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Teaching cases

Inflammatory myofibroblastic tumor: Clinical, morphological, immunohistochemical and molecular features of a pediatric case

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ABSTRACT

Inflammatory myofibroblastic tumor is an uncommon tumor regarded as "intermediate malignancy". We present the clinical, pathological and molecular features of a mesenteric inflammatory myofibroblastic tumor in a 9-month-old male infant. The patient was referred to Anna Meyer Children Hospital of Florence, Italy, for an asymptomatic abdominal mass measuring about 7 cm. The lesion was radically excised, and the postoperative course was uneventful. Histologically, the tumor was composed of spindle cells immunopositive for vimentin and desmin admixed with an inflammatory infiltrate. Rearrangement of *ALK* gene was demonstrated by FISH and immunohistochemistry (cytoplasmic, perinuclear and punctate immunocoloration). The peculiar punctate ALK immunocoloration suggested a possible unusual *ALK* gene rearrangement involving the CLTC gene.

Introduction

Inflammatory myofibroblastic tumor is a rare and poorly understood lesion. There is even disagreement about its inflammatory or neoplastic nature.

Currently, inflammatory myofibroblastic tumor is mostly considered a neoplasm of intermediate malignant potential as evidenced by molecular data and by a possible more aggressive clinical behavior burdened by local recurrence and even distant metastases [3,4,9].

The etiology of this tumor is also still unknown. Trauma, surgery, infections i.e. herpes and Epstein–Barr virus, radiotherapy, steroid use and autoimmune reactions have been suggested as etiological factors [3,4].

Inflammatory myofibroblastic tumor may occur in virtually every organ/system. However, the most common sites are lung, mesentery and omentum. Typically, it affects children and young adult although a broad age range may be affected. A slight female gender predilection has also been reported (male:female ratio, 3:4) [3,4].

Inflammatory myofibroblastic tumor consists of intersecting fascicles of spindle to fusiform and/or of epithelioid and stellate cells having prominent nucleoli. Tumoral cells are embedded in a variably collagenous or myxoid stroma with a conspicuous inflammatory infiltrate. Immunohistochemical analysis reveals constant reactivity for vimentin and variable reactivity for muscle-specific proteins [3,4].

In a significant percentage of cases, translocations involving the anaplastic lymphoma kinase (*ALK*) gene have been demonstrated [1–4,6,8,10–12].

Herein, we describe the clinical, morphological, immunohistochemical and molecular features of a mesenteric inflammatory myofibroblastic tumor in an infant.

Case presentation

A 9-month-old male infant was admitted to Meyer Hospital of Florence for an asymptomatic abdominal mass noted by his mother since two months. The baby was asymptomatic and was thriving well on normal feeds.

Clinical examination revealed a palpable abdominal mass occupying the right hypochondrium and the umbilical region of about 7 cm in maximum diameter. The lesion was mobile and had a firm elastic consistency without any skin alteration above.

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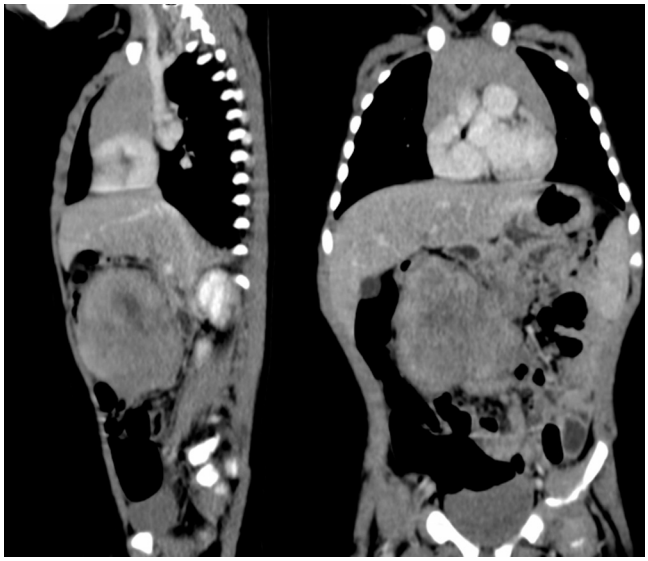

Fig. 1. Plain CT sagittal section (left) and coronal section (right).

Preoperative blood tests were within range. Tumor markers (βHCG, CEA, Ca19.9, αFP and NSE) resulted negative. Abdominal ultrasonography (US) and computed tomography (CT) of the abdomen and the thorax were performed. US and plain CT scan revealed the presence of a homogenous neoformation measuring 74 mm × 48 mm × 62 mm with distinct borders, in right median/paramedian sub hepatic space. After contrast enhancement, the mass was found to be having a heterogenous density with a hypoechoic central area with no obvious involvement of the surrounding abdominal organs; therefore, a primary surgical resection was planned (Figs. 1 and 2).

