J Ped Surg Case Reports 2 (2014) 290–293

Contents lists available at ScienceDirect

Journal of Pediatric Surgery CASE REPORTS

journal homepage: www.jpscasereports.com

Infantile fibrosarcoma of the intestine: A report of two cases and literature review☆

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ARTICLE INFO

Article history:
Received 4 April 2014
Received in revised form
28 May 2014
Accepted 1 June 2014

Key words:
Infantile fibrosarcoma
Small bowel tumor
Asymptomatic abdominal mass

ABSTRACT

Infantile fibrosarcoma (IF) is a rare tumor presenting in infants and young children that most commonly develops in the extremities. Intestinal IF has been rarely described, with all previous cases presenting with obstruction, perforation, or both. We present two cases of intestinal IF recently managed at our institution. The first was a previously healthy 4-month old female with an incidentally discovered abdominal mass. This tumor was found to arise from the jejunum with no additional visceral involvement. The second was a 15-day old female with a distal small bowel mass that resulted in an ileal obstruction. Pathologic evaluation of both tumors revealed a myxoid spindle cell histology, and genetic analyzes demonstrated the t(12; 15) translocation, confirming IF. We discuss the presentations of these patients, and describe the recommended treatment strategies for patients with intestinal IF.

Infantile fibrosarcoma (IF) is a rare, non-rhabdomyosarcomatous soft tissue sarcoma that typically presents in infants and children before 5 years of age [1–3]. Most tumors arise from the extremities or the trunk, although a small number of case reports describing IF arising from the bowel have been documented (Table 1) [4–13]. These cases have all presented with intestinal obstruction or perforation [5–13]. We present here two cases of small bowel infantile fibrosarcoma, one of which is the first reported presentation as an asymptomatic, incidentally discovered abdominal mass.

1. Case report–patient 1

A previously healthy 4 month old female was noted to have an abdominal mass by her primary care provider at a routine well-child visit. Her prenatal course had been unremarkable and she was delivered at term by Caesarean-section for breech position. She had been eating well and gaining appropriate weight. On physical examination she had a palpable, firm, non-tender mass occupying the right hemi-abdomen, without any identifiable lymphadenopathy or other pertinent findings. An ultrasound was obtained identifying a heterogenous mass of unclear origin, appearing to be distinct from the liver, kidneys, and spleen. Further cross-sectional imaging with computed tomography of the chest, abdomen, and pelvis identified a heterogenous right-sided abdominal mass measuring 6.9 × 8.1 × 8.3 cm in size, possibly arising from the ovary (Fig. 1). Tumor markers were obtained and included alpha fetoprotein (77.1 ng/mL; reference <87 ng/mL) [14], beta-human chorionic gonadotropin (<2.0 mIU/mL; reference <10 mIU/mL), and urine catecholamines (vanillylmandelic acid to creatinine ratio: 9.4; homovanillic acid to creatinine ratio: 24.6; references: <20.1 and <30.1, respectively), all of which were within age-based normal limits.

Surgical consultation was obtained, and she underwent exploration of her abdomen via a Pfannenstiel skin incision. Clear peritoneal fluid was collected and sent for cytology. Upon visual inspection, all solid organs including ovaries appeared normal. The small bowel was eviscerated and the mass was identified arising from the mid-jejunum along the anti-mesenteric surface. This portion of the jejunum was resected with 3 cm margins and a primary stapled anastomosis was performed (Fig. 2). Her post-operative course was uneventful and she was discharged home on post-operative day 3.

Subsequent pathology described a myxoid spindle cell neoplasm primarily invading the muscularis propria with a mitotic count of

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http://dx.doi.org/10.1016/j.epsc.2014.06.003

Table 1
Previously reported cases of intestinal infantile fibrosarcoma.

Study	Age	Gender	Presentation	Location	Treatment/outcome
Wright, 1971 [5]	16 months	M	Vomiting without obstruction	Duodenum	Pancreaticoduodenectomy, well at 24 months
Shearburn et al., 1975 [6]	1 day	F	Obstruction	Duodenum	Pancreaticoduodenectomy, well at 14 months
Sherman and Neustein, 1985 [7]	1 day	M	Obstruction	Ileum	Resection, well at 3 years
Shima et al., 2003 [8]	1 day	F	Perforation, meconium peritonitis	Jejunum	Resection, full recovery
Buccoliero et al., 2008 [9]	3 days	M	Perforation	Ascending colon	Ileocolectomy, well at 12 months
Islam et al., 2008 [10]	0 days	M	Meconium peritonitis	Descending colon	Resection, well at 5 years
van Niekerk et al., 2010 [11]	2 days	M	Obstruction	Ileum	Ileocecectomy, well at 9 months
Rizkalla et al., 2011 [12]	5 days	M	Perforation	Ileum	Resection, well at 12 months
Kim et al., 2013 [13]	2 days	M	Pneumo-peritoneum	Sigmoid colon	Sigmoid colectomy, well at 1 year
Present study – patient 1	4 months	F	Asymptomatic abdominal mass	Jejunum	Resection, full recovery
Present study – patient 2	15 days	F	Obstruction	Ileum	Ileocecectomy, full recovery

5 per 10 high power fields (HPF) and negative margins (Fig. 3). Immunohistochemistry (IHC) was weakly positive for smooth muscle actin (SMA) and CD117. Reverse transcriptase–polymerase chain reaction (RT-PCR) was positive for the t(12; 15) translocation that is commonly identified in IF. There were no tumor cells observed in the peritoneal fluid. Post-operative positron emission tomography computed tomography (PET-CT) did not identify residual tumor, involvement of lymph nodes, or metastatic disease. In the setting of negative margins and the absence of residual or metastatic disease, close observation without adjuvant therapy was deemed the most appropriate course. Her follow up will consist of physical exam and serial imaging for at least two years to monitor for tumor recurrence.

