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Pediatric colonic inflammatory myofibroblastic tumor presenting as colo-colonic intussusception: A case report and review of the literature

Jingjing Li Sherman [a], Armine Darbinyan [b], Margret S. Magid [b], Phat Ong [c], Barry Weissman [d], Keith Benkov [e], Aaron M. Lipskar [a,*]

[a] Division of Pediatric General and Thoracic Surgery, Department of Surgery, The Icahn School of Medicine at Mount Sinai, New York, NY, USA
[b] Department of Pathology, The Icahn School of Medicine at Mount Sinai, New York, NY, USA
[c] Hackensack Radiology Group, Hackensack, NJ, USA
[d] All Bergen Pediatrics, Hackensack, NJ, USA
[e] Department of Pediatric Gastroenterology, The Icahn School of Medicine at Mount Sinai, New York, NY, USA

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ABSTRACT

Inflammatory myofibroblastic tumors (IMTs) are rare tumors of intermediate biologic potential most often reported in children. Histologic characteristics of IMT include spindle cell proliferation with chronic inflammatory cell infiltrate. IMTs can occur anywhere in the body but are most commonly reported in lung, bowel mesentery, and liver. Nonmesenteric alimentary IMTs are exceedingly rare. We present the second case of colonic IMT presenting as colo-colonic intussusception in a child. The patient is a 12-year-old female who presented with vague abdominal pain for three months. Her workup revealed an intraluminal mass in the descending colon on both CT and colonoscopy. Intraoperatively, the mass was seen causing colo-colonic intussusception. Laparoscopic segmental colon resection was performed, and the patient did well postoperatively. Histology and immunohistochemistry of the mass confirmed IMT.

Inflammatory myofibroblastic tumors (IMTs) are solid tumors of intermediate biologic potential, most often reported in children [1]. They are referred to in the literature by many names, including inflammatory pseudotumors, pseudosarcomatous myofibroblastic tumors, fibromyxoid lesions, and plasma cell granulomas. Histologic characteristics of IMT include spindle cell proliferation with chronic inflammatory cell infiltrate.

IMTs can occur anywhere in the body but most commonly are found in the lung, bowel mesentery, and liver [1–4]. Nonmesenteric alimentary IMTs are rare; reported sites include gastroesophageal junction, small bowel, appendix, colon, and rectum [2,3,5–7]. First described in 1999, nonrectal colonic IMT is exceedingly rare, and only 23 have been reported in the literature [2–18]. Three colonic IMTs causing intussusception have been reported, only one of which was in a child [6,7,18]. We present the second case report of a colonic IMT presenting as colo-colonic intussusception in a child.

1. Case report

This is the case of a 12-year-old female with no significant past medical history and no significant past family history who had worsening abdominal pain for three months. Pain was periumbilical and episodic. She had constipation interspersed with bloody diarrhea and experienced a nine-pound weight loss. Her abdominal exam was nonspecific. Hemoglobin was normal at 12 g/dL, and all other laboratory workup was normal. She underwent an ultrasound of her abdomen and pelvis that revealed intussusception in the left abdomen (Fig. 1).

A CT scan was performed that showed a well-circumscribed, enhancing mass in the left colon (Fig. 2).

She then underwent an esophagogastroduodenoscopy that was normal, followed by a colonoscopy that showed nearly obstructing submucosal tumor, encompassing over half of the circumference of the descending colon (Fig. 3).

Attempts at endoscopic resection were unsuccessful due to the broad base of the lesion, so it was tattooed and biopsied. These limited endoscopic biopsies revealed normal mucosa with rare spindle cells.

* Corresponding author. Division of Pediatric Surgery, Department of Surgery, Icahn School of Medicine at Mount Sinai, 1 Gustave L. Levy Place, Box 1259, New York, NY 10029, USA. Tel.: +1 212 241 1602; fax: +1 212 410 0111.
E-mail address: Aaron.Lipskar@mountsinai.org (A.M. Lipskar).

