Journal of Cambodian Health
Volume. 4, No. 1, 2026, v4101
https://doi.org/10.70243/jch2026.4101

OPEN ACCESS

Kasabach-Merritt Phenomenon in a 7-month-old Cambodian Infant

Case Report

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Article history
Received: 7 May 2025
Revised: 9 October 2025
Accepted: 13 January 2026
Published: 20 January 2026

Keywords
Kasabach-Merrit Phenomenon
Kaposiform
Hemangioendothelioma
Vascular Tumor

Abstract: Kasabach-Merritt phenomenon (KMP) is a rare condition associated with vascular tumors such as kaposiform hemangioendothelioma and tufted angioma; it can be life-threatening due to its consumptive coagulopathy. Thrombocytopenia and hypofibrinogenemia are characteristic of KMP, and anemia and raised d-dimer levels can also be detected. Here, we report a 7-month-old Cambodian with the condition. The infant was admitted to the National Pediatric Hospital in Phnom Penh because of a mass on the right side of the neck that had been progressively enlarging. The patient had severe thrombocytopenia (8,000/μL), anemia (Hb 7.6g/dL) and reduced fibrinogen level (1.5g/L). CT scan suggested and histopathology of the lesion confirmed a diagnosis of kaposiform hemangioendothelioma. Kasabach-Merritt phenomenon was diagnosed, and the infant was treated with platelets and fresh frozen plasma infusions, prednisolone (2mg/kg/day) and propranolol (2.5mg/kg/day). After eight weeks of therapy, platelets raised to 102,000/μL. The infant developed Cushing's syndrome after 6 months of treatment, and prednisolone was scaled down to a maintenance dose of 0.5mg/kg/day. Fibrinogen levels went back to normal (2.14g/L) after seventeen months of treatment, and the tumor shrunk significantly. This case report shows that a combination of prednisolone and propranolol was effective for the treatment of KMP and kaposiform hemangioendothelioma. Timely recognition and treatment of Kasabach-Merritt phenomenon is essential.

1. Introduction

A case of capillary hemangioma with extensive purpura was first described by Kasabach and Merritt in 1940 (Kasabach, 1940); subsequently, the disease was named after them. Kasabach-Merritt phenomenon (KMP) is an extremely rare condition that can be life threatening because of its consumptive coagulopathy. It is associated with rare vascular tumors, particularly kaposiform hemangioendothelioma (where it occurs in up to 70% of cases) (Croteau et al., 2013) and, less frequently, tufted angioma (Mahajan et al., 2017). It can also be observed in complex vascular anomalies such as kaposiform lymphangiomatosis (Fernandes et al., 2015). KMP is estimated to affect .07 per 100,000 children/year (Lewis and Vaidya, 2025). When untreated, KMP has a mortality rate of 10%–30% (Kelly, 2010). It affects both males and females of any ethnicities and often presents in infancy; in one case series, the mean age at diagnosis was two months (Croteau et al., 2013). Marked thrombocytopenia, likely due to intralesional platelet trapping (Lyons et al., 2004) and hypofibrinogenemia characterize the condition; microangiopathic hemolytic anemia and raised d-dimer levels can also be found (Mahajan et

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al., 2017). Kaposiform hemangioendothelioma, in particular, generally affects the extremities, trunk or cervicofacial region (Croteau et al., 2013; Lyons et al., 2004; Sarkar et al., 1997). Kasabach-Merritt phenomenon occurs more frequently if the depth and infiltration of the tumor are higher, and retroperitoneum or thorax are involved (Croteau et al., 2013). We report a case observed in an infant in Cambodia, where only one case has been published previously to date (Chap & Bin, 2022). Our case report emphasizes the importance of a timely diagnosis. The combination of prednisolone and propranolol, used in the previous case reported from Cambodia (Chap & Bin, 2022) and in the current case, could be the most effective treatment.

2. Case Presentation

A 7-month-old male infant was admitted to the Surgical Ward of the National Pediatric Hospital in Phnom Penh, Cambodia because of a large, firm, poorly mobile mass on the right side extending to the back of the neck that had progressively increased in volume over four months (Figure 1a). The lesion was non-pulsatile and warm to touch, with overlying violaceous discoloration of the skin. Although there was no respiratory distress, the parents reported that the increasing size of the mass had begun to cause mild difficulty with feeding and swallowing. A computerized tomography (CT) scan confirmed a mass compatible with a hemangioma, of 105mm x 112mm x 136mm; the surrounding bones were intact. The patient weighed 6 kg; his height was 63cm. Respiratory rate was 28/min, pulse 115/min, temperature 36.3 ºC; oxygen saturation, SaO_2 97%; weight 6kg; height 63cm; blood pressure, BP was not recorded. Laboratory test showed patient had severe thrombocytopenia (8,000/μL; normal range 150,000–450,000/μL), and anemia (hemoglobin, Hb 7.6g/dL; normal range 10.5–13.5 g/dL); the fibrinogen level was reduced (1.5 g/L; normal range 1.8-4.0 g/L). Liver enzymes (aspartate transaminase, AST 28 IU/L; alanine transaminase, ALT 32 IU/L; normal range for 6-12 months: AST 20–60 IU/L, ALT 5–45 IU/L) were normal and kidney function tests showed a slightly increased creatinine level (urea 14mg/dL, creatinine 0.6 mg/dL; normal range for 6–12 months: urea 5–18 mg/dL, creatinine 0.2–0.4 mg/dL). Glucose level was not measured. An abdominal ultrasound ruled out a liver hemangioma.

