Inflammatory myofibroblastic tumor of the jejunum with extension to the mesentery associated with biliary ileus: Case report

Tumor miofibroblástico inflamatorio de yeyuno con extensión al mesenterio asociado al ileus biliar: Reporte de caso

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ABSTRACT
Inflammatory myofibroblastic tumor (IMT) is a very rare neoplasm that represents less than 1% of all soft tissue tumors. According to the National Cancer Institute of the United States, IMT occurs in fewer than 1 case per million people. Diagnosis is established by histopathological study. We present a case diagnosed with jejunal IMT associated with gallstone ileus in a sexagenarian patient who underwent complete surgical resection.
***Keywords**: Inflammatory myofibroblastic tumor; Plasma Cell Granuloma; Biliary ileus; jejunum; mesentery.*

RESUMEN
El tumor miofibroblástico inflamatorio (TMI) es una neoplasia muy infrecuente que representa menos del 1% de todos los tumores de tejidos blandos. Según el Instituto Nacional de Cáncer de los Estados Unidos el TMI se presenta menos de 1 caso por cada millón de personas. El diagnóstico se establece por estudio histopatológico. Presentamos un caso diagnosticado de TMI de yeyuno asociado al ileus biliar en una paciente sexagenaria en la que se realizó resección quirúrgica completa.
***Palabras claves:** Tumor miofibroblástico inflamatorio; Granuloma de células plasmáticas; ileus biliar; yeyuno; mesenterio.*

INTRODUCTION

The inflammatory myofibroblastic tumor (IMT) is a histologically distinctive spindle cell myofibroblastic neoplasm of borderline malignancy, which classically presents with a mixture of plasma cells and lymphocytes [1], with intermediate malignant potential and a low risk of metastasis [2]. It has had various denominations such as inflammatory pseudotumor, plasma cell granuloma, histiocytoma, xanthoma, fibroxanthoma, xanthogranuloma, fibrous xanthoma, xanthomatous pseudotumor, plasmacytoma, solitary mast cell granuloma, plasma cell pseudotumor, inflammatory myofibrohistiocytic proliferation, mesenteric omental myxoid hamartoma, inflammatory myofibroblastic sarcoma, or inflammatory fibrosarcoma, which reflects the complexity, significant histological heterogeneity, and behavior of this entity [2,3]. The 2020 World Health Organization (WHO) classification of soft tissue tumors (fifth edition) no longer recommends the use of these alternative terms [2]. The term "inflammatory myofibroblastic tumor" was coined by Pettinato in 1990 [4].

IMT is a rare mesenchymal neoplasm, representing less than 1% of all soft tissue tumors. Its exact prevalence is difficult to determine due to its rarity and the historical misclassification as a reactive or inflammatory process [2].

According to the United States National Cancer Institute, IMT is very rare and occurs in fewer than 1 case per million people. It is estimated that this tumor is diagnosed in 150 to 200 individuals each year in the U.S [5].

Inflammatory myofibroblastic tumor (IMT) commonly affects the lung [3,6], but it has been reported in almost every part of the body. The abdomen is the second most frequent site of presentation, whether in the abdominal cavity (particularly the mesentery and omentum), the gastrointestinal tract, the retroperitoneum, or the pelvis [2,3]. Gastrointestinal IMTs predominantly affect the small intestine and colon, followed by the stomach. Less frequently, they involve the esophagus, appendix, pancreas, and liver [2].

The primary treatment for IMT remains complete surgical resection, particularly in cases of localized disease. Achieving negative surgical margins is ideal, as this significantly reduces the risk of recurrence [2]. In general, IMT follows a benign course, with surgery being curative and associated with an excellent prognosis [7], although recurrences may occur and, in some cases, it can behave as a malignant entity [3].

The objective of this study is to report the clinical case of a 61 year old female patient with a finding of inflammatory myofibroblastic tumor of the jejunum with extension to the mesentery, associated with biliary ileus.