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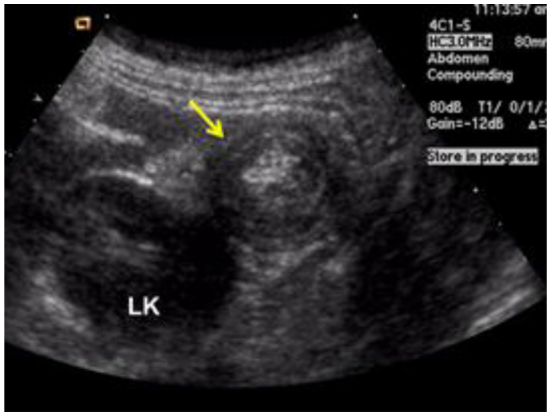

Fig. 1. Ultrasound image of the left abdomen revealing target sign (arrow) of intussusception just overlying the left kidney. LK is left kidney.

The patient then underwent diagnostic laparoscopy, which showed colo-colonic intussusception at the area of the mass in the proximal descending colon. The intussusception was reduced laparoscopically. A laparoscopic-assisted segmental colonic resection with a primary anastomosis was then performed. The mass and adjacent colon were resected with grossly negative margins. She recovered well postoperatively and was discharged home on postoperative day 6 tolerating regular diet and having bowel function. She was seen at follow-up at six months and eleven months with complete resolution of presenting symptoms. Eleven months post-operatively, she underwent surveillance colonoscopy with endoscopic mucosal biopsies at site of resection which were negative for tumor.

Macroscopically, the specimen measured 3.0 × 2.7 × 2.1 cm and was a firm, round mass with smooth, tan-pink mucosal surface. The lesion protruded into and obstructed 70% of the lumen of the colon (Fig. 4).

Microscopic evaluation showed a submucosa-based, well-circumscribed tumor composed of interweaving fascicles of spindle cells admixed with mild inflammatory infiltrates of lymphocytes, plasma cells, and eosinophils (Fig. 5).

Mitotic figures and areas of necrosis were not identified. The tumor projected under the focally necrotic and eroded mucosa, and infiltrated into the underlying muscularis propria. The surgical margins of resection and two pericolic lymph nodes were negative for tumor. On immunohistochemical studies, tumor cells were positive for smooth muscle actin, desmin, muscle specific actin, and calponin and negative for DOG1, c-kit, CD34, S100, ALK-1, pan-cytokeratin, and S100-consistent with IMT (Fig. 6).

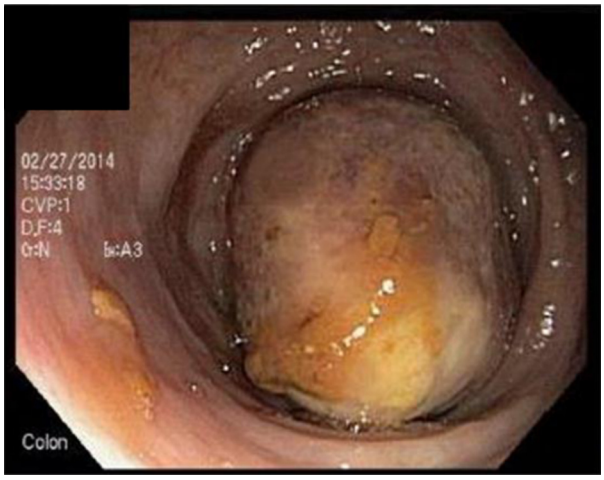

Fig. 3. Colonoscopy showing nearly obstructing submucosal mass in the descending colon.

Rearrangements of the *ALK* gene were not detected in this tumor by FISH.

2. Discussion

Intussusception causes abdominal pain and obstruction most commonly in young children between six months and three years of age [19–21]. Intussusceptions are often classified as primary (idiopathic) or secondary. The vast majority of pediatric intussusception cases are idiopathic, where the lead point is attributed to hypertrophied Peyer's patches in the terminal ileum, leading to ileo-colonic intussusception [20]. Hypertrophy of Peyer's patches has been associated with rotavirus immunization, adenovirus infections, and bacterial enteritis [22]. Secondary intussusception, caused by an identifiable lesion that serves as a lead point, is rare, constituting 1.5–12% of all intussuceptions. The most common lead points are Meckel's diverticula, appendices, hamartomas, lipomas, and polyps.

