

and normal liver and renal function. Peripheral blood smear showed normocytic normochromic erythrocytes and markedly reduced platelets. Bone marrow evaluation revealed normal erythropoiesis, myelopoiesis and megakaryocytic hyperplasia. Contrast enhanced computed tomography (CT) of chest and abdomen excluded deep seated visceral hemangiomas. She received platelet transfusions, oral tranexamic acid for control of gum bleeds and was also started on oral prednisolone 1mg/kg/day. After one month of starting steroid treatment, bleeding had stopped and platelet count had improved to $152 \times 10^9/l$. However, the hemangiomas had remained the same. Considering risk of traumatic bleeding, she was advised surgical excision of tongue hemangiomas. However, she was unwilling for surgery.

KMS is most commonly reported in infants and only a small percentage ($\approx 0.3\%$) of infants with hemangiomas develop KMS.¹ If not recognized and treated in time, KMS may be potentially fatal by causing disseminated intravascular coagulation and severe bleeding and large hemangiomas can cause high output cardiac failure and vital organ compression. Though rare, KMS has been reported among adults as well.^{2,3,4,5} The pathogenesis involves activation and consumption of platelets and clotting factors inside hemangiomas, giving rise to consumption coagulopathy and bleeding. However, no correlation has been reported between site, size and number of hemangiomas and development of KMS.¹ Cutaneous and visceral hemangiomas have both been implicated in KMS. Work-up in the index patient revealed probable low-grade disseminated intravascular coagulation, but there was no evidence of microangiopathic hemolysis. Histologically, kaposiform hemangioendotheliomas and tufted angiomas are the most frequent lesions reported in KMS.¹ Treatment options include compression therapy for hemangiomas, surgical excision of large solitary vascular lesions (whenever feasible) or ligation/embolization of feeder vessels, when lesions are inaccessible for surgery. Systemic steroids and interferon alfa have shown benefit when vascular lesions are extensive and not amenable to surgery or embolization.

References:

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Figure 1:

Two large purple hemangiomas of sizes 3 cm × 8 cm and 2 cm × 3 cm on the dorsum of protruded tongue (Figure 1A) and a smaller cutaneous hemangioma on the right side of face (Figure 1B)