CLINICAL CASE

Female patient, 61 years old, known hypertensive on regular treatment with enalapril 10 mg/day, with psoriasis treated

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with topical corticosteroids, allergic to procaine, underwent emergency appendectomy at age 15, and biliary ileus without cholecystectomy 1 year and 6 months prior to admission. She was admitted through the emergency department of the Hospital de Clínicas – San Lorenzo, with a 24 hour history of colicky abdominal pain, intense especially in the periumbilical region, accompanied by abdominal distension, cessation of stool and gas passage. She denied nausea and vomiting. As antecedent, 8 days earlier she had consulted for diffuse abdominal pain and was hospitalized with the diagnosis of incomplete intestinal obstruction, which resolved with medical treatment, and she was discharged after two days.

The patient was admitted lucid, afebrile, normotensive, tachypneic, and on physical examination presented with a globular, asymmetric abdomen due to a mass of approximately 6 cm in diameter located over the midline surgical scar, supra , para , and infraumbilical. On palpation, the abdomen was soft, depressible, painful in the periumbilical region, with muscular guarding, without signs of peritoneal irritation, and the mass over the midline scar was tense, painful, and irreducible. Tympany and increased bowel sounds were noted. Rectal and vaginal examination revealed no abnormalities.

The patient was hydrated with normal saline via peripheral access, and laboratory samples revealed leukocytosis with neutrophilia and mild anemia (white blood cells 13,000/mm^3, neutrophils 91.2%, hemoglobin 9.55 g/dl, hematocrit 29.2%) and albumin of 2.2 g/dl, with the rest of the studies within normal range. A surgical approach was decided upon, with the preoperative diagnosis of complicated incisional hernia M3W2R0.

During the surgical procedure, a supra , para , and infraumbilical midline incision was performed, followed by dissection of the hernia sac. Upon opening the sac, a small bowel loop with a violaceous coloration was found, containing a mobile stony mass of 4 cm *(Figure 1)*. A xipho pubic extension was then carried out, revealing a hernia ring of 4 cm in diameter and a plastic process of small bowel loops adherent at 90 cm from the Treitz angle. Resection of the jejunal loop was performed from 80 cm to 160 cm from the Treitz angle, including the mobile stony content within its lumen and the in-block mesenteric mass, after ligation and section of mesenteric vessels. A termino terminal anastomosis was carried out with manual two layer suturing using Vicryl 3 0. A tubular drain was placed adjacent to the anastomosis, exteriorized through a counter incision in the left flank, and primary closure of the abdominal wall was performed. Within the surgical specimen, a biliary stone measuring 3.4 cm was identified *(Figure 2)*. The specimen was sent to the Pathology Laboratory for study.

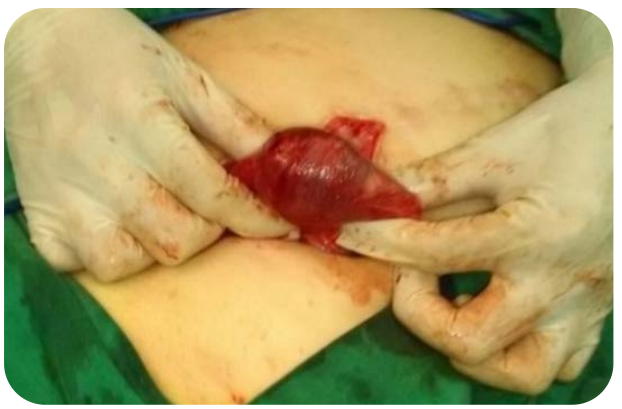

Figure 1. Small bowel loop with mobile stony content.

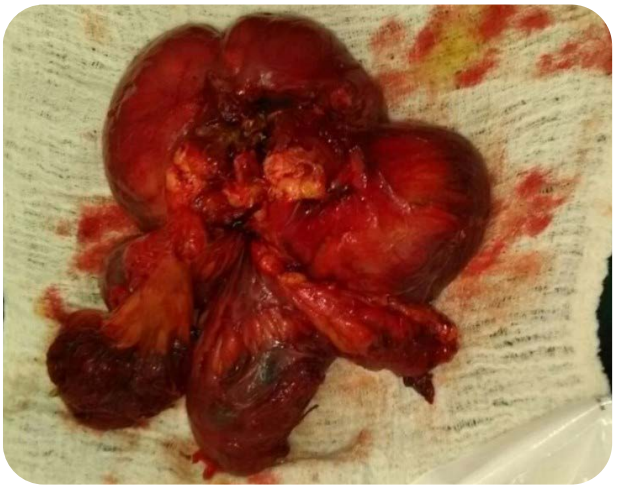

Figure 2. Surgical specimen with biliary lithiasis.

