CASE REPORT

Inflammatory myofibroblastic tumour masquerading as acute appendicitis in a child: Case report

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Abstract

Inflammatory myofibroblastic tumour (IMT) is an uncommon and enigmatic neoplasm that can imitate a range of other medical conditions, frequently making it difficult to reach an accurate diagnosis. It primarily affects children, though there is a shortage of documented cases in the medical literature. The lungs are the most common location for this tumour to develop and intra-abdominal lesions are rare. Tumours arising from the small intestine and mesentery are exceptionally uncommon. This report highlights a rare case involving a 10-year-old child who presented to the emergency department exhibiting signs and imaging features that closely resembled acute appendicitis. However, surgical exploration revealed acutely inflamed appendix attached to a mass originating from the ileal wall, six feet proximal to the ileocaecal junction, covered by necrotic omentum and another mass in the ileal mesentery just proximal to the ileocaecal junction. The child underwent surgical removal of the appendix along with both the abdominal masses. The excised tissues were analysed through histopathology, confirming the diagnosis of an inflammatory myofibroblastic tumour. The post-operative period was uneventful, and follow-up evaluations revealed no evidence of the recurrence of the tumour.

Keywords: Inflammatory myofibroblastic tumour, Appendicitis, Mesentery.

DOI: https://doi.org/10.47391/JPMA.30354

Introduction

Inflammatory myofibroblastic tumour (IMT) is an uncommon and typically low-grade tumour that primarily occurs in children. It shares clinical features with tumours that have uncertain malignant potential, which is why it is considered a neoplasm of borderline behaviour. Although the lungs are the most frequent site of origin, IMTs can also develop in other parts of the body. Among extra-pulmonary locations, the abdomen, particularly the retroperitoneum and pelvis, is the most affected. Less commonly, IMTs have been observed in areas such as the head and neck, respiratory tract, limbs, and uterus. However, the occurrence of IMT within the mesentery of the small intestine is exceptionally rare, with very few cases described in the existing medical literature.[1] This report presents a rare case of ileal inflammatory myofibroblastic tumour with an associated mesenteric lesion presenting as acute appendicitis in a child, highlighting the diagnostic challenge and surgical management.

Case Report

A 10-year-old boy presented to the Paediatric surgery emergency department, Mayo Hospital, Lahore, on April 24, 2024 with history of pain on the right side of the groin, fever, and vomiting since the previous day. He was haemodynamically stable. On examination of the abdomen, there was tenderness in the right lower quadrant. His haemoglobin level was 11 g/dl, his leukocyte count was 12,000/µl (Normal 4,500 to 11,000/µl). Based on the boy's symptoms and physical examination, acute appendicitis was suspected and an ultrasound of the abdomen and pelvis was performed for further evaluation. Ultrasound of the abdomen and pelvis revealed that right iliac fossa had a hypoechoic area measuring about 3.8 x 2.7cm with surrounding echogenic mesentery and streak of free fluid. Emergency exploration was done which showed acutely inflamed appendix attached with the mass. The mass had soft to firm consistency, about 8 x 3 x 2 cm in size, was originating from the lateral wall of the ileum six feet proximal to the ileocecal junction, it was covered by necrotic omentum. (Figure 1a,1b) Another 3 x 2 cm mass

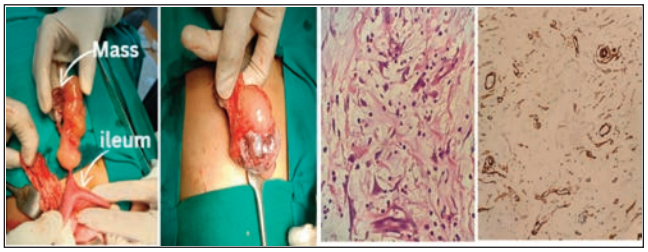

Figure: (a) Tumour attached to the lateral ileal wall, (b) Mass adherent to omentum and appendix (c) Lesion at 40X magnification: Elongated spindle-shaped cells with background inflammatory infiltrate, (d) CD 34: Positive in some lesional cells.

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Submission completed: 21-02-2025 **1st Revision received:** 08-07-2025
Acceptance: 03-01-2026 **Last Revision received:** 02-01-2026

with similar consistency was present 10 cm proximal to the ileocaecal junction on mesentery. Appendectomy and excision of both the masses was done. Histological analysis revealed the presence of spindle-shaped cells featuring eosinophilic cytoplasm and elongated nuclei, interspersed with inflammatory cells within a collagen-rich stroma. (Figure 1c,1d) Immunohistochemical testing indicated positive staining for smooth muscle antibodies and CD34, while other markers tested negative, supporting the diagnosis of an inflammatory myofibroblastic tumour (IMT). The patient recovered well after surgery and was discharged on the second post-operative day. Follow-up evaluations showed no evidence of tumour recurrence, and the child remained under regular clinical observation.

