Intestinal hemangioma presenting as recurrent hematochezia in a 6-week-old male

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1. Case report

We present a 6-week-old male, born at 35 weeks gestational age. He was on breast milk, reached his birth weight at the time of presentation, and typically had loose, brown stools every 1–2 days. One day prior to admission, he had 3 episodes of bright red blood per rectum. He was acting weak, tired, and had minimal non-bloody non-bilious spit up with feeds. Pertinent review of systems was negative for fever, abdominal distention, hematemesis, or melena. On exam, he was afebrile and hemodynamically stable; he was small, malnourished, and fussy. His abdominal exam was unremarkable, rectal exam demonstrated normal tone, no fissures, tags, or fistulas, guaiac positive.

Labs revealed hemoglobin 6.0 g/dL, mild leukocytosis and thrombocytosis, lactate 5.3, normal coagulation studies, normal ESR and CRP, and negative stool cultures. A technetium-99m pertechnetate (Meckel) scan was negative, barium enema was not suggestive of Hirschsprung’s disease, and abdominal ultrasound was not suggestive of intussusception. However, the abdominal ultrasound was significant for counter clockwise rotation of the superior mesenteric vein with associated dilatation and surrounding echogenic mesentery. A single contrast barium upper gastrointestinal series revealed anterior positioning of the proximal duodenum but normal location of the ligament of Treitz, making the study

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indeterminate for malrotation. Since suspicion for malrotation was low, as it typically does not present with hematochezia that leads to severe anemia, and given the strong family history of atopy, the patient was started on Neocate for presumed allergic colitis. His bowel movements improved and he began gaining weight appropriately. He was discharged, with hemoglobin 10.3 g/dL, but was readmitted seven days later with recurrent hematochezia and anemia (hemoglobin 6.3 g/dL). An abdominal MRI with and without contrast was completed on this admission, and showed an enhancing, infiltrating mesenteric mass with mass effect on the transverse colon and displacement of small bowel (Fig. 1). The infant was taken to the operating room and on exploratory laparotomy, diffusely edematous small bowel completely covered by what appeared to be a hemangioma versus arteriovenous malformation was found and biopsied (Fig. 2). Pathology revealed benign capillary hemangioma with diffuse GLUT1 positivity (Fig. 3) and focal areas of extension through the small intestinal wall, reaching peritoneal soft tissue.

Oral feeds were attempted and resulted in increased melenic stools, he was made nil per os, started on total parenteral nutrition, propranolol 1 mg/kg per dose twice a day, and methylprednisolone 0.5 mg/kg per dose twice a day. Two months later, CT imaging suggested response to pharmacotherapy with improvement compared to initial MRI. He was restarted on oral Pedialyte at about 5 and a half months of age and steroids were weaned off. At the age of 12 months, regular diet was introduced and was well tolerated, while the patient was gradually weaned off Neocate Junior. At the age of 17 months, he is on full oral feeds, and total parenteral nutrition was stopped. He continues on propranolol daily with no further episodes of hematochezia.

2. Discussion

Infantile hemangiomas are benign vascular tumors and have a female to male predominance of two to five to one [6]. Intestinal hemangiomas are rare, and typically represented only by case reports or series in the literature. If cutaneous lesions are present, especially if more than five lesions, there is an increased suspicion of the presence of a visceral lesion, with most common site being the liver [7]. Interestingly, this patient was male and did not have any cutaneous lesions.

Determining the source of hematochezia continues to be a diagnostic challenge. Some cases have been paired ultrasound and CT scan [4], used laparoscopy [8], or used scintigraphy with technetium-99m-labeled red blood cells and angiography [9] to identify the source of hematochezia as a gastrointestinal hemangioma. We used a combination of different imaging modalities, primarily ultrasound and MRI, which prompted exploratory laparotomy and therefore leading to the diagnosis. Soukoulis et al.
found there is often dilation of the superior mesenteric artery and vein in patients with hollow visceral GI hemangiomas, which our patient also had on ultrasound [3]. This may be due to increased blood flow to the tumor through feeder vessels. This case further supports high-resolution ultrasonography as the first line imaging when concerned for possible GI hemangioma.

Based on the radiographic findings, the patient underwent an exploratory laparotomy. Intraoperatively, on gross appearance the lesion appeared to be a hemangioma verses an arteriovenous malformation. Pathology revealed diffuse GLUT1 positivity. GLUT1 is an erythrocyte-type glucose transporter protein expressed in the endothelium of tissues. GLUT1 has been identified to distinguish infantile hemangioma from capillary proliferation in other vascular malformations [10].

The majority of children with infantile hemangiomas have a benign and uncomplicated course. However, there are certain indications for treatment, including functional impairment of vital organs or ulceration [11]. When necessary, propranolol, a non-selective beta-blocker, has become the first line treatment for infantile hemangiomas. The mechanism of action is not completely clear; however its effects can be divided into early, intermediate, and late effects [11]. The early effects of propranolol include vasoconstriction from decreasing the release of nitric oxide. Vascular endothelial growth factor and other angiogenic factors are blocked as part of the intermediate effects. Finally, apoptosis is induced in the proliferating capillary endothelial cells causing tumor regression as the long-term effects of the medication. The indication for corticosteroid therapy includes life-threatening infantile hemangioma or large hemangiomas [12]. The mechanism of action of corticosteroids is not entirely understood, but may be related to decreasing angiogenesis by inhibiting vascular endothelial growth factor and vasoconstriction.

This patient had a CT scan about two months after presentation which suggested response to pharmacotherapy compared to initial MRI. However, it was somewhat difficult to separate collapsed bowel loops from the mass. Therefore, MRI would be the preferred imaging for any additional follow-up as it could be used as direct comparison with the initial scan. There are no clinical guidelines that establish when, or if, to obtain follow-up imaging. Clinical judgment is used to determine the duration of therapy and should also dictate any plans for follow-up imaging.

3. Conclusion

Visceral vascular anomalies are a diagnostic challenge in the pediatric population. In the presence of dramatic drop in the hemoglobin and presence of hematochezia, it is recommended to consider this vascular anomaly in the differential diagnosis, as treatment options are drastically different. This patient’s initial diagnostic work up was inconclusive. However, imaging and histological evidence were paired to make the diagnosis, and subsequently initiate appropriate treatment. This case is unusual because, retrospectively, imaging was suggestive of malrotation; however, MRI was consistent with an infiltrating mesenteric mass that histologically was confirmed to be an intestinal hemangioma. Early recognition and treatment of this pathology may result in prompt initiation of medical treatment, resolution of symptoms, gradual advancement of enteral nutrition, and improvement of quality of life for the patient and family.

Conflict of interest

The authors have no conflicts of interest to disclose.

References