

An Uncommon Presentation of Inflammatory Myofibroblastic Tumor

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Inflammatory myofibroblastic tumor (IMT), also known as inflammatory pseudotumor or plasma cell granuloma, is a rare neoplasm that consists of spindle cell proliferation with a characteristic fibroinflammatory and even pseudosarcomatous appearance.^{1,2} The tumor presents most commonly in children and young adults, with a slight female gender predilection (male-to-female ratio, 3:4). These tumors most commonly occur in the lungs.¹ Although IMT has been reported in various extrapulmonary sites, its occurrence in the ileum is uncommon.³ Moreover, its presentation as intussusception is rare.³ In this article, we report the case of a woman who presented with intussusception and was found to have an IMT located in the ileum.

CASE PRESENTATION

Initial Presentation and History

A 32-year-old obese woman presented to an outpatient clinic for evaluation of severe (10/10), sharp, intermittent, worsening left lower quadrant abdominal pain of 3-months' duration that occasionally radiated to her back. There were no exacerbating/alleviating factors, no rectal bleeding, and no constitutional symptoms. Past medical history was significant for 2 cesarean sections. Menstrual history included menarche at age 12 years, with regular menses lasting 3 to 4 days with moderate flow. The patient was a nonsmoker, did not drink alcohol, and had no history of drug abuse. Family history was noncontributory.

Physical Examination and Laboratory Evaluation

Physical examination revealed a soft, nondistended abdomen, with left lower quadrant tenderness and normal bowel sounds but no organomegaly or palpable mass. The complete blood count was normal except for

a mildly elevated white blood cell count ($12.6 \times 10^3/\mu\text{L}$ [normal, $4.1\text{--}10.3 \times 10^3/\mu\text{L}$]). The differential count was as follows: neutrophils, 89% (normal, 40%–75%); lymphocytes, 8% (normal, 20%–53%); monocytes, 3% (normal, 0%–12%); eosinophils, 0% (normal, 0%–8%); and basophils, 0% (normal, 0%–2%). The basic metabolic profile was within normal limits. Computed tomography (CT) scan and small bowel series were suggestive of a distal small bowel mass (**Figure 1**). Based on this evaluation, a decision was made for the patient to undergo elective open laparotomy within a few days.

Treatment and Hospital Course

Laparotomy and segmental small bowel resection were performed to further evaluate the mass and revealed gross intussusception (**Figure 2**) and a palpable intraluminal pedunculated mass measuring $5.0 \times 3.5 \times 3.5$ cm, with surface necrosis and hemorrhage (**Figure 3**). The bulging, protuberant mass was protruding from but not penetrating the muscular layer of the bowel wall. Histologic examination of the mass showed loosely arranged myofibroblast-like plump and spindle-shaped cells with abundant small blood vessels and marked eosinophilic infiltration along with plasma cells and lymphocytes (**Figure 4**). Mitotic rate was very low. No ganglion-like cells or zones of calcification were observed. Immunoperoxidase stains were positive for vimentin and CD68 but

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Figure 1. Computed tomography scan of the abdomen showing right lower quadrant small bowel mass (arrow).

negative for CD117, desmin, S-100, CD34, and ALK-1. Based on the histologic findings, the patient was diagnosed with IMT.

Postoperatively, the patient's pain improved, and she had an uneventful recovery with complete resolution of her symptoms. She was discharged on the fourth hospital day. On outpatient follow-up, she continued to do well clinically and remained asymptomatic up to 6 months later.

INFLAMMATORY MYOFIBROBLASTIC TUMOR

IMT was first reported in the lungs in 1937 and was officially included in the World Health Organization classification of soft tissue tumors in 1994.⁴⁻⁶ IMT has been described as a neoplasm composed of spindle cells admixed with numerous inflammatory cells, including plasma cells and lymphocytes.⁶ IMT, which varies from 1 to 17 cm in size,^{1,7} occurs most commonly in the lungs. Extrapulmonary sites have also been reported and include the mesentery/omentum, genitourinary tract, gastrointestinal tract, retroperitoneum, pelvis, head and neck, trunk, and extremities.^{1,8} Although these tumors are typically benign, IMTs have an uncertain malignant potential and may show local recurrence, infiltrative growth, vascular invasion, and malignant sarcomatous transformation.^{1-3,5,9} Several terms have been used to refer to this entity in the literature, including inflammatory pseudotumor, inflammatory fibroid tumor, pseudosarcomatous myofibroblastic proliferation, plasma cell granuloma, xanthomatous pseudotumor, and eosinophilic granuloma.^{1-5,10,11}



Figure 2. Gross intussusception seen during surgery.

