

Neonatal Intestinal Obstruction Due to Kaposiform Hemangioendothelioma of the Jejunum

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ABSTRACT

Kaposiform hemangioendothelioma is a borderline vascular tumor usually seen in infants and children as cutaneous lesions classically associated with the Kasabach-Merritt phenomenon. Intestinal involvement is uncommon and can cause acute presentations, such as obstruction or gastrointestinal bleeding. A 5-day-old neonate presented with bilious vomiting for 2 days. The tumor was in the jejunum. Histopathological examination with immunohistochemistry of the resected jejunum showed CD34 positive endothelial lined vascular spaces infiltrating from submucosa to serosa, which is classical of kaposiform hemangioendothelioma. There was no deranged coagulation profile. This case forms an interesting cause for neonatal intestinal obstruction.

INTRODUCTION

Kaposiform hemangioendothelioma (KHE) is a rare type of vascular tumor occurring in infants and children.¹ KHE is classified as a borderline vascular tumor according to the International Society for the Study of Vascular Anomalies with the tendency of local infiltration.² These lesions usually present in cutaneous locations, especially in lower limbs. Other sites described were bones, mediastinum, and retroperitoneum.³ Intestinal (jejunum) occurrence was first described in 2012 and needs early diagnosis and management because of bleeding tendencies and obstruction.⁴ We describe a case of neonatal intestinal obstruction in a 5-day-old neonate due to KHE successfully treated by resection anastomosis.

CASE REPORT

A 5-day-old neonate was brought to the emergency department with multiple episodes of bilious vomiting for 2 days. On examination, abdomen distension was noted in the upper abdomen with a normal anal opening. The abdomen was nontender, and normal bowel sounds were heard. An abdominal x-ray showed proximal small bowel dilatation with air-fluid levels. An upper gastrointestinal contrast study showed a dilated proximal bowel up to the proximal jejunum; however, contrast flow into the distal bowel was present. The patient's laboratory parameters were normal, including normal platelet count and coagulation profile. A provisional diagnosis of neonatal intestinal obstruction was made supported by x-ray findings. Partial obstruction was inferred because of contrast follow-through beyond the dilated proximal small bowel.

Differential diagnoses of jejunal atresia, duplication cyst, and intussusception were kept in mind before exploration. Intraoperatively, there was thickening of the jejunum of an approximately 10-cm segment with a red vascular surface suggestive of a vascular lesion with lesion encroaching on the respective mesentery (Figure 1). The lesion was 10 cm distal to the duodenojejunal flexure, and the bowel proximal to the lesion was dilated. The rest of the bowel was normal. Resection of the abnormal jejunum containing the lesion along with the involved mesentery was performed, and end-to-end anastomosis was made. Histopathology showed an unremarkable mucosal lining with the presence of a diffuse, moderately cellular lesion composed of nodules of spindle-shaped cells extending from submucosa to serosa. These nodules were separated by interspersed dense stromal tissue and slit-like vascular spaces of variable caliber (Figure 2). No nuclear pleomorphism, atypia, or atypical mitotic figures were seen. Margins were free of any tumor involvement. Immunohistochemistry showed positivity for CD34 classical of KHE (Figure 3). The postoperative recovery was

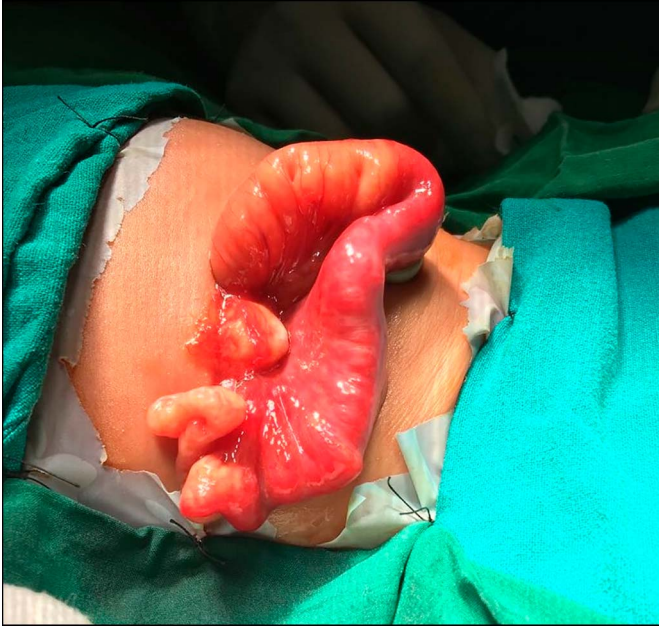


Figure 1. Thickening of the jejunum with a reddish surface including some part of mesentery.

uneventful and was discharged on full oral feeds. Every 6-month follow-up till 3 years was unremarkable with no evidence of any recurrence.

