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Case Report

Inflammatory Myofibroblastic Tumour of Ileo-Caecal Junction Presenting as Acute Intestinal Obstruction with Retrocolic Abscess in A Young Female- A Case Report

Vaibhav Raj Gopal[1*]; Mithlesh Bhargav[2]; Faraz Ahmad[3]; Neelam Kumari[4]

[1]Assistant Professor, Department of General Surgery, King George's Medical University Lucknow, Uttar Pradesh, India
[2]Assistant Professor, Department of Pathology, All India Institute of Medical Sciences Gorakhpur, Uttar Pradesh, India 2.
[3]Additional Professor, Department of General Surgery King George's Medical University Lucknow, Uttar Pradesh, India 3.
[4]Senior Resident, Department of General Surgery King George's Medical University Lucknow, Uttar Pradesh, India

Corresponding Author:

Vaibhav Raj Gopal
Assistant Professor, Department of General Surgery, King George's Medical University Lucknow, Uttar Pradesh, India

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ABSTRACT

Background: Inflammatory myofibroblastic tumour (IMT) is a very rare mesenchymal tumour with an unclear benign or malignant potential. Lung is the most common site of occurrence but less frequently it also occurs at various extra pulmonary sites. Common sites in abdomen include retroperitoneum and mesentery. Complete surgical excision is the standard of care and malignant variants need to be supported with adjuvant targeted therapy with ALK inhibitors.
Case report: We present here a case of IMT of ileo-caecal junction presenting as frank acute intestinal obstruction in an 18 year old female which highlights the potential of this tumour for causing life threatening emergencies. Patient underwent laparotomy with right hemicolectomy. Histopathology was consistent with IMT of ileocaecal junction with no atypia. Patient was kept on regular follow up and presently she is asymptomatic.
Conclusion: IMT is mainly a histopathological diagnosis and complete surgical excision with negative margins offers the best treatment. Owing to the risk of recurrence and malignant potential, regular follow up is mandatory and targeted therapy via ALK inhibitors is warranted in such situations

Keywords: *Inflammatory myofibroblastic tumour, ileocaecal junction, acute abdomen.*

INTRODUCTION

Inflammatory myofibroblastic tumour (IMT) is a very rare mesenchymal tumour with an unclear benign or malignant potential. It is mostly reported as of benign origin and its incidence is less than 1 % in the population.[1],[2] Common sites of its occurrence include lung, nose, throat, skin and extremities. Intraabdominal locations like omentum, mesentery and retroperitoneum have been reported in literature but overall incidence is extremely low.[3],[4],[5],[6]Clinical features of this condition are site specific and surgical excision is the most common treatment of choice. Intraabdominal IMT usually presents with abdominal pain, altered bowel habits, bleeding per rectum, intussusception and abdominal lump. We present here a case of IMT of ileo-caecal junction presenting as frank acute intestinal obstruction in a young female which highlights the potential of this tumour for causing life threatening emergencies.

Case presentation

An 18 year old female presented in surgical emergency with complains of pain in abdomen with distension and inability to pass flatus and feces for 5 days. She also had off and on low grade fever for 1 week and episodic vomiting. She had no history of similar complains in the past. Her menstrual history was within normal limits and she had no history of contact with any tuberculosis patient or anti tubercular drug intake.Her vitals at the time of presentation were: Blood pressure: 110/70 mmHg, Pulse rate: 100/ minute, respiratory rate: 18/ minute, temperature: afebrile. On examination her abdomen

was distended and bowel sounds were absent. She also had tenderness in the right upper and right lower quadrant of abdomen. Patient had a contrast CT scan done from somewhere else which was also suggestive of multiple pockets of collection in the right paracolic gutter with maximum volume of 322 ml and non-visualisation of appendix and dilated small bowel loops. Based on these clinical and radiological findings, diagnosis of acute large bowel obstruction with suspected appendicular perforation was made. Patient was planned for surgical exploration.

On exploration small bowel loops were grossly dilated, a hard obstructing mass was present in the ileo-caecal junction with approximately 350 ml pus in the retrocolic region with grossly dilated small bowel loops. No obvious perforation was found at the tumour site but the appendix could not be identified during dissection which raised the possibility of sealed appendicular perforation. Right hemicolectomy with evacuation of pus and ileo-transverse anastomosis was done. Specimen was sent for histopathology. Postoperative outcome was uneventful and she was orally allowed on 7^{th} day and abdominal drain was removed on 8th day. She was discharged on 9^{th} day and was advised follow up with the histopathology report for further management. On follow up after a week she had no fresh complains and her histopathology was suggestive of inflammatory myofibroblastic tumour of ileo-caecal junction with no evidence of malignancy. Patient was kept under regular follow up every month for one year and presently she is asymptomatic. No adjuvant treatment either medical or radiation was given.