Colo-colonic intussusceptions are extraordinarily rare and most commonly classified as secondary intussusceptions, where colonic masses act as lead points. They are more likely to be found in adults as they are due to well-defined masses such as malignancies or large polyps. Colo-colonic intussusceptions in the pediatric population are almost always due to a colonic mass [23]. The differential diagnosis for pediatric colonic masses includes polyps, duplication cysts, gastrointestinal stromal tumors, leiomyomas, inflammatory

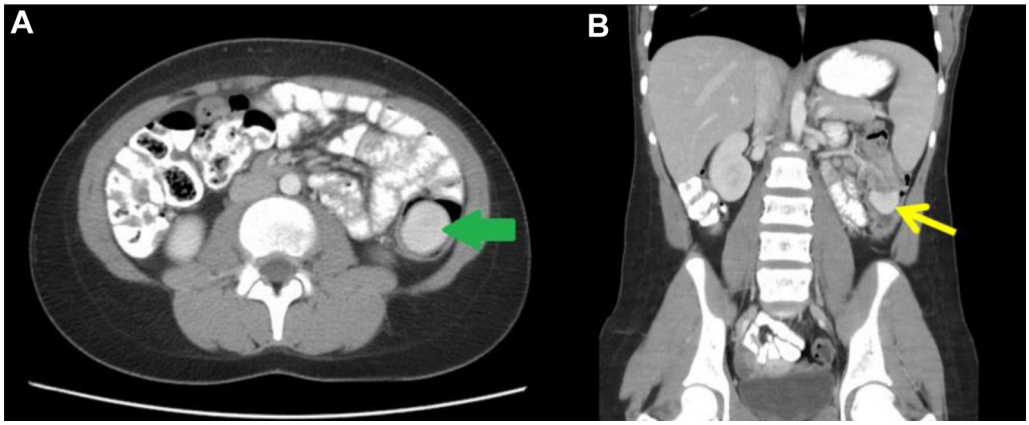

Fig. 2. Axial image (A) and coronal reconstruction (B) of an enhancing mass in the lumen of the descending colon (arrows).

