# Kaposiform Hemangioendothelioma With Kasabach-Merritt Syndrome Mistaken for Child Abuse in a Newborn

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## Practice Points

- Vascular tumors in dermatology may mimic child abuse.
- Even when all signs favor abuse, a nonresolving lesion should alarm clinicians to consider an alternate diagnosis.
- Kasabach-Merritt syndrome is a serious and potentially life-threatening condition that requires prompt evaluation.

*Kaposiform hemangioendothelioma is a rare vascular neoplasm of childhood that may have an alarming and potentially misleading clinical presentation. Awareness of this entity is important to provide appropriate and immediate medical care. We report the case of a 24-day-old female newborn who presented with a large bruiselike lesion on the left leg. A diagnosis of cellulitis suspected to be secondary to child abuse was made and the patient subsequently was placed in foster care; however, the lesion did not resolve after treatment and relocation. On reevaluation at our institution, physical examination revealed a round, 3×4-cm, violaceous, indurated, fixed, nonblanching, nontender plaque with an ivory center and peripheral erythema over the anteromedial aspect of the left leg. Biopsy demonstrated a vascular neoplasm consistent with kaposiform hemangioendothelioma (KHE), and laboratory evaluation revealed thrombocytopenia, low fibrinogen levels, and elevated D-dimer levels, confirming a diagnosis of Kasabach-Merritt syndrome (KMS).*

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## Case Report

A 24-day-old female newborn presented to a hospital with a large bruiselike lesion on the left leg. A diagnosis of cellulitis suspected to be secondary to child abuse was made and the patient subsequently was placed in foster care; however, the lesion did not resolve after treatment and relocation. At 69 days of age, the patient was readmitted, now to our hospital, after the lesion persisted and had progressively expanded. Physical examination revealed a round, 3×4-cm, violaceous, indurated, fixed, nonblanching, nontender plaque with an ivory center and peripheral erythema on the anteromedial aspect of the left leg (Figure 1). Ultrasonography showed hyperemia and infiltration of the soft tissues of uncertain significance, and a punch biopsy was unremarkable.

The patient was discharged to foster care, only to follow up 3 weeks later with the lesion unchanged. A deeper biopsy was performed and revealed a lobular vascular neoplasm composed of glomeruloid tufts of small blood vessels surrounded by ectatic spaces lined with thin-spindled endothelial cells extending from the mid dermis to the subcutaneous tissue; these findings were consistent with kaposiform hemangioendothelioma (KHE) and tufted

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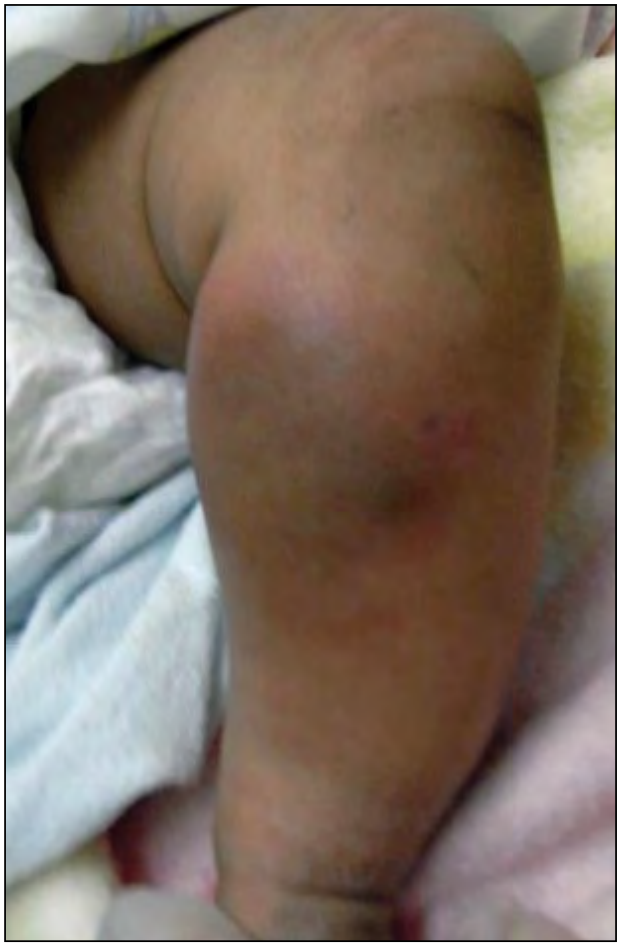

**Figure 1.** A round, 3×4-cm, violaceous, indurated, fixed, nonblanching, nontender plaque with an ivory center and peripheral erythema on the anteromedial aspect of the left leg.

angioma (Figure 2). Magnetic resonance angiography (Figure 3) and magnetic resonance imaging showed an infiltrative soft tissue mass with arterial phase enhancement and dilated feeding vessels characteristic of KHE. Abdominal sonography did not demonstrate additional angiomas. These features along with hematologic findings of severe thrombocytopenia (platelet count, 50,000/μL [reference range, 150,000–400,000/μL]), low fibrinogen levels, and elevated D-dimer levels led to a diagnosis of Kasabach-Merritt syndrome (KMS).

