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Inflammatory Pseudotumor of the Spleen

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Abstract

Background and Aims: Evaluation of effectiveness in the differential diagnosis of the very rare entity of inflammatory pseudotumor (IPT) of the spleen, with presentation of its pathology and pathogenicity.

Methods: The case is presented of a 60-year-old female who underwent ultrasound (US) examination of the upper abdomen, after an accidental fall from a height, which revealed a splenic mass. Meticulous clinical and laboratory examination and imaging confirmed the presence of the splenic mass, which mimicked malignant lymphoma.

Results: Splenectomy was performed and the histopathology report revealed a tumor consisting of inflammatory cells intermingled with spindle and mast cells, consistent with the diagnosis of IPT.

Conclusions: IPTs are very rare benign tumors which mimic other benign or malignant tumors. The clinical diagnosis is difficult, but they have a very good prognosis. Surgeons should be aware of the condition in order to avoid misdiagnosis and inappropriate treatment.

Key words: *Spleen; inflammatory pseudotumor; laparoscopic splenectomy; diagnostic evaluation*

Background

Inflammatory pseudotumor (IPT) is a very rare entity, and the splenic localation is particularly uncommon [1]. IPTs are characterized by proliferation of fusiform cells of unknown cause and origin and their histopathological appearance mimics that of a tumor. Many organs have been reported as the site of IPTs, including the eye, meninges of the spinal cord, digestive system, heart, soft tissues, the mesothelial membrane, bladder, lymph nodes, liver and spleen, but the lungs are the most frequently described location. Regarding the pathogenesis, IPT has been variously associated with the Epstein Barr virus (EBV), trauma and vascular disorders, and with an immune etiology [2-4].

We present the case of a 60-year-old female with the incidental finding of IPT of the spleen.

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Case report

A 60-year-old female underwent upper abdominal ultrasound (US) because of an accidental fall from a height. The US revealed splenomegaly with no other abnormal findings. She had a past history of arthritis.

Laboratory examination showed mild leucocytosis and raised inflammatory markers, including the erythrocyte sedimentation rate (ESR) 112 mm/hr (normal value 0-29 mm/hr), and C-reactive protein (CRP) 45mg/dl (normal value 0-10mg/dl).

Abdominal computed tomography (CT) revealed a heterogeneous splenic mass with slow uptake of iv contrast and a hypodense central area. Magnetic resonance imaging (MRI) showed delayed enhancement, with a rim between the splenic parenchyma and the mass and a hypodense central area, mimicking lymphoid tumor (Fig 1A, 1B).

The patient underwent open splenectomy along with complete resection of the tumor, which had infiltrated the splenic hilum and was 14 cm in its greatest dimension (Figure 2B).

The histopathology was characterized by a large number of inflammatory cells and desmoid-type tumor morphology, synoptically named as an inflammatory myofibroblastic tumor. The presence of spindle and mast or mast-like cells was pathognomonic of IPT (Figure 2A).

Triple immunization for *Streptococcus pneumoniae, Haemophilus influenzae type B,* and *Neisseria meningitides* was administered 14 days prior to the operation, to decrease the risk of overwhelming post-splenectomy sepsis from these organisms. Testing for EBV antibodies was negative