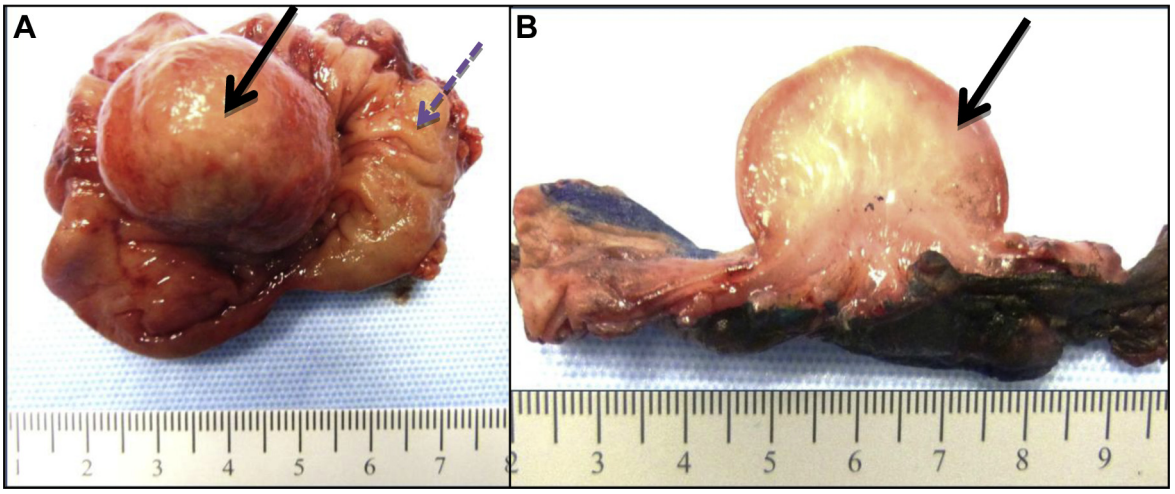

Fig. 4. (A) Gross image of the resected round sessile mass (3.0 cm greatest dimension) covered by a smooth tan-pink mucosal surface and protruding into the lumen of the colon (solid arrow). The mucosal surface of the adjacent bowel is a normal tan-pink with preserved folds (dashed arrow). (B) Following bisection, the tumor is seen projecting under the mucosa and infiltrating into the underlying muscularis propria (arrow).

masses, adenocarcinomas, IMTs, and ganglioneuromas [24–26]. Colo-colonic intussusception can present with vague symptoms including abdominal pain, nausea, emesis, constipation, diarrhea, and rectal bleeding [27].

IMTs can be found in the lung, mesentery, liver, and alimentary tract; nonmesenteric colonic IMTs are rare. After an extensive literature research, 23 case reports of nonrectal colonic IMT were found, of which only six cases were pediatric [3–7,9–16]. Fourteen of these 23 reported cases in the literature contain information on presentation, diagnosis, and treatment. Table 1 summarizes these 14 patients as well as our own, and these are the patients that will be used for discussion.

Diagnosis of colonic IMT is difficult as patients may present with vague abdominal symptoms [17]. In the 14 patients with colonic IMT in Table 1, the most common symptoms of colonic IMT include abdominal pain (57%), bloody stools (29%), and weight loss (29%). Two patients (14%) were asymptomatic, and IMT was picked up in these patients incidentally or on screening. Anemia was noted in eight of the twelve patients (67%) where hemoglobin was reported.

Imaging such as ultrasound, CT, or MRI may identify an abdominal mass of unclear origin or colonic mass. Barium enema and colonoscopy may reveal colonic mass. In the fourteen patients with colonic IMT, five were found in the ascending colon, four originated from the transverse colon, four from the descending colon, and one from the sigmoid colon.

IMT is considered a myofibroblastic tumor of intermediate biologic potential. Microscopically, IMT is composed of spindle-shaped cells resembling fibroblasts/myofibroblasts set in a fibromyxoid stroma with inflammatory cellular infiltrate composed of varying numbers of plasma cells, lymphocytes and histiocytes. Immunohistochemical studies show that most IMT can have strong diffuse cytoplasmic immunoreactivity for vimentin and variable reactivity for smooth muscle actin, muscle specific actin and desmin [8,10,17] The spindle cells do not exhibit reactivity for S-100. CD117 (c-kit) and CD34 are expressed in endothelial cells, highlighting angiogenesis. Inflammatory cells can be identified using specific markers, including CD3 (T-lymphocytes), CD20 (B-lymphocytes), CD138 (plasma cells) and CD68 (histiocytes) [10]. Approximately 60% of cases harbor rearrangement of the *ALK* gene on chromosome 2p23 and show positive cytoplasmic immunostaining for ALK-1 [28–30]. Interestingly, ALK-negativity has been associated with an increased likelihood of metastasis and less favorable prognosis as has a proliferative growth pattern with increased mitoses (which was not identified in this patient's tumor) [9,31].

Treatment of colonic IMT is surgical resection. There has not been a demonstrated benefit of chemotherapy or radiation in the literature [4]. All 14 of the reported patients were treated surgically. Six segmental resections were performed when a discrete mass was encountered. Five out of the six segmental resections were in pediatric patients. Anatomic resections were performed in six patients

