



## Kaposiform hemangioendothelioma with Kasabach-Meritt syndrome: life-threatening tumor treated with sirolimus

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### Background:

Kaposiform hemangioendothelioma (KHE) is a rare and aggressive, potentially life-threatening vascular tumor typically occurring in infancy. It is frequently associated with the Kasabach-Meritt syndrome (KMS), a serious consumptive coagulopathy with thrombocytopenia and hypofibrinogenemia. Because of this, morbidity rates are high. As of yet, no standardized guidelines exist for the treatment of KHE.

### Case presentation:

In January 2019 a male full-term newborn presented to the neonatal intensive care unit (NICU) with a 14x10 cm tumor on the left side of the neck. The pregnancy itself was well-controlled and the male newborn was delivered by an elective C-section, with an Apgar score of 9/9. The tumor, purple in color and covered in petechiae and ecchymoses, was extending from the base of the neck towards the mandibula. Upon the transfer to the NICU, laboratory findings (platelets 9,000/ $\mu$ L, fibrinogen 0.8 g/L, D-dimer >10 mg/L, aPTT 37.1 s) showed significant thrombocytopenia and hypofibrinogenemia. Initial assessment (MRI and TOF angiography) showed a hypervascular, heterogeneous structure suggestive of hemangioma. The initial treatment strategy consisted of administering blood derivatives, DSA embolization of the tumor, propranolol, antifibrinolytics, and corticosteroids – but no clinical or laboratory improvement was noted. The MRI, laboratory findings, and the clinical course of the disease were all indicative of KHE, hence treatment with peroral sirolimus (an mTOR inhibitor) in combination with parenteral corticosteroids was started. In the following two months, complete resolution of KMS was noted with a progressive reduction in tumor size. The coagulation parameters turned normal. The patient is still on sirolimus and demonstrates nearly complete involution of the lesion with no apparent side effects.

### Conclusion:

As there are no standardized guidelines for the treatment of KHE, this case strongly suggests the effectiveness of sirolimus treatment in severe KHE/KMS.

### Keywords:

Kaposiform hemangioendothelioma, Kasabach-Meritt syndrome, Sirolimus