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Kaposiform hemangioendothelioma causing intestinal obstruction

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ABSTRACT

A previously healthy toddler with bilious vomiting and erythematous gluteal rash over 2 weeks had intermittent pain, constipation and decreased appetite. All labs were negative with the exception of fecal occult blood. Abdominal x-ray and ultrasound revealed dilated air-filled loops of bowel and partial small bowel obstruction. After persistent worsening abdominal pain and vomiting a CT scan with IV contrast (Fig. 1) suggested small bowel obstruction. Emergent surgery was performed and diagnostic laparoscopy revealed about 61 cm of necrotic bowel causing stricture formation and mesenteric shortening in the distal small bowel. 56 cm of inflamed bowel was resected with end-to-end anastomosis. Final pathology report indicated diffuse intestinal angiomatosis with transmural involvement and focal erosion consistent with KHE (Fig. 2). Presentation is varied, consists of cutaneous lesion, retroperitoneal mass, intestinal obstruction, jaundice, intussusception, or multifocal neoplasms. Complete surgical resection with wide margins is the best therapeutic option and has achieved the best outcomes. If not treated in sufficient time, KHE has a relatively high mortality rate of 30%, with most deaths occurring due to its locally invasive effects [5]. There are limited reports of identifying features of KHE on imaging. Of 165 cases of KHE none were presented in the small bowel [5]. We report the unique case of KHE presenting as a hypervascular mass causing obstruction in the distal small bowel. Although extremely rare, KHE should be considered as a reason for severe GI stricture or obstruction in infants and children in obscure cases and included in the differential.

Abbreviations

CBC	Complete Blood Count
CMP	Comprehensive Metabolic Panel
CRP	C-reactive Protein
CT	Computed Tomography
ED	Emergency Department
KHE	Kaposiform Hemangioendothelioma
O &P	Ova and Parasites
VBG	Venous blood gas

A sixteen-month-old healthy boy with an initial presentation of bilious vomiting and abdominal colic underwent small bowel resection for obstruction. Pathology showed obstruction secondary to kaposiform hemangioendothelioma, a rare vascular tumor that affects mostly children and adolescents in the skin but can also involve the thymus, head, neck, colon, female genitalia, or the choledochal tract.

Kaposiform Hemangioendothelioma (KHE) is a rare, vascular tumor predominantly found in children and adolescents first described by Zuckerberg in 1993 [1]. It is locally extensive and aggressive and is not usually associated with distant metastasis. It most commonly presents as a superficial or deep soft tissue mass of the head and neck, mediastinum and retroperitoneum. In addition, KHE has been shown to involves other organs such as the thymus, pancreas, colon, female genitalia, and in one report, the choledochal tract [2,4,5]. Cases occurring in the small bowel are extremely rare. To our knowledge, there has never been a report of KHE originating in the small intestinal tract of a toddler. Thus, we present a unique case of significant intestinal involvement complicated by a stricture

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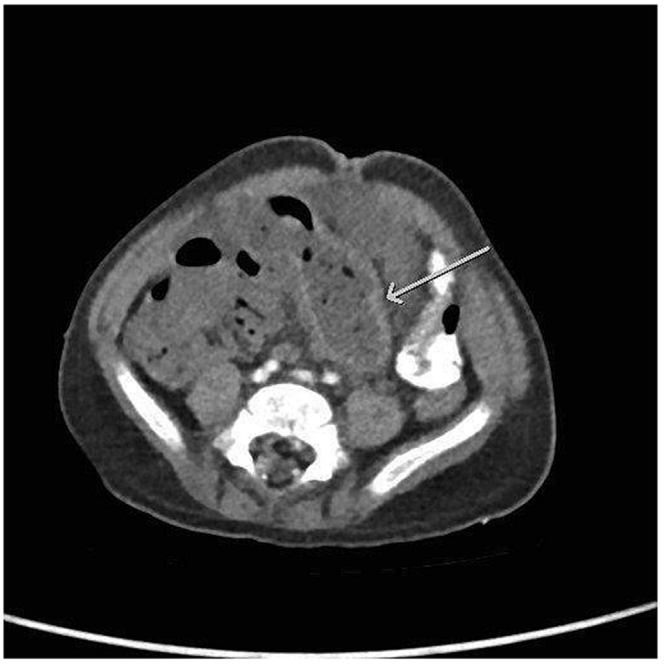

Fig. 1. CT scan with IV contrast showing severe stricture of small bowel.

causing clinical obstruction. We review the available literature and provide guidance regarding evaluation and treatment of this rare malformation.