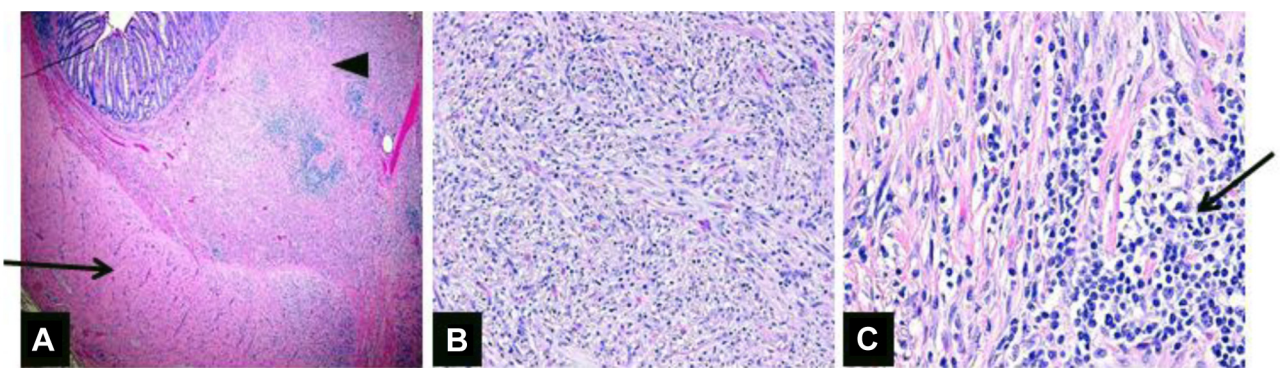

Fig. 5. Histological appearance of the inflammatory myofibroblastic tumor (IMT). (A) Low power view of the IMT (arrowhead) within the colonic wall (mucosa in upper left; muscularis propria below [arrow]). (B and C) IMT at higher magnification. Proliferating spindle cells arranged in an interweaving fascicular pattern. The nuclei are elongated and regular. Mitotic figures and areas of necrosis are not identified. Mild inflammatory infiltrates of lymphocytes and plasma cells are interspersed throughout the tumor (arrow).

