

Received: 2019.09.30

Accepted: 2019.12.17

Available online: 2020.01.24

Published: 2020.02.27

Acute Intestinal Obstruction Due to Ileocolic Intussusception in an Adult; A Rare Presentation of Inflammatory Myofibroblastic Tumor

Authors' Contribution:

Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

ABCDEF **Tariq Hameed**
BFG **Mohak Singh**
ADEF G **Adiba Nizam**
BDF **Rahul Bhatia**
CEFG **Gaurish Sawant**

Department of Surgery, Maulana Azad Medical College, New Delhi, India

Corresponding Author: Tariq Hameed, e-mail: drtariqhameed@gmail.com
Conflict of interest: None declared

Patient: Female, 40-year-old
Final Diagnosis: Inflammatory myofibroblastic tumor
Symptoms: Obstipation • pain • vomiting
Medication: —
Clinical Procedure: Exploratory laparotomy followed by right hemicolectomy
Specialty: Surgery

Objective: Unusual clinical course

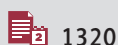
Background: Intussusception is not very common in adults, and acute intestinal obstruction with intussusception due to inflammatory myofibroblastic tumor (IMT) is extremely rare. IMT is an uncommon lesion and has no single defined cause. It predominantly affects the pediatric age group and commonly involves the lungs. Here we present a case of IMT causing ileocolic intussusception leading to acute intestinal obstruction in an adult.

Case Report: A 40-year-old female came to the emergency department with severe colicky pain in her abdomen, and reported 6 to 7 episodes of vomiting with bilious contents, along with an inability to pass feces and flatus for 3 days. An x-ray of her abdomen in erect posture revealed multiple air-fluid levels. Because she had a previous history of tuberculosis, a possible tubercular stricture as the cause of her acute obstruction was considered; an exploratory laparotomy was performed showing her bowel loops were dilated with ileocolic intussusception. The lead point of intussusception (a well-defined 4×4×3.5 cm solid mass), was found at 15 cm proximal to the ileocecal junction. A right hemicolectomy with ileo-transverse anastomosis was performed. The histopathological examination confirmed the presence of IMT.

Conclusions: IMT causing ileocolic intussusception with acute intestinal obstruction is an extremely rare presentation of an uncommon entity in adults. High index of suspicion, and appropriate investigations (x-ray abdomen, ultrasound, computed tomography, and colonoscopy) depending on presentation and clinical condition of the patient can result in prompt diagnosis and early management.

MeSH Keywords: Intestinal Obstruction • Intussusception • Neoplasms, Muscle Tissue

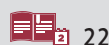
Full-text PDF: <https://www.amjcaserep.com/abstract/index/idArt/920438>



1320



5



22



Background

Inflammatory myofibroblastic tumor (IMT) is rare and has been reported at different anatomical sites with no single defined cause [1,2]. It is predominantly seen in the pediatric age group and rarely seen in adults [1]. It usually involves the lungs. The first case was described in the lungs; however, the gastrointestinal tract, retroperitoneum, orbits, urinary bladder, larynx, and testis may also be affected [3–6]. The entity now commonly referred to as IMT has been known in the past as plasma cell granuloma, plasma cell pseudo tumor, inflammatory myofibrohistiocytic proliferation, omental mesenteric myxoid hamartoma, and inflammatory pseudo tumor [7]. The diversity of nomenclature is due to the elusive etio-pathogenesis of this tumor. Due to its rare occurrence and presence at unusual sites, its presenting symptoms and signs are highly variable. Here we report a case of IMT which presented as acute intestinal obstruction due to ileocolic intussusception in a middle-aged female patient. It is an extremely rare presentation of an uncommon tumor in an adult.

Case Report

A 40-year-old Indian female came to surgical emergency department with complaints of severe colicky pain abdomen for 5 days which was followed 3 days later by 6 to 7 episodes of bilious vomiting and inability to pass feces and flatus. The pain had started in right lower abdomen becoming generalized after 3 days. She had a history of pulmonary tuberculosis 5 years ago, for which she had received complete treatment. She also had a history of colicky pain in her abdomen of 4 or 5 episodes in the last 1 year for which she consulted her primary treating physician. She had no history of addiction to alcohol or tobacco. Physical examination revealed abdominal distension with generalized tenderness. Bowel sounds were exaggerated.

