Inflammatory myofibroblastic tumor of the spleen in children

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Abstract

Inflammatory myofibroblastic tumor (IMT) is a rare benign neoplasm, which mainly involves lungs of the children. Extra-pulmonary locations are manifested mostly in viscera, but localization in the spleen is extremely rare with only few case reports in literature. Although it is commonly seen in children, the numbers of childhood cases are limited. We described a case of IMT of the spleen in a 16-year-old boy who visited our hospital with worsening fatigue symptoms and mild upper abdominal discomfort. The abdominal ultrasound was performed indicating a low echoic mass located in the spleen and measuring 18x10x10 cm. An abdominal CT scan was performed and demonstrated a large low-density hypovascular mass. Splenectomy was indicated and performed. Intra-operative and postoperative courses were uneventful.

Keywords: Children, Croatia, inflammatory myofibroblastic tumor, pediatrics, spleen.

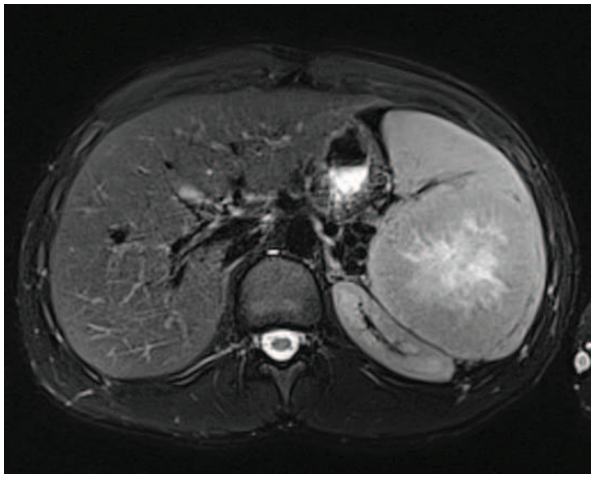
Introduction

Inflammatory myofibroblastic tumor (IMT) is a benign tumor of unknown etiology consisting of myofibroblastic spindle cells with an inflammatory cells infiltration. Their behavior is not aggressive, but sometimes can simulate malignant neoplasm, from which it impossible to distinguish before excision (1). A number of terms have been applied to this tumor including inflammatory pseudo-tumor, benign myofibroblastoma, plasma cell pseudo-tumor, hamartoma, fibrosarcoma, leiomyosarcoma and inflammatory myofibroblastic tumor (2). IMT was first observed in lungs and described by Bunn in 1939. It was named as IMT because it mimics malignant neoplasm clinically, radiologically and histopathologically. Recurrence is extremely rare if resection is complete and metastases occur in less than 5% (3).

Case report

A 14-year-old boy visited our hospital with worsening fatigue symptom, night sweats and mild upper abdominal discomfort. In a routine health evaluation ultrasound which was made incidentally, a splenic tumor was found. Abdominal ultrasound revealed an 18x10x10 cm solid splenic mass. These findings were confirmed by a computed tomography scan which showed a solid hypoechoic nodular, well-defined, round and with a smooth mass lesion (Figure 1).

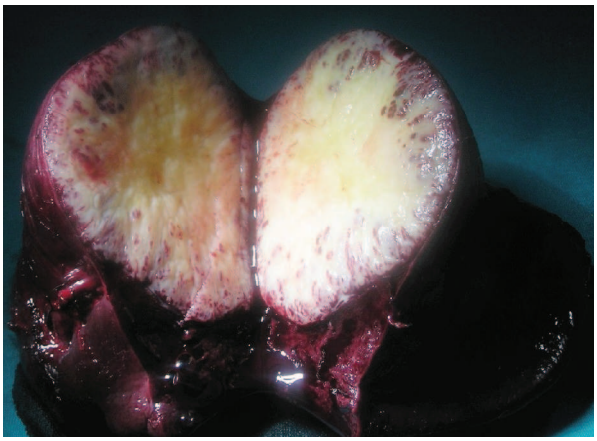
Figure 1. CT scan of the spleen with tumor



Images of other abdominal organs appeared normal. Physical evaluation showed no hepatomegaly, splenomegaly, or lymphadenopathy. The differential diagnosis included splenic hamartoma, hemangioma, lymphangioma or lymphoma. Laboratory findings were unremarkable with elevation of erythrocyte sedimentation rate (28 mm/ h), mild leukocytosis ($14.5x10^9$cells/ L) with mild neutrophilia and C-reactive protein (CRP) value was 15 mg/ dL.

Laparotomic splenectomy was indicated and performed. On abdominal findings, the mass was without adhesions to the surrounding organs or structures and spleen was removed without any accidents. Macroscopically, the serosal surface and the cut surface were both smooth. The cut surface revealed round, solid and well circumscribed mass. Upon cut, we could observe a whitish-yellowish sharply circumscribed tumor with a medium-to-firm consistency. The tumor did not permeate the capsule (Figure 2).

Figure 2. Macroscopic spleen with tumor



Histologically, the tumor was composed of sheaves of uniform, spindle-shaped cells with elongated nuclei and prominent nucleoli, and partially there were observed histiocytes, plasma cells, lymphocytes and eosinophils. Immunohistochemical analyses of single cells were positive for smooth muscle actin (SMA) and vimentin, with a part of lymphatic cells positive for CD20 and CD3, showing no restriction in expression in light chains (kappa, lambda). Endothelial cells were positive for CD 31. Tumor cells were negative for ALK, and Ki67 proliferation activity in the "hot spot" was less than 5%. Intra-operative and postoperative courses were uneventful and the patient is currently asymptomatic 4 years after surgery. He is now healthy and has no complaints.

Discussion

Although most commonly seen in the lungs, IMT are known to occur at several anatomic sites like orbit, spinal meninges, digestive system, heart, soft tissues, pancreas, liver, kidney, urinary bladder, mesothelial membranes and respiratory tract. Splenic involvement is extremely rare (4). A number of terms have been applied to the tumor, namely, inflammatory pseudotumor, benign myofibroblastoma, plasma cell pseudotumor, fibrous xanthoma, hamartoma, fibrosarcoma, leiomyosarcoma and inflammatory myofibroblastic tumor (2). Originally described in the lung, its occurrence is well-accepted to affect every organ. IMT can be found in adult spleen, whereas in children only rare cases have been reported. In a large review of splenic IMT, the average age was 53 years (range of 19 to 87 years) and there was no sex preference. IMT are most frequently detected incidentally. If symptomatic, most patients exhibit pain in the left upper quadrant, epigastric pain or very rare thrombocytopenic purpura (5). The cause of IMT of the spleen is unknown. Various hypotheses have been put forward in the pathogenesis: EBV, mycobacterium in immune-compromised patients, human immunodeficiency virus, auto immune and vascular causes. However, the exact cause remains uncertain (6,7).

Splenectomy is usually performed in patients with splenic tumors because imaging techniques cannot exclude malignancy. Due to the size of the tumor and the suspected malignancy, we performed an open splenectomy. Laparoscopic splenectomy may be a useful option for patients with smaller splenic tumors (8). Postoperatively, our patient had no clinical signs or symptoms, and the laboratory tests were normal. Two months later, an ultrasound examination was performed showing absence of any tumor in abdomen. No further treatment was considered necessary.

In conclusion, IMT of the spleen is extremely rare. This uncommon pathology is not associated with specific clinical findings and can be represented as a splenic neoplasm. Currently, no imaging techniques allow preoperative diagnosis. Splenectomy and histiopathologic studies of the specimen allow diagnosis and treatment.

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