Case Report

Inflammatory Pseudotumor of the Spleen in a 9-year-old Child: A Case Report at Hospital de Especialidades Fuerzas Armadas N°1, Quito

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Abstract

Inflammatory pseudotumor of the spleen is defined as a benign tumor of reactive origin that exhibits series of nonspecific histopathological changes of an inflammatory or fibrotic reparative type. This diagnosis encompasses other pathologies such as dendritic cell tumors, certain aggressive neoplasms (inflammatory myofibroblastic tumor) and postoperative reparative lesions.

A 9-year-old male patient who initially presented with 15-day history of diffuse, colicky abdominal pain associated with early postprandial vomiting. In CT angiography, hepatic perfusion disorder in venous phase with intraluminal filling defect in portal vein is reported in relation to thrombosis and irregular mass in spleen with reactive lymph nodes in retroperitoneum.

Keywords: Pseudotumor, inflammatory, spleen

Introduction

Inflammatory pseudotumor is considered a benign tumor of reactive cause that exhibits series of nonspecific histopathological changes of an inflammatory or fibrotic reparative type. This diagnosis encompasses other pathologies such as dendritic cell tumors, certain aggressive neoplasms (inflammatory myofibroblastic tumor) and postoperative reparative lesions [1].

This tumor is most common in lung, gastrointestinal tract, lymph nodes, and liver. It was first described in lung in 1939 by Brunn [2]. Splenic involvement is very rare, it was first described by Cotelingam and Jaffe in 1984 [1].

The etiology and pathogenesis of this entity are unknown, considering infectious (Epstein-Barr virus), autoimmune and even vascular causes as a possible origin. Clinical diagnosis of this entity can be complex due to its nonspecific symptoms; therefore, it requires imaging studies and histopathological confirmation. Its management is mainly surgical.

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mass in spleen with reactive lymph nodes in retroperitoneum is reported. Surgery was performed and histopathological analysis diagnosed an inflammatory pseudotumor of the spleen.

**Case Report**

A 9-year-old male patient, B RH+. History of right orchidopexy 7 and a half years ago and appendectomy in July 2018. He was admitted on September 28, 2018 with retroperitoneum’s uncertain behaviour tumor diagnosis. His mother reported diffuse colicky abdominal pain associated with early postprandial vomiting in the last 15 days. The patient was referred with a simple and contrasted CT scan that reported a 10x6cm mass of splenic-vascular component that displaces structures and thrombosis of the portal vein. CT angiography is performed in our hospital (Figure 1). A venous phase hepatic perfusion disorder with an intraluminal filling defect in the portal vein is reported in association with thrombosis; in the spleen, an irregular 6.6×5.9cm mass is observed, showing irregular central and peripheral enhancement in all phases; the retroperitoneum shows reactive-appearing lymph nodes in all chains. On physical examination, a 165 bpm tachycardia stands out.

![Figure 1: CT angiography showing a mass in the spleen](image)

Laboratory tests are requested in which negative tumor markers stand out; D-dimer: 6,074,000; alkaline phosphatase: 232. Laparoscopic splenectomy was performed, which was converted to laparotomy due to intraoperative difficult control bleeding. Among the findings are (Figures 2 and 3): spleen 15 cm long with an intraparenchymal tumor of 10 cm diameter, rounded with necrotic areas.

**Histopathological report**

**Pathological diagnosis**

- Negative for malignancy
- Inflammatory pseudotumor
- Negative lymph nodes for metastases with reactive hyperplastic characteristics.
Complements: immunohistochemistry

- Vimentin: positive in stroma
- K167: positive in 5%
- Desmine: negative
- CKAE1 / AE3: negative
- CD34 and CD31: positive in endothelial cells
- Trichomic: positive for fibrosis

**Figure 2**: Splenectomy product (15 cm spleen with intraparenchymal tumor).

**Figure 3**: 10 cm diameter intraparenchymal tumor with necrotic areas

**Discussion**

As we mentioned, this diagnosis encompasses other pathologies such as dendritic cell tumors, certain aggressive neoplasms and postoperative reparative lesions. Despite being classified as a benign tumor, some evidence has shown the presence of chromosomal abnormalities and aggressive proliferation, which would support the hypothesis that it is
a true neoplasm [3]. In contrast to inflammatory myofibroblastic tumors, inflammatory pseudotumors usually present abundant histiocytes and follicular dendritic cells instead of myofibroblasts [4], in addition to necrotic areas. Scientific evidence shows a low incidence of cases reported in the literature with a homogeneous distribution in both sexes and with more common range of presentation that rotates between the second and third decade of life [5]. The presentation of this tumor in childhood and adolescence is less common, with few cases being reported.