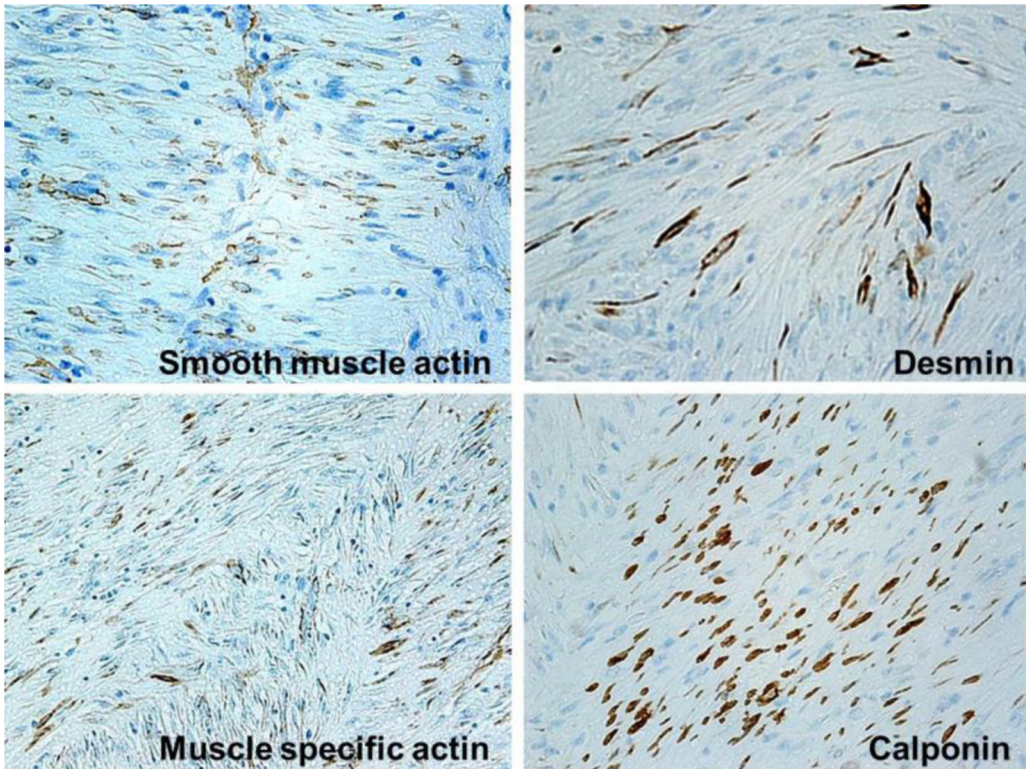

Fig. 6. Immunohistochemical results. Tumor cells are positive for smooth muscle actin (SMA), desmin, muscle specific actin and calponin, and are negative for DOG-1, c-kit, CD34, S100, ALK-1 and pan-cytokeratin.

Table 1
Summary of 14 case reports, including this case report, with colonic IMT.

Age (years) Sex	Source	Symptoms at presentation	Hemoglobin (mg/dL)	Significant diagnostic tests and results	Location of IMT	Treatment
2 M	Salameh et al. [6]	Abdominal pain, constipation, bloody stools	7.3	Colonoscopy–sigmoid mass CT–intussusception	Sigmoid colon	Open segmental resection
3 F	Velitchkov et al. [12]	Incidentally found at surgery for hydatid cyst	Normal	US and CT–5 cm hydatid cysts	Mid transverse colon	Open segmental resection
7 F	Cviko A et al. [15]	Abdominal pain	9	US–LLQ mass	Ileum, cecum, ascending colon	Open segmental resection
9 F	Salameh et al. [6]	Chest pain, lethargy	"Anemia"	CT & US–mass near bladder	Ascending colon	Open segmental resection
11 M	Karnak et al. [4]	Weight loss	n/a	CT–mass in pelvis	Descending colon anti-mesenteric wall	Open total excision
11 F	Sanders et al. [3]	Abdominal pain, distention, fevers	n/a	Initial CT–Pelvic mass, possible abscess Repeat CT at 4 months–mass in left upper quadrant	Appendix and left colon	• Initial surgery–drainage of abscess • Second surgery at 2 months–appendectomy, omentectomy • Third surgery at 4 months–Open left and sigmoid colon resection
12 F	Sherman et al.	Abdominal pain, bloody stools, weight loss	12	US–intussusception CT/colonoscopy–descending colonic mass	Proximal descending colon	Laparoscopic segmental resection
30 M	Aalbers et al. [5]	Abdominal pain	7.3	US–right cystic mass MRI–8 cm solitary mass next to right kidney and liver	Transverse colon	Open segmental resection
30 F	Jeong et al. [7]	Abdominal pain	9.2	Colonoscopy–5 cm mass PET/CT- SUV 7.8	Distal transverse colon	Laparoscopic left hemicolectomy
35 M	Kim et al. [11]	Bloody stools	14.5	Colonoscopy/CT–descending colon mass	Descending colon	Laparoscopic anterior resection

(*continued on next page*)

Table 1 (*continued*)

Age (years) Sex	Source	Symptoms at presentation	Hemoglobin (mg/dL)	Significant diagnostic tests and results	Location of IMT	Treatment
38 M	Velitchkov et al. [12]	Fevers Abdominal mass	10.0	US–cystic structure in left abdomen CT–18 cm mass in left upper abdomen	Distal transverse colon extending into stomach and spleen	Open extended left hemicolectomy, partial gastrectomy, splenectomy
56 M	Gurzu et al. [17]	Abdominal pain Bloody stool Weight loss	11	Colonoscopy–mass in ascending colon	Ascending colon	Open right hemicolectomy
68 M	Erkan et al. [16]	Abdominal pain, weight loss	10.5	CT/barium enema–mass in right colon	Ascending colon	Open right hemicolectomy
79 M	Tanaka et al. [13]	Positive screening fecal occult blood test.	"Normal"	Enema–ascending colon protrusion Colonoscopy–sessile mass	Ascending colon	Open right hemicolectomy

M: Male, F: Female, US: Ultrasound, LLQ: Left lower quadrant.

over age of 30 where adenocarcinoma was suspected. One patient underwent complete excision of the mass as it was on the anti-mesenteric side of the descending colon and did not require segmental resection to remove the tumor in its entirety [4]. Two patients who had extensive disease required en bloc resection of nearby structures [12]. One patient, an 11-year-old girl, had a complicated course in which three surgeries were required for diagnosis and treatment [3]. She also had a recurrence that resolved on its own without further adjuvant therapy or surgeries. No long-term recurrences were found in any of the 14 reported patients.

