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Inflammatory myofibroblastic tumor masquerading as perforated appendicitis

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ABSTRACT

Inflammatory myofibroblastic tumor (IMT) is a rare and histologically benign tumor that most commonly presents in pediatric patients. Abdominal IMTs often present with non-specific symptoms, and therefore they can masquerade as other more common abdominal conditions. This report describes two cases of IMT presenting as perforated appendicitis. Both cases are of young children who presented with over 48 hours of abdominal pain and who had peritonitis on examination. Pre-operative ultrasounds for both patients revealed complex free fluid. Laparoscopic evaluation demonstrated hemoperitoneum, and further exploration revealed mesenteric avulsion with abnormally thickened mesentery. Pathology confirmed IMT arising from the small bowel mesentery in both cases. Both children underwent limited bowel resection and were discharged in stable condition on a course of non-steroidal anti-inflammatory drugs (NSAIDs). Given the association of avulsed bowel to trauma, non-accidental trauma was suspected in both cases and demonstrated in one. Both patients showed complete resolution of tumor on follow-up MRI. The diagnosis of IMT must always be considered when an intra-abdominal mass is encountered as its treatment differs greatly from other neoplasms.

Inflammatory myofibroblastic tumor (IMT) is a rare and histologically benign tumor that most commonly presents in pediatric patients. These lesions were originally thought to be reactive, though current evidence suggests that IMTs are a neoplastic process. Contributing factors include trauma, prior surgery, infection, inflammation, or autoimmune processes. Overexpression of anaplastic lymphoma kinase 1 (ALK-1) has also been associated with development of IMT [1,2]. Complete resection is recommended for patients with discrete lesions. For patients with multifocal or diffuse tumors, or with tumors in difficult locations, adjuvant treatment with chemotherapy, radiation, corticosteroids, immune modulators, or non-steroidal anti-inflammatory drugs (NSAIDs) have been used, with limited supportive data. The local recurrence rate ranges from 14 to 40% and is higher for those with larger tumors, multilobulated lesions, and locally invasive tumors [2]. The most important factor in preventing recurrence is complete resection. Two studies have demonstrated no recurrence in patients who have undergone complete resection [3,4].

IMT is most often found in the lung followed by the mesentery and omentum, soft tissue, mediastinum, intestine, pancreas, oral cavity, skin, breast, nerve, bone, and central nervous system [5]. These tumors cause symptoms specific to their location. Abdominal lesions typically present with mass, fever, weight loss, pain, or obstruction [2,6]. However, they can also be found incidentally [7]. Given the non-specific symptoms related to abdominal IMTs, they can masquerade as other more common abdominal conditions. This report describes two cases of IMT presenting as perforated appendicitis.

1. Case 1

A 3 year-old previously healthy girl presented to the emergency department with three days of worsening abdominal pain that began in the peri-umbilical region and then migrated to the right lower quadrant. The patient had bilious emesis and low-grade fevers prior to arrival. Review of systems was otherwise negative. There was no history of recent trauma.

Abbreviations: IMT, Inflammatory myofibroblastic tumor; NAT, Non-accidental trauma.

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On examination, the patient was afebrile and normotensive. She appeared mildly ill. Abdominal exam was notable for distention and peritonitis at McBurney's point. Laboratory tests revealed a white blood cell count of 16,200 with 72% neutrophils. The remainder of her laboratory workup was unremarkable. An abdominal ultrasound revealed a moderate amount of fluid throughout the right hemi-abdomen and layering debris. The appendix was not visualized.

Given the patient's history, exam, laboratory values, and imaging results, a diagnosis of perforated appendicitis was suspected. The patient was taken to the operating room for definitive laparoscopy. A laparoscope was introduced through a 10 mm umbilical incision. Moderate hemoperitoneum was immediately encountered, of which the majority seemed to be old blood (Fig. 1a). A segment of necrotic bowel could be seen in the inferior abdomen. The appendix appeared normal. A limited laparotomy was then made to more completely assess the intra-abdominal pathology and perform definitive repair. With the laparotomy, 25 cm of deserosalized proximal jejunum was identified. The associated mesentery was completely avulsed and foreshortened down to the mesenteric root (Fig. 1b). This segment was resected en bloc and primary anastomosis was performed. The mesentery itself in this region appeared as a thickened, matted, and friable mass, and the abnormal tissue appeared to extend proximally to the ligament of Treitz. The mass had the appearance and texture of lymphoma. The extensive mesenteric involvement precluded complete resection. Therefore, a partial mesenteric resection was performed.

