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Neonatal kaposiform hemangioendothelioma of the spleen associated with Kasabach-Merritt phenomenon

Samuel A. Shabtaie [a], Bo Wang [a], Mark Owyong [a], Catalina Ruiz-Mesa [b], Fernando F. Corrales-Medina [b], Claudia P. Rojas [c], Juan C. Infante [d], Holly L. Neville [a], Eduardo A. Perez [a], Juan E. Sola [a], Anthony R. Hogan [a,*]

[a] Division of Pediatric Surgery, DeWitt Daughtry Family Department of Surgery, University of Miami Miller School of Medicine, Miami, FL
[b] Division of Pediatric Hematology-Oncology, Department of Pediatrics, University of Miami Miller School of Medicine, Miami, FL
[c] Department of Pathology, Holtz Children's Hospital, University of Miami Miller School of Medicine, Miami, FL
[d] Department of Pediatric Radiology, University of Miami Miller School of Medicine, Miami, FL

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ABSTRACT

Kaposiform hemangioendothelioma is a rare locally aggressive vascular tumor that usually manifests during early childhood. Typically the lesion presents with skin, soft tissue and bone involvement and is characterized histologically by ill-defined nodularity and the presence of spindle cells with resemblance to Kaposi's sarcoma. We report a rare neonatal case of a splenic kaposiform hemangioendothelioma associated with Kasabach-Merritt phenomenon that was diagnosed with radiographic imaging. Because of the rapid onset of thrombocytopenia and anemia, the patient required urgent splenectomy with subsequent resolution of the blood dyscrasias.

Kaposiform hemangioendothelioma (KHE) is an extremely rare vascular tumor that may be present at birth or develop early in childhood. Most commonly, KHE is a locally aggressive tumor that preferentially involves superficial soft tissues and extremities and rarely involves the mediastinum, retroperitoneum and internal organs. The lesion name was devised for its histologic resemblance to Kaposi's sarcoma with the presence of spindle cells and multiple discrete nodules. KHE is commonly mistaken for other vascular tumors [1]. Splenic involvement is highly uncommon, with few reported cases in the English literature to date. Additionally, more than 70% of patients with KHE develop Kasabach-Merritt phenomenon (KMP), a consumptive coagulopathy with associated thrombocytopenia [2]. In this case, we present a large solitary neonatal splenic KHE associated with KMP, which was successfully treated with splenectomy.

1. Presentation

The patient was a male infant born at 37 weeks gestation by cesarean section, weighing 2.93 kg. Prenatal ultrasound at 34 weeks gestation revealed a solid hypoechoic homogeneous lesion superior to the left kidney. The mass was measured to be 5.6 × 4.8 × 4.0 cm with positive Doppler flow. The mass was confirmed on postnatal ultrasound, located between the spleen and the upper pole of the left kidney (Fig. 1), with an initial differential diagnosis including neuroblastoma and adrenal hematoma. Further imaging was performed to assist with the diagnosis, while urine catecholamine metabolite levels were tested in light of the concern for neuroblastoma. MRI, on day of life 4, revealed a well-defined mass arising from the splenic hilum measuring 4.9 × 6.5 × 5.6 cm. The mass appeared isointense and enhanced homogeneously on T1 weighted post-contrast images, consistent with a vascular lesion of the spleen (Fig. 2).

On day of life 5, urgent surgery was performed because of rapid onset of anemia, thrombocytopenia and coagulopathy related to hypersplenic sequestration. The hemoglobin level decreased from 11 g/dL on postpartum day 1 to 8 g/dL on day 5, while the platelet level decreased from 111 × 10^3/ml to 50 × 10^3/ml. A prolonged coagulation profile (PT 27.2, INR 2.34, APTT >200) and a low fibrinogen level of 100 mg/dL (range 190–380) further highlighted the urgency for intervention. Of note, d-dimer and fibrin degradation product levels were not evaluated. Preoperative transfusions of blood and fresh frozen plasma were administered. Intraoperatively, a smooth, capsulated, homogenous splenic mass was identified and splenectomy was performed. The surgical specimen weighed 100 g and measured 7.5 × 6.2 × 4.8 cm in size with tumor replacing 90% of the parenchyma (Fig. 3). Microscopically, the mass appeared as an ill-defined lesion composed of fascicles of moderate plump and spindled endothelial cells with bland nuclei and eosinophilic-to-clear cytoplasm. The spindle

* Corresponding author at: Division of Pediatric Surgery, DeWitt Daughtry Family Department of Surgery, University of Miami Miller School of Medicine, 1120 N.W. 14th Street, Suite 450, Miami, FL 33136. Tel.: +1 305 243 5374.
E-mail address: ahogan@med.miami.edu (A.R. Hogan).