Inflammatory myofibroblastic tumors include cases of pulmonary (most common) and extrapulmonary (primarily abdominal) involvement characterized by proliferation of spindle cells whose main cellular component has the immunophenotype of myofibroblasts [6]. It is primarily a visceral and soft tissue tumor of children and young adults, although the age range extends into adulthood. The mean age is 10 years, and the median is 9 years, being more frequent in the first two decades of life with a slight female predominance [7].

The origin of this pathology is not clearly defined, including etiologies related to infectious diseases, vascular disorders and autoimmune disorders. The most associated infectious agent is the Epstein-Barr virus (EBV) [8], which is associated with cases of reactive pseudotumors that include follicular tumors with abundant dendritic cells in liver and spleen. This association has been proven, for example, in a clinicopathological and immunophenotypic study of 12 cases by Neuhauser et al. in 2004, whose results, based on an immunohistochemical panel, confirmed cellular immunoreactivity for EBV type 1 latent membrane protein in 2 cases, as well as positivity for EBV-encoded RNA in 6 of 10 cases [9]. Other associated microorganisms are: Legionella, Klebsiella pneumoniae, Pseudomonas veronii, Histoplasma capsulatum, Actinomyces, Corynebacterium equi, Bacteroides corredus, cytomegalovirus and hepatitis C, as well as scarlet fever and urinary tract infection [1,3]. Regarding vascular etiology, their relationship is presumed due to the presence of dilated veins and thrombi in the lesion. The relationship with an autoimmune disorder is given by the description of cases that were associated with processes such as idiopathic thrombocytopenic purpura [10].

Symptoms of this pathology can be non-specific and in some cases be absent. The most common manifestations are anorexia, abdominal pain, fever, lymphadenopathy and splenomegaly (sometimes it is the only finding and may be incidental), weight loss and asthenia or symptoms related to a compressive effect on adjacent structures.

In the specific case of splenic involvement, laboratory tests may show abnormalities related to splenic sequestration such as anemia and thrombopenia. Most of the time it is an incidental finding on imaging studies. There is currently no specific imaging modality that can distinguish this tumor from other splenic neoplasms or provide a definitive preoperative diagnosis. Performing a review of the different imaging methods, splenomegaly could be seen on plain radiographs and, in exceptional cases, calcifications at this level. Abdominal ultrasound may show a well-defined hypoechoic mass or a hyperechoic lesion due to partial calcification. On color flow Doppler, this lesion is commonly hypovascular. In CT without contrast, lesion is observed as an isodense mass, with IV contrast it appears as a hypodense mass with slow heterogeneous enhancement and sometimes with calcifications. MRI may show it as a well-defined hypo- or isointense lesion on T1 and with decreased or increased signal intensity on T2, compared to contiguous normal splenic tissue. The PET/CT scan usually shows variable uptake, sometimes intense, which would increase the suspicion for a malignant lesion [1,11].

Therefore, definitive diagnosis will be histopathological. Microscopically, non-specific inflammatory changes are observed, composed mainly of histiocytes, plasma cells, mature lymphocytes and rarely eosinophils in a fibroblastic stroma surrounding a central necrosis zone. Atypia or mitotic figures are not usually seen. Macroscopically, well-defined nodular or sometimes multinodular white lesion is identified [12]. Immunohistochemical analysis can help us to differentiate the variety of entities encompassed by the inflammatory pseudotumor. Thus, the inflammatory myofibroblastic tumor is commonly immunoreactive against vimentin and smooth muscle actin and negative against ALK1 [4].
Despite splenectomy has been considered the best method for diagnosis through the pathological analysis of the extracted piece, becoming at the same time treatment [13], ultrasound guided core splenic biopsy can be considered a quick and safe method to performing splenic biopsy, avoiding surgery to a definitive diagnosis and being a valid option in those patients whose treatment of choice is conservative therapy, dispensing with an unnecessary splenectomy. Despite this, in most cases traditional splenectomy is preferred, both for diagnosis and treatment of a mass in the spleen due to the risk of bleeding and the fear of spillage of tumor cells, if the tumor is malignant [10].

In most cases, splenectomy is chosen as treatment, with no reports of recurrences or subsequent development of hematological neoplasms. However, if diagnosis is made without prior splenectomy, one option is medical management of neoplasm through chemotherapy, immunosuppressants and / or radiotherapy. If symptoms persist despite medical treatment, tumor grows or malignancy’s possibility remains, surgical removal will be chosen [14].

**Conclusion**

Inflammatory pseudotumor of the spleen is an extremely rare entity that encompasses a variety of pathologies ranging from benign tumors to processes with potential malignant degeneration, which highlights the importance of determining an adequate diagnosis through histopathological and immunohistochemical study. Our case is one of the few reported in childhood, with no apparent association with infectious causes (no history) and with a clear vascular component that led to secondary portal venous thrombosis.

**References**


