

Tufted angioma with recurrent Kasabach–Merritt phenomenon

Sir,

A 5-month-old male child presented with an erythematous plaque on the right cheek extending to the mandibular area and neck. The lesion was noticed at birth and had gradually increased in size with rapid growth over the past 2 months. There was no history of ulceration or bleeding. He did not have any associated systemic complaints. Examination revealed a dusky erythematous, firm and tender plaque of 10 cm × 14 cm size with surface nodularity on the right cheek and extending to the right side of the neck and chest [Figure 1a and b].

The child was admitted with a clinical diagnosis of vascular tumor, probably infantile hemangioma. Routine hematological investigations were within normal limits. Cutaneous ultrasonogram of the lesion showed extensive arterial vascularity, diffuse infiltration and an echogenic volume of 73.2 ccs suggestive of a proliferative hemangioma. Magnetic resonance imaging of the affected area revealed a large proliferative hemangioma on the right side of the face and upper neck involving the skin, subcutaneous tissue and muscles of the right infratemporal fossa, right premaxillary and premandibular region, chin and upper cervical region. The brain and airways were normal [Figure 2]. Skin biopsy from the margin of the plaque showed nodular aggregates of proliferating endothelial



Figure 1a: Dusky erythematous plaque over the right cheek extending to the right side of neck

cells, capillaries and lymphatics in the upper, mid- and lower dermis giving a ‘cannon ball’ appearance, suggestive of tufted angioma [Figure 3a and b]. With these clinical findings and investigations, a final diagnosis of tufted angioma was made.

During his hospital stay, the infant developed a sudden increase in the size of the plaque with extension up to the infraorbital region. Moreover, there was an increase in erythema and firmness of the plaque along with some difficulty in feeding [Figure 1c and 1d]. A coagulation profile showed reduced platelets (23,000 cells/mcL) and fibrinogen (110 mg/dl), increased prothrombin time (15.1 s.) and D-dimer levels (>0.5 µg/ml). These findings along with the clinical features were suggestive of Kasabach–Merritt phenomenon. No episode of bleeding or



Figure 1b: Dusky erythematous plaque over the right cheek extending to the right side of neck and chest

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Figure 1c: Sudden increase in the size, erythema and firmness of the plaque, suggestive of Kasabach-Merritt phenomenon



Figure 1d: Extension of the swollen plaque up to the infra-orbital region

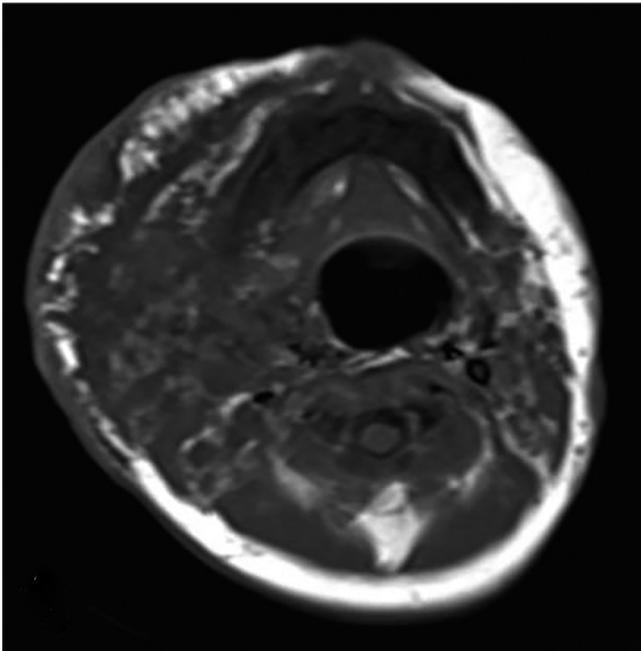


Figure 2a: Magnetic resonance imaging of head and neck: T1 weighted axial images at the level of the alveolar arch showed a large transcompartmental soft tissue mass lesion on the right side of face with infiltrative margins involving multiple neck spaces including the subcutaneous planes and muscles

ecchymosis was present. He was managed with weekly intravenous injections of vincristine (0.05 mg/kg) 0.35 mg/week, daily oral prednisolone (3 mg/kg/day) 20 mg, daily oral aspirin (10 mg/kg) 70 mg, one unit of fresh frozen plasma and platelet rich plasma. He recovered partially, however, he developed a similar episode within 3 days and hence daily oral propranolol was added at a dose of 2 mg/kg/day (14 mg). He continued to show similar fluctuation in clinical course in the next 14 days. Later, he was transferred to the pediatric intensive care unit. He showed partial clinical

