J Ped Surg Case Reports 3 (2015) 462–465

Contents lists available at ScienceDirect

Journal of Pediatric Surgery CASE REPORTS

journal homepage: www.jpscasereports.com

Surgical intervention for Kasaback-Merritt Syndrome: A case report

Morris Sasson [a,*], J. Alford Flippin [a], Gary Birken [a], Fuad Alkhoury [b]

[a] Department of Pediatric Surgery, Joe DiMaggio Children's Hospital, 1005 Joe DiMaggio Drive Hollywood, FL 33021, USA
[b] Department of Pediatric Surgery, Miami Children's Hospital, 3100 SW 62nd Ave., Miami, FL 33155, USA

ARTICLE INFO

Article history:
Received 1 July 2015
Received in revised form
9 September 2015
Accepted 12 September 2015

Key words:
Kasaback-Merritt Syndrome
Kaposiform hemangioendothelioma
Vascular tumor

ABSTRACT

Kasabach-Merritt Syndrome (KMS) is an uncommon phenomenon characterized by the presence of a vascular tumor in association with thrombocytopenia, consumption coagulopathy and hemodynamic instability. Typically presents in infancy and involve the skin and deep soft tissues of the arms and legs. Treatment options vary with clinical presentation, and ranges from medical management to immediate surgical intervention. We present an unusual case of a newborn with KMS and fatal hemodynamic collapse imminent, which was successfully treated with surgical resection at our institution.

Kasabach-Merritt Syndrome (KMS) is a rare and potentially life threatening phenomenon characterized by profound thrombocytopenia and consumption coagulopathy in the presence of a rapidly enlarging vascular tumor [1,2]. We report a case of a newborn male who presented with KMS resulting from a massive upper extremity vascular tumor.

1. Case

A 3.l kg male was born with an extensive upper extremity tumor. His prenatal evaluation included ultrasonography and an MRI (Fig. 1), which both revealed the mass was consistent with a vascular tumor. Moderate cardiomegaly and decreased biventricular function were also documented. Given the size of the mass, elective caesarian section was planned at term, but due to decreased fetal movement along with nonreactive Non-Stress test (NST), an urgent C-section was performed at thirty-seven weeks gestation. The baby was born with an APGAR score of 1, 1 at one and 5 min respectively. He was immediately noted to be severely bradycardic, hypotensive and in profound respiratory distress. Maximum cardio pulmonary resuscitation was undertaken without delay.

During the resuscitation period, the surgical team quickly recognized that the vascular tumor was enlarging at a precipitous rate, extending from the mid-upper right arm to the wrist, and was noted to measure 11 × 11 × 8 cm. Further examination of the limb showed significant ecchymosis and a pale, pulseless hand with no spontaneous movement. The mass was thought to be causing high output cardiac failure with resulting hemodynamic instability. Significant bleeding within the tumor with sequestration was also noted. Initial laboratory evaluation provided evidence of severe metabolic acidosis with a pH of 6.87, anemia (Hb 5.8 g/dL), thrombocytopenia (41,000 mm^3), and low fibrinogen (52 mg/dL). The neonate remained hypotensive despite multi-agent volume expansion and the administration of full pharmacologic hemodynamic support.

With the clinical presentation of hemodynamic instability, consumption coagulopathy, thrombocytopenia, and a presumptive diagnosis of KMS with fatal hemodynamic collapse imminent, it was decided to place a tourniquet proximal to the tumor (Fig. 2). This resulted in a marked hemodynamic improvement, and following the completion of essential resuscitative measures the baby was transferred to the operating room.

The procedure was initiated with dissection at the level of the superior aspect of the mass, by identifying and isolating the axillary artery and vein with vessel loops. After proximal vascular control was adequately achieved, we proceeded to remove the previously placed tourniquet. The dissection was then continued along the posterior and lateral aspect of the tumor where several large feeding arteries and draining veins were ligated (Figs. 3 and 4). The median and ulnar nerves were located and preserved. A surgical plane of dissection was established antero-medially, which

* Corresponding author.
E-mail address: morrissasson@gmail.com (M. Sasson).

http://dx.doi.org/10.1016/j.epsc.2015.09.005

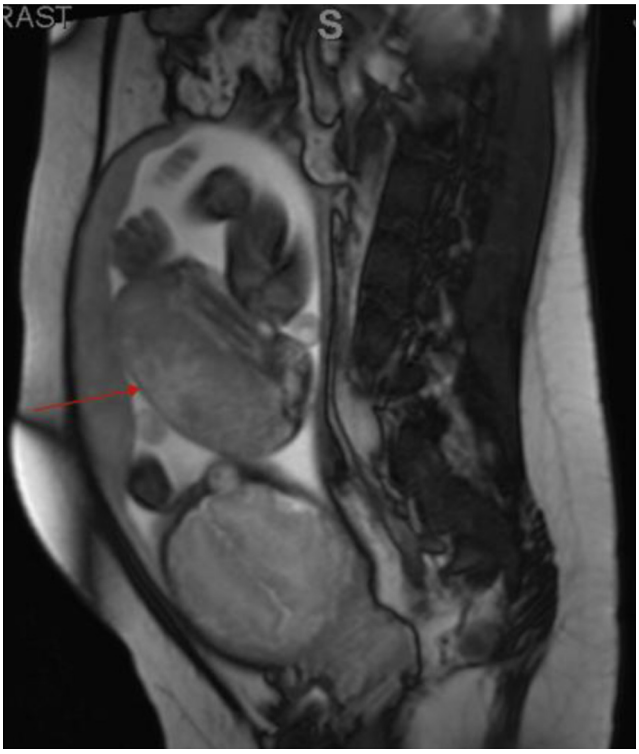

Fig. 1. Fetal MRI showing the vascular tumor.

