

Case Report

Inflammatory Myofibroblastic Tumor of the Small Intestine: A Case Report of a 2 Month-Old Infant

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Benign intestinal tumors are rare in children; however, the authors describe an inflammatory myofibroblastic tumor (IMT) of the terminal ileum in a 2-month-old infant who presented with an intestinal obstruction. During laparotomy, an annular mass around the terminal ileum was resected, from which a histological diagnosis of IMT was made. A review of the literature for this rare condition was done to delineate the natural history of this tumor and to do a histological confirmation of its benign nature. Because of the risk of local recurrence, IMT cases should have a long-term follow up.

Keywords: Inflammatory myofibroblastic tumor, Inflammatory pseudotumor, Small intestine, Infant

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The inflammatory myofibroblastic tumor (IMT), was originally termed as inflammatory pseudotumor or plasma cell granuloma, is a spindle cell proliferation with a characteristic fibroinflammatory appearance^(1,2). It was inflammatory in nature, to those that are clearly neoplastic⁽³⁾. Although neoplastic lesions generally pursue a benign clinical course, intraabdominal and retroperitoneal lesions of this type have typically shown higher local recurrence rates and even distant metastases. The authors report on a case of terminal ileal IMT presenting in a 2-month-old infant and review the literature to delineate the natural history of this entity in children.

Case Report

A 2-month-old male infant presented with a history of 11-day of abdominal distension and bilious vomiting. Pregnancy and delivery were uneventful, and there had been no history of delayed passage of the first meconium. Seven days before his hospital admission, the symptoms became constant and more severe.

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On physical examination, a visible bowel loop and markedly abdominal distension were found in the abdomen (Fig. 1). Plain abdomen, supine, upright, and lateral cross-table prone position were done and showed distal small bowel obstruction (Fig. 2A-2C). Barium enema revealed no colonic obstruction but the abnormal shape of colon indicated a chronic disease



Fig. 1 Markedly abdominal distension and a visible bowel loop of abdomen

of the large bowel (Fig. 2D). A small bowel follow through confirmed the diagnosis of distal small bowel obstruction. The patient did not respond to saline rectal irrigation and was presented with more bilious vomiting and a gradually increasing abdominal distension.

On exploratory laparotomy, an annular mass surrounding the terminal ileum was discovered and multiple mesenteric lymph nodes were enlarged (Fig. 3A). There was no evidence of metastasis. The mass was resected. The infant did well postoperatively. The resected specimen of small intestine measured