Exploratory laparotomy was planned after baseline blood investigations and an x-ray of her abdomen in erect posture. Her abdomen x-ray revealed multiple air fluid levels (Figure 1). Complete blood count revealed elevated WBCs (14 200/ μ L) with neutrophilia (83%). Blood urea nitrogen level was also raised. On laparotomy, bowel loops were distended with ileocolic intussusception (Figure 2). Lead point of intussusception was at distal ileum approximately 15 cm proximal to ileocecal junction (Figure 3). Right hemicolectomy and ileotransverse anastomosis were performed. Specimens were resected and a well-defined firm round mass measuring 4×4×3.5 cm in the ileum, approximately 15 cm from ileocecal junction, was observed (Figure 4). The post-operative period was uneventful, and the patient was transferred to the surgery ward after 24 hours of observation in the intensive care unit. She recovered well and was discharged on the 10th post-operative day.



Figure 1. Radiograph showing small bowel obstruction.



Figure 2. Intraoperative photograph of ileocolic intussusception.

Histopathological examination showed her tumor to be composed of mainly fibroblasts and myofibroblasts which were of spindle shape. The background was predominantly inflammatory in nature showing eosinophils and vascularized edematous stroma (Figure 5). Nuclear atypia and mitosis were not seen. Immunohistochemical study showed that spindle shaped cells (the fibroblasts and myofibroblasts) were positive for smooth muscle actin and negative for anaplastic lymphoma kinase (ALK), pancytokeratin, CD31, CD34 and S100 protein. Resected margins were found to be free of tumor deposits. Our patient was not given any adjuvant therapy and was asymptomatic at the end of her 1-year follow up. Abdominal sonography at 6 months and 12 months did not reveal any abnormality.

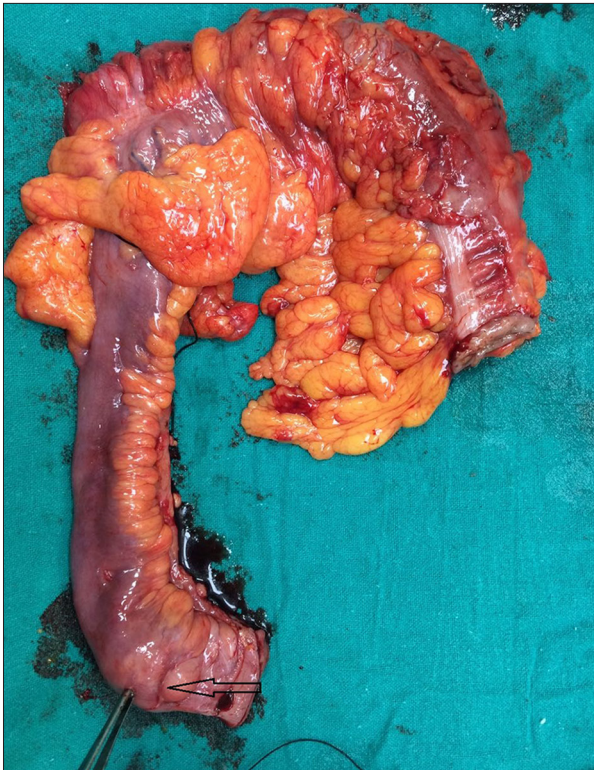


Figure 3. Gross image of resected specimen showing lead point of intussusception marked by arrow.



Figure 4. Cross section of ileum showing solid, rounded tumor.

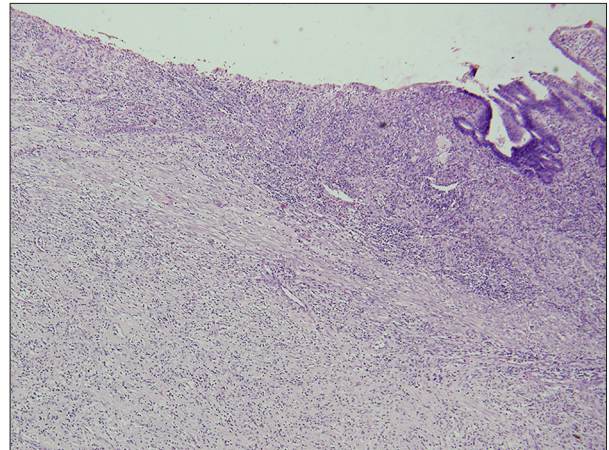


Figure 5. Proliferation of spindle cells – myofibroblasts and fibroblasts in a background of mixed inflammatory infiltrate with eosinophils and edematous vascularized stroma.