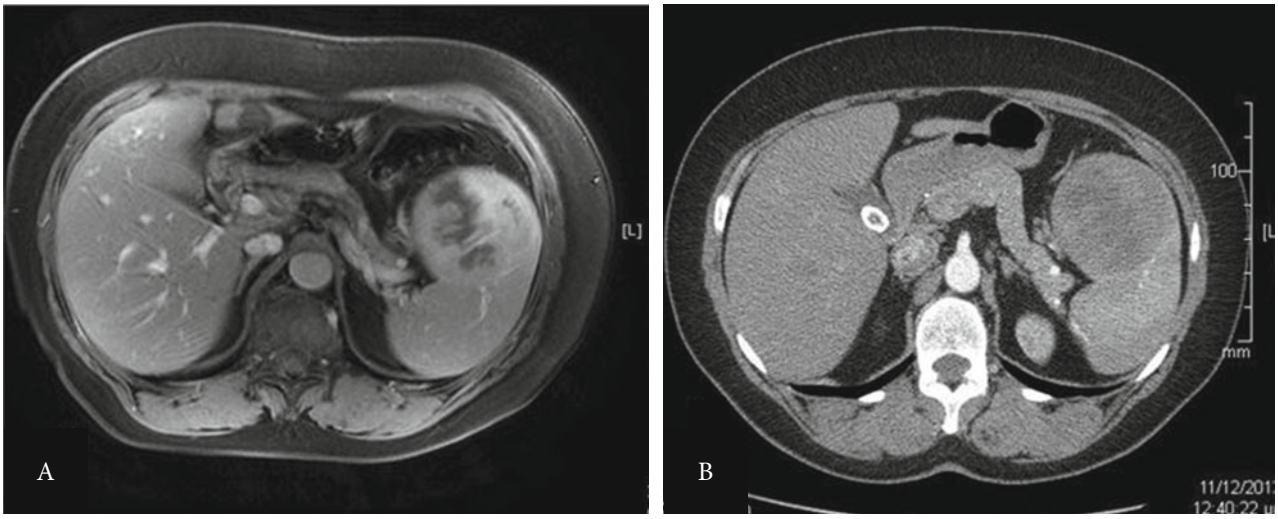

Figure 1. Abdominal computed tomography (CT) in a 60-year-old female, showing an inflammatory pseudotumor of the spleen with (A) hypodense, low attenuation in the non-enhanced CT, and (B) heterogeneity in the enhanced CT.

and the etiology of the IPT was unclear.

The postoperative course was uneventful and the patient is in very good condition 6 months after surgery.

Discussion

The incidence of primary malignant tumors located in the spleen accounts for 0.007% of malignancies. IPT is a very rare entity. It combines specific histopathological, consisting of a variety of inflammatory cells. The first 2 cases of splenic pseudotumors were described by Cotelingam and Jaffe in 1984, although the term "inflammatory" had been recorded in the medical literature in this context in 1954. IPT is a benign lesion found in various locations, including the orbit, the liver, and the respiratory and digestive tracts [5-7].

The etiology and pathogenesis of IPT are unknown, but possible causes are previous Epstein- Barr infection, immune disorders and vascular causes, focal parenchymal necrosis with hemorrhage and a granulomatous inflammation process with the activation of such inflammatory mediators as interleukin-1 (IL1) [8].

IPT has also been reported following a ruptured splenic hemangioma, and some authors postulated ineffective antibiotic therapy, or an abnormality of lipid metabolism as the etiology, but the pathogenesis remains unclear. [4,7,9].

The main symptoms and signs of splenic IPT are ab-

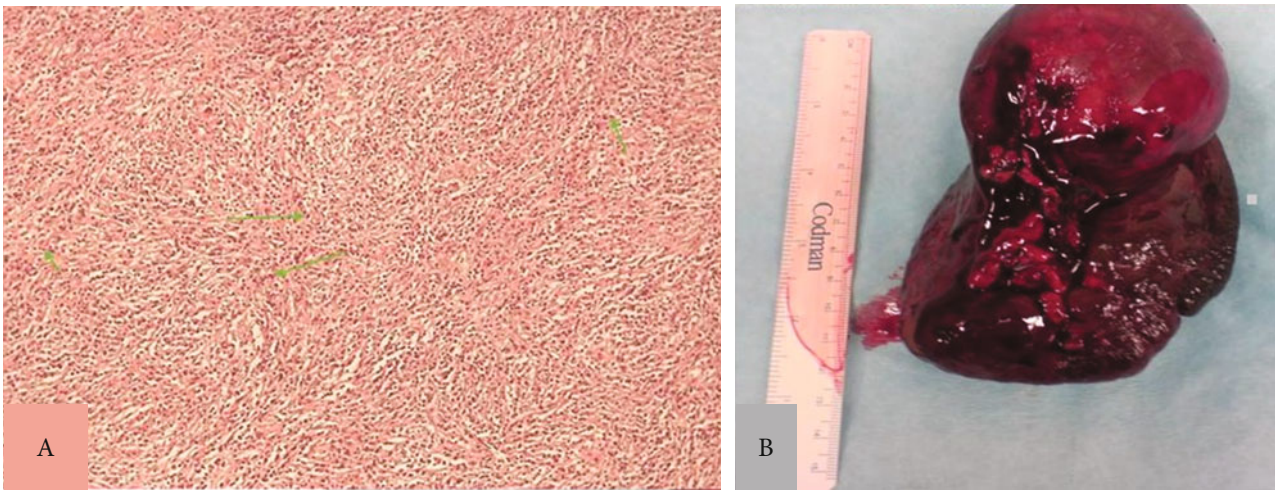

Figure 2. Inflammatory pseudotumor of the spleen in a 60-year-old female. A: Histopathology of the splenic lesion (HE x100) showing inflammatory myofibroblastic tumor: The desmoid morphology of the lesion is evident, with a large number of inflammatory cells (mainly lymphocytes and plasma cells) (green arrows). B: Gross appearance of the splenic pseudotumor.

dominal discomfort, left upper quadrant pain with the Kehr sign and a palpable mass, but it may be an incidental finding, as in the case presented here.

The main US features of the splenic mass are partial calcification with a hypoechoic appearance, presenting as a well-defined encapsulated tumor.

