

# 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>

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