



Inflammatory myofibroblastic tumor of the greater omentum in a 7-year-old girl

Juan P. Pérez Rossini¹ , Eugenia Perié¹, Cyntia Soriano¹, Santiago Caparrós¹, Nicolás López¹, Claudio Yamahachi¹, Luis A. D'Alú¹

ABSTRACT

Inflammatory myofibroblastic tumor is a rare neoplasm. Its preferred location is intrathoracic; there are few reports of intra-abdominal involvement, where it may affect the mesentery and retroperitoneum.

A 7-year-old girl was brought in for evaluation due to fever, nonspecific lower abdominal pain, fatigue, and progressive weight loss. Laboratory tests revealed anemia, thrombocytosis, and elevated CRP. An abdominal ultrasound showed a hypoechoic mass with well-defined borders and heterogeneous content, measuring approximately 67 × 52 × 79 mm. Complete excision was performed via laparotomy.

Its low incidence, varied location, nonspecific histology, and clinical heterogeneity make diagnosis extremely difficult. Only histopathology allows for a definitive diagnosis, and the cornerstone of treatment remains complete removal of the tumor. This rare condition should be included in the pediatric differential diagnosis because of its nonspecific clinical presentation, intra-abdominal mass, and high recurrence rate after incomplete resection.

Keywords: abdominal neoplasms; myofibroblasts, pathology; omentum.

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¹ Pediatric Surgery Department, Hospital Materno Infantil Don Victorio Tetamanti, Mar del Plata, Argentina.

Correspondence to Juan P. Pérez Rossini: jpprossini@gmail.com

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INTRODUCTION

Inflammatory myofibroblastic tumor (IMT) is a rare, intermediate-grade neoplasm characterized by the proliferation of myofibroblasts and spindle-shaped fibroblasts. Although it most commonly occurs in the chest (where it appears to have a particular predilection for lung tissue), sporadic reports describe its occurrence in the abdomen, urinary tract, and musculoskeletal system, among other sites.¹⁻⁷

This neoplasm has a low potential for distant metastasis and a high recurrence rate after incomplete resection. It primarily affects children, adolescents, and young adults,¹ predominantly females, with a mean age of 9.5 years.^{2,8-10} Its clinical presentation is nonspecific, with symptoms related to the inflammatory response it causes or to mass effect.^{1,2,8,9}

An IMT located in the greater omentum is difficult to distinguish from other abdominal tumors based solely on imaging studies; histopathology is necessary to establish a definitive diagnosis.^{1,11}

We present this case because we believe this condition should be included in the differential diagnosis of any pediatric intra-abdominal mass.

CLINICAL CASE

A 7-year-old girl with no significant medical history presented with lower abdominal pain,

fatigue, and progressive weight loss. Initially, these symptoms were accompanied by nausea and vomiting, which later resolved spontaneously. Laboratory results showed decreased hematocrit and hemoglobin, thrombocytosis, and elevated C-reactive protein (CRP). An abdominal ultrasound revealed a pelvic mass measuring 67 × 52 × 79 mm, with well-defined margins and no evidence of infiltration into adjacent structures. The mass had heterogeneous content, with scattered images showing posterior acoustic shadowing. Isolated internal Doppler signal foci were noted. The internal genitalia showed no pathological findings (*Figure 1*).

On physical examination, the patient appeared pale, febrile, tachycardic, and tachypneic. The abdomen was soft, depressible, and tender in the hypogastrium and left iliac fossa, where a mobile mass with well-defined borders was palpable. We administered a red blood cell transfusion and adopted a watch-and-wait approach. During her hospitalization, she remained hemodynamically stable, persistently febrile, with preserved bowel movements and urine output. Serum tumor markers (alpha-fetoprotein [AFP] and B-HCG) showed no pathological findings. Given the tumor's easily accessible anatomical location, combined with the lack of evidence of invasion into adjacent structures and the need to obtain

FIGURE 1. Transabdominal ultrasound



A hypoechoic, heterogeneous image with well-defined borders is seen (red arrow).

tissue for a histological diagnosis, it was decided to perform an exploratory laparoscopy.