3. Conclusions

Colonic IMT is very rare. Symptoms associated with colonic IMT include abdominal pain, bloody stools, and weight loss. Patients may also present with anemia and colo-colonic intussusception. Treatment is segmental resection with grossly negative margins and debulking of local invasion. There is no proven role for chemotherapy or radiation therapy.

Prognosis seems to be excellent and resection curative, but longer follow-up is needed to accurately know if colonic IMTs can recur after segmental resection. Close follow-up is also indicated in this ALK-negative tumor, which has been associated with a less favorable prognosis.

Financial disclosure and conflict of interest statement

The authors have no financial disclosures or conflicts of interests.

References

[1] Souid AK, Ziemba M, Dubansky AS, Mazur M, Oliphant M, Thomas FD, et al. Inflammatory myofibroblastic tumor in children. Cancer 1993;72:2042–8.
[2] Kovach SJ, Fischer A, Katzman PJ, Salloum RM, Ettinghause SE, Madeb R, et al. Inflammatory myofibroblastic tumors. J Surg Oncol 2006;94:385–91.
[3] Sanders BM, West K, Gingalewski C, Engum S, Davis M, Grosfeld JL. Inflammatory pseudotumor of the alimentary tract: clinical and surgical experience. J Pediatr Surg 2001;36:169–73.
[4] Karnak I, Senocak M, Ciftci AO, Caglar M, Bingol-Kologlu M, Tanyel FC, et al. Inflammatory myofibroblastic tumor in children: diagnosis and treatment. J Pediatr Surg 2001;36:908–12.
[5] Aalbers AG, De Wilt J, Zondervan PE, Ijzermans JN. A colon-derived inflammatory pseudotumor. Dig Dis Sci 1999;44:578–81.
[6] Salameh M, Sultan I, Barbar M, Al Hussaini M, Jameel A, Ghandour K, et al. Inflammatory myofibroblastic tumor causing unexplained anemia in a toddler: a case report. J Med Case Rep 2011;5:69.
[7] Jeong JH, Cho I, Kong EJ, Chun KA, Kim YJ, Kim JH. (18)F-FDG PET/CT in inflammatory pseudotumor of the colon causing intussusception. Ann Nucl Med 2011;25:447–50.
[8] Makhlouf HR, Sobin L. Inflammatory myofibroblastic tumors (inflammatory pseudotumors) of the gastrointestinal tract: how closely are they related to inflammatory fibroid polyps? Hum Pathol 2002;33:307–15.
[9] Chun YS, Wang L, Nascimento AG, Moir CR, Rodeberg DA. Pediatric inflammatory myofibroblastic tumor: anaplastic lymphoma kinase (ALK) expression and prognosis. Pediatr Blood Cancer 2005;45:796–801.
[10] Coffin CM, Alaggio R. Fibriblastic and myofibroblastic tumors in children and adolescents. Pediatr Dev Pathol 2012;15:127–80.
[11] Kim EY, Lee I, Lee YS, Yang N, Chung DJ, Kim KI, et al. Inflammatory myofibroblastic tumor in colon. J Korean Surg Soc 2012;82.
[12] Velitchkov N, Losanoff J, Kjossev K, Michaylova V. Inflammatory pseudotumor of the colon. Dig Dis Sci 2004;2000:515–6.
[13] Tanaka A, Hirabayashi K, Sadahiro S, Maeda Y, Suzuki T, Ogoshi K. Inflammatory myofibroblastic tumor of the ascending colon in adults manifested by positive fecal occult blood test. Gastrointest Endosc 2010;71:214–6.
[14] Biselli R, Ferlini C, Fattorossi A, Boldrini R, Bosman C. Inflammatory myofibroblastic tumor (inflammatory pseudotumor): DNA flow cytometric analysis of nine pediatric cases. Cancer 1996;77:778–84.
