A rapidly growing violaceous tumor in a neonate

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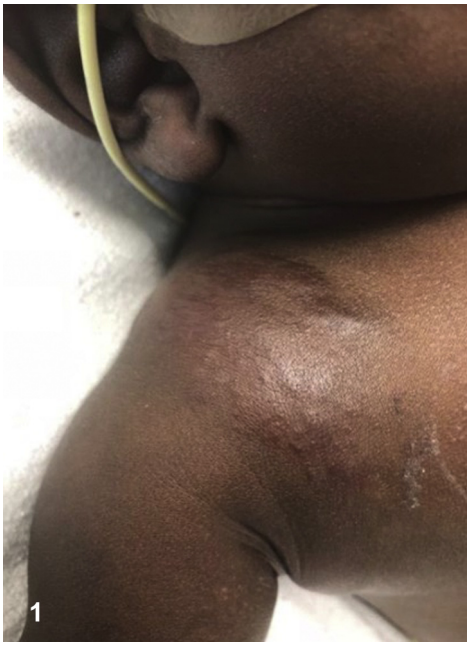

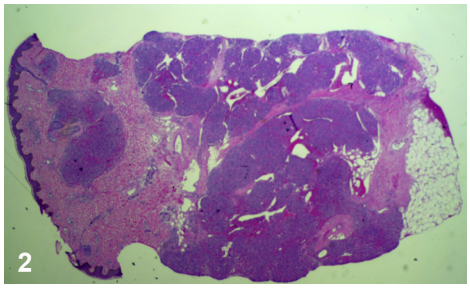

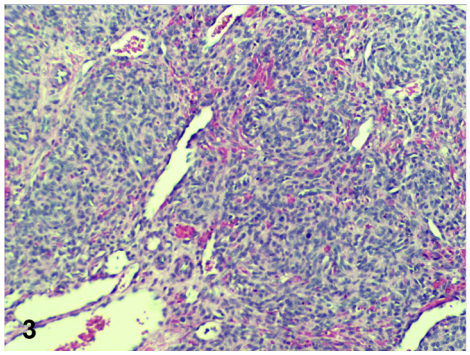

A 7-month-old boy with a complex medical history presented for a bump on his right shoulder. His mother attributed the lesion to an attempt at venous access with multiple pokes 5 months earlier, adding that it had rapidly grown over the previous week. Examination found a solitary, firm, violaceous plaque (Fig 1). A 4-mm punch biopsy specimen showed large lobules of spindled and epithelioid endothelial cells involving the dermis and extending to the superficial subcutis (Fig 2). Slitlike and staghorn vascular spaces were visible (Fig 3).

Question 1: What is the most likely diagnosis?

A. Blue-rubber bleb nevus (BRBN) syndrome

B. Infantile hemangioma

C. Kaposiform hemangioendothelioma (KHE)

D. Kaposi sarcoma

E. Tufted angioma

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Answers

A. BRBN — Incorrect. BRBN presents as small, multifocal venous malformations rather than as a large, unifocal lesion as seen in this patient.

B. Infantile hemangioma — Incorrect. Infantile hemangiomas usually present as red to crimson papules, plaques, or nodules. Deeper lesions have a bluish violaceous hue. They express an unusual endothelial phenotype that stains positive for GLUT-1, a feature shared only by placental capillaries, and lack fascicles of spindle cells.[1-3]

C. KHE — Correct. KHE is a rare vascular tumor that usually involves the skin but can also be found within deeper structures (retroperitoneum, thoracic cavity, and muscle). Lesions appear as large (>5 cm), unifocal plaques with ill-defined borders, involving multiple planes of tissue. KHE typically presents in infants and children, although cases are reported in adults. The tumors are firm to palpation and violaceous in color, with a predilection for the proximal extremities, trunk, and retroperitoneal regions. Histopathology reveals confluent lobules of neoplastic, spindled endothelial cells that stain positively for Podoplanin/D2-40 and negatively for GLUT-1 and HHV-8.[1-3] Although rare, there are other reports of antecedent trauma, as seen in our patient.[4]

D. Kaposi sarcoma — Incorrect. Kaposi sarcoma is distinct pathologically in that it lacks the lobularity of KHE and contains plasma cell infiltrates. HHV-8 immunostaining is also highly sensitive and specific for Kaposi sarcoma.[2]

E. Tufted angioma — Incorrect. Although the histologic features of KHE and tufted angioma overlap, tufted angioma is thought to be a more superficial form of KHE, involving only one plane of tissue. Tufted angiomas typically present with firm, erythematous macules. They are distinguishable from KHE histologically by their scarcity of endothelial spindling and an absence of pericyte-rich epithelioid nodules.

Question 2: What is the most feared complication in this patient?

A. Intussusception and volvulus

B. Kasabach-Merritt phenomenon

C. LUMBAR/PELVIS/SACRAL syndrome

D. Metastases

E. PHACE syndrome

Answers

A. Intussusception and volvulus — Incorrect. Intussusceptions are associated with gastrointestinal involvement of BRBN; they are not associated with KHE.[1]

B. Kasabach-Merritt phenomenon — Correct. KHE is associated with Kasabach-Merritt phenomenon, a complication characterized by profound thrombocytopenia, microangiopathic hemolytic anemia, and hypofibrinogenemia (<100 mg/dL). Endothelial cell abnormalities and vascular stasis within the tumor lead to intralesional platelet trapping and clotting factor activation, setting off the coagulation cascade. Microangiopathic hemolytic anemia then results as erythrocytes undergo mechanical trauma travelling through the tumor's partially thrombosed vascular channels. If not quickly diagnosed and treated, this complication of KHE can be life threatening.[1-3]

C. LUMBAR/PELVIS/SACRAL syndrome — Incorrect. Infantile hemangiomas located over the lumbar or sacral spine may be associated with genitourinary or anorectal anomalies or neurologic issues such as a tethered cord. These anomalies are not associated with KHE.[2]

D. Metastases — Incorrect. KHE does not metastasize but may be locally destructive and disfiguring.[2]

E. PHACE syndrome — Incorrect. PHACE syndrome encompasses large, segmental infantile hemangiomas and brain malformations as well as arterial, cardiac, and eye anomalies. This constellation of findings is not seen with KHE.[1,2]

Question 3: What is the first-line treatment for the complication mentioned in question 2?

A. Endoscopic sclerotherapy

B. Propranolol therapy

C. High-dose systemic corticosteroids

D. Interferon-α

E. Vincristine

Answers

A. Endoscopic sclerotherapy — Incorrect. This procedure might be an appropriate treatment for blue-rubber bleb nevus syndrome complicated by gastrointestinal lesions with hemorrhage.

B. Propranolol therapy — Incorrect. Although this would be an appropriate approach to an infantile

hemangioma at risk for growth, ulceration, or disfigurement, it is not the first-line treatment for KHE because of mixed reports on efficacy.[2]

C. High-dose systemic corticosteroids — Incorrect. KHE lesions may not respond to steroid treatment.[2]

D. Interferon-α — Incorrect. Interferon-α is responsible for spastic diplegia in infants and should be avoided whenever possible in this age group, particularly as the data supporting its efficacy are insufficient.

E. Vincristine — Correct. Vincristine is a vinca alkaloid antimitotic agent that blocks formation of microtubules in cells. Intravenous vincristine has been found to decrease tumor size, reduce pain, and improve the aesthetic appearance of KHE.[1-3,5]

Abbreviation used:

KHE: kaposiform hemangioendothelioma

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