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Inflammatory pseudotumor of spleen: a case report



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ABSTRACT

INTRODUCTION: Inflammatory pseudotumor of spleen is an extremely rare benign condition of uncertain etiology that presents with nonspecific symptoms or as an incidental finding in patients studied by other processes. Since the first description in 1984 by Cotelingam and Jaffe, only 114 cases have been reported. **PRESENTATION OF CASE:** We present a case of a fifty-six years old woman with a splenic injury in ultrasound and computed tomography. The patient undergoes laparoscopic splenectomy and the histologic study of the specimen revealed findings consistent with inflammatory pseudotumor of spleen.

DISCUSSION: This rare entity whose pathogenesis is still unknown, can present with nonspecific symptoms. Radiologic studies may lead the diagnosis being useful CT and MRI. The definitive diagnosis is established with the histological findings, characterized by the presence of inflammatory cells with areas of necrosis and fibrosis. There are multiple differentials diagnoses: metastasis, lymphoma, splenic infarction, hemangiomas, vascular malformations, lymphangioma, plasmacytoma, reactive lymphoid hyperplasia, abscess and infectious granulomatous processes; therefore suspicion of malignant neoplasm must be considered, being indicated splenectomy to confirm the diagnosis.

CONCLUSION: Inflammatory pseudotumor of spleen is a benign disease, in which diagnostic approach must bear in mind the possibility of a malignant lesion. For this reason, the surgical approach is appropriate to confirm the diagnosis and rule out malignancy with histology.

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1. Introduction

Inflammatory pseudotumor of spleen is an extremely rare benign condition of uncertain etiology that presents with nonspecific symptoms or as an incidental finding in patients studied by other processes.

We present a case of inflammatory pseudotumor in a woman of fifty-six years old with a splenic injury in ultrasound and computed tomography (CT).

The definitive diagnosis is established with the histological findings, characterized by the presence of inflammatory cells with areas of necrosis and fibrosis.

There are multiple differentials diagnoses: metastasis, lymphoma, splenic infarction, hemangiomas, vascular malformations, lymphangioma, plasmacytoma, reactive lymphoid hyperplasia, abscess and infectious granulomatous processes; therefore suspicion of malignant neoplasm must be considered, being indicated splenectomy to confirm the diagnosis.

2. Presentation of case

Fifty-six year old woman was referred to our service with a splenic nodule found in the studies for renal colic. The patient complained of epigastric and left lower back pain since childhood. She did not present acute or chronic abdominal pathology. Physical examination shows good overall appearance with a normal abdominal exploration with no palpable masses or organ enlargement. Ultrasound described a splenic cyst of 65 mm with internal calcifications which could correspond with hydatid cyst. Radiological studies were completed with CT, identifying a mass in the splenic middle third of 72 × 62 × 61 mm with well-defined margins, heterogeneous density with a peripheral hyperdense area and central hypodense zones, without criteria of cystic lesion and suspicious for malignancy (Fig. 1). The analytical before surgery (blood count, biochemistry) were normal as well as echinococcus granulosus serology.

The patient undergoes laparoscopic splenectomy with removal of the entire piece in endobag and placing a suction drainage in splenic cell. Postoperative elapses without complications being discharged on sixth day. Three years after surgery the patient remains asymptomatic without evidence of recurrence.

The splenectomy piece weighed 650g and measured 12 × 11.5 × 7cm. Macroscopically the capsule was intact with

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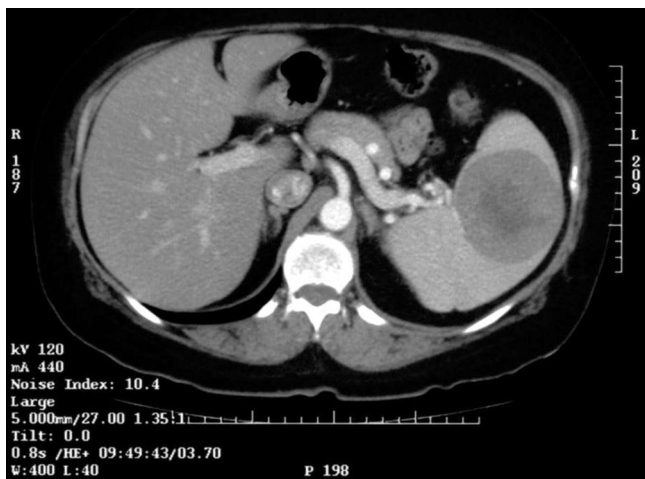


Fig. 1. Computed tomography.



Fig. 2. Surgical specimen.

yellowish lesions which correspond to a well-defined nodular lesion, unencapsulated of 7 cm with a necrotic center. The surgical margin in the splenic hilum was not affected (Fig. 2).

Microscopic findings were a well circumscribed cell proliferation of mixed cell population, predominated by plasmatic cells and lymphocytes with isolated eosinophils and multinucleated giant cells. There are necrotic areas “pseudogranulomatous” with cellular detritus and some with calcifications (Fig. 3).