Discussion

IMT is classically defined as proliferation of fibroblasts-myofibroblasts and lymphocytic infiltration, with a background which is inflammatory in nature, exhibiting eosinophils and vascularized stroma. Proposed factors for development of IMT include trauma, surgery, and infection like Epstein Barr virus (EBV), human herpes virus (HSV)-8, and human papillomavirus (HPV) which are associated with reactive cytokine production [8–10]. IMT has been associated with raised levels of interleukin (IL)-6, spindle myofibroblast cells and lymphocytic cells mingled with spindle cells that expressed IL-6 on immunohistochemistry [10]. Some cases have been linked to translocations involving the ALK gene [11]. However, the exact mechanisms responsible for development of IMT remain unclear.

Our patient did not express the ALK gene but approximately half of all IMTs express ALK translocation [12,13]. It has been reported that those IMTs which are ALK-negative are aggressive with a higher propensity for metastasis [12]. Coffin et al. reported 6 patients of metastatic IMT, all were ALK negative [12], and older patients are more likely to have ALK-negative tumors [12].

IMTs are usually benign tumors. Local aggressiveness and malignant change are rare [14].

It usually develops in children or young adults but can affect people of any age, and these tumors might be asymptomatic. Constitutional non-specific symptoms such as fever, weight loss, and malaise are also seen in patients. When history and symptomatic and clinical presentations are determined by the anatomical location of IMT, and it is mostly due to the mass effects of the tumor. The tumor might occur at any anatomical site.

IMT has been described as present in multiple anatomic sites, but typically is located in the liver and biliary tract (32%), lung (27%), and gastrointestinal tract (10%). In gastrointestinal system involvement, omentum and mesentery are the most involved. Bowel involvement is infrequent. Acute intestinal obstruction with intussusception is an extremely rare presentation of IMTs.

Histologically, IMTs are characterized by a cellular spindle cell proliferation in a myxoid to collagenous stroma with a prominent inflammatory infiltrate composed primarily of plasma cells and lymphocytes, with occasional admixed eosinophils and neutrophils [2]. Diagnosis of IMTs is histopathological, following surgical resection of a tumor producing symptoms due to mass effect.

Three basic histological patterns have been described: 1) fasciitis-like, with elongated myofibroblasts, vascular, myxoid, and inflamed stroma, including plasma cells - the inflammatory infiltrate in these areas often contains abundant neutrophils and eosinophils and fewer plasma cells as compared to other pathological variants. 2) Compact spindle cell pattern is characterized by compact arrangement of myofibroblasts and fibroblasts in collagenous stroma and show numerous plasma cells and mitotic figures. 3) Fibromatosis-like pattern is relatively hypocellular, with elongated rather than plump spindle cells in a densely collagenous background containing scattered lymphocytes and plasma cells [1,15]. Pleomorphism is moderate, with little to no evidence of any mitotic activity.

Immunohistochemistry can confirm myofibroblastic origin of tumor spindle cells, which are typically reactive to vimentin (99%), smooth muscle actin (92%), and muscle specific actin (89%). Other myogenic markers include desmin (69%), cyto-keratin (36%), and CD68 (25%). IMTs are typically negative for myoglobin and S-100 protein [16].

Complete surgical excision is the mainstay of treatment and helps in establishing definitive diagnosis [17,18]. Complete resection with negative margins should be the surgical aim,

References:

1. Coffin CM, Watterson J, Priest JR, Dehner LP: Extrapulmonary inflammatory myofibroblastic tumor (inflammatory pseudotumor). A clinicopathologic and immunohistochemical study of 84 cases. *Am J Surg Pathol*, 1995; 19(8): 859–72
2. Gleason BC, Hornick JL: Inflammatory myofibroblastic tumours: Where are we now? *J Clin Pathol*, 2008; 61(4): 428–37
3. Voelker H-U, Kuehn D, Strehl A, Kircher S: Testicular inflammatory myofibroblastic tumor: A known entity at a very rare site. *Case Rep Urol*, 2017; 2017: 1410843
4. Strianese D, Tranfa F, Finelli M et al: Inflammatory myofibroblastic tumor of the orbit: A clinico-pathological study of 25 cases. *Saudi J Ophthalmol*, 2018; 32(1): 33–39
5. Guilemany JM, Alós L, Alobid I et al: Inflammatory myofibroblastic tumor in the larynx: clinicopathologic features and histogenesis. *Acta Otolaryngol*, 2005; 125(2): 215–19
6. Etani T, Naiki N, Nagai T et al: Inflammatory myofibroblastic tumor of the urinary bladder: A case report. *Case Rep Oncol*, 2016; 9(2): 464–69
7. Fletcher CDM, Unni KK, Mertens F: World Health Organization classification of tumours pathology and genetics of tumours of soft tissue and bone. *Cancer*, 2002; 177: 1365–76
8. Gurzu S, Bara T, Jung I: Inflammatory myofibroblastic tumor of the colon. *J Clin Oncol*, 2013; 31: e155–58
9. Dalton BG, Thomas PG, Sharp NE et al: Inflammatory myofibroblastic tumors in children. *J Pediatric Surg*, 2016; 51(4): 541–44

as it limits risk of local recurrence and malignant transformation [9]. Chemotherapy, steroids and radiation have been used as adjuncts to surgery, but no definitive guidelines exist [19]. Adjunctive treatments may be used in locally recurrent or margin positive disease.

