
<td><strong>Splenic cyst</strong></td>
<td>Well-defined hypoechoic lesion with fluid component and no calcification.</td>
<td>Hypodense mass with fluid attenuation and no calcification. No enhancement.</td>
<td>Low intensity on T1WI, high on T2WI. No enhancement.</td>
<td>No uptake.</td>
</tr>
<tr>
<td><strong>Hemangioma and angiosarcoma</strong></td>
<td>Hypoechoic lesion with no calcification and no fatty elements.</td>
<td>No fatty elements</td>
<td>No calcification.</td>
<td>Hypoattenuating lesion with uptake.</td>
</tr>
<tr>
<td><strong>Metastasis</strong></td>
<td>Well-defined hypoechoic lesion.</td>
<td>Hypodense mass with occasional calcification with heterogeneous enhancement.</td>
<td>Low to iso-intense mass on T1WI and high intense mass on T2WI.</td>
<td>Intense uptake.</td>
</tr>
</tbody>
</table>

Table 1: Differential diagnoses for Pseudotumor of the spleen
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**Etiology**
Unknown.
Speculated etiologies: infections, focal parenchymal necrosis, autoimmune processes

**Incidence**
Extremely rare. Less than 50 cases reported.
0.007% among all subjects on whom an operation or autopsy is performed

**Gender Ratio**
Male = Female

**Age predilection**
Middle age to older age (range of 20-81 y).

**Risk Factors**
Unknown.

**Treatment**
Splenectomy. No treatment needed in some cases.

**Prognosis**
Excellent prognosis.

**Differential diagnosis**
Splenic cyst, hamartoma, hemangioma, angiosarcoma, lymphangioma, plasmacytoma, metastasis, lymphoma, abscess

**Finds on imaging**
CT scan = hypodense mass with occasional calcification with heterogeneous slow enhancement.

MRI = low to iso-intense mass on T1-weighted images and high intense mass with surrounding low intensity on T2-weighted images.

Ultrasound = well-defined hypoechoic lesion or a partially calcified hyperechoic mass; hypovascular on color flow Doppler ultrasound.

PET/CT scan = occasional intense uptake.

**Table 2: Summary table for Pseudotumor of the spleen**

<table>
<thead>
<tr>
<th>Anti SM</th>
<th>Anti smooth muscle</th>
</tr>
</thead>
<tbody>
<tr>
<td>Anti RNP</td>
<td>Anti Ribonucleoprotein</td>
</tr>
<tr>
<td>Anti SSA</td>
<td>Anti Sjögren's Syndrome A</td>
</tr>
<tr>
<td>Anti SSB</td>
<td>Anti Sjögren's Syndrome B</td>
</tr>
<tr>
<td>Anti Scl-70</td>
<td>Anti Scleroderma-70</td>
</tr>
<tr>
<td>ANA</td>
<td>Antinuclear antibody</td>
</tr>
<tr>
<td>ESR</td>
<td>Erythrocyte sedimentation rate</td>
</tr>
<tr>
<td>HGB</td>
<td>Hemoglobin</td>
</tr>
<tr>
<td>MRI</td>
<td>Magnetic resonance imaging</td>
</tr>
<tr>
<td>PET/CT</td>
<td>Positron emission tomography/Computed tomography</td>
</tr>
<tr>
<td>T1WI</td>
<td>T1 weighted images</td>
</tr>
<tr>
<td>T2WI</td>
<td>T2 weighted images</td>
</tr>
</tbody>
</table>

**ABBREVIATIONS**

**Online access**
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**KEYWORDS**
inflammatory pseudotumor; spleen; image guided; ultrasound; core needle biopsy