3. Discussion

Inflammatory pseudotumors are benign conditions of unknown etiology that may present in different locations (orbit, liver, respiratory and digestive tracts) [1,2]. Inflammatory pseudotumors of the spleen are extremely rare. Since the first description in 1984

by Cotelingam and Jaffe, only 114 cases have been reported [3,4] (Table 1).

According to data released, most patients are middle – advanced age and some reports suggest a greater tendency of affection in woman [2,4]. There is no typical clinical presentation and symptoms may be disparate from abdominal pain or discomfort, fever, splenomegaly or showing signs and symptoms suggesting malignancy to incidental findings in the study of other diseases [1,4,5]. Laboratory data are usually normal, although hypercalcemia, monoclonal peaks in the proteinogram and polyclonal hypergammaglobulinemia have also been reported [1,6].

There are different theories about its etiology: bleeding or rupture of hemangioma, bacterial infection, Epstein- Barr virus and herpes infection, inflammatory granulomatous process, vascular causes and other immune disorders. However, the real pathogenesis remains unknown [1,2,4,5,7,8].

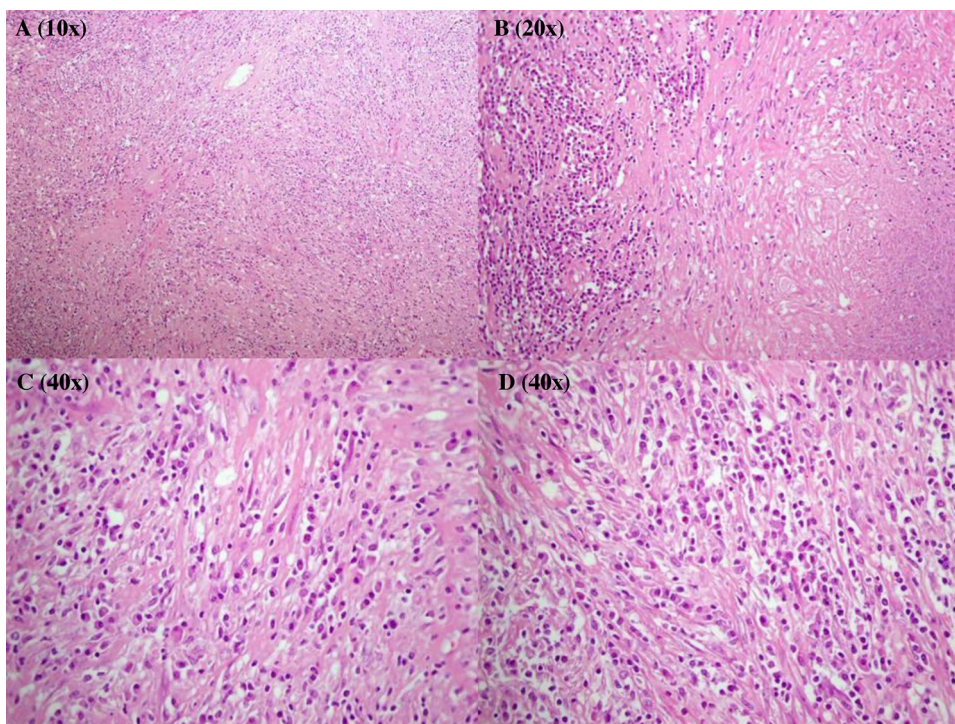


Fig. 3. A, B, C, D – Tumor composed of mature plasma cells, small lymphocytes and variable proportion of spindle cells and collagen bundles (hematoxylin-eosin). B – Necrosis area to the right of the image.

Table 1
Clinic, radiology and pathology of the reported cases.

