Kaposiform hemangioendothelioma is a rare, potentially life-threatening vascular tumor often associated with a coagulopathy known as Kasabach–Merritt phenomenon (KMP). We report the case of an infant with satellite lesions around a large vascular tumor and associated KMP.

Key words: Hemangioma, hemagioendothelioma, kaposiform, satellitosis

INTRODUCTION

Kaposiform hemangioendothelioma (KHE) is a rare vascular tumor that usually affects soft tissues of newborns and infants. The prognosis of this neoplasm is related to the risk of development of severe consumptive coagulopathy or Kasabach–Merritt phenomenon (KMP), which is associated with high mortality. Although it lacks metastatic potential, KHE is locally aggressive because of its tendency to infiltrate surrounding tissues.

CASE REPORT

An 8-month-old child presented with a 5 months history of slowly enlarging painful swelling over left leg and thigh. It was followed by development of few small lesions in vicinity. Swelling was associated with occasional bleeding on local trauma and the child had difficulty in extending left leg. There was no history of purpura, petechiae, hematuria, hematemesis, melena, bleeding from gums, or epistaxis. On examination, an erythematous, well demarcated tumor of approximately 10 cm × 15 cm was seen over lower part of left thigh, knee, and upper part of leg [Figure 1]. The tumor had regular border, smooth surface and was associated with hypertrichosis. On palpation, it was warm, tender, and indurated. Few similar nodular lesions of smaller size were present in its vicinity. Auscultation of the lesion revealed no bruit or abnormal pulsations.

The differential diagnosis included various vascular tumors such as congenital hemangioma, KHE, tufted angioma, hemangiopericytoma, angiosarcoma, arteriovenous malformations, and hemangioblastoma.

Routine hematological examination revealed a low platelet count (50,000/µL) which decreased to 15,000/µL over next 4–5 days. Other hematological and biochemical investigations were normal. Coagulation profile including bleeding time, clotting time, prothrombin time, activated partial thromboplastin time, d-Dimer and fibrinogen levels were normal. Skin biopsy was not performed due to the risk of bleeding. Ultrasound whole abdomen was normal. Magnetic resonance imaging (MRI) of left
thigh revealed an ill-defined marginated infiltrative fairly large heterogenous signal intensity (increased T2 and decreased T1) lesion involving the inter and intramuscular compartment of anterior, lateral, and medial aspect of thigh associated with dermal and subcutaneous thickening and infiltration extending into the adjacent muscles with edema, suggestive of KHE [Figure 2]. The patient was put on oral steroid 1 mg/kg/day and intravenous vincristine 0.05 mg/kg weekly after the consultation with pediatrician. After 2 weeks, there was a slight decrease in the size of tumor and the platelet count raised to 80,000/µL. The child was then lost to follow-up.

DISCUSSION

KHE involves a spectrum of lesions from small, superficial tumors to large, infiltrative lesions with life-threatening complications including KMP.[4] It usually occurs as a solitary lesion[5] though cases with multiple cutaneous lesions[6] and involving different organs[7] have been described. Metastatic disease has not been reported.[8] Satellitosis has been described with other forms of hemangioendothelioma, esp. composite with epithelioid pattern being predominant,[4] but it has not been described with KHE. Although, KHE lacks metastatic potential, it is classified within hemangioendothelioma because of its potential to cause death of the patient.[9]

In 1940, Kasabach and Merritt described an infant with a “capillary hemangioma” with extensive purpura and profound thrombocytopenia.[2] KMP is associated exclusively with either KHE or tufted angioma.[4] It is a serious clinical condition, with a mortality of 24%. Maximum cutaneous diameter >8 cm and extension into multiple anatomic regions confer an increased risk of KMP.[10] The large tumor and inter and intramuscular invasion in our patient predisposed him to the risk of KMP.

The diagnosis of KHE is confirmed by either histopathology or by characteristic MRI findings.[8] In our case because of possible risk of bleeding, we relied on MRI and did not opt for biopsy. The pharmacologic treatment of KHE, although not always curative, aims to decrease tumor size and correct coagulopathy. The various treatment options include oral and intravenous corticosteroids, vincristine, cyclophosphamide, proparanol, sirolimus, IFN-α, and anti-fibrinolytic agents. For cases of KHE associated with KMP, first-line therapy with intravenous vincristine 0.05 mg/kg once weekly and oral prednisolone 2 mg/kg/day or intravenous methylprednisolone 1.6 mg/kg/day is recommended.[10]

We report this case for its relative rarity, the presence of satellite lesions and the fact that a missed diagnosis can have catastrophic consequences for the patient.

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Conflicts of Interest
There are no conflicts of interest.

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