Prognosis of IMT remains favorable [20]. Though IMTs have a tendency for local recurrence, it has a risk of distant metastasis and many cases have been described in literature [21,22]. The recurrence rate of IMT depends on the anatomical site of tumor; it is 2% for tumors confined to the lungs and up to 25% for extra pulmonary lesions where recurrence is mostly related to incomplete surgical removal or multinodular intra-abdominal tumors [2]. The main prognostic indicator is the adequacy of the primary excision [9]. Mortality rate is low, with only a few cases described in literature [7,9].

Because IMTs can recur locally especially during the first post-operative year, regular follow-up is advised even when surgical resection was adequate [9].

Conclusions

A middle-aged female presenting with acute intestinal obstruction due to IMT, which is an extremely rare presentation of an uncommon entity. Proper pre-operative evaluation, prompt diagnosis, and adequate excision are important prognostic factors.

The approach for each patient should be personalized as no uniform guidelines currently exist for follow-up of patients of IMT. Diagnosis is histological, and the treatment is complete surgical excision with microscopically tumor negative margins. Prognosis is favorable as tumor recurrence and metastasis is rare.

Conflicts of interest

None

10. Gómez-Román J, Ocejo-Vinyals G, Sánchez-Velasco P et al: Presence of human herpesvirus-8 DNA sequences and overexpression of human IL-6 and cyclin D1 in inflammatory myofibroblastic tumor (inflammatory pseudotumor). *Lab Invest*, 2000; 80: 1121–26
11. Cessna MH, Zhou H, Sanger WG et al: Expression of ALK1 and p80 in inflammatory myofibroblastic tumor and its mesenchymal mimics: A study of 135 cases. *Mod Pathol*, 2002; 15: 931–38
12. Coffin CM, Hornick JL, Fletcher CD: Inflammatory myofibroblastic tumor: Comparison of clinicopathologic, histologic, and immunohistochemical features including ALK expression in atypical and aggressive cases. *Am J Surg Pathol*, 2007; 31: 509–20
13. Butrynski JE, D'Adamo DR, Hornick JL et al: Crizotinib in ALK rearranged inflammatory myofibroblastic tumor. *N Engl J Med*, 2010; 363: 1727–33
14. Salehinejad J, Pazouki M, Gerayeli MA: Malignant inflammatory myofibroblastic tumor of the maxillary sinus. *J Oral Maxillofac Pathol*, 2015; 17(2): 306–10
15. Arora K: Inflammatory myofibroblastic tumor of soft tissue. *PathologyOutlines.com* website. <http://www.pathologyoutlines.com/topic/softtissueinflammmyofibro.html>
16. Marioni G, Bottin R, Staffieri A, Altavilla G: Spindle-cell tumours of the larynx: Diagnostic pitfalls. A case report and review of the literature. *Acta Otolaryngol*, 2003; 123: 86–90
17. Lee HJ, Kim JS, Choi YS et al: Treatment of inflammatory myofibroblastic tumor of the chest: the extent of resection. *Ann Thorac Surg*, 2007; 84(1): 221–24
18. Janik JS, Janik JP, Lovell MA et al: Recurrent inflammatory pseudotumors in children. *J Pediatr Surg*, 2003; 38: 1491–95
19. Kovach SJ, Fischer AC, Katzman PJ et al: Inflammatory myofibroblastic tumors. *J Surg Oncol*, 2006; 94(5): 385–91
20. Melloni G, Carretta A, Ciriaco P et al: Inflammatory pseudotumor of the lung in adults. *Ann Thorac Surg*, 2005; 79: 426–32
21. Al-Sindi KA, Al-Shehabi MH, Al-Khalifa SA: Inflammatory myofibroblastic tumour of paranasal sinuses. *Saudi Med J*, 2007; 28: 623–27
22. Deshingkar SA, Tupkari JV, Barpande SR: Inflammatory myofibroblastic tumor of the maxilla. *J Oral Maxillofac Pathol*, 2007; 11: 76–79