

Abstract: Kaposiform Haemangioendothelioma Complicated by Kasabach-Merritt Phenomenon

Authors

Elijah Aaron Sischy¹  | Johannes Petrus Potgieter¹  | Alireza Dehghan-Dehnavi²  | Mothusi Mathibe³ | Elliot Motloun⁴ 

¹Department of Paediatric Surgery, School of Clinical Medicine, Faculty of Health Sciences, Sefako Makgatho Health Sciences University

²Acting Head of Department, Department of Diagnostic Radiology, School of Clinical Medicine, Faculty of Health Sciences, Sefako Makgatho Health Sciences University

³Head of Clinical Unit: Paediatric Oncology, Department of Paediatrics, School of Clinical Medicine, Faculty of Health Sciences, Sefako Makgatho Health Sciences University

⁴Head of Department, Department of Paediatric Surgery, School of Clinical Medicine, Faculty of Health Sciences, Sefako Makgatho Health Sciences University

Correspondence to:

EA Sischy: eliyah@sischy.co.za

E Motloun: elliot.motloun@smu.ac.za

Sefako Makgatho Health Sciences University, Department of Paediatric Surgery, P. O. Box 231, MEDUNSA, 0204, Tel (+2712) 521 4921, Fax (+2712) 521 4467

Abstract

Background: Kasabach-Merritt Phenomenon (KMP) is a rare, life-threatening coagulopathy of infancy. In contrast to infantile haemangiomas, KMP is almost exclusively associated with Kaposiform Haemangioendothelioma (KHE). Clinical cases of KMP are rarely reported in the literature, and it poses diagnostic and therapeutic challenges due to its rapid growth and potentially fatal complications of thrombocytopenia and consumptive coagulopathy.

Aim and Objectives: This study aims to describe the clinical presentation, diagnostic approach, management and outcomes of a patient diagnosed with KMP at our institution. We identify key diagnostic features, discuss the interventions made by a multidisciplinary team, and aim to contribute to the limited literature on this rare condition.

Methodology: We conducted a retrospective case report of an 11-month-old child diagnosed and treated for KHE complicated by KMP. Clinical presentation, laboratory findings, imaging results, multimodal intervention, and outcomes were assessed.

Results: The patient was referred to our facility with a rapidly growing buttock mass. The referring centre initially thought it to be a congenital haemangioma that wasn't responding to propranolol. The acute presentation of a thrombocytopenia and coagulopathy, in conjunction with diagnostic imaging, confirmed KHE complicated by KMP. Multidisciplinary care with interventional radiology and oncology was crucial to our management plan.

Conclusion: KHE and KMP are rare but important differentials in infants and children presenting with locally aggressive vascular tumours. Early recognition and multidisciplinary care are essential as KHE holds significant morbidity and mortality risks. Further research and discussion are needed to establish strong evidence-based guidelines of treatment.

Keywords: Kaposiform haemangioendothelioma; Kasabach–Merritt phenomenon; Paediatric; Oncology