eventually allowed for complete removal of the mass. Proximal vascular control was then released confirming improved perfusion and a viable hand (Fig. 5). Owing to the extensive dissection required to remove the tumor, the defect in the baby's arm was sizeable. For this reason, the wound was left open and wrapped with sterile gauze.

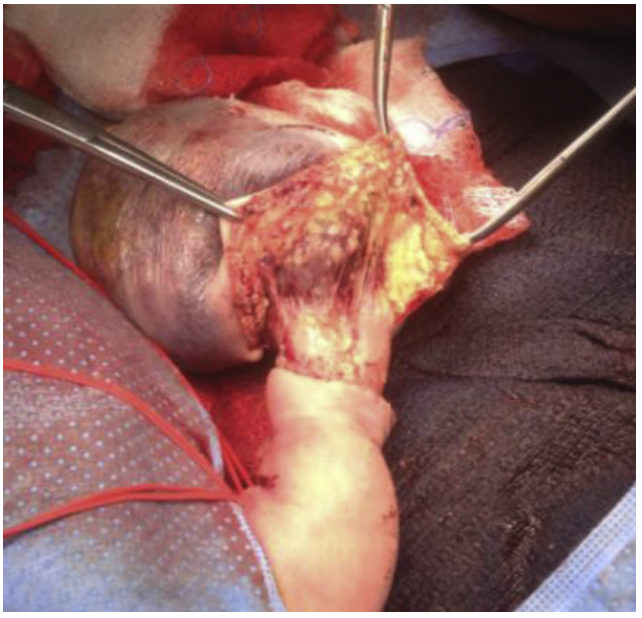

Fig. 3. Initiation of the dissection.

The baby's post-operative course was largely uneventful with marked hemodynamic and metabolic improvement. Daily improvement in the perfusion of his hand was also noted. Plastic Surgical consultation was obtained to assist with definitive wound closure. On post-operative day four, debridement of the wound and negative pressure wound therapy were initiated (Wound V.A.C. system). One week following the procedure, the patient was noted to have flexion and extension of his fingers. Split thickness skin grafting was successfully carried out, and at two months of age, he was discharged. At one year of age, he continues to develop normally and has full range of motion of the right arm and hand (Fig. 6).

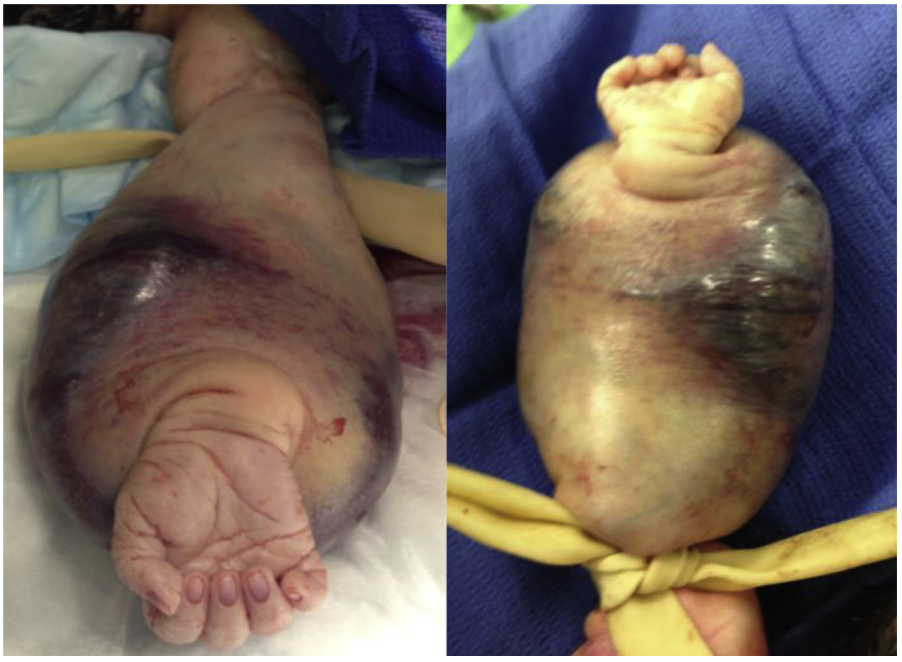

Fig. 2. Preoperative image showing large mass with tourniquet (Penrose drain) in place.