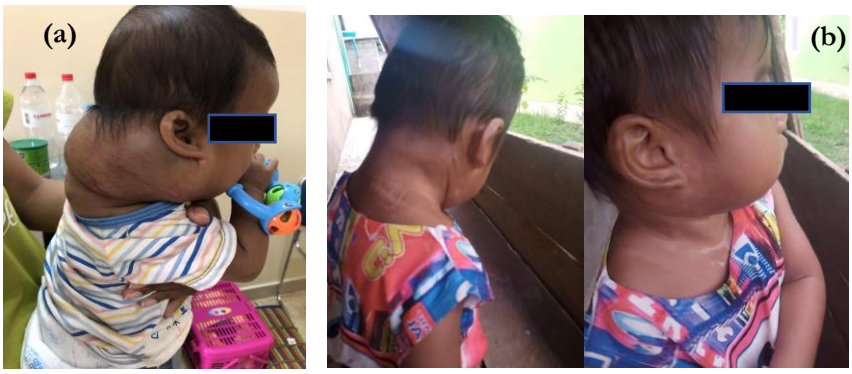

Figure 1. 7-month-old male (a) with a large, firm, poorly mobile mass on the right side extending to the back of the neck (b) after 17 months of therapy

A biopsy was performed and histopathology revealed nodules of spindle-shaped endothelial cells arranged in sheets and short fascicles with slit-like vascular channels, consistent with kaposiform hemangioendothelioma. A diagnosis of Kasabach-Merritt phenomenon was made, and the patient received two doses of platelets infusion (10 mL/kg) and one dose of fresh frozen plasma (10 ml/kg over 2hrs) due to active bleeding at the biopsy site; prednisolone (2 mg/kg/day) and propranolol (2.5 mg/kg/day) were started. One month later, platelets were 120,000/μL and fibrinogen was 1.5g/L. After

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eight weeks of treatment, platelets were 102,000/μL and, because of the insurgence of Cushing's syndrome, prednisolone was scaled down by 0.5mg every month until a maintenance dose of 0.5mg/kg/day was reached. Fibrinogen levels became normal (2.14 g/L) only after 17 months of therapy, when the tumor had shrunk considerably (Figure 1b).

3. Discussion

We have reported the second case of Kasabach-Merritt phenomenon observed in Cambodia. Due to the rarity of KMP, treatment of KMP is not standardized. In 2013, a regimen of systemic corticosteroids and weekly intravenous vincristine was recommended by a multidisciplinary expert panel in North America (Chiu et al., 2012; Tlougan et al., 2013); in the same year, the results of a phase II study of sirolimus which had included 10 patients with kaposiform hemangioendothelioma and KMP showed a complete and rapid resolution of KMP in all patients (Adams et al., 2016). Recently, a low dose of sirolimus monotherapy (0.06 mg/day) given over 12 months to a neonate reportedly reduced the size of the tumor and stabilized the hematological parameters (Nakamura et al., 2024). In Spain, vincristine, aspirin, and ticlopidine have been used with good reported outcomes (Fernandez-Pineda et al., 2010, 2013). Steroids are generally weaned after two-three weeks to avoid side effects such as metabolic and renal issues, when combined with other medications, such as vincristine or sirolimus. In this patient, vincristine was not used, despite its availability in Cambodia. Vincristine requires intravenous administration and careful monitoring in a hospital setting, which was not possible with this infant. An oral-only regimen with prednisolone and propranolol made an outpatient management feasible.

All the drugs mentioned above have side effects. Vincristine can cause constipation, peripheral neuropathy, irritability, and syndrome of inappropriate antidiuretic hormone secretion. Sirolimus can cause mucositis and dyslipidemia, and decreases immune responses (Fernandez-Pineda et al., 2010, 2013). Corticosteroids also have important side effects (growth retardation, hyperglycemia, immunosuppression and gastrointestinal disturbances) (Chiu et al., 2012; Tlougan et al., 2013)., as shown in our case.