The patient showed good postoperative progress and was discharged on the 6th postoperative day after drain removal, with good tolerance to oral intake, no spontaneous pain, preserved physiological habits, laboratory values within normal range, and with instructions for outpatient follow up.

The Pathology report described, macroscopically, a mass measuring 8 × 6 × 5.5 cm located at the mesentery, 25 cm from the nearest surgical margin. On sectioning, it revealed a heterogeneous beige yellowish surface with some hemorrhagic areas, appearing to form adhesions with certain intestinal loops.

Microscopically: the jejunal mucosa was largely preserved, with foci of ulceration covered by fibrin and granulation tissue. A spindle cell proliferation infiltrating the muscular and submucosal layers of the intestinal wall was observed *(Figure 3)*. At the mesenteric level, a spindle cell proliferation composed of activated fibroblasts separated by loose edematous stroma with plasma cells, lymphocytes, and neutrophils was noted *(Figure 4)*. Numerous small caliber blood vessels and microabscess formation were present.

The histopathological appearance was compatible with an inflammatory myofibroblastic tumor measuring 8 cm.

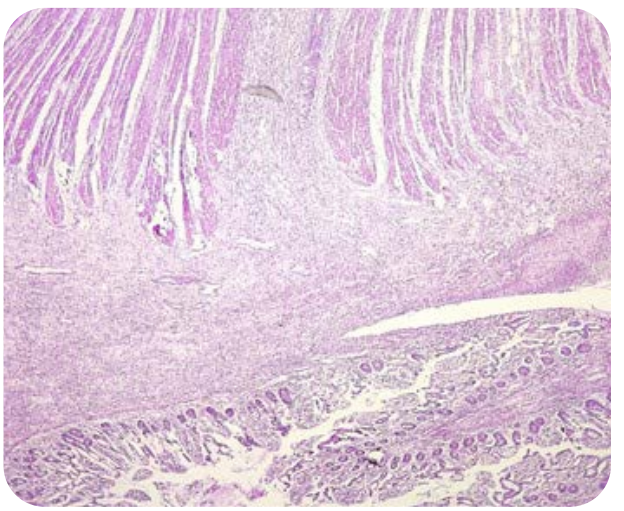

Figure 3. Histological section stained with Hematoxylin and Eosin at 40X magnification showing small intestine with a spindle cell proliferation infiltrating the muscular layer and the submucosal layer of the intestinal wall.

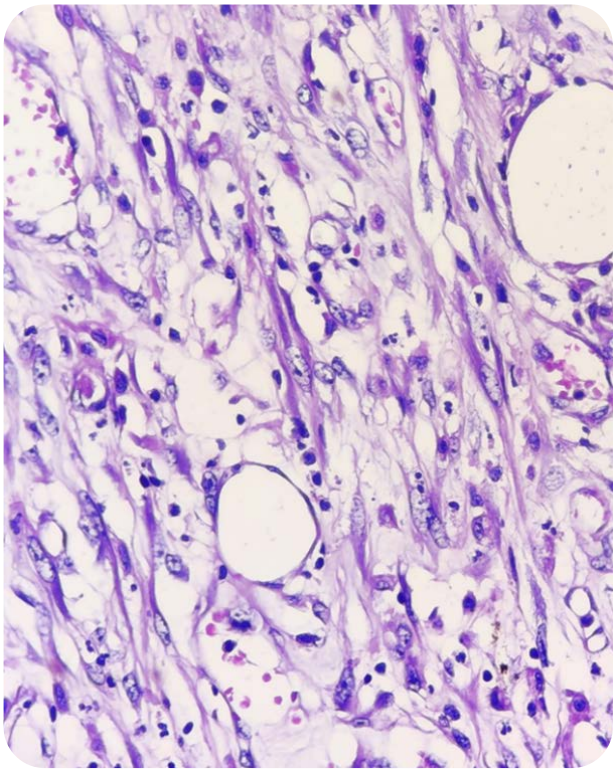

Figure 4. Histological section stained with Hematoxylin and Eosin at 400X magnification showing spindle cell proliferation composed of activated fibroblasts, separated by loose edematous stroma with plasma cells, lymphocytes, and neutrophils. Numerous small caliber blood vessels are observed.