Authors	Number of cases	Age	Sex	Symptoms	Laboratory	Radiological findings	Preoperative biopsy	Histology	Follow up
Yan J, et al.	2	60–77	M–F	Abdominal discomfort. Weight loss.	Elevated immunoglobulin G and B2-microglobulin.	CT: Low density hypovascular mass. Central necrosis.	No	Necrotic areas. Mix of inflammatory cellular elements, predominantly plasma cells and lymphocytes.	3–10 months. Asymptomatic
Martinez Celada, et al.	1	37	F	Abdominal discomfort. Weight loss. General syndrome.	Normal	CT: Focal splenic injury: hypodense, homogeneous, with peripheral uptake.	No	Plasma cells, lymphoid elements and occasional eosinophils.	Asymptomatic
Ma ZH, et al.	1	77	M	Asymptomatic.	Normal	CT-MRI: Splenic mass with diffuse heterogeneous enhancement.	No	Necrotic focus in the center, with admixture of inflammatory cellular elements, predominantly plasma cells and lymphocytes with hyalinization, fibrosis, lymph follicles and multinuclear giant cells.	4 months. Asymptomatic
Rosenbaum, et al.	2	33–60	M–F	Abdominal pain. Weight loss. Night sweats.	NA	CT: Splenic mass.	No	Necrotic foci. Mixed inflammatory infiltrate with abundant mature plasma cells and a proliferation of spindled cells. Abundant EBV-infected cells that included the proliferating oval and spindle cells.	17–9 months. Asymptomatic
Noguchi H, et al.	1	72	F	Nausea.	Normal	CT: Partially calcified, low-density, hypo-vascular, well-defined, smooth mass. MRI: Splenic mass with low to iso-intensity on T1 and high intensity with surrounding low intensity on T2. Low intensity in the center of the lesion. Angiography: Hypovascular area in the arterial phase.	No	Focus of necrosis in the center, with admixture of inflammatory cellular elements, predominantly plasma cells and lymphocytes with hyalinization, fibrosis, lymph follicles and multinuclear giant cells.	24 months. Asymptomatic
Matsubayashi H, et al.	1	61	F	Nasal bleeding.	Pancytopenia, Hepatitis C virus antigen-antibody (+)	CT: Splenomegaly. Tumor with nodule-in-nodule pattern, with low density outer nodule and high density inner nodule. High contrast enhancement in early phase in inner nodule. MRI: Low intensity inner nodule on T1. On T2 low intensity outer nodule with a highly intense inner nodule. Angiography: Cotton-wool appearance.	No	Granulomatous component, with large amount of giant cells, plasma cells, lymphocytes and fibroblast. Inner mass with histology of cavernous hemangioma.	NA
Fonguerat M, et al.	2	56–67	F–M	Abdominal pain. General syndrome.	Anemia	CT: Splenic injury with low attenuation, homogeneous and without well-defined borders. MRI: Splenic mass with low intensity on T2.	Yes	NA	
Bhatt S, et al.	1	31	M	NA	NA	CT: Hypodense splenic lesion with early enhancement. MRI: Isointense on T1. Low intensity on T2.	No	Mixed inflammatory infiltrate of lymphocytes, plasma cells and occasional eosinophils.	NA
Hamdi I, et al.	1	48	F	Abdominal pain.	Normal	US: Hypoechoic, heterogeneous splenic lesion. MRI: Isointense on T1. Heterogeneous mass with multiple hypointense partitions in radial layout on T2.	No	Admixture of inflammatory cellular elements represented for plasma cells, eosinophils and histiocytes.	NA
Yano H, et al.	2	51–51	F–M	Abdominal pain. Diarrhea. Asymptomatic.	Leukocytosis. Elevated CRP.	CT: Low density splenic mass. MRI: Low to isointensity on T1. Irregular intensity on T2.	No	Infiltration of plasma cells, lymphocytes and histiocytes.	NA

Radiological tests could orientate but there are not pathomonic images. Ultrasound can show a hypoechoic splenic mass with or without calcifications [1,9–13]. CT can show a low density injury that usually has an attenuated central zone corresponding with fibrosis in the histological findings. Calcifications may be present [6,9,10,12–15].

MRI may reveal a well-defined mass, isointense on T1-weighted images, and with either an increased or decreased signal intensity on T2-weighted images, with respect to the surrounding normal spleen [1,4,10,16].

Preoperative biopsy of spleen is not recommended because of poor specificity, the risk of possible bleeding or tumor spread in case of malignancy, so it is recommended splenectomy and evaluation of the surgical specimen [1,2,17].

Differential diagnoses are broad as metastasis, malignant lymphoma, inflammatory granuloma, splenic infarction, hemangiomas, hamartomas, lymphangioma, plasmacytoma, hemangioendotheliomas, angiosarcomas, reactive lymphoid hyperplasia, abscess, infectious granulomatous processes and sarcoidosis, so the final diagnosis of pseudotumor is established with the pathological anatomy study [2,4,6,12,13,15].

The cellular composition can be remarkably heterogeneous and show some resemblance to granulation tissue. Normal lymphocytes and plasma cells are constant features, although variable in mixture and number. Neutrophilic and eosinophilic leukocytes are also present in certain cases. Although there have been attempts to classify these lesions, the terminology used when referring to this entity is confusing. Somere classified these lesions into 3 histopathologic subtypes: xanthogranuloma type, plasma cell granuloma type and sclerosing pseudotumor [6,17,18].

The prognosis is good after splenectomy. Local invasion, recurrence or metastases have not been described. However, clinical follow up is recommended, since some patients with inflammatory pseudotumors of the liver are reported probably to have died as result of the disease [1,2,6] (Table 1).

4. Conclusion

Inflammatory pseudotumor of spleen is a benign lesion of uncertain origin with multiple differential diagnoses. Currently it is not possible to establish a definitive diagnosis of this entity preoperatively, therefore suspicion of malignant neoplasm must be considered, being indicated splenectomy to confirm the diagnosis by the pathological anatomy of the surgical piece.

Conflicts of interest

All authors declare that there are not conflicts of interest.

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Ethical approval

Not necessary for case report.

Author contribution

Paúl Ugalde – Design, data collection, writing the paper. Carmen

García Bernardo – Design, data collection, writing the paper, surgeon of the case. Pablo Granero – Review, contributor. Alberto Miyar – Review, contributor. Carmen González – Pathologist of the case, microscopy photos, contributor. Ignacio Gonzalez-Pinto – Review, contributor. Luis Barneo – Review, contributor. Lino Vazquez – Review, contributor.

Consent

The authors confirm that written informed consent was obtained from the patient for publication of this case report and accompanying images.

Guarantor

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