Figures

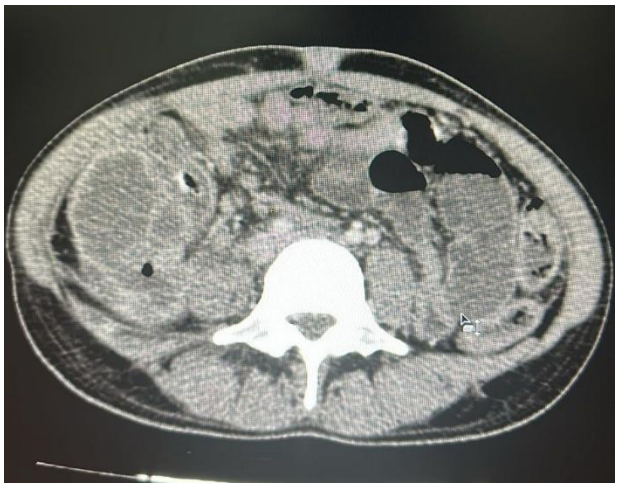

Figure 1a

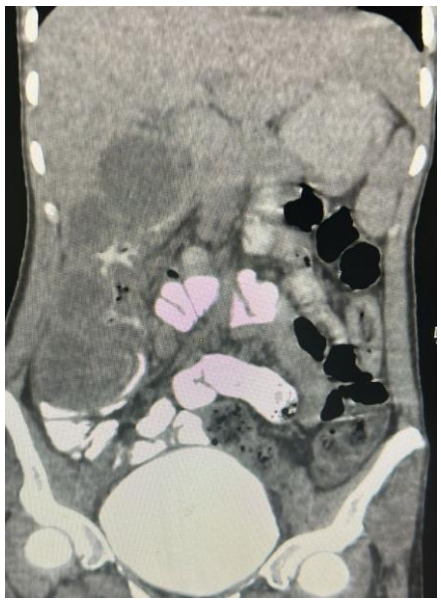

Figure 1b

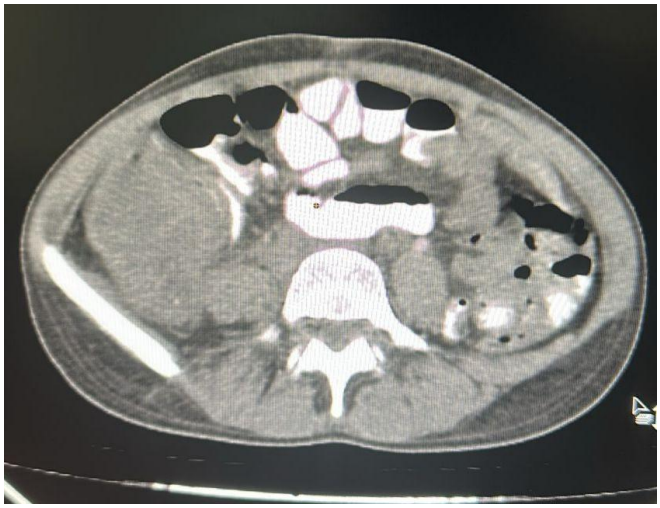

Figure 1c

Figure 1 a,b,c : CECT scan suggestive of multiple pockets of collection with air foci in right paracolic gutter with diffuse long segment thickening of ascending colon and caecum with obstructing lesion at ileo-caecal junction and dilated small bowel loops.

