# Kasabach-Merritt syndrome complicating femur fracture in a 16 year old boy with Klippel Trenaunay Syndrome

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## Abstract

Kasabach-Merritt Syndrome (KMS) is a rare and life threatening complication presenting as thrombocytopenia with consumptive coagulopathy in patients with large capillary or cavernous hemangioma. It is associated with a mortality of 30-40%[1,2]. We report a case of a 16 year old boy with Klippel-Trenaunay Syndrome (KTS) who sustained a right supra condylar femur fracture complicated by KMS. His thrombocytopenia and coagulopathy was managed followed by rush rodding of femur fracture. The preoperative management of KMS was demanding and needed constant monitoring due to the risk of haemorrhage and thrombosis. This is the first case of surgical management of a long bone fracture complicated by KMS in children to be reported.

**Keywords:** Kasabach-Merritt Syndrome, Klippel-Trenaunay Syndrome, Hemangioma, Coagulopathy.

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## Introduction

Kasabach-Merritt Syndrome (KMS) is a very rare and fatal form of consumptive coagulopathy along with purpurae seen in hemangiomata, both cutaneous and visceral. It is seen alone in hemangioma or as part of syndromes like KTS which is a triad of limb hypertrophy, vascular naevi and venous malformations. Fractures and its surgical management in patients with KMS pose a great challenge for the surgeon and the supporting staff due to its unpredictable nature and increased mortality[1]. The site and size of hemangioma are not good predictors of development of KMS. Massive hemangiomata inherently have an increased risk. Majority of hemangioma appear at birth or in the neonatal period during the first one-two years of life and then undergo regression by around 8-10 years of age. There are no clear guidelines for the management of KMS in literature. We report a case of 16 year old boy who is having Klippel-Trenaunay Syndrome (KTS) with right supra condylar femur fracture complicated by KMS with focus on the pre-operative management of the coagulopathy.

## Case Report

A 16 year old boy presented to us with pain over the right distal thigh since 3 days following a fall at home. On examination he also had multiple large hemangioma over the right lower limb, with hypertrophy of right leg and foot which was present since childhood (Fig. 1). He had inability to move his right knee since 4 years of age following septic arthritis of right knee which was evident by fixed flexion deformity of $90^0$ and bony ankylosis of right knee in X-ray along with multiple phleboliths over the right thigh and leg (Fig. 2). Radiographs confirmed a supracondylar fracture of right femur (Fig. 2). His bleeding parameters were deranged with high PT of 21 seconds, high APTT of 44 seconds, decreased fibrinogen of 147 milligram per decilitre, increased D-Dimer of 11.9 micro grams per millilitre and a very low platelet count of 29000 cells per micro litre. Fibrin degradation products test was positive.

**Fig. 1: Right lower limb appears to be larger compared to left with varicosities over the thigh and calf**

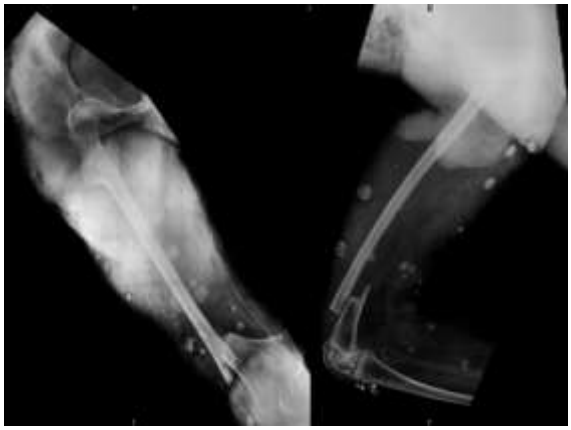

**Fig. 2: Plain x-ray AP and lateral of right femur: Multiple pheboliths are seen throughout the thigh along with closed supracondylar femur fracture and bony ankylosis of knee**

He was diagnosed as a case of Klippel-Trenaunay Syndrome with right supracondylar femur fracture complicated by Kasabach-Merritt syndrome in the form of consumptive coagulopathy and thrombocytopenia.