The patient was started on high-dose prednisone (24 mg and 3 mg/kg daily) and was discharged to follow-up with the pediatric hematology and oncology department. The investigation of child abuse was closed. Within 1 month, steroid therapy resolved the coagulopathy, though minimal changes to the vascular neoplasm were noted. Steroids were continued for months with no further impact on

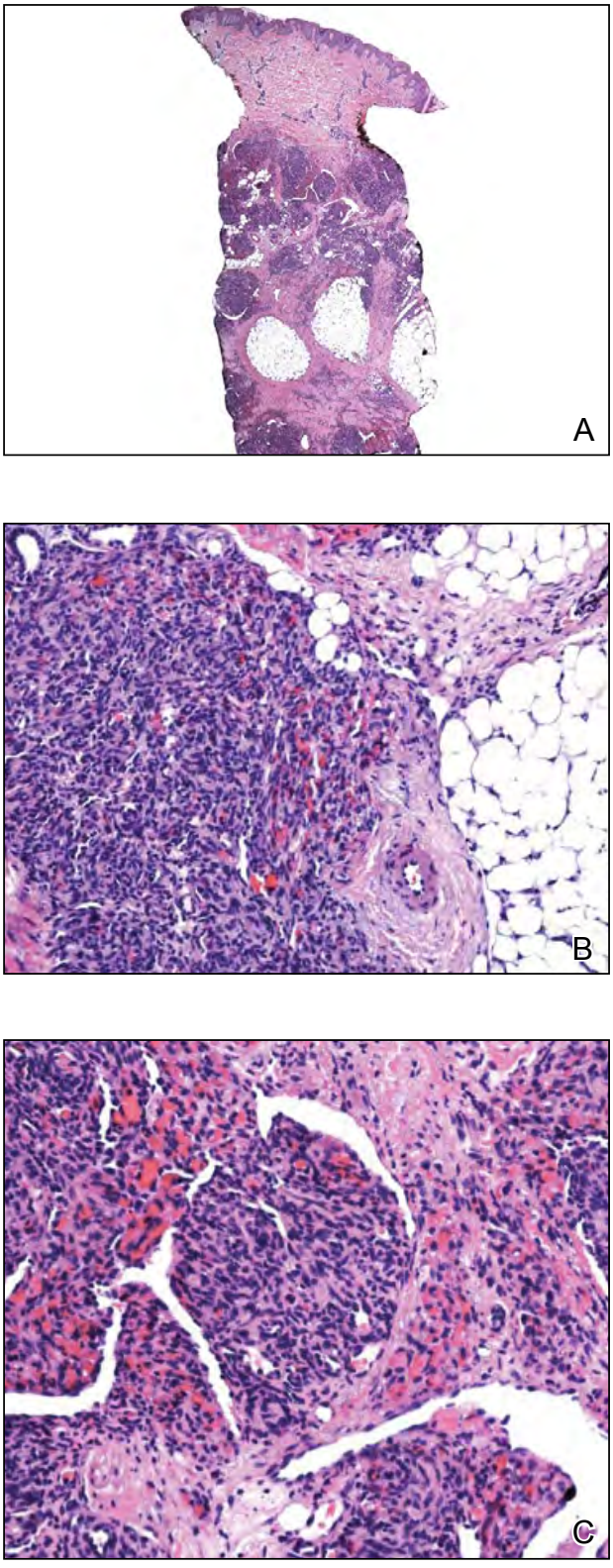


**Figure 2.** Low- and high-power views demonstrating a lobular vascular neoplasm composed of glomeruloid tufts of small blood vessels surrounded by ectatic spaces lined by thin-spindled endothelial cells extending from the mid dermis to the subcutaneous tissue (H&E; original magnifications ×4 [A], ×10 [B], and ×20 [C]).





