

# Inflammatory Myofibroblastic Tumor of the Small Bowel: A Case Report

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## ABSTRACT

Inflammatory Myofibroblastic Tumor (IMT) is a rare neoplastic lesion with tendency toward local aggressive behavior and recurrence. The tumor most commonly occurs in the pulmonary system of children and young adult, although it may rarely develop in older patients and other organs. Symptoms are non-specific and depend on the location of the tumor. The gastrointestinal tract is rarely the primary site of origin for this lesion. We report an unusual presentation of this rare lesion in a 58 year old woman with intussusception and partial intestinal obstruction.

## KEYWORDS

Inflammatory myofibroblastic tumor; Ileum; Gastrointestinal

## INTRODUCTION

Inflammatory Myofibroblastic Tumor (IMT), previously called inflammatory pseudotumor and plasma cell granuloma, belongs to a class of rare spindle cell lesions showing a rather unpredictable biological behavior with occasional tendency toward invasion to the surrounding tissue and local recurrence.<sup>1-3</sup> The lesion, as primarily described by Bahadori and Liebow in 1973, is a reactive/inflammatory process in the pulmonary system mostly occurring in children and young adults.<sup>4</sup> Since then, many cases have been reported in older patients in addition to extra-pulmonary sites.<sup>5, 6</sup>

## CASE REPORT

A 58 year old female was admitted to the hospital with a long standing history of dyspepsia and abdominal pain. The pain character changed in the last month prior to admission. She described the pain as cramping in the lower abdomen starting 1-1.5 hours after food accompanied by bloating and nausea but no vomiting and without radiation to other locations. It usually subsided after 4-5 hours with no treatment. She also complained of constipation. She was reluctant to eat because of the pain and as a result lost 3 kg weight in one month. From two days prior to admission, she started to feel an intensified and generalized abdominal pain associated with belching, nausea and vomiting. She also gave a history of decreased gas passing during this period.

On physical examination at admission, she was not in acute

distress. The abdomen was slightly distended with mild generalized tenderness on palpation. Bowel sounds were present. The rectal examination was normal. There were no other positive physical findings noted. Laboratory results were normal and included Hb: 13 gr/dl; WBC: 9000/mm<sup>3</sup>; platelets: 220000/mm<sup>3</sup>; ESR: 25 mm/h; FBS: 99 mg/dl; BUN: 26 mg/dl; Cr: 1.1 mg/dl; Na: 138 meq/l; and K: 3.8 meq/l.

CT scan of the abdomen and pelvis revealed an intraluminal space occupying lesion in the terminal ileum. There was no lymphadenopathy. The lesion was also demonstrated in the small bowel barium study and noted to have a maximum diameter of 5.5 cm (Figure 1).



**Fig 1:** Small bowel series shows intraluminal mass with pressure effect.

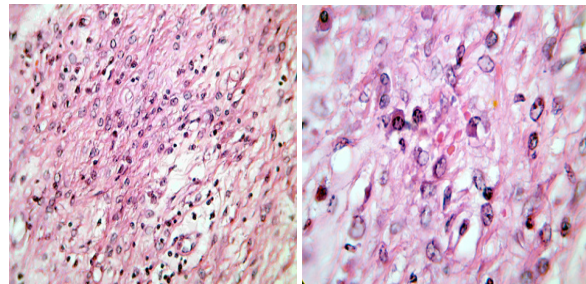
Colonoscopy did not show any gross pathology in the colon. The patient underwent laparoscopic exploration that showed a pedunculated mass in the terminal ileum accompanied by intussusception (Figure 2).



**Fig 2:** Pedunculated tumoral mass removed along with a segment of the ileum.

Thirty cm from the ileo cecal valve including the mass, was surgically removed. The ileocecal valve was preserved. In gross examination the mass was found to be creamy-white in color with a rubbery consistency, measuring 5.5 x 4.5 x 4.0 cm with smooth surface and patchy erythema of the covering mucosa.

Cut sections showed homogenous tan cut surfaces. Histological examination of the submucosal mass showed a rather cellular lesion composed principally of bland-looking spindle cells with a collagenous and myxoid background accompanied by various diffusely scattered inflammatory cells, including plasma cells, lymphocytes and eosinophils (Figure 3).



**Fig 3:** Histological findings of the tumor consist of proliferation of spindle-shaped myofibroblastic cells and scattered inflammatory cells, including numerous plasma cells (H&E: 100x left panel and 400x right panel).

No mitosis, necrosis or hemorrhage was present. Immunohistochemical studies showed positive reactions of the spindle cells for smooth muscle actin, human muscle actin, vimentin and calponin and was negative for ALK-1, desmin, CD34, CD117 (C-kit) and P63. Ki-67 was positive in 5% of the tumor cell nuclei. The final pathologic diagnosis was IMT.

