Successful treatment of visceral infantile hemangioma of the omentum and mesentery with propranolol

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Infantile hemangiomas (IH) are the most common tumors of infancy. In the typical cutaneous presentation, they follow a predictable and benign clinical course requiring medical intervention only for growth that interferes with vision or respiration, or, rarely, in cases of intractable bleeding. Hemangiomas arising from within visceral structures or surfaces are far less common, but can be associated with increased morbidity and mortality secondary to gastrointestinal hemorrhage, abdominal compartment syndrome, high output cardiac failure, and hypothyroidism. Here we present the case of a three-week-old boy with acute abdomen caused by hemorrhage of a hemangioma of the omentum and mesentery. He underwent operative exploration with debulking of the lesion and was subsequently treated with propranolol. Serial imaging demonstrated gradual involution and he remained free of further life-threatening events.

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Infantile hemangioma (IH) is the most common tumor of infancy with an incidence between 1 and 9%, varying by race [1]. The molecular mechanisms underlying pathogenesis remain incompletely understood, but the clinical course follows a stereotyped pattern: a phase of early vascular proliferation over the first year of life followed by a gradual phase (1–7 years in duration) of spontaneous involution and replacement of vascular channels by fibro-fatty tissue. Most cutaneous lesions follow a benign course. In the absence of bleeding, ulceration, or impairment of vision or respiration, treatment consists of observation and reassurance [2]. It has been suggested that the presence of five or more cutaneous hemangiomas should arouse suspicion for the possibility of visceral lesions [3], which are associated with higher rates of morbidity. Visceral hemangiomas, most common in the liver, may be associated with high output cardiac failure, coagulopathy, abdominal compartment syndrome, or respiratory distress [4,5]. Cases of small bowel perforation, intussusception, and gastrointestinal hemorrhage from hemangiomatosis have been reported [6–8].

Here we describe a unique presentation of an omental and mesenteric IH in a newborn boy, which hemorrhaged and caused an inflammatory obstruction of the small bowel. After operative exploration, lysis of adhesions, and partial debulking of the lesion, post-operative treatment with propranolol was initiated and the patient was followed with serial imaging. One year after initiation of treatment, the hemangioma had largely regressed and he remained symptom free.

1. Case report

A three-week-old boy born via emergent caesarian section (36 days, 6 weeks gestation) due to maternal HELLP syndrome and pre-eclampsia, was noted shortly after birth to have multiple hemangiomas on his lower lip, chin, and bilateral pre-auricular regions in addition to one sublingual lesion. He was discharged home without incident, but on day of life nineteen had decreased oral intake, irritability, and non-bilious emesis. He became febrile and lethargic with a distended, firm abdomen and was admitted to the intensive care with presumed sepsis. An abdominal x-ray demonstrated dilated loops of small bowel and he had multiple heme-positive small bowel movements. His infectious work-up remained negative while his abdomen became increasingly firm and bowel movements ceased. Laboratory values were significant only for a normocytic anemia, with a hematocrit of 30.1%. Ultrasound demonstrated a 4.3 × 1.9 cm complex collection in the upper
abdomen with multiple internal septations, adjacent thickened and hyperemic loops of bowel, and a large amount of free fluid with echogenic debris (Fig. 1). No solid organ visceral hemangiomas were appreciated.

Given a differential diagnosis that included visceral hemangioma, macrocystic lymphatic malformation, and bowel perforation with adjacent collection, the patient was taken for operative exploration. Dark red-brown, clear, non-purulent fluid was encountered upon entering the abdomen. There was no evident bowel ischemia, but there was an inflammatory process with a resultant band causing obstruction of several loops of adjacent small bowel. Further dissection revealed a complex, beefy red, bosselated soft tissue mass arising from the omentum between the greater curvature of the stomach and transverse colon, extending laterally along the splenic flexure and inferiorly along the descending colon. This tissue was dissected off of the adjacent colon and resected posteriorly to the retroperitoneum on the anterior surface of the pancreas. It was felt that continuing the dissection to include stripping the peritonealized surface of the colonic and small bowel mesentery as well as the pancreas and splenic hilum was too risky and the decision was made to leave a small amount of tissue abutting the pancreas, splenic hilum and mesenteric pedicles to the colon and small bowel. There was a small (sub-centimeter) hemangioma present on segment 2 of the liver, but no evidence of diffuse hepatic involvement.

Pathology revealed an aggregate of irregular, pink-red, diffusely hemorrhagic and necrotic soft tissues with well-formed capillary spaces grouped in a lobular fashion, consistent with hemangioma. The cells were subsequently shown to be positive for GLUT-1, but not D2-40 further supporting the diagnosis of IH and making a combined vascular-lymphatic malformation less likely (Fig. 2). The patient was started on propranolol prior to discharge home. He was readmitted one week later with abdominal distension, increased fussiness, and difficulty sleeping. Ultrasound showed a large, complex fluid collection in the left abdomen and Interventional Radiology placed a drain with successful resolution of the cavity. Studies of the drain fluid were not consistent with chylous ascites.

The patient remained on propranolol with surveillance MRI performed two months after initial presentation revealing persistent hemangioma along the root of the mesentery, encasing the splenic vein, celiac artery, SMA, and IVC (Fig. 3a). Six months after initial presentation, repeat MRI showed substantial decrease in the size of the mesenteric hemangioma with slight decrease in the extent of the retroperitoneal component (Fig. 3b). Clinically, he continued to do well, meeting all developmental milestones with appropriate weight gain. MRI one year after initial presentation revealed near complete resolution of the lesion (Fig. 4).

2. Discussion

This is an unusual case of visceral IH presenting not with gastrointestinal bleeding, but rather as intra-abdominal

![Fig. 1. Abdominal ultrasound demonstrating 4.3 x 1.9 cm complex fluid collection with multiple, thick, internal septations and adjacent thickened loops of bowel.](image1)

![Fig. 2. Immunohistochemistry showing GLUT-1 positivity (20×).](image2)

![Fig. 3. a) T2 weighted, gadolinium enhanced MRI of the abdomen two months after initial presentation demonstrating hemangioma along the root of the mesentery, encasing the splenic vein, celiac artery, SMA, and IVC. b) T2 weighted, non-contrast enhanced MRI six months after initial presentation demonstrating a significant decrease in the amount of mesenteric hemangioma and a small decrease in the size of the retroperitoneal components.](image3)

The effect has been proposed to result from vasoconstriction due to decreased nitric oxide release, down-regulation of vascular growth factors, blocking of GLUT-1 receptors, and induction of apoptosis [22]. The drug is well tolerated, exists in liquid preparation, and has a proven safety record with a long history of use in pediatric cardiology. In the absence of large-scale clinical studies to define evidence-based guidelines for its use in IH, a 2011 American consensus conference reviewed the existing data and produced a set of recommendations for propranolol dosing and monitoring [23]. Although its efficacy has not been formally defined in large-scale randomized controlled trials, a growing body of evidence, including this case of successful treatment of an omental and mesenteric lesion, supports the use of propranolol in visceral infantile hemangiomas.

**Conflict of interest statement**

The authors have no conflicts of interest.

References


**Fig. 4.** Repeat T2 weighted, contrast enhanced MRI 14 months after presentation demonstrating near complete resolution of the hemangioma along all previously involved surfaces.