Surgical intervention for Kasaback-Merritt Syndrome: A case report

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A B S T R A C T

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 3.1 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 mm3), 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...
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.

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).
Final pathologic analysis classified the extensive mass as a congenital benign vascular tumor without the classic features of either a congenital hemangiomas or a Kaposiform hemangioendothelioma.

2. Discussion

The infant we present was born with a large vascular tumor of his right upper arm in association with a consumption coagulopathy and thrombocytopenia. These findings are pathognomonic for Kasabach-Merritt Syndrome [1-3]. KMS is a potentially life threatening phenomenon seen in association with large vascular tumors [1,2]. First described in 1940 [4], it remains a rare condition with less than 300 cases described in the literature [5]. The most common vascular tumor associated with KMS are kaposiform hemangioendothelioma (KHE) and tufted angioma. Approximately 70 percent of patients with KHE and 10 percent of those with tufted angiomas will develop this phenomenon [1,6].

Vascular tumors associated with KMS may be present at birth or develop in early infancy and childhood. These neoplasms typically involve skin and deep soft tissues of the arms and legs. The trunk, retroperitoneum and thoracic cavity are less commonly affected [6,7]. Superficial lesions will often show a well-circumscribed mass with purpura, while severe KMS will present with vascular limb tumors that are painful and rapidly enlarging. They are frequently indurated with a red-purple hue and may involve all or a significant portion of the affected extremity. These cases are frequently associated with thrombocytopenia, severe anemia, hypofibrinogenemia and elevated fibrin degradation products [8,9]. Mortality rates from KMS have been reported to be as high as 10–37% [6,10]. The most important factors that negatively affect survival are the location of the tumor, its size and delay in diagnosis.

In 2011, a multi-disciplinary expert panel developed a consensus-derived treatment protocol that resulted in a comprehensive guideline for the management of complicated KHE [2]. The committee strongly recommended a multi-disciplinary approach to

Fig. 4. Mass at near completion of removal. Note the large venous channels.

Fig. 5. Appearance of arm following complete removal of the vascular tumor.

Fig. 6. Arm at 1 year follow up.
treatment. Proposed strategies included pharmacologic therapy, embolization, and in patients who are hemodynamically unstable or those with threatened limb loss, urgent surgical intervention [1,2,11].

Although complete surgical resection has been considered the gold standard in the treatment of KHE, this option is oftentimes associated with an unacceptably high morbidity and mortality and can preclude total excision. However, for patients with life threatening tumors and for those who have failed non-operative management, surgical resection becomes the only viable option [3,12,13].

The medical management of KMS is aimed at decreasing the size of the tumor and correction of the coagulopathy. Specific therapeutic modalities widely utilized for KMS include systemic corticosteroids, antiplatelet agents, vincristine and interferon-α [2,14]. More recently, the mTOR inhibitor sirolimus has been used for KHE with KMS [15,16], and a Phase II clinical trial is underway to evaluate the safety and efficacy of the medication in complicated vascular anomalies (ClinicalTrials.gov NCT00975819).

However, pharmacologic therapy oftentimes requires weeks to months to be effective rendering this option unacceptable for the patient with a worsening consumption coagulopathy and hemodynamic instability [2,17,18]. The infant we report is unique because he was profoundly symptomatic at birth. Although, benign vascular tumors may be present at birth, KMS is rare. More typically, the syndrome presents in infancy but after the first month of life [19]. It is also noteworthy that infants who are diagnosed prenatally or in the immediate neonatal period characteristically develop a more advanced disease severity [2].

In conclusion, this case lends credence to the concept that a multi-disciplinary approach coupled with prompt surgical intervention are key elements in achieving a favorable outcome in infants stricken with KMS. The immediate postnatal resuscitative measures coupled with rapid surgical intervention and subsequent collaboration with our Division of Plastic Surgery were directly responsible for both salvaging the baby’s arm in the proximate neonatal period and the subsequent excellent outcome from both a functional and cosmetic standpoint.

Conflicts of interest
None.

References