

## Kasabach- Meritt Syndrome- A Rare Cause of Bleeding In a Neonate

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### Abstract

Kasabach-Merritt Syndrome (KMS) is a rare disorder that can affect infants from the time of birth or later in infancy. Diagnosis of KMS is made based on the constellation of hemangioma, thrombocytopenia, and coagulopathy. Our case presented at birth with hemangioma on perineal area and bleeding tendency. On evaluation thrombocytopenia and coagulopathy were found and diagnosis KMS was established supported by histopathology. Baby was treated with oral prednisolone and showed complete resolution of hemangioma and normalization of lab parameters.

**Keywords:** Kasabach- Meritt syndrome; Hemangioma; Bleeding neonate

### Case Report

A 3 day old male presented with bluish red swelling involving perineal gluteal regions and upper part of right thigh since birth. He was born to 29 years old primigravida by normal vaginal delivery in a local hospital and was a product of nonconsanguous marriage. There was no history of any adverse antenatal, natal and postnatal events. On examination baby was active, alert with good cry. His vital signs were within normal limit and systemic examination was unremarkable. In local examination there was bluish red swelling involving lower back, perineal and gluteal regions and upper part of right thigh (Figure 1). Prolonged bleeding from puncture sites were noticed although there was no clinically evident bleeding. Differential diagnoses of soft tissue tumour/ hemangioma/ bleeding diathesis were made.



Figure 1: Before treatment.

His laboratory test revealed thrombocytopenia (platelet count of 19000/cmm), increased PT/INR (23.8 seconds/2.18), increased APPT (43.3 seconds), d- dimer of 4.6 mg/dl. Other investigations including CBC, KFT, LFT, ultrasonography abdomen were normal. His sepsis work up was also negative. His coagulation profile did not improve even after vitamin K administration. Biopsy from swelling showed lobulated tumour extending deep into dermis and subcutaneous tissue with interspersed dilated capillary channels. Lobules mainly had large capillary channels having single endothelial lining. These features were diagnostic of mature capillary hemangioma. Based on thrombocytopenia, coagulation profile and biopsy confirming capillary hemangioma; diagnosis of Kasabach-Merritt syndrome was established. Patient was started on prednisolone @ 2 mg/kg/day along with blood component therapy for bleeding tendency. Platelet count and coagulation profile improved and hemangioma resolved in around eight weeks. Steroids were then tapered off over next three weeks (Figure 2).

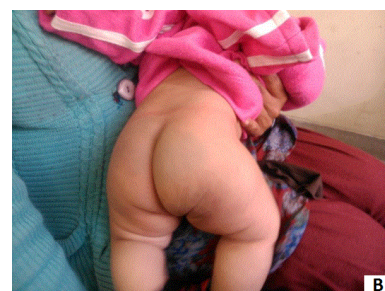


Figure 2: After treatment.

The association of hemangioma, thrombocytopenia, and hypofibrinogenemia was first described in 1940 by Kasabach and Merritt [1], who took care of an infant with a giant capillary hemangioma and thrombocytopenic purpura. Kasabach-Merritt syndrome (KMS) is a rare disorder that can affect infants from the time of birth or later in infancy. Diagnosis of KMS is made based on the constellation of hemangioma, thrombocytopenia, and

coagulopathy. KMS is an infrequent but potentially fatal complication of rapidly growing distinctive vascular lesions in infants. Unlike true capillary hemangiomas that regress in childhood, KMS are distinctive vascular tumours [2]. The pathophysiology is believed to be exposure of subendothelial elements or abnormal endothelium within the haemangioma resulting in aggregation and activation of platelets with a secondary consumption of clotting factors [2] and formation of intralesional thrombosis [3]. The lesions are typically superficial and solitary, but may involve internal structures such as the liver. Cardiac failure may result from high-volume arteriovenous shunting; shock, intracranial bleeding, or other internal hemorrhages may result in mortality rates as high as 30% [3]. Investigative workup shows thrombocytopenia, abnormal coagulation profile i.e. Prolonged Prothrombin Time (PT) and activated Partial Thromboplastin Time (aPTT), decreased fibrogen level, increased fibrin degradation product (FDP), and D-dimer levels. Diagnostic imaging is obtained as appropriate and may include the following: radiography, Computed Tomography (CT), Magnetic Resonance Imaging (MRI).

Kasabach-Merritt syndrome shows wide variation in its response to different treatment modalities. Currently, there are no known treatment guidelines [3]. Different interventions are recommended including compression, embolization, and use of interferon, propranolol, steroids, laser therapy, sclerotherapy, chemotherapy, radiation or surgery [1,2]. In each case the treating physician must decide the most suitable treatment to achieve maximum involution of the lesion and preservation of organ function.

Several researchers agree that most patients with Kasabach-Merritt syndrome respond to steroids within a few days of treatment [4]. However, one third will not respond to conventional dose of prednisolone (2 mg/kg/day) and mega dose 5 mg/kg/day may be effective. The angiogenetic character of Kasabach-Merritt syndrome

indicates that chemotherapy is also a logical treatment. While monitoring the effects of above treatments, the outcome measures were an increase in platelet count and fibrinogen level, and decrease in tumor size.

Surgical excision may be required for single cutaneous lesions or multiple lesions in the spleen (splenectomy) or liver (wedge resection/hepatectomy) [2,5]. However, before each surgical intervention the patient must be stabilized.

### Conclusion:

Kasabach- Meritt syndrome is a rare cause of coagulopathy in neonate. It should be suspected in any neonate with hemangioma and bleeding tendency. If untreated the condition is usually fatal. However most of the cases can be effectively treated with oral steroids.

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