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Atypical Presentation of Intestinal Intussusception Caused by Insidious Inflammatory Myofibroblastic Tumor of the Bowel Wall

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Introduction: Inflammatory myofibroblastic tumor (IMT) is a rare but real tumor, which is histologically characterized by myofibroblastic spindle cells proliferation with inflammatory infiltrate. The lung is the most common affected organ, and extrapulmonary IMTs are less common. However, IMT seldom presents in the gastrointestinal tract, and intussusception is a rare complication of this tumor.

Case presentation: We documented this rare case of a 12-year-old Chinese girl presenting with abdominal pain. The clinical and radiologic impression was bowel intussusception and bowel obstruction. No sign of abdominal mass was found before surgery, neither physical examination nor radiologic images. Operative findings revealed intestinal intussusception secondary to a little mass. Histopathlogic evaluation of this mass revealed IMT.

Conclusion: In conclusion, IMT may present with bowel intussusception. However, at the intestinal location, the tumor may be found as an abdominal mass or may be insidious; hence, detailed history, physical examination, and imaging studies are necessary for early recognition and diagnosis.

Key words: Intussusception – Inflammatory myofibroblastic tumor – Ileum – Childhood

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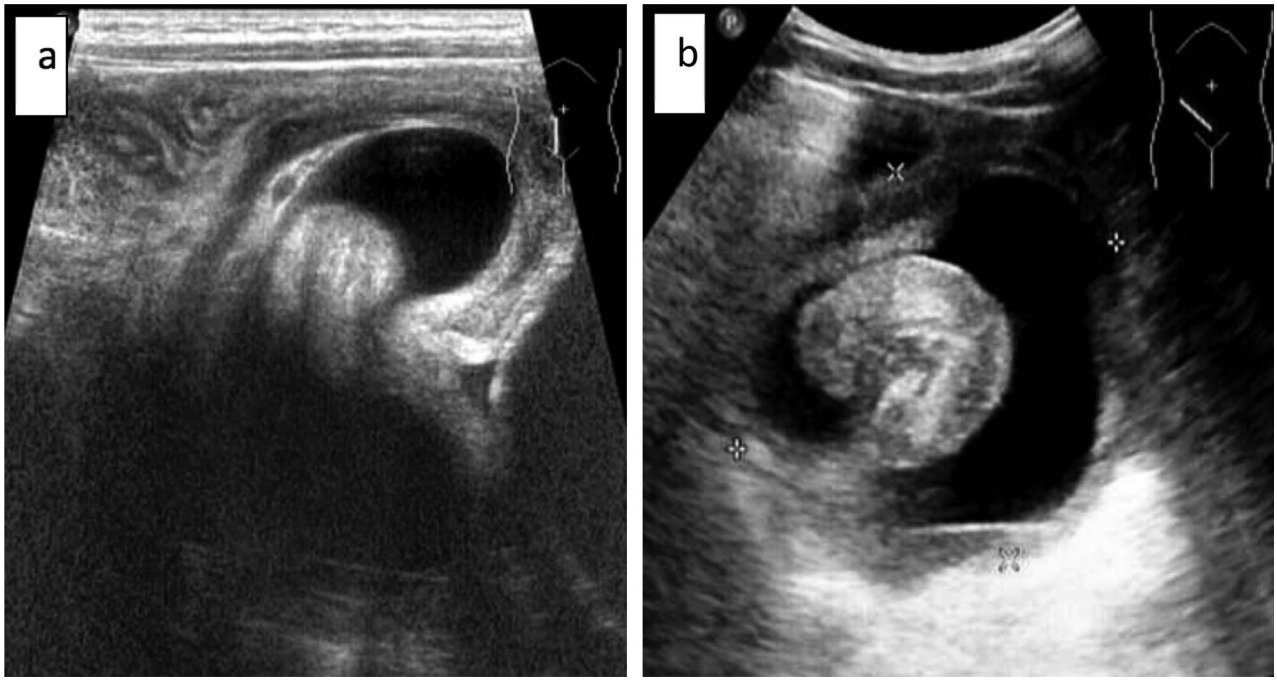

Fig. 1 Abdominal ultrasonic images of intestinal intussusception. Longitudinal (a) and transverse (b) planes views showed a "huge cyst;" inside was the peristaltic hyperechoic bowel. No abdominal mass was found.