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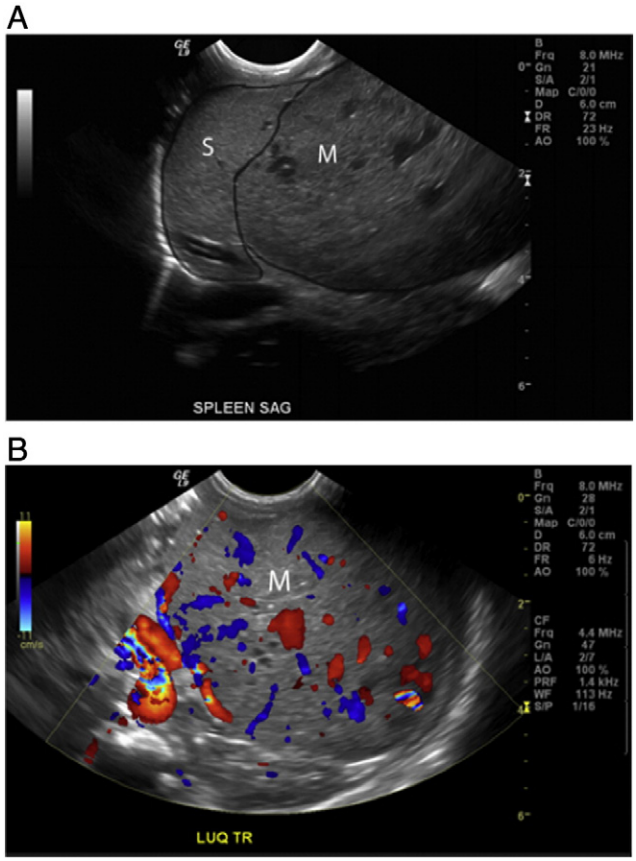

Fig. 1. A) Grayscale sonographic image of the abdomen shows the lateral posterior border of the mass (M) adjacent to and inseparable from the spleen (S). Structure has been outlined for clarity. Because the spleen forms a "claw sign" with the mass and no tissue plane was seen between the mass and the spleen, it was suspected that the mass was splenic in origin. The mass demonstrates variable echogenicity with hypoechoic serpiginous areas that could be areas of necrosis or large vascular channels. B) Color Doppler image of the mass (M) showing that the hypoechoic serpiginous regions were mostly vascular and that the majority of the mass is vascularized.

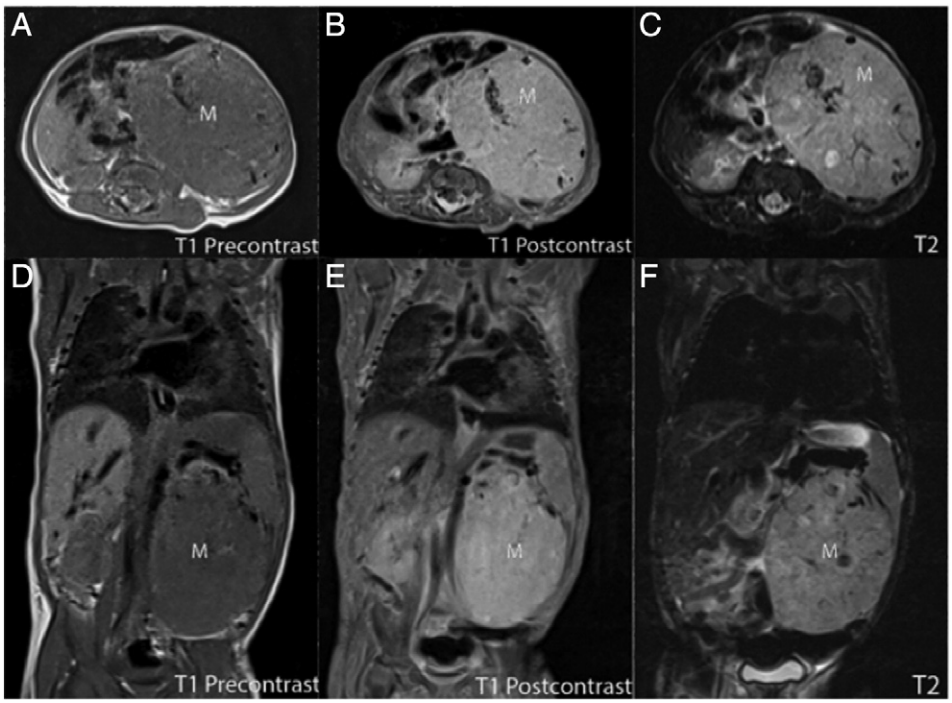

Fig. 2. Multiple MRI sequences of the abdomen localize the mass to the splenic hilum and characterize the tissue. On T1 precontrast images in axial (A) and coronal (D) planes the mass is low intensity with black areas that are either flow voids or hemosiderin from hemorrhage. The mass enhances fairly homogeneously after administration of contrast in both axial (B) and coronal (E) planes. On T2 weighted unenhanced images in axial (C) and coronal (F) planes the mass is heterogeneously bright in appearance.