1. Case report

A 16-month-old previously healthy boy of Middle Eastern descent presented to the E.D. with bilious vomiting and an erythematous gluteal rash. Over the course of 2 weeks the patient was in and out of the unit for abdominal colic, intermittent constipation, and poor oral intake. Labs for CBC, CMP, VBG, lactic acid, stool O&P, stool culture, and CRP were negative. Fecal occult blood and C. Diff toxin results came back positive. Abdominal x-ray series and abdominal ultrasound revealed dilated air-filled loops of bowel and evidence of partial small bowel obstruction. After persistent worsening of abdominal pain and vomiting a CT scan with IV contrast (Fig. 1) suggested small bowel obstruction secondary to an inflammatory process. Stricture was suspected, thus emergent surgery was performed which confirmed severe stricture. Diagnostic laparoscopy revealed about 61 cm of inflamed and thickened bowel causing stricture formation and mesenteric shortening in the distal small bowel (Fig. 2). About 56 cm of inflamed bowel was resected with primary enteroenterostomy with end-to-end anastomosis. Final pathology report indicated diffuse intestinal angiomatosis with transmural involvement and focal erosion consistent with kaposiform hemangioendothelioma (Fig. 3). The patient tolerated the procedure well and was discharged. On follow-up, the patient has shown no signs of tumor recurrence. KHE should be considered in any unexplained small bowel stricture/obstruction on CT scan in the absence of other etiologies such as mass, malrotation or previous surgery so treatment including surgery can be performed on timely manner.

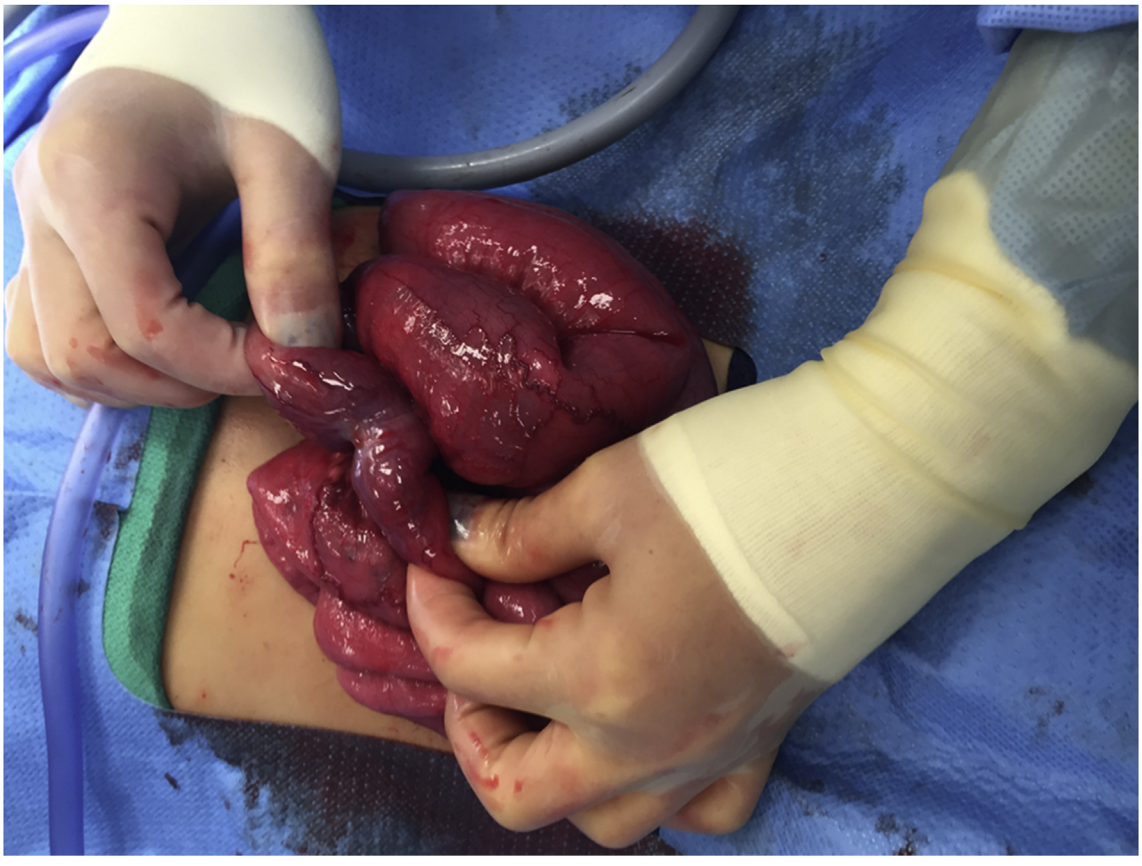

Fig. 2. Laparoscopy revealed about 61 cm of inflamed and thickened bowel causing stricture formation.