

Kasabach-Merritt phenomenon (KMP) is a rare condition that is associated with a coagulopathy with features including profound thrombocytopenia (low platelets), hypofibrinogenemia (low fibrinogen), and anemia. This phenomenon is only associated with two rare vascular tumors: kaposiform hemangioendotheliomas and tufted angiomas. This condition can be life threatening secondary to the risk of bleeding and progression to DIC (disseminated intravascular coagulopathy). Initially a vascular lesion is noted on the skin which can be firm, indurated and purpuric. Areas of petechiae (tiny red dots) can appear around the lesion or on other parts of the body. If the vascular lesion is internal, these petechiae can be seen on the skin. Bruising and spontaneous bleeding can also occur. These tumors are not hemangiomas. They usually present in young infants, less than three months of age, but have rarely been reported in older children. These tumors occur in the extremities, chest, neck, abdomen and pelvis. They infiltrate across tissue planes and can be aggravated by interventions, infection and trauma. When these tumors with KMP are internal such as in the pleural or retroperitoneum, they can cause significant morbidity and mortality. The morbidity and mortality is caused by bleeding. The cause of Kasabach-Merritt phenomenon is unknown. It is believed to be secondary to sequestration or trapping of platelets into the tumor. These tumors are made up of abnormal endothelial cells (spindle cells) and also lymphatic malformation. It is unclear why the KMP occurs and if it is caused by the spindle cells or the lymphatic component. Kasabach-Merritt phenomenon is a rare disorder that affects males and females equally. The diagnosis is most often made during infancy but older children have been reported with this phenomenon. KHE and TA tumors can occur without KMP. The reason for this is still unknown and may be secondary to a smaller size of the tumor, an older age at presentation or other clinical features. The diagnosis of Kasabach-Merritt phenomenon is based on the diagnosis of Kaposiform hemangioendothelioma/tufted angioma and this coagulopathy as noted above. If this diagnosis is suspected blood work including a CBC with differential and platelets, fibrinogen, D-dimer, PT, and PTT should be ordered. The best imaging modality to assess the extent of the lesion is a MRI with contrast. A biopsy will confirm the diagnosis.