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Case Report

Inflammatory myofibroblastic tumor of the spleen: a case report ☆

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ABSTRACT

Inflammatory myofibroblastic tumors (IMTs), otherwise known as the inflammatory pseudotumor, is a rare solid mesenchymal tumor, simulating malignant neoplasms, histologically characterized by the proliferation of spindle cells in a fibrous myxoid stroma containing inflammatory cells. CT and MR imaging are the most used tools in their assessment. Clinical features are nonspecific and depend on the localization of the tumor, radiologic findings are polymorphic and no-conclusive and present a diagnostic challenge to the radiologist. Although histology remains obligatory for the final diagnosis. Heren, we report a case of splenic IMT with histological correlation.

Introduction

Inflammatory myofibroblastic tumors (IMTs) are a rare solid mesenchymal tumor commonly documented in children and young adults. It is often observed in the lungs, the extra pulmonary location is rare and mainly affects the intestinal mesentery and the omentum. These tumors have nonspecific clinical features and various radiological appearances. Histology remains fundamental for the final diagnosis. Heren, we report a case of splenic IMT with histological correlation.

Case report

A 45-year-old female, consulted for left upper quadrant (LUQ) pain for over 9 months coupled with unexplained weight loss and intermittent fever. Laboratory results revealed a white blood cell count of 10 000 /mL with segmental neutrophilia (70 %) and slightly elevated levels of C- reactive protein (CRP, 109 mg/L) and fibrinogen (5.11 1 g/L); the erythrocyte sedimentation rate (ESR) and hemoglobin level (9 g/dL) were normal.

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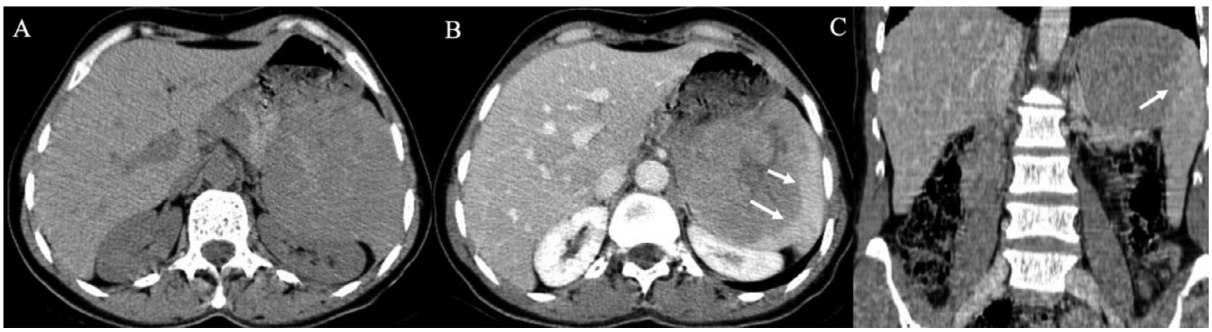

Fig. 1 – CT Scan images in axial and coronal views before and after injection of contrast agent showing superior polar mass lesion of the spleen, well limited, with heterogeneous enhancement in portal venous phase; the tumor presented a contact with the posterior face of the stomach and the left kidney without sign of invasion.

Ultrasound showed that there was a well limited hypoechoic splenic mass with no vascular signal on Doppler. An abdominal contrast- enhanced computed tomography (CECT) confirmed tumor localization in the superior polar of the spleen, and showed a heterogeneous enhancement in portal venous phase; the tumor presented a contact with the posterior face of the stomach and the left kidney without sign of invasion, the masse size was 6,5 cmx 7 cm. In front of this appearance, a sarcomatous origin was evoked (fig. 1). An Ultrasound-guided percutaneous splenic biopsy was done. A microscopic examination revealed a myofibroblastic proliferation. total splenectomy was recommended in patient. Microscopic study of the resected spleen was composed of a proliferation of spindle-shaped cells arranged in hyaline material with chronic inflammatory cells, composed mainly of plasma cells and lymphocyte. Immunohistochemical findings were compatible with an IMT (Fig. 2).

Discussion

Inflammatory myofibroblastic tumors (IMTs), otherwise known as inflammatory pseudotumors, are rare mass lesions, simulating a malignant tumor process, with a tendency to local recurrence. They affect soft tissues and visceral organs; the most frequent localizations are pulmonary and intra-orbital. Histologically characterized by a proliferation of spindle cells in an inflammatory stroma composed mainly of plasma cells and lymphocytes, with eosinophilic and neutrophilic mixtures. [1,2]

These spindle cells are myofibroblastic with an eosinophilic cytoplasm, and a vesicular nucleus containing one or two small nucleoli. immunohistochemistry analysis is very important. Tumor cells are characteristically positive for vimentin and do not express CD117 and CD34 in IMT.1 12 The cells are positive for smooth muscle actin (SMA) with or without desmin expression and S100 positivite.13 .Their mitotic activity is low (0 to 2 mitoses / 10 HPF) and atypical mitoses are rare. [1,2,3,4]. Necrosis and vascular invasion have been reported in typical IMT, they have been shown to express chromosomal rearrangements explaining their locally aggressive character, about 50% of IMTs express the ALK gene rearrangement found frequently in younger patients. [1,5,6].