Final pathology from the specimen was consistent with an IMT arising from the small bowel mesentery. The specimen stained positive for smooth muscle actin and desmin. Interestingly, the specimen was negative for ALK-1. The mass did not invade the submucosa of the involved small bowel. The patient's post-operative course was uncomplicated, and she was discharged home on a course of NSAIDs. She had Magnetic Resonance Imaging (MRI) performed of her abdomen and pelvis at 6 months postoperative, and this did not show any recurrence or persistence of disease.

2. Case 2

A 20-month-old boy was transferred to the emergency department with two days of non-bloody diarrhea, emesis, abdominal pain, and decreased activity. He was afebrile. There were no sick contacts or family members with similar symptoms. Initial evaluation revealed the patient to have low grade fevers and tachycardia. His abdomen was soft, non-distended and diffusely tender. The tenderness was associated with rebound in the lower quadrants. White blood cell count was elevated to 22,000. An abdominal X-ray was unremarkable. Abdominal ultrasound demonstrated complex fluid throughout the abdomen, edematous mesentery, and thickened loops of bowel. The appendix could not be visualized. Based on these findings, perforated appendicitis remained at the top of the differential diagnosis.

The patient was taken to the operating room for laparoscopic exploration. Laparoscopy visualized moderate hemoperitoneum and ecchymosis with torn peritoneum along the anterior abdominal wall in the right inguinal region (Fig. 2a). A source of bleeding could not be identified, and the umbilical incision was lengthened to 3 cm to enable bowel evisceration. Exploration revealed obvious avulsion of the mesentery from a segment of mid-small bowel. The associated 15 cm small bowel segment was ischemic (Fig. 2b). Three hundred milliliters of sanguinous fluid was evacuated. There were also several areas of serosal disruption in the small bowel immediately proximal and distal to the mesenteric avulsion. The small bowel mesentery was abnormally thick. No discrete mass was seen. Frozen section of the mesentery demonstrated fibrotic changes without evidence of malignancy. The avulsed and ischemic small bowel was resected with subsequent primary anastomosis. The remaining thick mesentery was left in place. Exploration of the remainder of the abdomen demonstrated no other solid or hollow viscus injuries. The appendix was normal.

The findings of mesenteric avulsion and torn peritoneum of the abdominal wall were concerning for non-accidental trauma (NAT). Questioning of family members revealed the patient had suffered blunt trauma to the abdomen by one of his caretakers. He was discharged in the custody of the state's child protective services.

At the same time as disposition arrangements were being made, the pathology specimen returned. Pathology was consistent with IMT arising from small bowel mesentery, and revealed a cytologically bland proliferation of spindle and stellate-shaped cells arranged in a myxoid/hyaline stroma with scattered inflammatory cells. Staining was again positive for smooth muscle actin and desmin, but negative for ALK-1. This was thought to be a coincidental finding that had perhaps exacerbated the effects of blunt trauma to the abdomen. The patient's postoperative course was otherwise unremarkable. He was discharged on a 6-week course of NSAIDs. He also had an MRI at 4 months postoperative – this did not show any recurrence or persistence of disease.

3. Discussion

In this article, we describe two cases of IMT of the mesentery presenting with acute abdomen. Both patients were taken to the operating room with a preoperative diagnosis of perforated appendicitis based upon their symptoms and imaging findings. Intra-abdominal IMT typically presents with a more sub-acute course related to the inflammatory response, including fever, impaired growth, weight loss, and anemia [2,6,8,9]. These findings

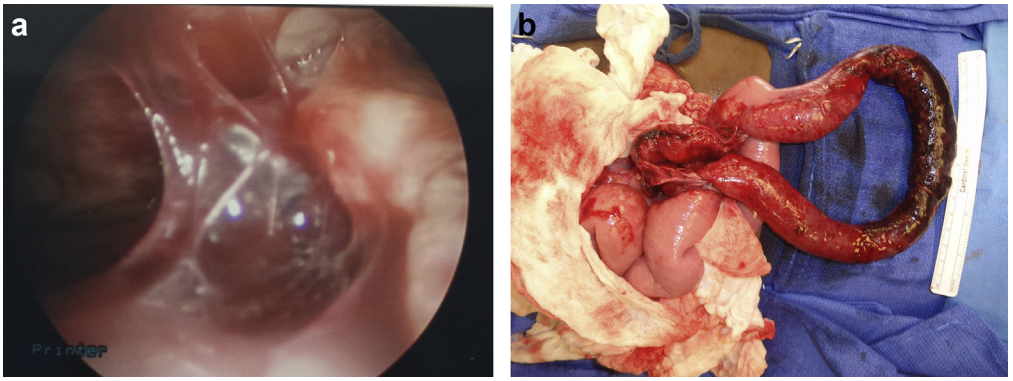

Fig. 1. a) Moderate hemoperitoneum encountered upon placement of laparoscope. b) 25 cm segment of deserosalized proximal jejunum.