Discussion

Inflammatory Myofibroblastic Tumour (IMT), categorised under soft tissue neoplasms,[2] is primarily identified through histological examination. Since Dr Harold Brunn first described the condition in 1939, there has been a growing body of case reports and research expanding our clinical insight into this rare and intriguing tumour. Over the years, IMT has been known by several different terms, such as inflammatory pseudo-tumour, plasma cell granuloma, fibrous histiocytoma, solitary mast cell tumour, and fibroxanthoma, highlighting the tumour's diverse presentation and histological complexity.[2]

The aetiologic factors are not clearly understood.[2,3] It may be an immunological reaction to any infectious or non-infectious agents, or alternatively, a true neoplastic process. Some associated organisms are Campylobacter jejuni, Epstein-Barr Virus, and E. coli. Other factors may be trauma, abdominal surgery, steroid use, and genetic factors. However, the pathogenesis remains unclear.[3]

The clinical signs of IMT differ widely, based on where the tumour develops. Although it most commonly arises in the lungs, it can also be seen in several other parts of the body, such as the mesentery, omentum, upper airways, urinary system, retroperitoneal space, pelvic region, head, neck, spleen, brain, pancreas, liver, and various parts of the digestive system. Because of this wide range of possible locations, the symptoms can vary greatly, making each case unique and requiring a location-specific approach for both diagnosis and treatment.[1,4] Notably, documented cases of abdominal IMT have typically involved mesentery of the small intestine unlike the current patient, whose tumour originated from the lateral wall of the ileum.[5] Patients with intra-abdominal tumours typically experience intermittent abdominal pain, abdominal distension, and weight loss, accompanied by feelings of general discomfort, loss of appetite, and vomiting. These symptoms are often caused by the solid mass and abdominal distension. In some cases, the presentation can be more complex, with complications such as intestinal obstruction, intussusception, or acute abdomen, which can mimic acute appendicitis, as seen in the current case.[2,6,7] Only a few patients with intra-abdominal tumours exhibit laboratory abnormalities, which may include hypochromic microcytic anaemia, elevated serum immunoglobulins, and increased thrombocyte counts.[4,5] Additionally, leucocytosis, a rare haematologic finding, was also present in this patient. Notably, all these laboratory abnormalities, as well as the patient's symptoms, completely resolved after the tumour was surgically removed.

A recent study on inflammatory myofibroblastic tumours identified three distinct histological features. The first type is characterised by an abundance of inflammatory cells and a myxoid stroma. The second type exhibits a dense proliferation of spindle-shaped cells.[8] The third type is distinguished by a plate-like pattern of abundant collagen deposition, which may also include lymphoplasmacytic infiltration with germinal centres in lymphoid follicles as reported by Yamamoto et al.[1] In the current case, IMT displayed fascicles and bundles of plump spindle-shaped cells with admixture of inflammatory cells, including lymphocytes and plasma cells. Individual plump spindle cells have moderate amount of eosinophilic cytoplasm and elongated nuclei with finely dispersed chromatin, pinpoint nucleoli, pseudo inclusions and occasional mitotic activity. Background stroma is collagenised and exhibits hyalinised blood vessels. There is no evidence of increased mitotic activity or necrosis. The tumour cells showed immunoreactivity to anti-smooth muscle antibodies and CD34, while they did not express cytokeratin AE1/AE3, Desmin, ALK protein, ALK DF53, DOG-1, or STAT-6.

Identifying inflammatory myofibroblastic tumours (IMTs) can be challenging, even for highly skilled pathologists. However, immunohistochemistry serves as a valuable tool in distinguishing IMTs from other similar tumours, such as gastrointestinal stromal tumours, leiomyosarcomas, and inflammatory malignant fibrous histiocytomas.[9]

Inflammatory myofibroblastic tumours (IMTs) are typically non-aggressive in nature, with recurrence or spread to other areas being uncommon. However, when these tumours occur in the mesentery or retroperitoneal region, they can recur locally in approximately 15% to 37% of cases.[4] There is currently no established guideline on additional treatment.

Surgery to completely remove the tumour remains the standard treatment approach for IMTs and further research is needed to determine the optimal management strategies in these cases.[4,7,10]

Conclusion

This report describes a rare case of inflammatory myofibroblastic tumour originating from the ileal wall with an associated mesenteric lesion, presenting clinically as acute appendicitis in a child. The diagnosis was established following surgical exploration and confirmed on histopathology and immunohistochemistry. Complete surgical excision resulted in an uneventful postoperative course, with no evidence of recurrence on follow-up. This case highlights the need to consider uncommon intra-abdominal tumours in children presenting with acute abdomen.

Consent to Publish: Written consent was provided by the patient's parents for publishing the case report.

Disclaimer: None.

Conflict of Interest: None.

Funding Sources: None.

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MRW: Writing, editing, literature review, gathered relevant information and coordinated the submission and revision process.
MNUH: Writing, editing, concept and design.
SSHZ: Writing and editing.
BF & MA: Supervision and relevant guidance.
ZZ: Data collection and interpretation.