A large, mobile, hypervascularized mass that was difficult to manipulate but without adhesions was observed. The procedure was converted to a laparotomy via a Pfannenstiel incision. The mass, located in the greater omentum (*Figure 2*), was brought out through the incision, and complete excision was performed along with a partial omentectomy due to the presence of multiple lymph nodes in the adenomegaly range within the omentum.

The rest of the abdominal cavity was examined, with no significant findings.

Pathological examination revealed an inflammatory myofibroblastic tumor with loose connective tissue, abundant myofibroblasts, and inflammatory cells (lymphocytes, plasma cells, and leukocytes) with a low mitotic count. In some areas, the stroma was edematous and myxoid. It exhibited a rich, congested vascular network. Complete resection was reported. No

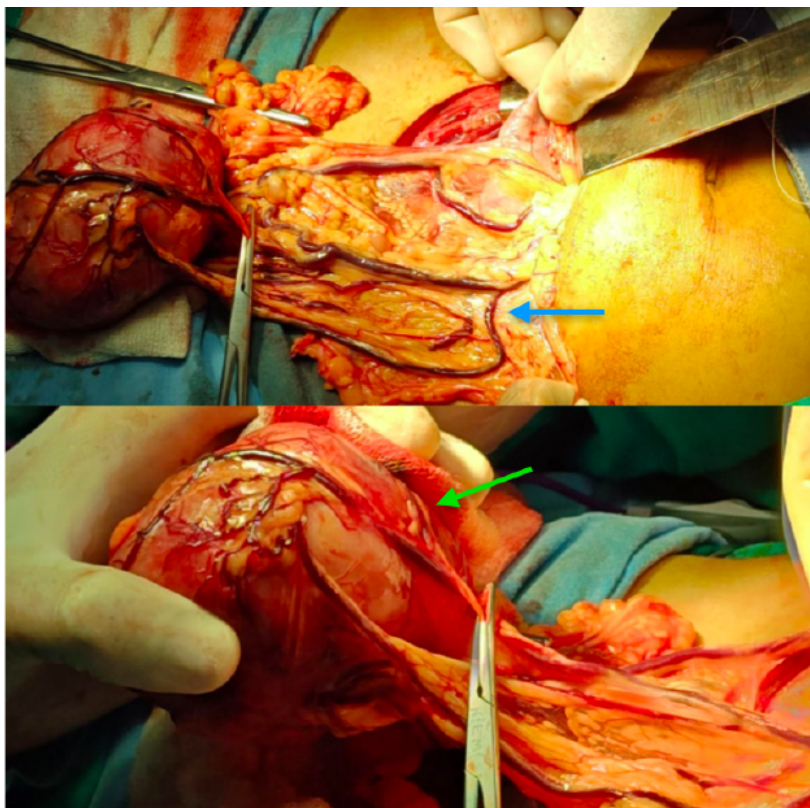
immunohistochemical techniques were performed. The patient had a favorable outcome and was discharged on the fourth postoperative day. She presented no complications. The patient is currently being monitored jointly by the Pediatrics, Pediatric Surgery, and Oncology departments.

DISCUSSION

The former names for IMT (such as plasma cell granuloma or inflammatory pseudotumor, among others) are no longer in use.¹ Today, it is defined as a true neoplasm with the potential for local invasion and a high risk of recurrence following incomplete resection.^{1,3,8,9,12}

Histologically, they show a proliferation of spindle cells in a stroma with variable characteristics and fibrosis, with an inflammatory cell infiltrate.^{1,2,8,13} Its mitotic rate is usually low.^{1,2,14} Histologically, IMT is classified into three patterns: myxoid (with scattered proliferation of myofibroblasts, myxoid edematous tissue, and abundant blood vessels and lymphocytic/

FIGURE 2. Intraoperative images



Note the dilation and engorgement of the omental vessels (blue arrow), as well as the macroscopic appearance and the tumor's close relationship with the greater omentum (green arrow).

eosinophilic infiltrates, as observed in our patient's pathology), hypercellular (more compact cellular proliferation), and hypercellular.¹

An IMT located in the greater omentum presents as a smooth-surfaced, grayish or yellowish, well-circumscribed, solid, multilobed, and highly vascularized mass, with an average diameter of 7 cm (1-20 cm).^{3,6,9,11}