Non-enhanced CT usually reveals a rounded hypodense mass with low attenuation, while enhanced CT is characterized by heterogeneity with slightly intense enhancement [4,10,11]. MRI T2 sequence shows analogous heterogeneity, but with a discrete periphery delineating it from the surrounding normal spleen. Dynamic MRI reveals a low signal in the early phase and a high signal intensity lesion in the delayed phase.

Preoperative needle biopsy of a splenic lesion encompasses the risk of bleeding and possible dissemination in the case of a malignant tumor, and it is not recommended for suspected splenic IPT [12]. The same inflammatory reaction is present in the periphery of malignant tumors, but even if the needle biopsy could establish the diagnosis, the patient would not avoid surgery [13,14].

In other locations, such as lung and liver, however, it has been reported that fine needle aspiration (FNA) or core needle gun biopsy can play a valuable role in the diagnosis of IPT [14].

The differential diagnosis of splenic IPT is between malignant lesions, such as malignant lymphoma, malignant fibrous histiocytoma, plasmatocytoma, hemangiosarcoma, and other benign lesions, such us hemangioma, hamartoma, cysts and lymphangioma [1].

Recently, the use of laparoscopic splenectomy for splenic IPT has been reported to be suitable for small tumors and tumors with a low preoperative malignant potential [14].

The 3 main histopathological subtypes of IPT reported in the literature are: i. xanthogranuloma type, ii. plasma cell granuloma type, and iii. sclerosing pseudotumor [8]. These types may reflect the variation of the disease process, since IPT constitutes a variety of solid space occupying lesions mimicking benign or malignant tumors [8].

Although the prognosis of splenic IPT is good after splenectomy, follow-up is highly recommended, as septic complications have been described, with associated mortality, following excision of IPTs located in other organs [13,14].

Ethical Approval: *All human studies have been approved by the appropriate ethics committee and the patient gave her informed consent prior to inclusion in this study.*

Conflict of Interest: *The authors declare that they have no conflict of interest.*

References

1. Tsugawa K, Hashizume M, Migou S, et al. Laparoscopic splenectomy for an inflammatory pseudotumor of the spleen: Operative technique and case report. Hepatogastroenterology 1998;45:1887-91.
2. Yamaguchi M, et al. Specific detection of Epstein-Barr virus in inflammatory pseudotumor of the spleen in a patient with a high serum level of soluble IL-2 receptor. J Gastroenterol 2000;35:563-6.
3. Kutok JL, Pinkus GS, Dorfman DM, et al. Inflammatory pseudotumor of lymph node and spleen: An entity biologically distinct from inflammatory mycofibroblastic tumor. Hum Pathol 2001;32:1382-7.
4. Chen WH, et al. Inflammatory pseudotumor of the spleen. J Chin Med Assoc 2004;67:533-6.
5. Cotelingam JD, Jaffe E. Inflammatory pseudotumor of the spleen, Am J Surg Pathol 1984;8:375-80.
6. Martinez Celada M, et al. 37 year old woman with abdominal discomfort and splenic focal lesion. Rev Clin Esp 2007 March:135-7.
7. Yafei Zhang, Hongwei Lu, Hong Ji, et al. Inflammatory pseudotumor of the liver: A case report and literature review. Intractable Rare Dis Res 2015;4:155-8.
8. Ugalde P, García Bernardo C, Granero P, et al. Inflammatory pseudotumor of spleen: A case report. Int J Surg Case Rep 2015;7:145-8.
9. Matsubayashi Hiroyuki, Mizoue Toshiroh, Mizuguchi Yasuhiro, et al. A case of hemangioma accompanied by inflammatory pseudotumor of the spleen. J Clin Gastroenterol 2000;31:258-61.
10. Franquet T, Montes M, Aizcorbe M, et al. Inflammatory pseudotumor of the spleen: Ultrasound and computed tomographic findings. Gastrointest Radiol 1989;14:181-3.
11. Glazer M, Sagar V. SPECT imaging of the spleen in inflammatory pseudotumor: correlation with ultrasound, CT, and MRI. Clin Nucl Med 1993;18:527-9.
12. Jiqi Yan, Chenghong Peng, Weiping Yang, et al. Inflammatory pseudotumour of the spleen: Report of 2 cases and literature review. Can J Surg 2008; 51:75-6.
13. Jae Hak Kim, Jae Hee Cho, Moo Suk Park, et al. Pulmonary Inflammatory Pseudotumor. A report of 28 cases. Korean J Intern Med 2002;17:252-8.
14. Zhen-Hai Ma, Xiao-Feng Tian, Jinhui Ma, et al. Inflammatory pseudotumor of the spleen: A case report and review of published cases. Oncol Lett 2013;5:1955-7.