Under general anesthesia, a supraumbilical transverse incision was performed. After mobilization of the omentum, a bulging floating mass was found. The lesion had an irregular shape with a pedicle arising from the anti-mesenteric aspect of transverse colon near the splenic flexure (Fig. 3). The vascular pedicle was ligated and divided to excise the mass. A wedge of adherent transverse colon was resected and was later closed primarily. The postoperative course was uneventful.

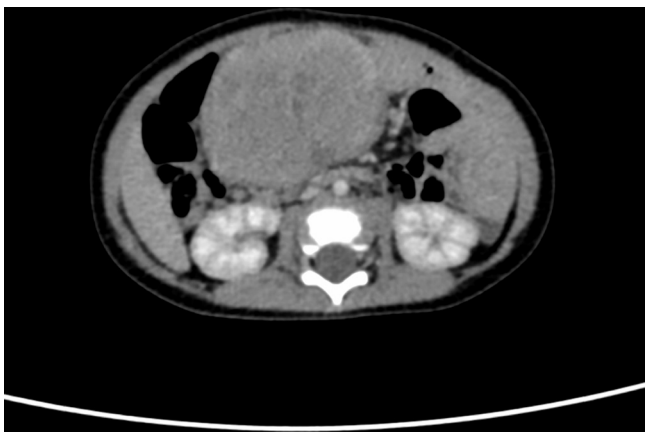

Fig. 2. CT scan after administration of contrast medium showed a disomogeneous enhancement of the abdominal mass.

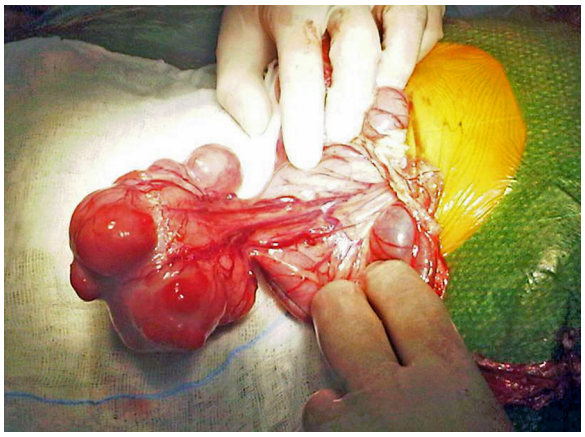

Fig. 3. After mobilization of the omentum, a bulging floating mass was found. The lesion had an irregular shape with a pedicle arising from the anti-mesenteric aspect of transverse colon near the splenic flexure.

Pathological findings

The surgical specimen was routinely fixed in neutral buffered formol, generously sampled and embedded in paraffin.

Five-micrometer sections of each sample were stained with hematoxylin and eosin. Additional sections of the most representative samples were mounted on electrostatic slides and used for the immunohistochemical and fluorescence in situ hybridization (FISH) studies. Immunohistochemical staining was accomplished with commercially available antibodies (Vimentin clone V9, Desmin clone DE-R-11, Actins clones HHF35 and 1A4, Myogenin clone F5D, CD34 clone QBEnd/10, S-100 clone 4C4.9 and anaplastic lymphoma Kinase (ALK)-1 protein clone ALK01, Ventana-Roche, Tucson, Arizona; KI67 clone MIB-1, DAKO, Carpinteria, CA) and the standard streptavidin–biotin technique. For FISH analysis, we used a dual-color break-apart FISH probe (Vysis Inc., des Plaines, IL, USA).

Macroscopically, the mass was solid, multi-lobulated and encapsulated. The cut surface was gray-white and firm in consistency.

Microscopically, the tumor was adherent to the serosal side of the intestinal wall that was not infiltrated (Fig. 4). It was composed of spindle cells admixed with plama cells, lymphocytes, neutrophils and eosinophils. A stroma of abundant hyalinized collagen was interposed between the cells. The spindle cells had a moderate

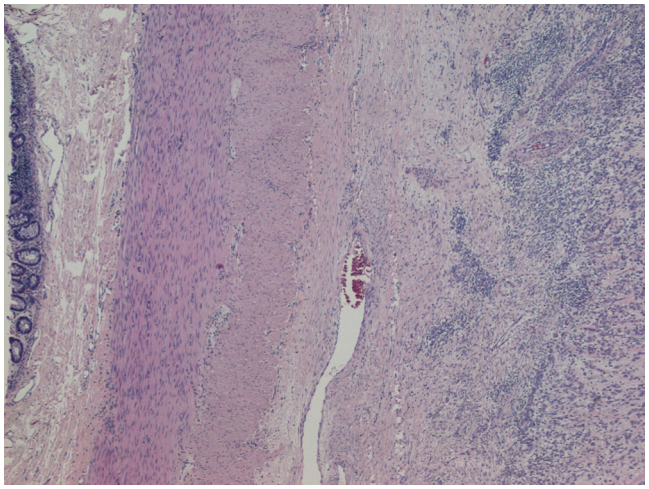

Fig. 4. The tumor was adherent to the serosal side of the intestinal wall that was not infiltrate.