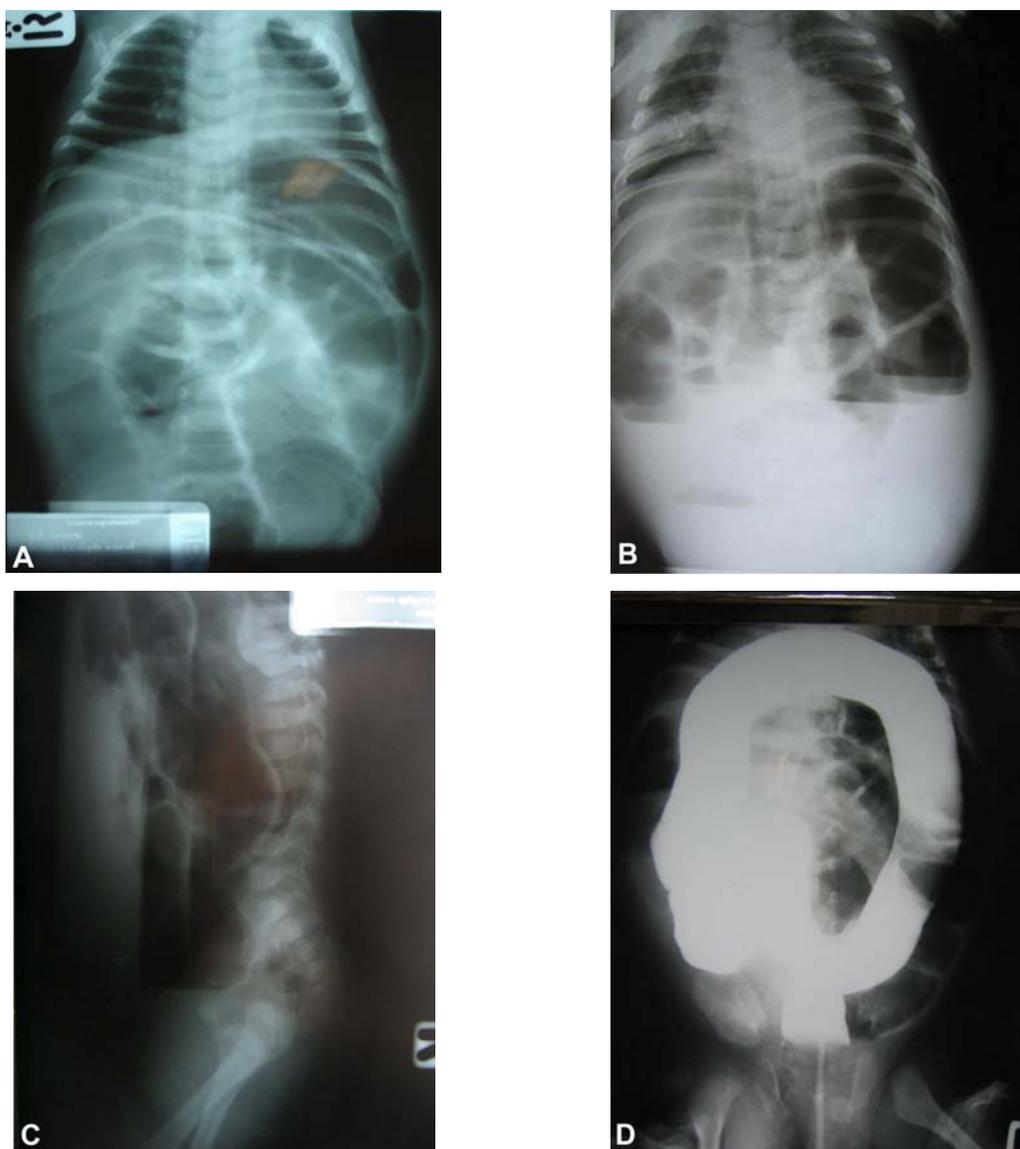


Fig. 2 Plain abdominal radiographs and radiocontrast studies

- A) Abdominal supine showing moderate dilatation of the entire small bowel. The colonic and rectal gas shadows are not definitely visible
- B) Abdominal upright showing of step ladder pattern of bowel
- C) Prone cross table lateral view showing the gas-filled rectum appears smaller than small bowel
- D) Barium enema showing the entire colon appears tubular, shortened, slightly narrowed, with loss of normal haustral folds, but corrugated in some areas. No reflux of barium into terminal ileum or appendix

3 cm in length, 2 cm in diameter showing an intramural nodule 1.5 cm in diameter, situated 2.5 cm from one resected margin (Fig. 3B).

Histologically, the tumor was covered by mucosa and was composed of fascicular arrangement of spindle cells with large oval nuclei occupying submucosa, muscularis propria, serosa, and lamina propria (Fig. 4A). There was some myxoid change of the stroma with infiltration of lymphocytes and plasma cells, mainly perivascularly. Neither nuclear atypia nor mitoses were present. Immunohistochemical study revealed positive to actin, desmin and vimentin, negative CD 117 and ALK protein. (Fig. 4B). Follow-up clinical status at 6 months and one year postoperatively revealed no evidence of recurrent tumor.

Discussion

IMTs have been described in a wide variety of anatomic sites and a broad spectrum of patient ages⁽¹⁻³⁾. They are the currently accepted terminology for tumors which have been previously referred to with a myriad of different names, the most common being inflammatory pseudotumor, plasma cell granuloma and inflammatory fibroid tumor⁽⁴⁾. They are not uncommon and typically arise in children between the ages of 2 years and 16 years⁽⁵⁾. Infantile cases have also been described⁽⁶⁾. Within the gastrointestinal tract, they most commonly arise from the wall of the small bowel and stomach. In the study by Coffin et al⁽⁷⁾, 36 of 84 cases (43%) arose in these sites. Although the age range is broad, extrapulmonary tumors show a predilection for