Key differential diagnoses include leiomyosarcoma, liposarcoma, inflammatory pseudotumor, Epstein-Barr virus-positive inflammatory follicular dendritic sarcoma, gastrointestinal stromal tumors (GIST), and others such as lymphoma, mesothelioma, and peritoneal carcinomatosis/metastases.¹⁻³

Patients generally present with nonspecific symptoms such as fever, weight loss, malaise, diffuse abdominal pain, and vomiting, along with a palpable abdominal mass and symptoms related to mass effect.^{3,9,12} There are no specific laboratory parameters, although most cases show elevated CRP, erythrocyte sedimentation rate (ESR), and leukocytosis. They may present with anemia, thrombocytosis, and hypergammaglobulinemia.^{4,9} There are also no pathognomonic radiological findings. Ultrasound reveals a rounded, well-defined, heterogeneous, solid mass that is hypo- or hyperechoic. Hypoechoic regions related to necrosis may be seen centrally. On computed tomography (CT) or contrast-enhanced magnetic resonance imaging (MRI), it may appear as a "target-like" lesion; on CT, it appears isodense or hypodense compared to skeletal muscle, and on MRI, it appears hypointense on T1- and T2-weighted images.^{1,9,15} On both CT and MRI, omental IMT appears as a well-defined, homogeneous or heterogeneous, multilobulated mass, with or without calcifications, and has been described as presenting with both hypovascularization and hypervascularization.^{5,8-10,14} Contrast uptake varies, with early peripheral enhancement due to vascular tissue or central necrosis and late central enhancement due to its fibrotic components.^{1,8,14,15}

Given these characteristics, the diagnosis will always be histopathological. In the absence of official guidelines, the biopsy method is left to the surgeon's or treating team's discretion.¹¹ The recommendation remains complete excision whenever possible without damaging adjacent structures,¹⁰ as this represents the cornerstone of treatment for this tumor.⁹ Signs and symptoms, along with laboratory parameters, return to normal following resection.¹³

For staging, the tumor-node-metastasis (TNM) system can be used based on CT or MRI findings, with T1 or T2 depending on local invasion and TA or TB if the tumor diameter is less than or greater than 5 cm, respectively.^{2,9} Classification according to the IRS (Intergroup Rhabdomyosarcoma Study) has also been proposed, based on residual tumor following initial surgery.¹⁰

Factors such as age, histology, stage, and location determine the prognosis and the feasibility of a complete resection. For metastatic, unresectable, or recurrent tumors, the first-line treatment is ALK inhibitors (monoclonal antibodies, primarily crizotinib) as neoadjuvant therapy.^{1,2} In the absence of specific guidelines, chemotherapy follows the regimens used for small-cell sarcomas with anthracyclines. At the same time, radiation therapy is limited to patients with positive margins or contraindications to surgery.²

Pathological anatomy plays a fundamental role in both the diagnosis and oncological treatment of these tumors. Overexpression of the *ALK-1* gene (atypical lymphocyte kinase 1) correlates with less aggressive tumor behavior and a low recurrence rate following complete resection.^{4,6,9,14} In contrast, an ALK-negative tumor tends to be more locally invasive and carries a higher risk of metastasis. Recent advances have made it possible to identify various gene rearrangements; therefore, for ALK-negative tumors, fluorescent in situ hybridization (FISH) testing or RNA fusion analysis is recommended to identify fusions involving *ROS1*, *NTRK3*, or other less common genes such as *RET* or *PDGFRB*.¹ This would open the door to identifying new targets for monoclonal antibodies, as well as to characterize each tumor and its behavior better.^{1,2}

Overall, the prognosis is favorable, with recurrence rates of up to 6%,^{3,4} and a reported mortality rate of up to 7% in cases with multiple recurrences.⁶

In conclusion, IMT of the greater omentum poses a diagnostic challenge due to its nonspecific clinical and radiological presentation. Its neoplastic nature and invasive potential require a thorough differential diagnosis, as well as complete surgical excision and accurate histological characterization, to properly guide any subsequent cancer treatment. ■

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