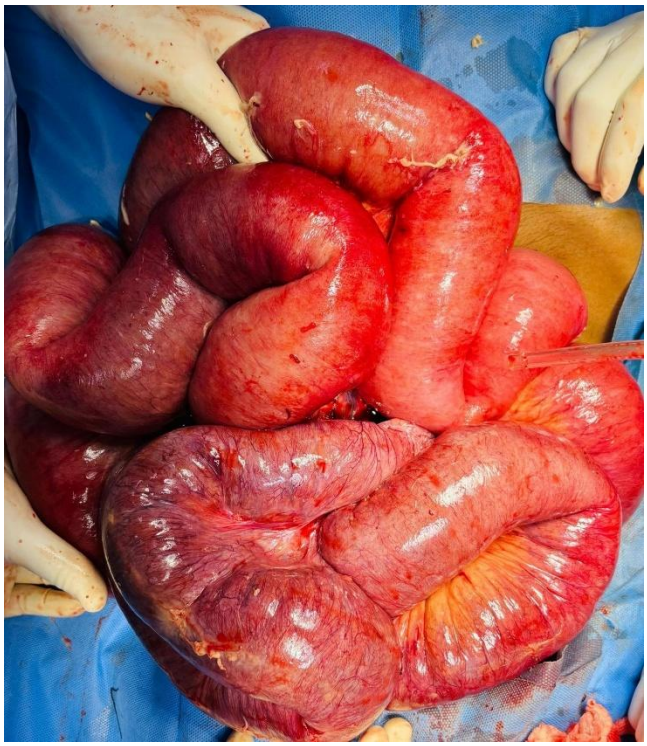

Figure 2 : Grossly dilated bowel loops with pus flakes

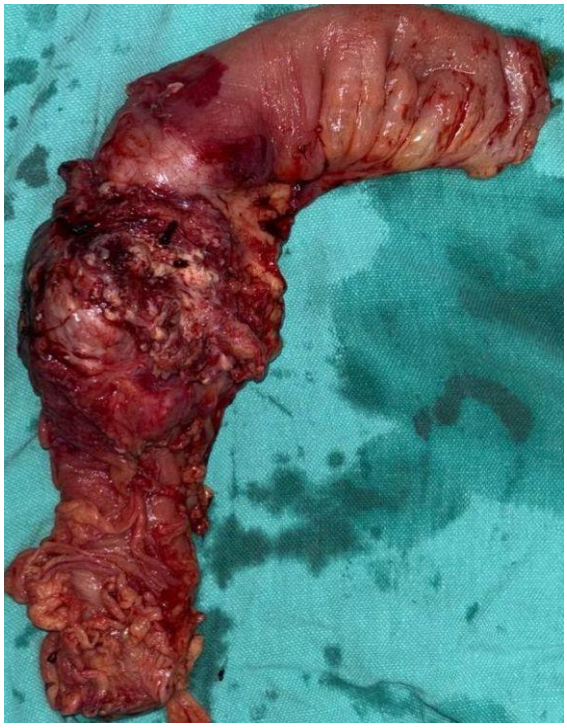

Figure 3: Right hemicolectomy specimen with a part of terminal ileum showing growth at the ileocaecal junction