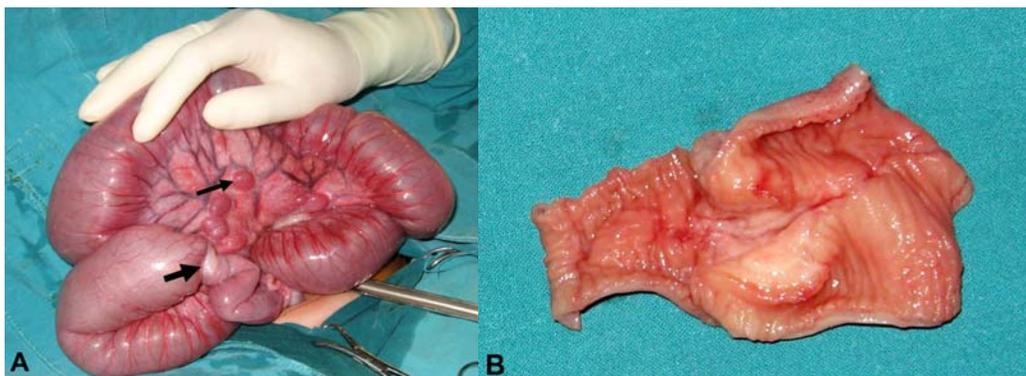


Fig. 3 A) Annular mass at terminal ileum (large arrow) with multiple mesenteric lymphadenopathy (small arrow)
B) Cut surface of specimen, homogeneous, tan-white color

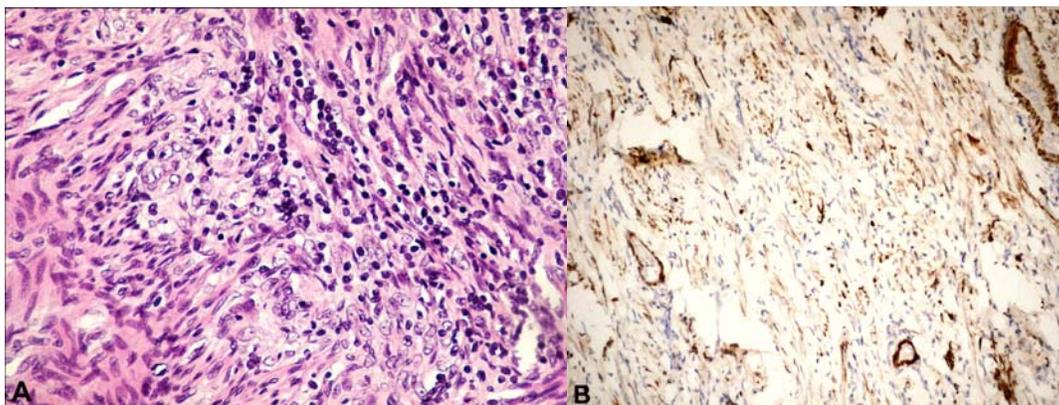


Fig. 4 A) Fascicles of spindle cells with thick-walled blood vessels in the muscularis propria and the submucosa (H&E, x20)
B) Immunoreactive to desmin of the tumor between the muscularis propria and muscularis mucosa (H&E, x20)

children, with a mean age of approximately 10 years. Females are affected slightly more commonly than males. Presenting symptoms depend on the site of primary tumor involvement. Patients with intra-abdominal tumors most commonly present with abdominal pain, abdominal mass with increased girth, or occasionally, with intestinal obstruction. Some patients have prominent systemic manifestations, including fever, night sweats, weight loss, and malaise. Laboratory abnormalities are present in a minority of patients and include an elevated erythrocyte sedimentation rate, anemia, thrombocytosis, and hypergammaglobulinemia, which often resolve with excision of the lesion⁽⁷⁻⁹⁾.

Because of the rarity of this proliferation in the terminal ileum, its natural history and biological potential are still uncertain and there have been no other reports of IMT in Thailand before. Intra-abdominal IMT has a propensity for more aggressive clinical behavior than their extraabdominal counterparts. A recent report by Sanders et al⁽¹⁰⁾ documented four cases of IMT of the alimentary tract, one at the gastroesophageal junction, one in the rectum, one involving the appendix and sigmoid colon. One patient (25%) had two recurrences; the first occurred within two months and was treated by surgical resection. The second recurrence was discovered six months later and regressed without surgical intervention, while the patient was on oral inflammatory therapy. In the study of extrapulmonary tumors by Coffin et al⁽⁷⁾, of 53 cases with follow-up 13 patients (25%) had one or more recurrences from 1 to 24 months after initial excision. Nine of the 13 patients who developed recurrence had retroperitoneal or intraabdominal tumors, including two tumors that underwent histologic transformation into a higher grade sarcoma. Although none of the patients developed metastatic disease, including the two patients whose tumors showed histologic progression, two patients with mesenteric tumors died as a direct result of their tumors.