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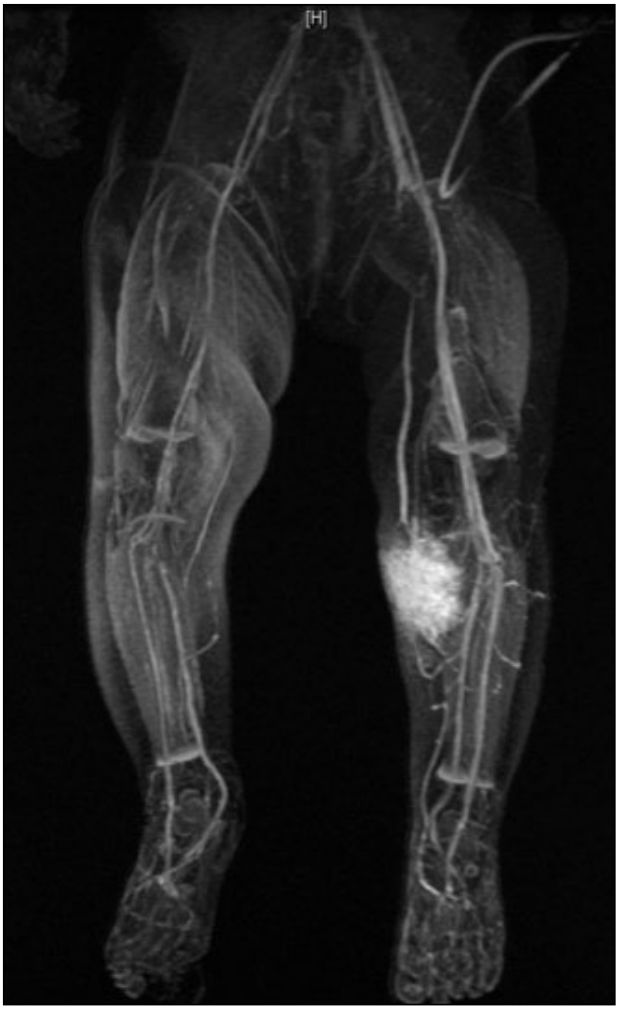


**Figure 3.** Magnetic resonance angiography demonstrated a vascular neoplasm.

the lesion. Treatment with vincristine (0.4 mg or 0.05 mg/kg weekly) was initiated for a total of 14 doses and the steroids were tapered with an excellent clinical response. Specifically, the patient was able to ambulate and there was no evidence of discoloration, induration, or pain; however, despite clinical improvement, the only notable radiologic changes on magnetic resonance angiography and magnetic resonance imaging included decreased vascularity of the tumor and slight decrease in size. The patient was referred to an institution specializing in vascular birthmarks for further treatment options.

## Comment

Kaposiform hemangioendothelioma is a rare vascular tumor that presents almost exclusively in childhood, with approximately half of cases arising during the first year of life alone.[1] Most commonly, lesions are blue-red in color, are located on the extremities, and are found in both the superficial or deep soft tissue. Most cases of KHE, especially deep tumors and retroperitoneal variants, are associated with a thrombocytopenic consumption coagulopathy, also known as KMS.

Kaposiform hemangioendothelioma traditionally has been distinguished from tufted angioma, another rare vascular tumor; however, because of their overlapping clinical, histologic, and immunophenotypic features, as well as their unique association with KMS, it is believed these entities are closely related, if not identical tumors, with tufted angioma representing a mild superficial form of KHE.[1-4]

Histopathologically, KHE shares features with capillary hemangioma and Kaposi sarcoma.[1,4] Both glucose transporter 1 and Lewis-Y antigen, 2 common markers expressed in juvenile hemangiomas, are absent in KHE. In further contrast to juvenile hemangiomas, KHE does not regress. The outcome largely depends on the site, clinical extent, and presence of KMS.[4] Single cutaneous lesions tend to exhibit a more benign clinical course in contrast to KHE found in additional locations.[2] Mortality generally is associated with local aggressiveness and presence of KMS,[2,5] occurring in approximately 10% of patients.[1]

Therapies for KHE are suboptimal; they vary depending on the size and location of the lesion as well as the presence of KMS and are associated with a range of adverse outcomes and success rates.[1,2,5,6] Complete surgical excision and arterial embolization have been effective but often are impractical. Surgery should be considered the first-line treatment of KHE; however, technical difficulties from locally aggressive lesions and the presence of KMS can limit its feasibility.[1,2,6] Embolization is minimized by the hardship of cannulating small feeder vessels.[2] Corticosteroids (2–3 mg/kg daily) can be effective when treating KMS[6] but often do not lead to resolution of KHE, ultimately requiring higher doses with a host of negative steroid sequelae such as limited growth and osteoporosis.[2] Interferon alfa has been used with limited success and is associated with neurologic effects such as spastic diplegia[2,6]; thus it should be reserved for severe cases, used for shorter periods of time, and monitored with close neurologic follow-up. Vincristine has been reported to improve KHE and KMS, especially after treatment failures with corticosteroids and interferon alfa.[2,5] Factors to consider when using vincristine include its hepatic metabolism, the need for a central venous catheter, and the risk for hematologic and neurologic complications (eg, myelosuppression, peripheral neuropathy).[2] However, if dosed appropriately, it appears to be well tolerated in the pediatric population.[2]



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## Conclusion

Kaposiform hemangioendothelioma is a rare vascular neoplasm of childhood that may have an alarming and potentially misleading clinical presentation. Awareness of this entity is important to provide appropriate and often immediate medical care.

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