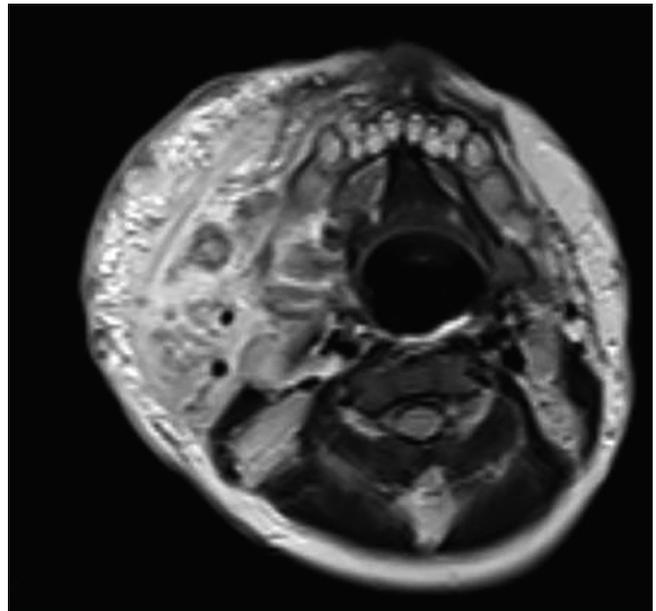


Figure 2b: Magnetic resonance imaging of head and neck: T2 weighted axial images at the level of the alveolar arch showed changes similar to those noted on the T1 weighted image

response and was discharged after 2 weeks on weekly injection vincristine and daily oral prednisolone, aspirin and propranolol. The child was lost to follow-up thereafter.

Tufted angioma is a rare vascular tumor that presents as a solitary, firm, dusky erythematous nodule. It shows an initial growth phase followed by a static course. Kasabach-Merritt syndrome/phenomenon, also known as “platelet trapping syndrome,” is an important complication of tufted angioma. It is characterized by the triad of consumptive coagulopathy, microangiopathic hemolytic anemia and rapidly enlarging vascular tumor with darkening of the lesion.^{1,2}

Kaposiform hemangioendothelioma and tufted angioma are the common primary causes of Kasabach–Merritt phenomenon. Infantile hemangiomas are not associated with this complication. In a retrospective review of 19 cases of Kasabach–Merritt syndrome, Kaposiform hemangioendotheliomas were seen in 18 (94.7%) cases and tufted angioma in 1 (5.3%).³

The treatment of choice is complete surgical excision or embolization by an interventional radiological procedure, limiting the tumor's blood supply.⁴ As surgical options may not be feasible in all cases, systemic treatment may be required. Systemic corticosteroids (prednisolone 1–5 mg/kg/day) and vincristine (0.05 mg/kg/week intravenous) are the mainstay of