DISCUSSION

In 1993, Zukerberg et al⁵ first described KHE as an entity different from infantile hemangioma because of its locally invasive nature and its focal Kaposi-like appearance. Since then, 183 cases have been reported, but the actual incidence can be more because some asymptomatic cases are less likely to be reported or diagnosed by pathology.⁶ Small intestinal involvement has

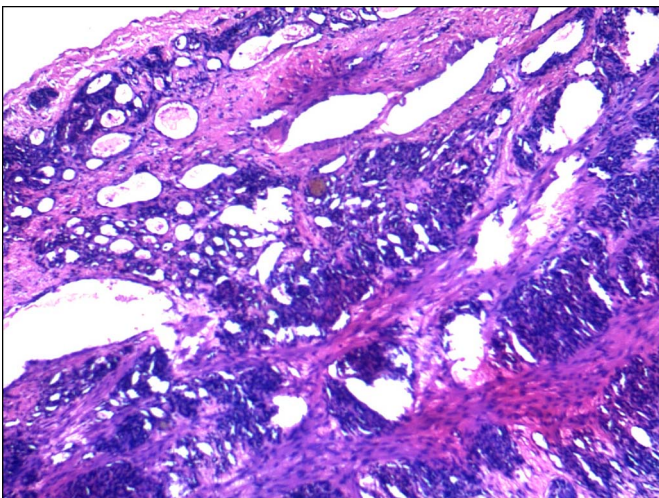


Figure 2. Microscopic section shows spindle-shaped cells extending from submucosa to serosa separated by interspersed dense stromal tissue and slit-like vascular spaces of a variable caliber.

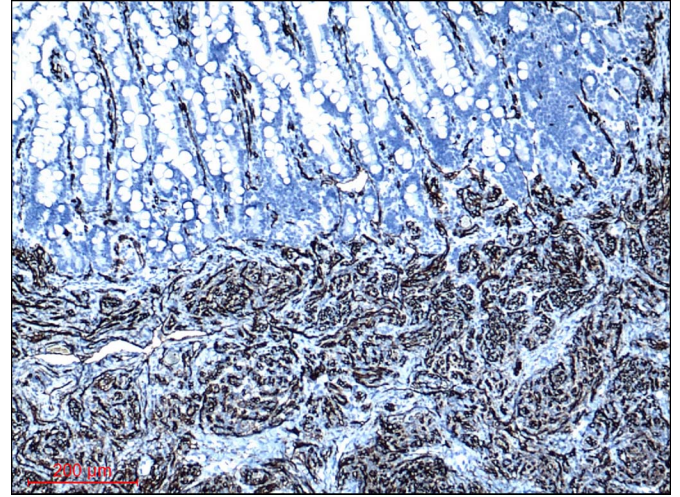


Figure 3. Tumor cells strongly express CD34 (immunohistochemistry, 3,3'-diaminobenzidine as chromogen, 40 \times).

been reported in less than 20 cases, with only 7 cases reported in neonates. Among those 7 neonatal cases of intestinal KHE reported, 4 had the lesion in the jejunum, 1 had in the duodenum, and 2 had in the colon. Six of them presented with intestinal obstruction and 1 with mass per abdomen. One had the antenatal finding of fetal hydrops, and the baby could not survive the surgery. All others were treated by surgery successfully.⁷⁻¹³

KHE origin is multifactorial. It occurs because of the dysregulation of both angiogenesis and lymphangiogenesis. Complications in KHE are common where it can cause pressure symptoms at the site of involvement similar to the index case, causing intestinal obstruction. The specific complication described in KHE is the Kasabach-Merritt phenomenon (KMP), with an estimated incidence of 42 to 71%.¹⁴ However, KMP was not reported in all the intestinal cases reported in the literature, probably because of the early age of diagnosis and management.

Ultrasonography is the investigation of choice for the skin and superficial lesions. However, a magnetic resonance image forms an essential asset in defining the infiltrative nature of KHE in deeper areas, characterized by multiplanar involvement with diffuse enhancement. Magnetic resonance imaging could not be performed in the index case because of acute presentation. There was no suspicion of KHE with availed investigations. Histopathology with immunohistochemistry remains the gold standard for the diagnosis of KHE. The histologic hallmark of KHE is infiltrating, confluent nodules, which are composed of spindle endothelial cells which align to form malformed lymphatic channels and slit-like vascular lumina containing erythrocytes, along with platelet thrombi, eosinophilic hyaline bodies. Immunohistochemical staining shows that endothelial cells in KHE lesions are positive both for vascular endothelial markers CD31 and CD34, lymphatic endothelial marker vascular endothelial growth factor receptor 3, D2-40, lymphatic endothelial hyaluronan receptor-1, and PROX1, but negative for glucose transporter-1 and human herpes virus-8 staining.¹⁵

The management of KHE depends on the extent of the lesion. Surgical excision is curative in localized lesions. However, widely dispersed lesions cannot be excised. In such cases, medical management with steroids, interferons, and chemotherapeutic agents is used in combinations rather than monotherapy because of the locally invasive nature.⁹ The biological behavior of KHE can be unpredictable; it can resolve spontaneously or can progress to unresectable large masses with a reported mortality of approximately 10%, especially when associated with KMP.¹⁶ However, KHE cases that occurred in the intestine showed a good prognosis probably because of complete excision.

KHE can occur in the small intestines and can present as neonatal intestinal obstruction. Exploratory laparotomy and excision of the involved intestine and primary anastomosis is the treatment of choice. Small intestinal KHE was observed to have a better prognosis because of the complete excision of the lesion.

DISCLOSURES

Author contributions: V. Maddileti wrote and approved the article and is the article guarantor. A. Mammen and V. Maddileti reviewed the literature and revised the article for intellectual content. S. Kuruvilla provided the images and edited the article. All authors approved the submission of the revised version.

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