Inflammatory pseudotumor of the spleen

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Abstract

Objective: Inflammatory pseudotumor (IPT) is a rare process which occurs in a number of different organ systems.

Methods: This 40 year-old male had no significant past medical history and presented to his primary medical doctor with worsening hypertension.

Results: Inflammatory pseudotumor of the spleen (IPT) is a rare disease process which continues to be a challenge to diagnose.

Conclusions: Inflammatory pseudotumor (IPT) is a rare process which occurs in a number of different organ systems. The spleen is an unusual location for IPT. Due to non-specific clinical findings at presentation and on radiographic imaging, splenic IPT is challenging to diagnose pre-operatively.

Key words
Spleen, Tumor, Inflammatory

Introduction

Inflammatory pseudotumor (IPT) is a rare process which occurs in a number of different organ systems ranging from the lung, respiratory tract, gastrointestinal tract, orbit, liver, lymph nodes, heart, and brain [5]. In general, this tumor is benign in nature; however it is a disease which poses much difficulty in diagnosing. Inflammatory pseudotumor of the spleen falls into the spectrum of an uncommon location. The first case of splenic IPT was reported in 1984 by Cotelingam. Since that initial presentation of this rare tumor, there have been several reported cases in the literature. Due to non-specific clinical findings on presentation and imaging, the diagnosis of this rare tumor becomes difficult. Modalities including sonography, CT scanning, MRI’s, and even angiography have all been implemented in working up patients with splenic lesions, however no single diagnostic study has been successfully able to diagnose this rare disease. Through the use of interventional radiology, the possibilities of less-invasive biopsy options become available. Defining a robust pre-operative differential is essential, as the surgical approach may change whether the disease process is characterized as benign or malignant.
Case presentation

This is a 40 year-old male with no significant past medical history, who presented to his primary medical doctor with worsening hypertension. This was initially diet controlled, however became progressively worse requiring medications. On his subsequent work-up, he underwent a kidney sonogram which revealed a lesion in the anterior pole of the spleen.

During this time period he denied any associated symptoms such as fevers, chills, nausea, changes in bowel habits or significant weight loss. He also denied any family history of cancers, lymphoma’s or similar early onset hypertension. A CT scan of the abdomen and pelvis was performed which revealed a mass in the anterior pole of the spleen measuring 4.4 cm × 4.0 cm with non-specific enhancement pattern (Figure 1). With a benign physical exam, and an indeterminate lesion in the spleen, interventional radiology was notified to evaluate for possible biopsy. Differential diagnoses at the time included a splenic cyst, hemangioma, lymphoma or a solid tumor.

Using ultrasound guidance, an 18-gauge core biopsy of the splenic mass was performed using a sub-costal approach. A total of four passes were made, and specimen placed in formalin. Pathology revealed inflammation and spindle cell proliferation. Immunostains for numerous soft tissue markers were checked, including CD34, Factor XIIIa, smooth muscle actin, desmin, S-100, and CD 31; all of which were suggestive of a non-neoplastic process (Figure 2; Figure 3). Favored diagnoses at this time were of a reactive lesion such as a periphery of infarct or inflammatory pseudotumor. With our pre-operative biopsy results of an inflammatory pseudotumor, a laparoscopic splenectomy was the favored approach, as this lesion was consistent with an underlying benign process.

Operative technique

The patient was placed in the right lateral decubitus position with the left side up. Port placement was in the following locations: 12-mm supra-umbilical, 5-mm sub-xiphoid, 5-mm mid-clavicular, 12-mm anterior axillary, and another 5-mm mid-axillary port.

Dissection began by taking down adhesions around the abdominal wall and the anterior pole of the spleen with the harmonic scalpel. The dense adhesions encountered were likely secondary to the previous biopsy site. The splenic flexure and the left colon were then mobilized, which allowed the take down of the greater omentum from the spleen. Once completed, the retroperitoneal attachments were also freed. The short gastric vessels were identified, and carefully taken using the harmonic scalpel. This left the spleen attached at the hilum, which was then taken using two Endo-GIA 60 vascular loads. The specimen was placed into an endoscopic retrieval bag, spleen morcellized, and removed through the umbilical port which was slightly extended (Figure 4).
Figure 3. Low-power photomicrograph showing the circumscribed, hypocellular tumor mass adjacent to the splenic parenchyma.

