

# Denouement and Discussion

## Kaposiform Hemangioendothelioma With Kasabach-Merritt Phenomenon

**K**aposiform hemangioendothelioma (KHE) is a locally aggressive rare vascular tumor that was described in 1979 by Pearl and Mathews<sup>1</sup> and further characterized and named *kaposiform hemangioendothelioma* by Zukerberg et al<sup>2</sup> in 1993.

The clinical features of KHE are quite distinguishable from other pediatric endothelial tumors. Kaposiform hemangioendothelioma is evident at birth or appears shortly after in the early infantile period. Rarely, KHE develops in adults.<sup>3</sup> Unlike the more common infantile hemangioma that has a definite predilection to affect female infants, KHE has no significant sex predilection.<sup>3-5</sup>

The histological features of KHE are characterized by a predominant Kaposi sarcoma-like, fascicular spindle cell growth pattern of epithelial cells, infiltrating nodules with slitlike or crescentic vessels that are poorly canalized.<sup>2</sup> Clinically, KHE presents with an infiltrative ecchymotic induration of the skin and underlying soft tissue. The lesion is solitary with predilection to the trunk, extremities, retroperitoneum, and head and neck.<sup>6</sup> Kaposiform hemangioendothelioma has a mortality rate of approximately 24% related to the coagulopathy or complication of local tumor infiltration.<sup>3</sup>

Severe thrombocytopenia (Kasabach-Merritt phenomenon) is a major cause of morbidity and mortality and is encountered in the majority of patients. Kasabach-Merritt phenomenon can also be associated with hypofibrinogenemia/fibrinolysis and anemia with normal or minimally elevated prothrombin and activated partial thromboplastin times.<sup>7</sup> Low-flow vascular anomalies (such as venous and extensive lymphatic malformations) may also cause consumptive coagulopathy characterized by elevated D-dimer level, low fibrinogen level, and normal to mildly decreased platelet count.<sup>3</sup> The combination of clinical, laboratory, and imaging findings in KHE is usually sufficient to establish the diagnosis. The patient presented herein was considered to have KHE with Kasabach-Merritt phenomenon on the basis of the classic cutaneous findings, anatomical location, age at presentation, and severe thrombocytopenia.

Chemotherapy, including the use of vincristine, cyclophosphamide, corticosteroids, interferon alfa-2a or -2b, and other agents, has been reported to be effective in Kasabach-Merritt phenomenon. However, no single treatment has given consistently reproducible results (defined as normalization of the platelet count and shrinkage of the tumor).<sup>4</sup>

**Accepted for Publication:** December 29, 2009.

**Correspondence:** Ahmad I. Alomari, MD, MSc, Division of Vascular and Interventional Radiology, Children's Hospital Boston, 300 Longwood Ave, Boston, MA 02115 (ahmad.alomari@childrens.harvard.edu).

**Author Contributions:** *Study concept and design:* A. K. Alomari and A. I. Alomari. *Acquisition of data:* A. K. Alomari and A. I. Alomari. *Analysis and interpretation of data:* A. K. Alomari and A. I. Alomari. *Drafting of the manuscript:* A. K. Alomari and A. I. Alomari. *Critical revision of the manuscript for important intellectual content:* A. K. Alomari and A. I. Alomari. *Study supervision:* A. K. Alomari and A. I. Alomari.

**Financial Disclosure:** None reported.

### REFERENCES

1. Pearl GS, Mathews WH. Congenital retroperitoneal hemangioendothelioma with Kasabach-Merritt syndrome. *South Med J.* 1979;72(2):239-240.
2. Zukerberg LR, Nickoloff BJ, Weiss SW. Kaposiform hemangioendothelioma of infancy and childhood: an aggressive neoplasm associated with Kasabach-Merritt syndrome and lymphangiomatosis. *Am J Surg Pathol.* 1993;17(4):321-328.
3. Sarkar M, Mulliken JB, Kozakewich HP, Robertson RL, Burrows PE. Thrombocytopenic coagulopathy (Kasabach-Merritt phenomenon) is associated with Kaposiform hemangioendothelioma and not with common infantile hemangioma. *Plast Reconstr Surg.* 1997;100(6):1377-1386.
4. Enjolras O, Mulliken JB, Wassef M, et al. Residual lesions after Kasabach-Merritt phenomenon in 41 patients. *J Am Acad Dermatol.* 2000;42(2, pt 1):225-235.
5. Lyons LL, North PE, Mac-Moune Lai F, Stoler MH, Folpe AL, Weiss SW. Kaposiform hemangioendothelioma: a study of 33 cases emphasizing its pathologic, immunophenotypic, and biologic uniqueness from juvenile hemangioma. *Am J Surg Pathol.* 2004;28(5):559-568.
6. Konez O, Burrows PE. Magnetic resonance of vascular anomalies. *Magn Reson Imaging Clin N Am.* 2002;10(2):363-388.
7. Mulliken JB, Anupindi S, Ezekowitz RA, Mihm MC Jr. Case records of the Massachusetts General Hospital: weekly clinicopathological exercises. Case 13-2004: a newborn girl with a large cutaneous lesion, thrombocytopenia, and anemia. *N Engl J Med.* 2004;350(17):1764-1775.