## DISCUSSION

IMT is a rare, but distinctive spindle cell tumor that contains a variable number of inflammatory cells, including plasma cells. This is the reason for previous designation of mass as plasma cell granuloma.<sup>4</sup>

Plasma cell granuloma as well as other alternative terms are discouraged to be used including inflammatory pseudotumor, inflammatory myofibroblastoma and inflammatory myofibrohistioblastic proliferation.<sup>1, 2, 4, 5, 7</sup>

Multiple studies have shown expression of p80 and the clonal rearrangement of the anaplastic lymphoma kinase (ALK) gene on chromosome 2p23 leading to the over expression of the oncoprotein in the spindle cell components of some of these tumors.<sup>8-13</sup>

Abnormalities in chromosome 2p is seen in up to 60% of patients younger than 10 years of age.<sup>11,13</sup> This finding indicates a true neoplastic nature for the tumor cells.<sup>10,14</sup> DNA aneuploidy and association of the lesion with oncogenic viruses such as Epstein-Barr virus, Human Herpes virus type 8 and over expression of IL-6 have also been demonstrated and proposed to be involved in the pathogenesis of the tumor by some investigators.<sup>14,15</sup>

The lesion usually occurs in children and young adults but can develop in older ages with no predilection for any sex.<sup>2, 12, 16, 17</sup> The lung is the most common site of involvement but the lesion has also been reported in other organs including the stomach, mesentery, omentum, retroperitoneum as well as the kidneys, renal pelvis, liver, spleen, esophagus and lymph nodes.<sup>5, 6, 7, 15, 18</sup>

In a case series of 38 tumors, stomach was found to be the most common extra pulmonary site observed in 34% of the cases.<sup>2, 19</sup> Intra-abdominal lesions usually present with non-specific signs and symptoms including abdominal pain, gastric and intestinal mass with occasional obstruction and growth retardation in children.<sup>1, 2, 17</sup> Constitutional symptoms may occur and include fever, night sweats, weight loss and malaise. Laboratory abnormalities are rarely present and could include anemia, thrombocytosis, an elevated ESR and hypergammaglobulinemia. These abnormalities often resolve with excision of the lesion.<sup>20</sup>

IMT has a well known tendency for local invasive behavior and recurrence.<sup>1, 2, 21</sup> Only a small risk of distant metastasis has been reported by some authors.<sup>6, 18, 20, 22</sup>

Characteristic histopathological findings in typical IMT are a fasciitis-like, compact spindle cell proliferation with areas of myxoid

change and hypocellularity showing a collagenous back ground.<sup>20</sup> Various numbers of mixed inflammatory cells including polyclonal plasma cells, lymphocytes, eosinophils and rarely foamy macrophages are invariably seen.<sup>18, 22</sup> Typically the spindle cells of the IMT express vimentin, smooth muscle actin and other markers which correspond to the myofibroblastic nature of these cells.<sup>12, 14, 22</sup>

Histological features usually can not predict the biological behavior of the tumor. The presence of aneuploidy may, however, indicate the possibility of a local aggressive behavior and recurrence.<sup>11</sup> In pulmonary lesions atypia of the spindle cells are believed to indicate aggressiveness.<sup>22</sup>

The treatment of choice is believed to be complete surgical excision and long term follow up with physical examination, imaging techniques and serial monitoring of the erythrocyte sedimentation rate.<sup>23</sup> Radiotherapy and chemotherapy (cisplatin, doxorubicin and methotrexate) have also been tried as an adjunct to surgery with no evidence for substantial benefit for the patient.<sup>24, 25</sup> Steroid therapy achieved regression of a case of renal IMT. The benefits of non-steroidal anti-inflammatory medications are controversial.<sup>26</sup>

The case we present showed some of the typical features of IMT, namely the clinical presentation with intestinal obstruction. The tumor also showed the typical histomorphology of proliferating spindle cells and a considerable number of inflammatory cells, including plasma cells. Immunohistochemical studies were not in favor of any other spindle cell tumors of the gastrointestinal tract.

Although ALK-1 protein could not be demonstrated in the tumor cells, negative results are not strictly against the diagnosis of IMT. On the other hand, age of the patient and site of the lesion are not usual for most IMT cases.

Intussusceptions in adults always denote intraluminal pathology as was the case in our patient. The patient underwent laparoscopic

surgery with complete removal of the obstructing tumoral mass. No adjuvant therapy was employed and the patient remained asymptomatic after three years of follow-up without any evidence of recurrent disease.

In summary, IMT is a neoplastic lesion with the tendency for local aggressive behavior and recurrence. This illness should be considered in the list of the differential diagnoses of spindle cell tumors in any part of the body of patients at any age, particularly children and young adults.

### CONFLICT OF INTEREST

The authors declare no conflict of interest related to this work.

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