RESEARCH ARTICLE

INFLAMMATORY PSEUDOTUMOR OF THE SPLEEN: A CASE REPORT

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Manuscript Info

Abstract

Inflamatory pseudotumor of the spleen (IPTS) is an extremely rare benign condition of uncertain etiology. Preoperative diagnosis of this entity is usually difficult, only the histological study allows the diagnosis to be made. We report an observation of a 35-year-old women patient, hospitalized for left hypochondrium pain with vomiting. Physical examination noted tenderness of the left hypochondrium and splenomegaly, CT scan showed a large tissue mass, measuring 14x15x9.6 cm, probably at the expense of the upper pole of the spleen, containing central calcifications. Exploratory laparotomy revealed a splenic tumor. A splenectomy was performed. IPTS diagnosis was made after histological and immunohistochemical study of the splenectomy specimen.

Introduction:

Inflammatory pseudotumor is a benign and rare tumor. Composed of polymorphous inflammatory cell infiltrates, fibrosis, necrosis, granulomatous reaction, and myofibroblastic spindle cells. Its etiology is unknown, that often may imitate a malignant tumor [1].

Occurring preferentially in adults, with a predominantly female and can sit in any organ with a predilection for the eye orbit and the respiratory system. Splenic localization is extremely rare [2]. Inflammatory pseudotumor of the spleen (IPTS) presents with nonspecific symptoms or as an incidental finding in patients studied by other processes and only the histological study allows the diagnosis to be made [3].

Case Presentation:

A 35-year-old women with type II diabetes, was hospitalized for left hypochondrium pain with vomiting. Physical examination noted tenderness of the left hypochondrium and splenomegaly, without abnormal peripheral lymphadenopathy. Laboratory blood tests showed mild anemia with hemoglobin of 10 g/dl, platelet count of 584000/µL, a plasma C-reactive protein level of 217.2 mg/l and negative hydatid serology.

Abdominal ultrasound revealed a mass in the left hypochondrium, measuring 12x9 cm, hypoechoic, containing central calcifications. Abdominal computed tomography (CT) scan (figure 1) showed a large tissue mass, measuring 14x15x9.6 cm, probably at the expense of the upper pole of the spleen, occupying the interhepatosplenic space, pushing the stomach inside, homogeneously enhanced by contrast, containing non-enhanced central calcifications. There was no detectable lymphadenopathy.

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Abdominal CT scan showed a large tissue mass measuring 14x15x9.6 cm, containing central calcifications. Midline exploratory laparotomy revealed a splenic tumor, without invasion of neighboring organs, or locoregional lymphadenopathy. A splenectomy was performed (figure 2).

In the anatomopathological study, the resection specimen weighed 1240g, had an oval mass of 16/13/11cm with a smooth and calcified external surface and a fairly well limited greyish appearance, continued with a spleen of 12/6/3 cm. The mass corresponds to a splenic parenchyma of modified architecture and dissociated by moderate fibrosis with calcifications and strewn with inflammatory elements essentially mononuclear with polynuclear cells, macrophage histiocytes and some epithelioid-looking cells and rare cells with an irregular nucleus sometimes polypoid and nucleolus. The cytoplasm is quite abundant sometimes poorly limited. After the immunohistochemical study the diagnosis of inflammatory pseudotumor of the spleen was made. The patient had simple operative suites.

Discussion:-
Since the publication of the first two cases in 1984 by Cotelingam and Jaffe, a hundred cases have been reported concerning IPTSs. Inflammatory pseudotumors are benign tumors of unknown etiology [4]. Several hypotheses have been put forward: bleeding or rupture of hemangioma, bacterial infection, Epstein-Barr virus and herpes infection, inflammatory granulomatous process, vascular causes and other immune disorders [3]. The IPTS would be considered as a non-specific response of the spleen to an attack of various etiologies [5].

It occurs preferentially in the adult subject, with a female predominance. Clinical signs are nonspecific and vary from person to person, the symptomatic patient may present with left upper quadrant abdominal pain or discomfort,
nausea, vomiting, fever, anemia, weight loss, splenomegaly [6,7]. Nevertheless a fortuitous discovery of an asymptomatic mass during an examination for another condition or during a laparotomy has already been described [4].

On routine abdominal investigations, splenic pseudotumor mimics with other splenic neoplasms. Abdominal ultrasonography is the initial investigation to diagnose morphological abnormalities of the spleen, can show a hypoechogenic splenic mass with or without calcification [3]. On color flow Doppler ultrasound this mass is typically hypovascular [7]. CT scan reveals an isodense mass on unenhanced studies. Following IV contrast administration, they appear as a hypodense solid mass with mild delayed enhancement [7]. The central stellate hypodense zone is related to a fibrous plaque [2]. Magnetic resonance imaging may reveal 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 [7]. The tumor mass shows progressive inhomogeneous enhancement on gadolinium-enhanced images [7]. Positron emission tomography usually demonstrates variable uptake, with occasional intense uptake [7].

Needle biopsy is not recommended for splenic masses due to of poor specificity, the risk of metastases in the event of a malignant tumor and the potential bleeding complications of the procedure [3]. Therefore, histological examination of surgical resection specimens is the standard for the diagnosis of spleen tumors [8].

The typical macroscopic appearance of an IPTS lesion is that of a well-circumscribed and unencapsulated mass varying in size between 4 and 19 cm, with a large amount of ocher-white necrotic tissue in the center. The multinodular form has also been described [4,8].

The cellular composition of IPTS may be remarkably non-specific and heterogeneous. The commonest finding is reactive nature of benign cells, and shows some resemblance to granulation tissue, normal lymphocytes and plasma cells are constant features, although in variable mixtures and numbers. Neutrophilic and eosinophilic leukocytes are also sometimes present. Someren classified these lesions into 3 histopathologic subtypes: xanthogranuloma type, plasma cell granuloma type, and sclerosing pseudotumor [6,9].

The diagnosis of malignant lymphoma was initially made in a number of cases of IPTS reported in the literature, as occurred in the present case prior to immunohistochimy [8]. The differential diagnosis for IPTS also considers other lesions, including metastasis, inflammatory granuloma, splenic infarction, hemangiomas, hamartomas, lymphangioma, plasmacytoma, hemangioendotheliomas, angiosarcomas, reactive lymphoid hyperplasia, abscess, infectious granulomatous processes and sarcoidosis [3].

Treatment of IPTS is surgical, with total splenectomy in the majority of cases. Partial splenectomy is rarely indicated [2]. Using laparoscopic or open surgery is a relatively suitable surgical approach [8]. However, laparoscopic splenectomy is safe and beneficial treatment for this tumor and should be performed in cases in which it is diagnosed or is strongly suspected before surgery [10].

The prognosis of IPTS is good after splenectomy. There have been no reports of local invasion, metastatic disease or recurrence. However, clinical follow up is recommended [8].

**Conclusion:-**

Inflammatory pseudotumor of the spleen is a very rare benign lesion of uncertain origin with multiple differential diagnoses. Currently no imaging modality can distinguish these lesions. Only splenectomy and histopathological study of the specimen enable a definitive diagnosis, therefore splenectomy is diagnostic and curative.

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