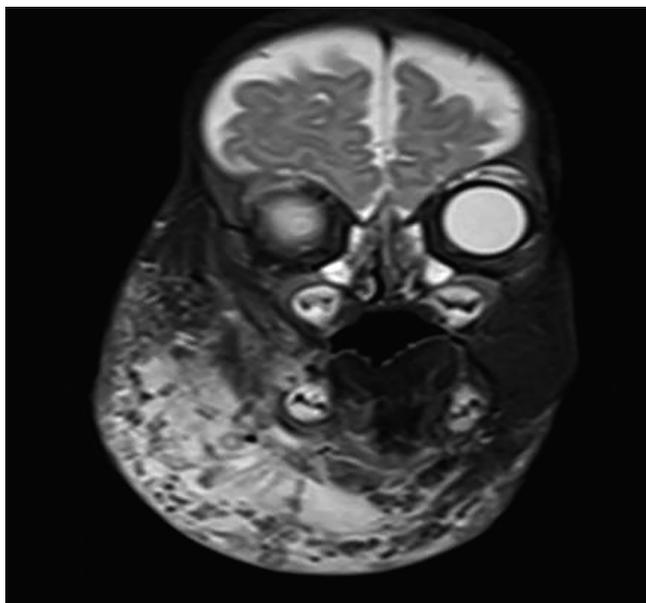


Figure 2c: Magnetic resonance imaging of head and neck: Coronal T2 weighted MR image better shows the extent of the lesion

medical therapy for Kasabach–Merritt syndrome.⁴ Other useful drugs are interferon- α and antiplatelet drugs. Interferon- α is used at a dose of 0.5–10 million units/day or 3–4 times a week for 2–7 months. Ticlopidine (10 mg/kg/day) and aspirin (10 mg/kg/day) are the most commonly used antiplatelet drugs, although the risk of bleeding should be considered. Propranolol, sirolimus, human angiostatin and pegylated human megakaryocyte growth and development factors have also been tried. Coagulopathy and thrombocytopenia are managed by transfusion of platelets and cryoprecipitate/fresh frozen plasma.^{4,5}

The interesting feature in the present case was the multiple episodes of Kasabach–Merritt syndrome, which is quite rare. A combinational and multidisciplinary management approach is required due to the risk of serious, life-threatening complications such as shock and embolism.

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Conflicts of interest

There are no conflicts of interest.

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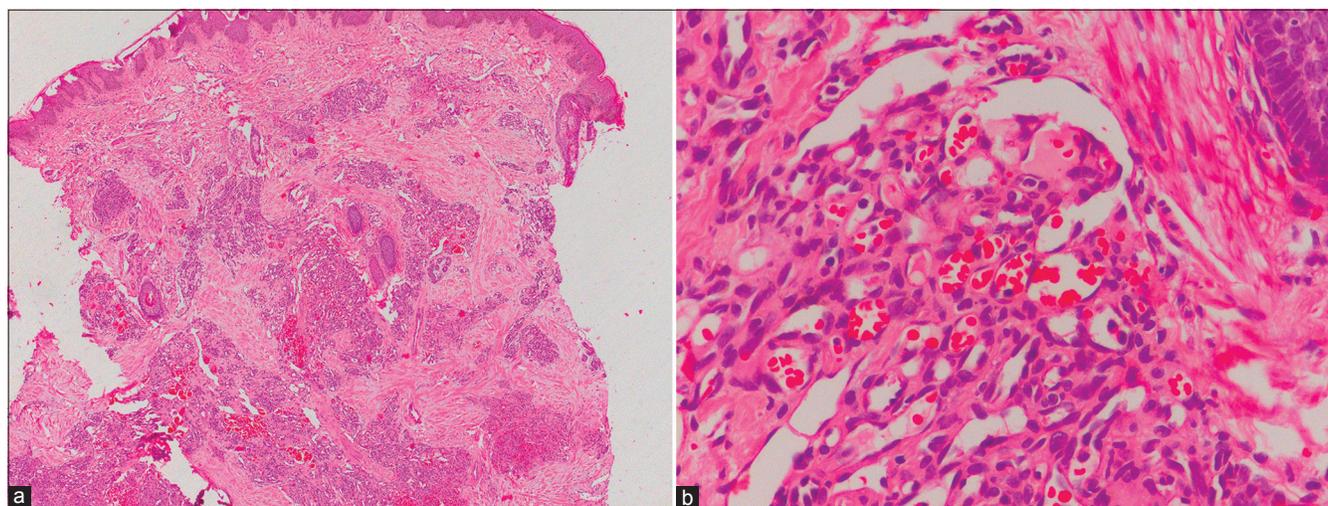


Figure 3: Skin biopsy shows (a) nodular aggregates in the upper, mid and lower dermis giving a cannon ball appearance (H and E, $\times 100$). (b) These nodular aggregates are composed of proliferating endothelial cells, capillaries and lymphatics (H and E, $\times 400$)

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