2. Case report–patient 2

A 12-day old term infant with no pregnancy or vaginal delivery complications was admitted for poor feeding, abdominal distension, and dehydration. An abdominal x-ray demonstrated a distal small bowel obstruction, which was confirmed by an upper gastrointestinal contrast study. She was taken to the operating room for abdominal exploration, and was found to have a distal ileal mass resulting in obstruction and focal intestinal ischemia. An ileocecectomy was performed with a primary anastomosis. No other masses or abnormalities were observed during complete exploration of the abdomen. Her post-operative course was marked by slow progress tolerating oral feedings and she was discharged home on post-operative day 9.

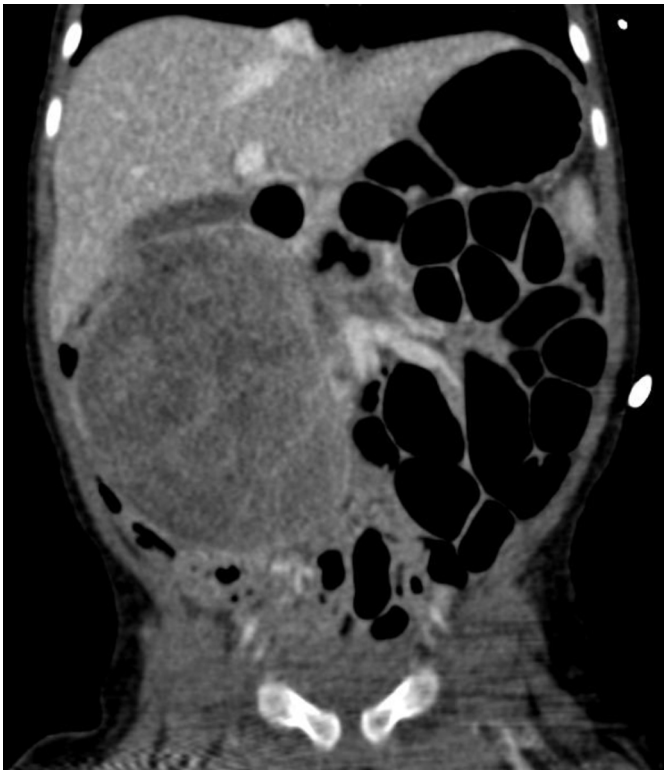

Fig. 1. Computed tomographic image of intra-abdominal mass seen on pre-operative evaluation for Patient 1.

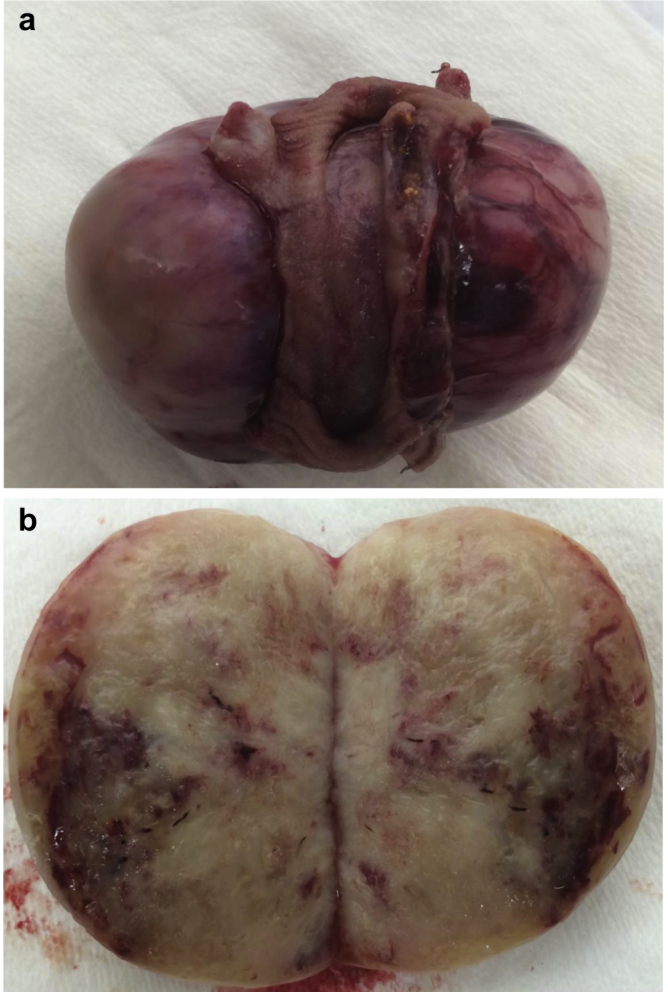

Fig. 2. (a) Segment of jejunum from Patient 1 with tumor [8 × 5.5 × 5.5 cm] arising from anti-mesenteric surface. (b) Cut surface showing a bulging surface composed of firm white-tan whorled fibrous tissue.