Inflammatory myofibroblastic tumor (IMT) is a rare but real tumor, which is histologically characterized by myofibroblastic spindle cells proliferation with inflammatory infiltrate. IMT may occur throughout the body in both children and adults, but the lung is the most common affected organ.[1–3] Its clinical symptoms and signs are also diverse according to the location of the tumor. Patients with intestinal IMT (I-IMT) usually present with nonspecific symptoms, such as fever, anemia, abdominal pain, abdominal mass, loss of appetite and weight, altered bowel habits, fecal occult blood positive, and occasionally bowel obstruction. However, the tumor in the gastrointestinal tract accompanied by intussusception appears to be rare.[3–15]

We documented this rare case of a 12-year-old girl presenting with abdominal pain. The clinical and radiologic impression was bowel intussusception and bowel obstruction. However, no sign of abdominal mass was found before surgery, neither physical examination nor radiologic images. Operative findings revealed intestinal intussusception secondary to a little mass. Histopathlogic evaluation of this mass revealed IMT, which originated in the bowel wall of the ileum.

Case Report

A 12-year-old girl was admitted to our hospital with a 9-day history of vomiting interrupted by abdominal pain and bloating, and no fever. At admission, clinical examination revealed a slightly distended abdomen and mild tenderness on palpation, particularly surrounding the umbilical area. There were no other positive physical findings noted. Laboratory results of routine examination were normal.

Abdominal plain film demonstrated bowel dilation with air fluid levels. Ultrasonographic images of abdomen showed a "huge cyst," and inside was the peristaltic hyperechoic bowel (Fig. 1). An enhanced computed tomography (CT) scan of the abdomen and pelvis revealed extensively dilated small bowel loops with air fluid levels and a multiple concentric ring sign change of the small bowel (Fig. 2). However, all radiologic examinations did not show any mass in both the abdomen and pelvis.

The clinical and radiologic impression was bowel intussusception and bowel obstruction. Air-contrast enema for treatment was initially tried but was not successful. The patient therefore underwent abdominal exploration. The intraoperative findings revealed an intussusception of the small bowel

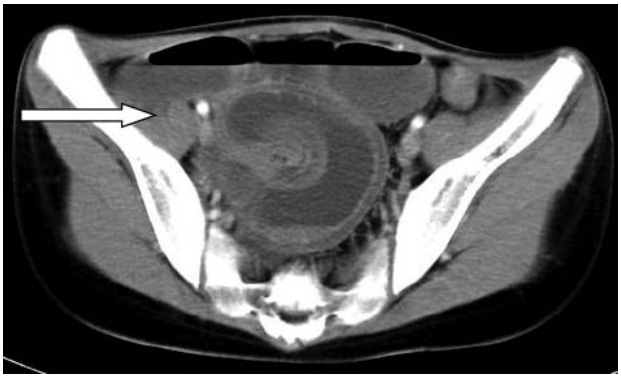

Fig. 2 An enhanced computed tomography (CT) scan of the abdomen and pelvis revealed extensively dilated small bowel loops with air fluid levels and a multiple concentric ring sign change of the small bowel. No abdominal mass was found (arrow).

located in the ileum, 80 cm proximal to the ileocecal junction. The involved segment was black, necrotic, partly perforated, and 30 cm in length. Surprisingly, there was a little mass at the antimesenteric bowel wall of the intussusceptum. On gross examination the mass was found to be hard and have a maximum diameter of about 2 cm (Fig. 3).

The whole mass, including the necrotic bowel, were surgically excised and ileoileal anastomosis was performed in an end-to-end manner. Gross findings of the mass showed gray-white, firm, nodular features measuring 1.5 cm × 1.3 cm ×