MRI imaging was done to determine the extent of lesion and also to identify occult lesions and later biopsy of lesion for tissue diagnosis was done along with definitive management of femur fracture. His cardiology evaluation was normal. Fresh Frozen Plasma (FFP) was transfused at 15 ml/kg body weight and cryoprecipitate at 5 ml/kg bodyweight. Transfusions were started overnight and PT, APTT and fibrinogen levels repeated 2 hours after transfusion. He was effectively optimized with 3 units of cryoprecipitate, 2 units platelet, and one unit FFP prior to surgery. PT, APTT, fibrinogen assay and platelet count was found to be satisfactory and rush rodding of right femur was done along with biopsy of the soft tissue structure (Fig. 3). Post operatively bleeding parameters were repeated and 1 unit cryoprecipitate and 2 units platelet was transfused. Two units platelet were transfused each on post op day 3 and 5. Histopathology report came as intramuscular hemangioma. The boy did not develop any complications in the post-operative period. He was discharged on post op day 12. Three month post-surgery radiographs showed complete union of fracture.

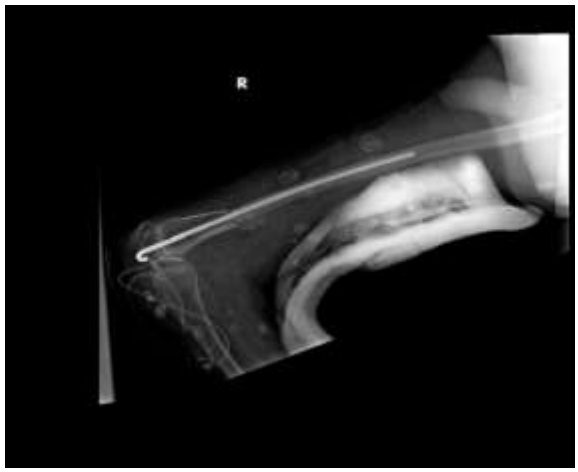


**Fig. 3: Fixation of right femur done using rush rod**

## Discussion

The pathogenesis of KMS is not clearly understood. It is presumed to be due to platelet trapping by abnormally proliferating endothelium within the hemangioma leading to platelet activation and secondary consumption of various clotting factors[3]. It results in massive thrombocytopenia with count usually less than 20,000 cells per micro litre with a shortened half-life of platelets. This along with fibrinolysis, results in intralesional bleeding leading to enlargement of hemangioma, thus forming a vicious cycle. There is very little literature evidence available on treatment of KMS in children with fractures or major trauma.in our knowledge this is the first case of KMS complicating the surgical treatment of a long bone fracture in children to be reported. Chen et al has reported about KMS complicating femoral shaft fracture in adult and Memendez et al has reported of closed reduction and cast application in case of KMS complicating treatment of a closed femur fracture in a 46 year old woman[4,5]. There is also no clear cut guideline for management of KMS with bleeding. Usual diagnosis is by detecting thrombocytopenia usually count less than 20,000 cells per micro litre, hypofibrinogenemia, Fibrin degradation products (FDP). Occasionally hemangiomas of liver and spleen are detected. Histology will reveal the subtype of hemangioma. MRI of the lesion helps in knowing the extent of lesion and helps in deciding whether the lesion is amenable to surgery or not. No single modality of treatment exists but a vigorous management will help in minimizing the mortality.

The current management of KMS involves securing haemostasis before planning for a surgical procedure. Small hemangioma can be ablated but resection of large hemangioma can be fatal. Supportive therapy is mainly by administering platelets, FFP (15 ml/kg), and cryoprecipitate (5-10ml/kg) if severe hypofibrinogenemia is not corrected with FFP alone. Aim is to keep the prothrombin time within 2 to 3 seconds of control; a fibrinogen level > 150 mg/dl; and a platelet count of >50,000/$cm^2$[5].

Surgical treatment involves excision for single cutaneous lesions. Wide local excision and even amputations has been performed in some cases[6]. Vascular embolization, compression therapy, radiotherapy, corticosteroids, interferon alpha, chemotherapy, anticoagulants, antiplatelets, anti-fibrinolytic agents, has all been tried for the treatment of KMS[1].

## Conclusion

Kasabach-Merritt Syndrome can manifest alone in hemangioma or associated with syndromes like Klippel-Trenaunay Syndrome. KMS should be detected and managed effectively before and after any surgical procedures. A standard and specific management protocol for transfusion for control of the thrombocytopenia and coagulopathy should be formulated in view of the severity of this complication.

## Disclaimer

No benefits in any form has been received or will be received related directly or indirectly to the subject of this article.

## Compliance with Ethical Standards

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**Informed consent:** Informed consent was obtained from the parents of the patient mentioned in this article.

**Conflicts of interest:** Both the authors declare that we have no conflicts of interest.

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