Case Report: A Case of Kasabach Merritt Requiring Lower Limb Amputation in a 4 Years Old Child

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Abstract
Kasabach merritt syndrome is a very rare disorder where the infant usually suffers a giant hemangioma, thrombocytopenia and consumption coagulopathy.

Here we describe a very difficult case of kasabach merritt that has been progressing over the years with failure of medical treatment and non-feasibility of the surgical option or the embolization option due to the complexity of the vasculature of the tumor, eventually the only option left was above knee amputation in a 4 years old girl.

Keywords: Kasabach Merritt; Amputation

Abbreviations
KMP: Kasabach Merritt Phenomenon; KHE: Kaposiform Hemangioendothelioma; TA: Tufted Angioma

Introduction
Kasabach-Merritt phenomenon (KMP) is a life-threatening thrombocytopenic coagulopathy associated with rare vascular tumors, such as kaposiform hemangioendothelioma (KHE) and less frequently with tufted angioma (TA). It was first described by Kasabach and Merritt in 1940 [1].

It may result in life-threatening multi-organ hemorrhage. About 80% of patients present within 1st year after birth, and the reported mortality rate ranges from 10% to 37% [2]. However, the optimal therapy for neonatal KMP is currently unclear.

Kaposiform hemangioendothelioma most commonly appears as an enlarging, firm, solitary, purpuric cutaneous or soft tissue lesion. Often multiple tissue planes are involved and tumor borders are ill-defined [3].Locally invasive solitary lesions that most commonly manifest on the extremities, trunk, and face or neck. Retroperitoneal and intrathoracic lesions are less common but are frequently associated with KMP.

Kaposiform hemangioendothelioma arise from capillary and lymphatic endothelium. It is an infiltrative tumor, typically involving the dermis, subcutaneous fat, and muscles. Histologically, KHE is marked by irregular sheets of spindle-shaped endothelial cells and characteristic slit-like vascular channels; tufted angioma is characterized by irregularly sized nodules or tufts of capillaries in a “cannonball” pattern [4].

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The intravascular coagulopathy is likely secondary to platelet trapping, given the distinct endothelial architecture of the associated vascular tumors. Platelet trapping has been directly illustrated by positive immunohistochemistry for CD61, a marker of platelets and megakaryocytes, within the vascular lumen [5].

Similarly, studies also demonstrate localized consumption of fibrinogen when radiolabeled fibrinogen is infused [6].

**Lab evaluation**

This includes a complete blood count, fibrinogen, d-dimer, prothrombin time (PT) and activated partial thromboplastin time (aPTT).

Kasabach-Merritt phenomenon is characterized by very low platelet levels, commonly ranging from 3000 to 60 000 per microliter. Fibrinogen levels are significantly decreased and d-dimer and fibrin degradation products are elevated. The PT and the aPTT are typically normal to slightly elevated [7].

Significant anemia can occur due to intralesional bleeding, coagulopathy, sequestration of blood within the tumor and/or hemolytic anemia secondary to the sheering of red blood cells in the abnormal vasculature of the tumor.

**Management**

The significant morbidity and mortality associated with KMP necessitates aggressive treatment, but the management of these tumors varies widely among different academic centers with expertise in vascular anomalies. Surgical removal may result in definitive treatment of KMP, but KHE and TA are often unrespectable due to the large size and infiltrating nature.

**Supportive care**

Despite profound thrombocytopenia, severe bleeding is rare and platelet transfusions should be avoided, except for active bleeding and prior to or during surgery [8].

Adequate pain management is also critical in managing patients with vascular tumors associated with KMP as tumor engorgement by blood elements can cause severe pain and mass effect on the surrounding tissues.

**Drug options**

Invasive interventions within the presence of coagulopathy are associated with the potential for risk for bleeding; therefore, medical management is often first line and can achieve hemostatic stability in KMP.

These tumors are uncommon, and therefore, there is a lack of controlled studies, which makes it difficult to directly compare treatment modalities. Furthermore, patients with KMP show wide variation in response to different therapeutic options.

Many drug options have been used including steroids, vincristine, interferon-alpha, antiplatelet agents, propranolol, and sirolimus with variable outcomes and long-term side effects [9,10].

**Surgical management**

Surgical management, endovascular intervention, and radiation are some options for KMP.

Complete surgical resection offers the most definitive cure for small, localized tumors; for tumors that have decreased in size with medical therapy; or for life-threatening tumors [11].

However, surgical intervention at initial presentation with KHE is rarely feasible, given the infiltrative nature of the associated tumor and existing coagulopathy. Use of radiation therapy has been reported in KMP [12].

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Successful use of embolization has also been described in patients with KMP [13].

Case Report and Discussion

We describe a 4 years old girl presented to us with a large hemangioma involving the whole left leg till above the knee, also she presented with bleeding per gum and epistaxis. The lesion was very painful despite strong pain management. There was also severe pallor.

Lab evaluation revealed severe thrombocytopenia and anemia, Hb was 5 gm/dl and platelets were 6,000 per microliter, low fibrinogen and high d dimer.

The patient received packed RBCs and platelets to stop the bleeding several times.

CT angiogram revealed distal leg and foot subcutaneous branching serpiginous venous vascular malformation, acting like a shunt with consequent hypertrophy and edema of left lower limb from the pelvis down to foot with consequent overgrowth of the bony element.

The condition started at birth with small lesion over the dorsum of the foot that was cherry red, despite early it was very painful interfering with sleep and feeding.

As the family sought medical advice the exact diagnosis was as late as the child was 6 months with the lesion spreading over her left leg.

Surgical intervention was not a possibility even at that early age so patient was started on medical treatment.

She received IV vincristine 0.05 mg/kg weekly and oral aspirin 10 mg/kg/d for a total of 3 months but there was no improvement. The limb is nonfunctioning and pain was very severe the child could not sleep at night.

As the child reached 2 years of age a trial of interferon alpha was given at dose of 3 million U/m2/d a course very expensive for also one month yet no response was noted. Tragedy continued.

As regards pain management the child has received many courses of strong pain killers up to opiates in trial to make her sleep at night yet it was useless, the pain was so severe causing personality changes, irritability and insomnia. She never had the chance for proper education, school attendance or even sharing the daily activities with other children.

In a course over 4 years the condition has been progressing the lesion enlarging in size darken in color, pain increases and causing severe distress for the child and her family interfering with any aspect of normal life, yet she received no blood transfusion and she did not bleed till this last hospital admission.

The child did crawl for a while that was later lost.

Surgical intervention was also deemed impossible by neurovascular surgeons.

As the pain increased, bleeding and thrombocytopenia developed with anemia requiring transfusion, nonfunctioning limb causing harm more than benefit, amputation was decided.

The surgery was very high risk considering the wide spread hemangioma of the left leg reaching above knee but not extending to the pelvis, the current coagulopathy and thrombocytopenia which carries the risk of bleeding till death after surgery, adding to all that the psychological trauma that the child and her family would have to deal with after the surgery.

The patient received packed RBCS, platelets every 6 hours for 36h before surgery and for 48h after, kept under close observation in PICU after the surgery where she spent about 2 weeks.
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Conclusion
This case represents failure of both medical and interventional options in a 4 years old kasabach merritt phenomenon.

Bibliography