Etiology

The etiologic factors responsible for the development of IMT are not clearly understood, and it is unclear whether IMT is a reactive process or a true neoplasm. IMT may be caused by either immunologic triggers or an aberrant inflammatory response to tissue injury, such as infectious agents (eg, Epstein-Barr virus, human herpesvirus, *Mycobacterium avium intracellulare*, *Corynebacterium equi*, *Campylobacter jejuni*, *Bacillus sphaericus*, *Coxiella burnetii*, *Escherichia coli*), abdominal surgery, trauma, radiotherapy, steroid use, and overexpression of interleukin 6.^{2-5,9,10,12-14} Recent cytogenetic research, however, has revealed recurrent translocations involving chromosome band 2p23 associated with activation of the anaplastic lymphoma kinase (ALK) gene (a part of the insulin receptor family of receptor tyrosine kinases generally expressed in the nervous system). Fusion of the ALK gene with tropomyosin genes TPM3 and TPM4 and the clathrin heavy chain gene CLTC suggest neoplastic pathology.^{3,7,9,10,14} ALK is present more frequently in patients aged 40 years or younger (positivity rate, 21.7%) and is associated with better prognosis.^{3,14}

Clinical Features

The clinical presentation of the tumor depends to some extent on its site of origin. It may present as a mass, weight loss, fever, pain, or vomiting. The presentation of IMT as intussusception is rare and may be further complicated by intestinal obstruction. Signs and symptoms of intussusception may include vomiting, constipation, diarrhea, bloody stools, abdominal pain, or intestinal obstruction. Laboratory abnormalities associated with IMT may include hypochromic microcytic anemia, thrombocytosis, elevated erythrocyte sedimentation rate, and polyclonal hypergammaglobulinemia.^{1-5,7,11} Radiographic presentation of intra-abdominal IMT is nondiagnostic. Abdominal films



Figure 3. Gross specimen showing intraluminal pedunculated mass measuring 5.0 × 3.5 × 3.5 cm.

may reveal displacement of bowel segments due to a soft tissue mass and amorphous calcification within the tumor. Ultrasound and CT scan may show a solid, occasionally heterogeneous, well-demarcated spherical mass.^{5,12}

Histology

The differential diagnosis for spindle cell tumors includes IMT, leiomyoma, leiomyosarcoma, rhabdomyosarcoma, malignant fibrous histiocytoma, inflammatory fibrosarcoma, intra-abdominal fibromatosis, and gastrointestinal stromal tumor.^{4,5} Three basic histologic patterns have been recognized for IMT: (1) myxoid-vascular-inflammatory pattern resembling nodular fasciitis, (2) compact spindle cell pattern admixed with inflammatory cells (eg, plasma cells, lymphocytes, and eosinophils) resembling fibrous histiocytoma, and (3) dense collagen pattern resembling desmoid or scar tissue.^{1,9,10} Immunohistochemistry results also may be positive for vimentin, actin, desmin, cytokeratin, CD68, CD31, CD34, S-100, CD117, and neurofilament protein.^{1,3}

Treatment

The mainstay of treatment for this tumor is surgical resection with wide margins. Radiotherapy, immunosuppression, and chemotherapy have not been proven to have any definitive benefit.^{2–6,10–12,15} In the case patient, the tumor was completely resected. Incompletely resected tumors may have local recurrence within 1 year.¹² As noted earlier, because IMT exhibits uncertain malignant potential ranging from local recur-

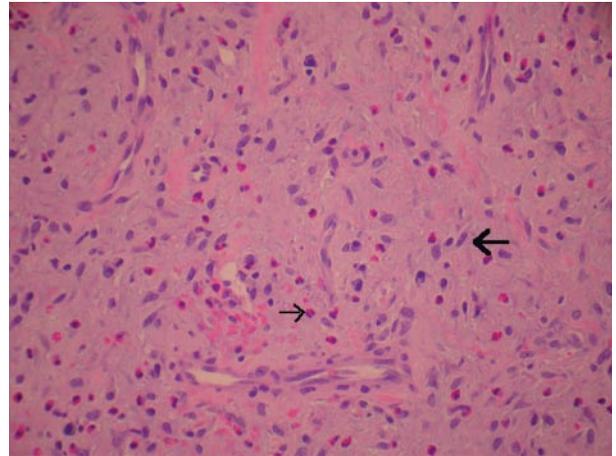


Figure 4. Histology slide showing abundant eosinophils (thin arrow) and myofibroblastic cells (thick arrow; hematoxylin and eosin stain, 400x).

rence to malignant transformation, it should be closely followed over a long-term period. Recurrence has been seen up to 9 years after resection of the primary tumor.¹⁵ Patients can be followed clinically or radiologically. In this case, the patient was followed clinically because she had complete resection of the tumor and resolution of all her symptoms.

CONCLUSION

IMT is a rare spindle cell tumor with an uncertain malignant potential. Thus, early recognition and complete surgical resection are necessary to avoid recurrences and prevent spread of locally aggressive tumors. In addition, early detection of signs of intussusception may prevent complications. Close follow-up is recommended to prevent recurrence. **HP**

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