Figure 4. High-power photomicrograph on inflammatory pseudotumor showing fibrotic stroma containing myofibroblastic type and inflammatory cells including numerous plasma cells.

The patient was extubated uneventfully, and sent to the PACU in stable condition. On POD 2, he spiked a temperature to 102, WBC increased to 20, and the patient underwent a CT of his chest, abdomen and pelvis. Besides post-surgical changes, there was left lower lobe atelectasis, with a possible superimposed pneumonia. He improved over the next several days, however was placed on albuterol, ipratropium bromide, and pulmicort inhalers, and subsequently discharged on POD [6]. At his follow up appointments, he has been doing well. He has been followed by a pulmonologist, and has since developed asthma, requiring the use of inhalers. Although no definitive correlation to the splenectomy and developing respiratory issues can be made, a question raised could be if a reactive airway disease developed secondary to his decreased immune response or possibility of a recurrent IPT developing in his respiratory tract. He underwent surveillance CT imaging at his 6 months follow-up, and there is no sign of further disease. It is important to note that IPT does affect various other parts of the body including the liver, GI tract and lungs, so continued close surveillance is essential.

Discussion

Inflammatory pseudotumor of the spleen (IPT) is a rare disease process which continues to be a challenge to diagnose. It is formed on the basis of irregular proliferation of inflammatory cells [2]. Generally these tumors have benign behavior with spontaneous regression, but occasionally they have been reported to recur, metastasize and undergo sarcomatous transformation [4]. The clinical symptoms are diverse in nature, with some patients complaining of left flank or abdominal pain, with or without fever and splenomegaly [8]. Due to the vague clinical picture oftentimes encountered, it becomes extremely important that a thorough pre-operative assessment be performed.

Although imaging is not definitively diagnostic, it offers the possibility to rule out other splenic lesions such as cysts, hemangiomas, lymphoma’s, and other solid tumors. Various modalities can be used to image the spleen such as ultrasound, CT, MRI, and even angiography. Splenic inflammatory pseudotumors may appear as homogenous, low-density solid tumors with delayed contrast enhancement on CT [7]. This still leaves a wide array of possible diagnoses of the mass, and further evaluation is often needed. In our patient, CT imaging confirmed that there was non-specific enhancement pattern. Objective diagnosis requires histologic evaluation, characterized by a mixed population of inflammatory and spindle cells, sometimes clustered around a necrotic avascular central area [10].

With the use of interventional radiology, the possibilities of less-invasive biopsies become an option. Defining an accurate pre-operative differential diagnose is essential, as the surgical approach to an invasive biopsy is determined based on the
presumption of a benign or malignant process. The FNA biopsies obtained showed components that were described by Yano; inflammatory cells such as plasma cells, lymphocytes and histiocytes \[^{10}\]. Although this can be interpreted as non-specific, a constellation of these findings on pre-operative biopsy directs us to a benign process, rather than an aggressive malignant tumor. For example, Tanaka et al. reviewed three cases of primary splenic diffuse large B-cell lymphoma, all of which were diagnosed with pre-operative ultrasound guided biopsies \[^{9}\]. Through the use of interventional radiology, the splenic mass was categorized as malignant, and the appropriate treatment plan performed. Generally for malignant lesions of the spleen, a laparoscopic approach is not recommended due to the possibility of seeding port sites with cancer. This further supports the use of less-invasive pre-operative biopsies to guide proper treatment plans.

With the advent of minimally invasive surgical techniques, proper pre-operative differential diagnoses must be evaluated. Through less-invasive biopsy techniques, we are now able to categorize splenic lesions as benign or malignant, and proper treatment plans formulated. As radiological imaging becomes more detailed and accurate, we still rely on histopathology to direct our hands in which operative or non-operative approach serves the patient best.

**Conclusion**

Inflammatory pseudotumor (IPT) is a rare process which occurs in a number of different organ systems. The spleen is an unusual location for IPT. Due to non-specific clinical findings at presentation and on radiographic imaging, splenic IPT is challenging to diagnose pre-operatively.

**Conflict of interest**

The author declares that there is no conflict of interest statement.

**References**