The clinical manifestations of IMTs depend on the localization, however an inflammatory syndrome may be associated involving fever, weight loss, anemia, hyperplateletosis, polyclonal hypergammaglobulinemia and high sed rate. This is due to a group of secreted inflammatory mediators [7].

The radiological features of IMTs are polymorphic and inconclusive; on ultrasound these tumors can appear hypoechoic circumscribed mass lesion, with non-systemic vascularization. Computed tomography (CT) and magnetic resonance (MR) are the most used imaging tools in their assessment. CT scan shows a hypodense or isodense lesion, with variable contrast due to the fibrous contingent. It is heterogeneous in the case inflammatory fibrosis, delayed and homogeneous in the case of collagenous fibrosis. The same semiological aspects are found in MRI [8]. They present a diagnostic challenge to the radiologist, although histology remains obligatory for the final diagnosis.

Splenic IMTs are extremely rare, their incidence is about 0.0007% including all surgical and autopsy series. according to our research, 120 cases have been reported in the literature including our case. The true pathogenesis of IMTs remains undetermined, some investigators believe that it represents an immunologic response to an infectious or noninfectious agent 1,5 a history with EBV, HIV, and HHV-8 infections, of surgery, trauma, bleeding or rupture of hemangioma, or disorders immunological was reported [9,10,11].

The clinical manifestations of splenic localization signal pain of HCG, splenomegaly. The imaging techniques show various aspects. According to a study by Hayasaka et al, ultrasound shows in the majority of cases a hypoechoic hypovascularized mass on color Doppler, the hypovascular character was confirmed on angiography [12]. On CT scan, the mass is almost always hypodense in spontaneous contrast and in the venous portal phase, with a weak enhancement in delayed phase [12,13]. on MRI the tumor shows a low signal T1, a low signal T2 and the enhancement is heterogeneous and depends on fibrosis condition [13]. The differen-

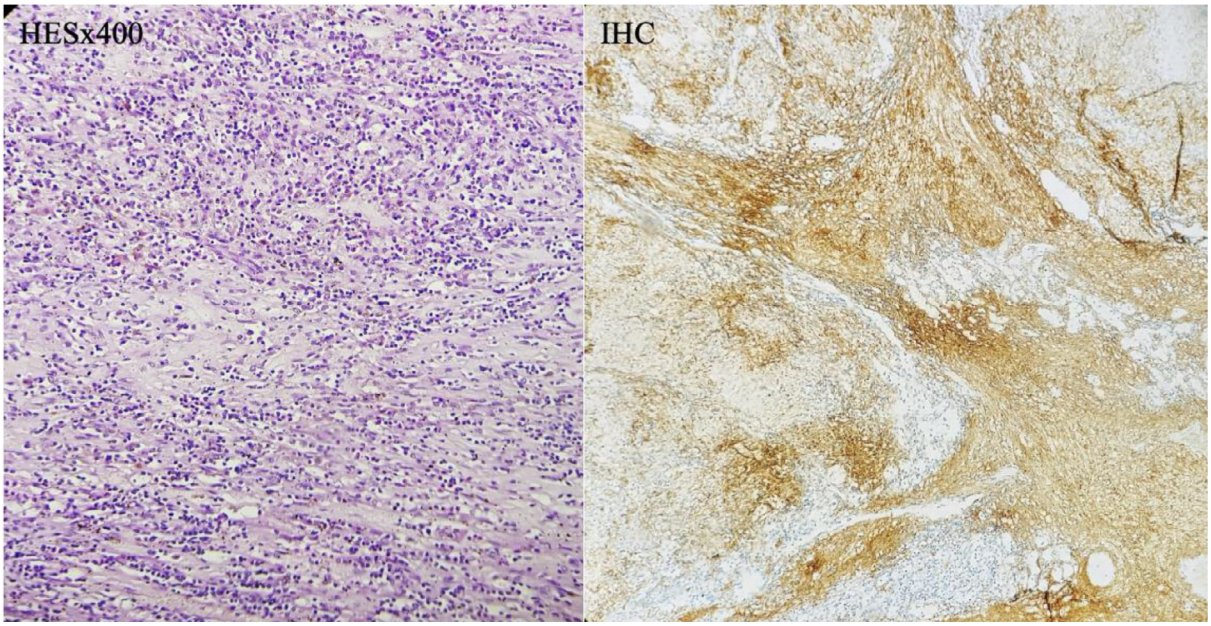

Fig. 2 – Proliferation of spindle-shaped cells arranged in hyaline material with chronic inflammatory cells. Immunohistochemical findings are compatible with an IMT.

tial diagnosis is made mainly with lymphoma and hamartoma [12]. The classic treatment for splenic localization is total splenectomy [12].

Conclusion

IMTs are rare lesions, simulating a malignant tumor process. Radiologic features are polymorphic and no conclusive and present a diagnostic challenge to the radiologist. It should be considered in the list of differential diagnoses of masses, although histology remains obligatory for the final diagnosis.

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