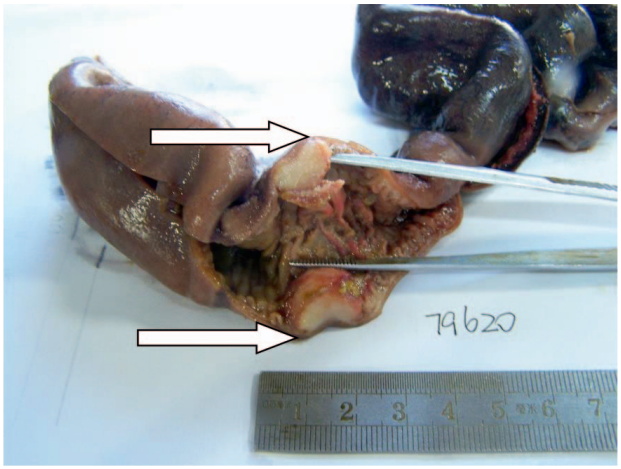

Fig. 3 Gross findings of the mass showed gray-white, firm, nodular features, measuring1.5 cm × 1.3 cm × 1 cm, on the submucosa of the antimesenteric bowel wall (arrow). Involved bowel was black, necrotic, partly perforated, and 30 cm in length.

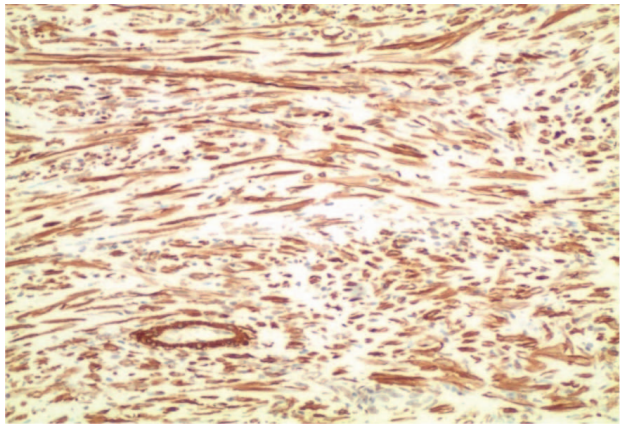

Fig. 4 The expression of smooth muscle actin (SMA) by immunohistochemistry in the spindle cells. Antismooth muscle actin (×100).

1cm, on the submucosa of the antimesenteric bowel wall, 10 cm from the cut margin of the small bowel (Fig.3). Histopathologically, this tumor was composed of spindle-shaped myofibroblastic cells interspersed with inflammatory cells, predominantly lymphocytes. No tumor cells were found in 2 cut margins. The tumor also did not exhibit atypia or mitotic activity. Immunohistochemical analysis revealed that the spindle-shaped cells were positive for smooth muscle actin (SMA) and vimentin (Fig. 4). None were positive for anaplastic lymphoma kinase (ALK), desmin, CD34 and CD117 (C-kit), and revealed a low proliferative index (Ki-67 = 3%).

Discussion

Intestinal intussusception is a common condition in children that is usually primary and benign. In all intussusceptions, frequencies of secondary cases were 1%–3% for children under 5 years old; frequency increased to 14.9% in 5–12 years old, and 37.1% in 13–18 years old. The 3 major causes of secondary intussusception were found to be Schönlein-Henoch purpura, Meckel's diverticulum, and polyps.[16] However, intestinal intussusception caused by inflammatory myofibroblastic tumor (IMT) was rare.[3–15] Intestinal IMT (I-IMT) can be served as a lead point of intussusceptum, cause bowel disorders, and result in intestinal intussusception.

IMT was first described in the lung, and multiple extrapulmonary manifestations have been reported.[2–15] Coffin *et al* showed that the most common sites of extrapulmonary IMT were the mesentery

Table 1 Clinicopathologic features of intestinal inflammatory myofibroblastic tumor with intussusception