The etiology of IMT remains unclear. Several reports have focused on the potential relationship associated with and infectious⁽¹¹⁻¹⁵⁾ or other clear inflammatory process. Defined as such, cases of IMT clearly are not neoplastic. However, there is sufficient evidence to support the fact that the IMT lesions described are true neoplasms, albeit typically benign. Clonal cytogenetic abnormalities have been demonstrated in some cases,⁽¹⁶⁻¹⁸⁾ particularly defined abnormalities on chromosome 2. Biselli et al⁽³⁾ recently found that almost 50% of pediatric extrapulmonary IMT is aneuploid.

IMT has a wide variation in histologic appearance, with three major subtypes: fibromyxoid and vascular pattern, proliferating pattern, and sclerosing pattern⁽¹⁾. In general, the tumors are ill-defined, with a variable admixture of inflammatory cells and spindle-shaped myofibroblasts set in a collagenous or edematous hypocellular stroma. Plasma cells usually dominate over other inflammatory cells with lymphocytes, eosinophils and macrophages present in more variable proportions⁽¹⁹⁾. The investigations to highlight the origin of the spindle cells in IMTs using immunohistochemistry have revealed myofibroblastic differentiation⁽¹⁾. The spindle cells have often been found to be reactive with antibodies against vimentin, smooth-muscle actin and muscle-specific actin. Differential diagnoses of IMT include the gastrointestinal stromal tumor (GIST) and myofibroma for the benign lesion. Myofibroma shows nodular pattern of myoid cells around thin-walled blood vessels. GIST can be excluded by histologic features and negative to CD 117. The gastrointestinal autonomic nerve (GAN) tumors are histologically similar to IMT, but they are uniformly negative to muscle-specific actin and CD 68, in contrast with IMT⁽²⁰⁾. Moreover, GAN tumors have been reported to show a predilection to affect middle-aged patients.

Clinical behavior of IMT is unpredictable, with approximately 25% of cases recurring locally⁽²⁾. In the present case, the barium enema revealed abnormal radiological features of the colon. The authors assume that IMT may have been involved in the colon but with close long-term follow-up, no clinical symptoms of colonic involvement were found.

Conclusion

IMT cannot be distinguished clinically from highly malignant neoplasm, and some other conditions, and they may mimic malignant tumor radiologically. Surgical resection of this tumor is advisable, and patients with IMT should be informed that there is a small but definable possibility of aggressive definable behavior and emphasizes the importance of long-term clinical and radiological follow-up.

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Inflammatory myofibroblastic tumor ของลำไส้เล็ก: รายงานผู้ป่วยทารกอายุ 2 เดือน

ภิเชก ยิ้มแย้ม, สุรัชย์ สราญฤทธิชัย, พูนศิริ สีนะวัฒน์, ตูลา เทียนศิริ

เนื้องอกชนิดไม่ร้ายแรงของลำไส้ในเด็กนั้นพบได้น้อย อย่างไรก็ตาม ผู้นิพนธ์ได้อธิบายถึงเนื้องอก myofibroblastic tumor ของลำไส้เล็กส่วนเทอร์มินัล อีเลียม ซึ่งพบในเด็กทารกอายุ 2 เดือน ที่มาด้วยอาการของลำไส้อุดตันจากการผ่าตัดพบก้อนเนื้องอกอยู่โดยรอบลำไส้ส่วนนี้และได้ถูกตัดออกไปจนหมด ร่วมกับส่งตรวจทางจุลพยาธิวิทยาเพื่อการวินิจฉัย นอกจากนี้ผู้นิพนธ์ยังได้ทบทวนวรรณกรรมเพื่อให้ทราบถึงธรรมชาติ และการวินิจฉัยทางจุลพยาธิวิทยาของเนื้องอกชนิดนี้ เนื่องจากมีโอกาสเสี่ยงที่จะเกิดซ้ำ และจำเป็นต้องติดตามการรักษาเป็นเวลานาน