DISCUSSION

IMT has been described across a wide age range; however, it is more common in young adults and in children between 2 and 16 years of age [3]. There is a slight predominance in females [2]. In our case, the neoplasm occurred in a sexagenarian patient.

Generally, it is a solitary neoplasm as in our patient, being multiple in only 5% of cases [3]. The clinical presentation of IMTs varies according to the site of origin. Abdominal IMTs may present with vague abdominal pain, gastrointestinal obstruction, or bleeding [1]. In 15 to 40% of cases, they may be asymptomatic [2].

Approximately 20% to 30% of patients with IMT present with a systemic inflammatory syndrome characterized by fever, general malaise, weight loss, and abnormal laboratory findings. These abnormalities include anemia, thrombocytosis, leukocytosis, polyclonal hypergammaglobulinemia, and elevated erythrocyte sedimentation rate and C reactive protein levels. This syndrome usually resolves after tumor resection [2,3]. Our patient presented with abdominal pain, leukocytosis, and anemia.

The clinical picture of biliary ileus may present acutely (which is the most common), sub-acutely, or even chronically with recurrent episodes of pain caused by the passage of the stone through the intestine without producing a complete obstructive event, also known as Karewsky's syndrome [3,4]. Reisner and others described that in biliary ileus, stone impaction occurs most commonly in the ileum (60.5%), jejunum (16%), stomach (14.2%), colon (4.1%), and duodenum (3.5%) [8].

Macroscopically, IMT typically presents as a well defined multinodular mass, ranging in color from white to gray, yellow, or tan, and may exhibit a whorled, fleshy, or myxoid texture. Tumor size varies considerably, from 1 cm to more than 20 cm, with a median of 5–6 cm [2]. In our case, it measured 8 cm, with a heterogeneous beige yellowish coloration.

Microscopically, three histological patterns of IMT are described: myxoid, hypercellular, and fibrous hypocellular. The first is characterized by plump or spindle shaped myofibroblasts loosely arranged in a myxoid background, with abundant blood vessels and a prominent inflammatory infiltrate of plasma cells, lymphocytes, and eosinophils, resembling granulation tissue or a reactive process; the second includes a dense and compact proliferation of fascicular spindle cells in a variably myxoid and collagenous stroma, accompanied by an inflammatory infiltrate; and the third is characterized by a hyalinized stroma, rich in collagen, with a reduced density of spindle cells and a relatively scant inflammatory infiltrate, similar to scar tissue or desmoid fibromatosis [2,3]. The lesion in our patient shows a hypercellular appearance. Multiple histological patterns may coexist within the same tumor [2]. If there are doubts in the diagnosis, immunohistochemical studies may be used.

The etiology of IMT is unknown, though it is believed that there may be predisposing factors such as surgeries, trauma, immunologic reactions, steroids, radiotherapy, and infections. Infectious agents are thought to play a role in the early stages, triggering a series of reactions in the cells and the inflammatory stroma, through which the tumor eventually becomes autonomous. Associated organisms include Mycobacterium; Epstein Barr virus; Escherichia coli, Klebsiella, Pseudomonas, Actinomycetes, and Mycoplasma [3]. Our patient had a history of previous surgeries, and since the case was associated with biliary ileus with recurrent abdominal pain, the remaining question is whether the chronic presence of biliary lithiasis within the intestinal lumen could act as a predisposing factor for IMT, as it was an incidental intraoperative finding and the lithiasis was located in the jejunal lumen near the tumor.

According to the reviewed literature, IMT is not usually considered before or during surgery [3], as occurred in our case. The diagnosis is established through anatomopathological study [3].

CONCLUSION

IMT is an uncommon neoplasm of intermediate behavior and has been reported in almost all parts of the body. Its clinical manifestations vary depending on the affected anatomical site. In general, it follows a benign course, with surgery being curative, although in some cases it may recur. Diagnosis is made through histopathological study, with immunohistochemical studies required in certain situations. In this work, we presented a case of jejunal IMT with mesenteric extension associated with biliary ileus, for which complete surgical resection was performed.

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