Case no.	Study	Sex	Age	Localization of tumor	Diameter of tumor (cm)	Types of intussusception
1–4	Makhlouf[3] *et al*, 2002	NA	NA	NA	NA	NA
5	Milne[4] *et al*, 2006	F	29 y	Ileum	2.5	Ileocolic
6	Zuccarello[5] *et al*, 2006	M	4.5 y	Ileum	2	Ileoileal
7	Ambiru[6] *et al*, 2009	F	36 y	Ileum	4.0	Ileoileal
8	Salameh[7] *et al*, 2011	M	32 mo	Transverse colon	NA	Colocolic
9	Rezaii[8] *et al*, 2011	F	58 y	Ileum	5.5	NA
10	Ntloko[9] *et al*, 2011	M	49 y	Ileum	1.5	Ileoileal
11	Ida[10] *et al*, 2013	M	79 y	Ileum	3.5	Ileal
12-13	Gupta[11] *et al*, 2013	F	51 y	Descending colon	NA	NA
		F	28 y	Ascending colon	NA	NA
14	Walia[12] *et al*, 2014	F	10 y	Ascending colon	5	Ascending colon
15	Appak[13] *et al*, 2014	F	7 y	Ascending colon	5.5	Colocolic
16	Waszak[14] *et al*, 2015	M	57 y	Ileum	6.3	Ileocolic
17	Dulskas[15] *et al*, 2016	F	42 y	Jejunum	5.1	NA

NA, data not available.

and omentum in 36 of 84 cases (43%); only 1 case(1.2%) exhibited an IMT of ileal origin.[2] In the presented case, a tumor arose from the bowel wall of the ileum and appeared to be rare.

There were few cases of I-IMT with intussusception in the literature, as listed in Table 1. These tumors occur in both male and female patients ranging in age from 32 months to 79 years. An I-IMT with intussusception can involve the ileum and colon, the same location as the primary intussusception. The tumors range in size from 1.5 cm to 6.3 cm in diameter. Among the I-IMTs with intussusception in the literature, the tumor for our case was one of the 2 smallest IMT,[3–15] which measured 1.5 cm in its largest diameter. The types of intussusception include ileoileal, ileocolic, and colocolic intussusception.[3–15]

IMT located in the abdominal cavity often can become quite large in size and can be detected as an abdominal mass upon palpation or radiologic imaging, though often nonspecific.[3–15] Unfortunately, before surgery, all means of routine examinations we performed, including abdominal plain film, enhanced computed tomography (CT), ultrasonography, and other laboratory analysis, failed to detect this insidious tumor of the abdomen. Ntloko *et al* reported 5 patients with I-IMT, ranging in size from 1.5 cm to 20 cm in their largest diameters, and 1 patient was an incidental finding during laparotomy for a traumatic insult (tumor size 1.5 cm × 1 cm × 0.5cm).[9] Although speculative, the missed diagnosis of this tumor in our patient may be a function of the smaller size (1.5 cm × 1.3 cm × 1 cm) and the predominant submucous-based growth.

Additionally, in our young case, ultrasonographic images did not show the classic appearance of an intussuscepted bowel, including the "target sign" (transverse plane), the "pseudo-kidney sign" or "sandwich appearance" (longitudinal plane), only a "huge cyst" (extensively dilated bowel with full of fluid) and inside was the peristaltic hyperechoic bowel (compressed bowel of the intussusceptum). These differed from previously described patients with the same diagnosis.[3–15]

The case we present showed some of the typical histomorphology of IMT, namely myofibroblastic proliferation and a varying degree of inflammatory cells infiltrate, predominantly of lymphocyte type.[1–3] Tumor cells were limited to the submucosa of the bowel wall and did not invade into the mucosa, muscularis propria, or serosa. Immunohistochemistry analysis showed strong positive for SMA, vimentin, and negative for ALK, desmin, CD34 and CD117 (C-kit), corresponding with the myofibroblastic nature of these cells, and essentially ruled out other gastrointestinal stromal tumors. These findings and immunohistochemistry were strongly favoring the diagnosis of IMT in our case.

World Health Organization (WHO) classified IMT as a tumor of intermediate biologic potential due to a tendency of local recurrence and mild risk of distant metastasis.[17] Intraabdominal IMT has a highest recurrence rate of approximately 40%.[2] It is mainly agreed that surgery is the predominant

treatment modality for IMT. The present patient underwent a clear margin surgical excision with no further adjuvant therapy. There was no clinical evidence of local recurrence or distant metastasize on follow-up for 1 year.

In conclusion, the rare site of the IMT (ileum), the rare complications of this tumor (intussusception), and the atypical presentation of this case (nonclassical sign of intussusception, insidious abdominal mass) were the most interesting findings in our case. It should always be kept in our mind is that I-IMT may present with bowel intussusception; hence, detailed history, physical examination, and imaging studies are necessary for early recognition and diagnosis.

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