[15] Cviko A, Milic Z, Cizmic A, Seiwerth S, Kruslin B. Inflammatory myofibroblastic tumor with extensive involvement of the bowel in a 7-year-Old child. Croat Med J 1999;40:550–3.
[16] Erkan N, Yildirim M, Yilmaz C, Yagci A. Inflammatory pseudo-tumor as an unusual case of colonic obstruction: a case report. Acta Chir Belg 2004;104.
[17] Gurzu S, Bara T, Jung I. Inflammatory myofibroblastic tumor of the colon. J Clin Oncol 2013;31:e155–8.
[18] Memba IR, Lamas MS, Ramos RE, Climent EMJ. Adult colonic intussusception secondary to inflammatory myofibroblastic tumor. Cir Esp 2007;81.
[19] Mandeville K, Chien M, Willyerd FA, Mandell G. Intussusception: clinical presentations and imaging characteristics. Pediatr Emerg Care 2012;28:842–4.
[20] Stringer MD, Pablot SM, Brereton RJ. Paediatric intussusception. Br J Surg 1992;79:867–76.
[21] Fischer TK, Bihrmann K, Perch M, Koch A, Wohlfahrt J, Kare M, et al. Intussusception in early childhood: a cohort study of 1.7 million children. Pediatrics 2004;114:782–5.
[22] Montgomery EA, Popek E. Intussusception, adenovirus, and children: a brief reaffirmation. Hum Pathol 1994;1994:2.
[23] Mahmudloo R, Gheibi S, Vahed SN. Colocolic intussusception without lead point; a case report and literature review. Iran J Pediatr 2008;18:373–6.
[24] Arthur AL, Garvey R, Vaness DG. Colocolic intussusception in a three-year-old child caused by a colonic polyp. Conn Med 1990;54:492–4.
[25] Abrahams RB, Franco A, Lewis KN. Pediatric colocolic intussusception with pathologic lead point: a case report. J Med Cases 2012;3:84–8.
[26] Soccorso F, Puls F, Richards C, Pringle H, Nour S. A ganglioneuroma of the sigmoid colon presenting as leading point of intussusception in a child: a case report. J Pediatr Surg 2009;44:e17–20.
[27] Yakan S, Caliskan C, Makay O, Denecli AG, Korkut MA. Intussusception in adults: clinical characteristics, diagnosis and operative strategies. World J Gastroeneterol 2009;15:1985–9.
[28] Coffin CM, Patel A, Perkins S, Elenitoba-Johnson KS, Perlman E, Griffin CA. ALK1 and p80 expression and chromosomal rearrangements involving 2p23 in inflammatory myofibroblastic tumor. Mod Pathol 2001;14:569–76.
[29] Cook JR, Dehner L, Collins MH, Ma Z, Morris SW, Coffin CM, et al. Anaplastic lymphoma kinase (ALK) expression in the inflammatory myofibroblastic tumor: a comparative immunohistochemical study. Am J Surg Pathol 2001;25:1364–71.
[30] Sirvent N, Hawkins A, Moeglin D, Coindre JM, Kurzenne JY, Michiels JF, et al. ALK probe rearrangement in a t(2;11;2)(p23;p15;q31) translocation found in a prenatal myofibroblastic fibrous lesion: toward a molecular definition of an inflammatory myofibroblastic tumor family? Genes Chromosomes Cancer 2001;31:85–90.
[31] Coffin CM, Hornick J, Fletcher CD. Inflammatory myofibroblastic tumor: comparison of clinicopathologic, histologic, and immunohistochemical features including ALK expression in atypical and aggressive cases. Am J Surg Pathol 2007;31:509–20.