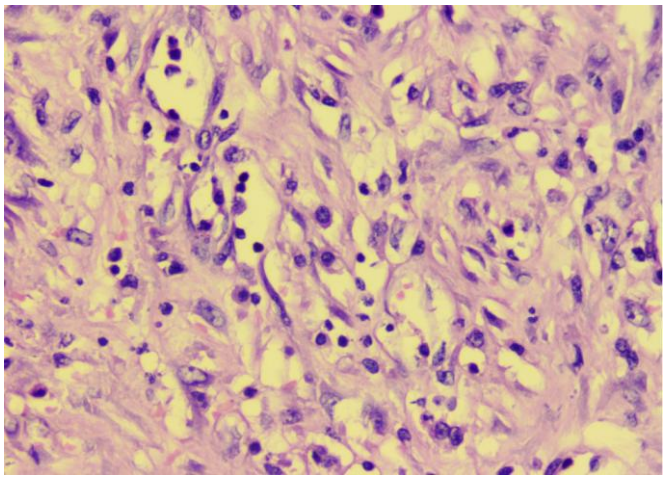

Figure 4 a

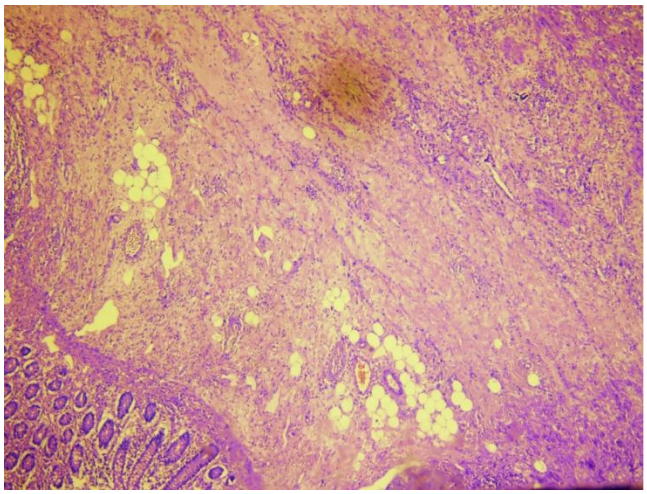

Figure 4 b

Figure 4 a,b : Sections from growth show diffuse areas of ulcerated mucosa and displaying panmural proliferation of myofibroblasts and fibroblastic spindle cells with intense mixed inflammatory cell infiltrate comprising of lymphocytes, plasma cells, histiocytes and polymorphs. These myofibroblasts are oval to spindle shaped, having bland nuclear chromatin, micro-nucleoli having ill-defined eosinophilic cytoplasm. Fair number of proliferating blood vessels and congested vascular channels are also seen. No evidence of granuloma or malignancy.

Discussion

Acute intestinal obstruction (AIO) is a surgical emergency necessitating surgical exploration and cause directed treatment. Common causes of AIO in adolescent population in India include adhesions, herniation, tuberculosis[7], intussusception, and less commonly neoplasia. Most of the times the diagnosis is clinical and the surgeon does not wait for the sophisticated radiological tests to substantiate the clinical finding. An erect abdominal X-ray is sufficient to aid in the diagnosis and head for surgical exploration. Hence a proper pre-operative diagnosis is not possible in majority of AIO. In our case also the clinical signs and symptoms were suggestive of acute bowel obstruction and she had a CT scan done at some other centre which was also consistent with the clinical diagnosis hence we proceeded for exploratory laparotomy.

IMT is mainly a histopathological diagnosis and is mostly regarded as a benign tumour with a very low chance of recurrence when completely excised. Though it usually occurs in young age and is gender non-specific, its presentation in old age has also been reported.[8] Some risk factors like trauma, association with Epstein Barr virus, Human Herpes - 8 virus, various autoimmune processes, IgG-4 related diseases have been identified but they don't hold a one to one causal relationship with the disease process.[9],[10] Lung is the most common site and mesentery and omentum constitute the most common site in the abdomen. Complete surgical excision of the tumour with negative margins is the best treatment till date. IMTs of small bowel, sigmoid colon and transverse colon presenting as AIO have also been reported in literature but the overall frequency is scarce.[11],[12] Owing to the rarity of this disease, standardised guidelines for management and follow up still lack in the literature.

Regarding the malignant potential of IMTs, it is reported to be of less than 5% and prognosis depends on the grade of tumour, metastasis and ALK gene positivity. [13]Tumours with ALK gene positivity have a good prognosis and this gene expression is seen in a fair number of IMTs.[14], [15]Crezotinib, an ALK gene (anaplastic lymphoma kinase) inhibitor is the targeted therapy drug which is approved for IMT with a fair response rate of 60-70%. It is very commonly used in lung IMT and in cases of locally aggressive lesions and metastasis.[16] There is a growing body of evidence in literature regarding the steroid therapy for IMTs of different sites like lung and maxillary sinus with a positive response through unknown mechanisms but this intervention will require consolidation and authentication via further studies.[17] Malignant gastric IMT with diffuse peritoneal and liver metastasis has also been reported in literature and hence malignant nature of IMT cannot be overlooked and even after complete excision, regular follow up of the patient with local examination and radiological imaging is mandatory.[13] Epithelioid inflammatory myofibroblastic sarcoma (EIMS) is also a malignant and aggressive variant of IMT which carries a high metastatic potential and poor prognosis and it presents in older age with a male preponderance. On immunostaining it is differentiated from the regular IMT by perinuclear ALK gene staining and RANBP-2/ALK fusion gene arrangement as compared to cytoplasmic ALK gene staining in the later. EIMS of ileocaecal junction has also been reported in literature.[18]

Conclusion

Ileo-caecal junction IMT presenting as AIO is a very rare case scenario and surgical management is in accordance with the management protocol of AIO and should be guided by the general condition of the patient, overall extent of disease, feasibility of resection with or without stoma creation or anastomosis. Preoperative histopathological diagnosis should only be reserved for stable patients not requiring emergency surgery. Since malignant potential and recurrences have also been described, regular follow up is necessary. Further research and studies are required for better understanding of IMT pathogenesis and genetics for innovation of refined diagnostic tests, novel chemotherapeutic agents for targeted therapy and prediction of prognosis. Presently, complete excision of the tumour with negative margins, if possible is considered to be the standard of care.

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