Propranolol promotes regression of cutaneous infantile hemangiomas (Léauté-Labrèze et al., 2008; McGee et al., 2013); the responses to this drug of kaposiform hemangioendothelioma, tufted angioma, and Kasabach-Merritt phenomenon are reportedly variable (Chiu et al., 2012). The side effects of propranolol include bradycardia, hypotension, bronchospasm and hypoglycemia. In our case, a combination of prednisolone and propranolol was effective. Finally, we need to mention that, although platelet transfusions are often ineffective and can cause tumor engorgement (Croteau et al., 2013), and platelets have shorter half-life in KMP (Koerper et al., 1983; Mulliken et al., 2004), platelets were infused to our infant because of active bleeding from the biopsy site. Given the significant risk of hemorrhage, the biopsy may have been avoidable. Indeed, tissue biopsy is frequently contraindicated in suspected cases of Kasabach–Merritt phenomenon due to the associated consumptive coagulopathy. In such cases, the diagnosis is typically established on the basis of clinical presentation, characteristic laboratory abnormalities, and supportive imaging findings.

Author contributions

Conceptualization, writing-original draft and visualization, Prom.V.; resources, investigation, Chinith.P. and Nheb.A.; expert advisor, and scientific review, Vento.S; content review and editing, Pol.S

Data availability

All data relevant to the case are included in the report.

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Ethical considerations

The case study was reported in the context of the routine standard of care and treatment that patients and families can expect, which includes respect, confidentiality, and informed consent for any treatment. The parents were provided with information as to the purpose of publishing this case, including that confidentiality would be maintained, and that their child would receive treatment regardless of their decision to allow for this case to be published. Fully informed written consent was requested and received on this basis"

ICMJE and CARE guidelines for a single case report do not require a formal ethical review; however, this case report does uphold ethical and legal standards of clinical practice (BMJ Case Reports, 2025; Gagnier et al., 2013). This case report was not submitted to Cambodia's National Ethics Committee for Health Research (NECHR), as it involved no experimental treatment and falls under the category of a single case report. Moreover, this case is reported for clinical medical education purposes due to the rarity of the condition. This report has been approved by the University of Puthisastra Research Committee (UPRC), Approval number [018UPRC].

Acknowledgment

We thank the University of Puthisastra and the National Pediatric Hospital for providing the necessary support for the data collection process. We also thank the parents of the sick child who consented to use his related information.

Conflicts of interest

Authors declare no conflicts of interest.

References

Adams, D. M., Trenor, C. C., Hammill, A. M., Vinks, A. A., Patel, M. N., Chaudry, G., Wentzel, M. S., Mobberley-Schuman, P. S., Campbell, L. M., Brookbank, C., Gupta, A., Chute, C., Eile, J., McKenna, J., Merrow, A. C., Fei, L., Hornung, L., Seid, M., Dasgupta, A. R., … Azizkhan, R. G. (2016). Efficacy and safety of sirolimus in the treatment of complicated vascular anomalies. *Pediatrics*, *137*(2), e20153257. https://doi.org/10.1542/peds.2015-3257

BMJ Case Reports. (2025). *Instructions for Authors-Patient Consent and Ethical Approval*. BMJ. Retrieved on May 7, 2025, from https://casereports.bmj.com/pages/authors

Chap, C., & Bin, S. (2022). Kasabach-Merritt syndrome: a rare cause of swelling thigh in a Cambodian newborn. *Oxford Medical Case Reports*, *2022*(2), omac004. https://doi.org/10.1093/omcr/omac004

Chiu, Y. E., Drolet, B. A., Blei, F., Carcao, M., Fangusaro, J., Kelly, M. E., Krol, A., Lofgren, S., Mancini, A. J., Metry, D. W., Recht, M., Silverman, R. A., Tom, W. L., & Pope, E. (2012). Variable response to propranolol treatment of kaposiform hemangioendothelioma, tufted angioma, and Kasabach–Merritt phenomenon. *Pediatric Blood & Cancer*, *59*(5), 934–938. https://doi.org/10.1002/pbc.24103

Croteau, S. E., Liang, M. G., Kozakewich, H. P., Alomari, A. I., Fishman, S. J., Mulliken, J. B., & Trenor, C. C. (2013). Kaposiform hemangioendothelioma: atypical features and risks of Kasabach-Merritt phenomenon in 107 referrals. *The Journal of Pediatrics*, *162*(1), 142–147. https://doi.org/10.1016/j.jpeds.2012.06.044

Fernandes, V.M., Fargo, J.H., Saini, S., Guerrera, M.F., Marcus, L, Luchtman-Jones, Adams, D., Meier, E.R. (2015). Kaposiform lymphangiomatosis: unifying features of a heterogeneous disorder. *Pediatric Blood & Cancer*, *62(5)*, 901–904. https://doi.org/10.1002/pbc.25278

Fernandez-Pineda, I., Lopez-Gutierrez, J. C., Chocarro, G., Bernabeu-Wittel, J., & Ramirez-Villar, G. L. (2013). Long-term outcome of vincristine-aspirin-ticlopidine (VAT) therapy for vascular tumors associated with kasabach-merritt phenomenon. *Pediatric Blood & Cancer*, *60*(9), 1478–1481. https://doi.org/10.1002/pbc.24543

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Fernandez-Pineda, I., Lopez-Gutierrez, J. C., Ramirez, G., & Marquez, C. (2010). Vincristine-ticlopidine-aspirin: an effective therapy in children with Kasabach-Merritt phenomenon associated with vascular tumors. *Pediatric Hematology and Oncology*, *27*(8), 641–645. https://doi.org/10.3109/08880018.2010.508299

Gagnier, J. J., Kienle, G., Altman, D. G., Moher, D., Sox, H., & Riley, D. (2013). The CARE guidelines: consensus-based clinical case reporting guideline development. *BMJ Case Reports*, *2013*, bcr2013201554. https://doi.org/10.1136/bcr-2013-201554

Kasabach, H. H. (1940). Capillary hemangioma with extensive purpura. *American Journal of Diseases of Children*, *59*(5), 1063. https://doi.org/10.1001/archpedi.1940.01990160135009

Kelly, M. (2010). Kasabach-Merritt phenomenon. *Pediatric Clinics of North America, 57(5),* 1085–1089. https://doi.org/10.1016/j.pcl.2010.07.006

Koerper, M. A., Addiego, J. E., de Lorimier, A. A., Lipow, H., Price, D., & Lubin, B. H. (1983). Use of aspirin and dipyridamole in children with platelet trapping syndromes. *The Journal of Pediatrics*, *102*(2), 311–314. https://doi.org/10.1016/S0022-3476(83)80550-2

Léauté-Labrèze, C., de la Roque, E. D., Hubiche, T., Boralevi, F., Thambo, J.-B., & Taïeb, A. (2008). Propranolol for severe hemangiomas of infancy. *New England Journal of Medicine*, *358*(24), 2649–2651. https://doi.org/10.1056/NEJMc0708819

Lewis, D., Vaidya, R. (2025). Kasabach-Merritt syndrome. In: StatPearls [Internet]. Treasure Island, FL: StatPearls Publishing. https://www.ncbi.nlm.nih.gov/books/NBK519053/

Lyons, L. L., North, P. E., Mac-Moune Lai, F., Stoler, M. H., Folpe, A. L., & Weiss, S. W. (2004). Kaposiform Hemangioendothelioma. *The American Journal of Surgical Pathology*, *28*(5), 559–568. https://doi.org/10.1097/00000478-200405000-00001

Mahajan, P., Margolin, J., & Iacobas, I. (2017). Kasabach-Merritt phenomenon: classic presentation and management options. *Clinical Medicine Insights: Blood Disorders*, *10*, 1179545X1769984. https://doi.org/10.1177/1179545X17699849

McGee, P., Miller, S., Black, C., & Hoey, S. (2013). Propranolol for infantile haemangioma: a review of current dosing regime in a regional paediatric hospital. *The Ulster Medical Journal*, *82*(1), 16–20.

Mulliken, J. B., Anupindi, S., Ezekowitz, R. A. B., & Mihm, M. C. (2004). Case 13-2004. *New England Journal of Medicine*, *350*(17), 1764–1775. https://doi.org/10.1056/NEJMcpc049002

Nakamura, S., Ozeki, M., Hayashi, D., Yasue, S., Endo, S., & Ohnishi, H. (2024). Sirolimus monotherapy for Kasabach-Merritt phenomenon in a neonate; Case report. *International Journal of Surgery Case Reports*, *117*, 109497. https://doi.org/10.1016/j.ijscr.2024.109497

Sarkar, M., Mulliken, J. B., Kozakewich, H. P. W., Robertson, R. L., & Burrows, P. E. (1997). Thrombocytopenic coagulopathy (Kasabach-Merritt phenomenon) is associated with kaposiform hemangioendothelioma and not with common infantile hemangioma. *Plastic and Reconstructive Surgery*, *100*(6), 1377-1386. https://journals.lww.com/plasreconsurg/fulltext/1997/11000/thrombocytopenic_coagulopathy__kasabach_merritt.1.aspx

Tlougan, B. E., Lee, M. T., Drolet, B. A., Frieden, I. J., Adams, D. M., & Garzon, M. C. (2013). Medical management of tumors associated with Kasabach-Merritt phenomenon. *Journal of Pediatric Hematology/Oncology*, *35*(8), 618–622. https://doi.org/10.1097/MPH.0b013e318298ae9e

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