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Case Report

Kaposiform hemangioendothelioma of the thigh: A case report ☆

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ABSTRACT

Kaposiform hemangioendothelioma is a rare, locally aggressive or borderline vascular tumor that typically affects infants. It presents as a purpuric cutaneous lesion and may be associated with life-threatening coagulation disorders, such as the Kasabach-Merritt phenomenon. The differential diagnosis can be challenging based on clinical presentation alone. Imaging plays a crucial role in the diagnostic workup, particularly magnetic resonance imaging.

We present a case report of a 4-month-old patient with an enlarging vinous cutaneous mass on the thigh and coagulation abnormalities. Magnetic resonance imaging revealed a large, infiltrative, soft-tissue lesion with poorly defined margins and heterogeneous enhancement, that involved all muscle compartments of the thigh and was associated with lymphedema, stranding of the subcutaneous fat and cutaneous thickening. These findings were consistent with kaposiform hemangioendothelioma of the thigh and the diagnosis was confirmed by histopathological characterization.

Abbreviations: KHE, Kaposiform hemangioendothelioma; GLUT-1, Glucose transporter-1; HHV-8, Human herpesvirus-8; KMP, Kasabach-Merritt phenomenon; MRI, Magnetic resonance imaging; T2WI, T2-weighted images; T1WI, T1-weighted images; KLA, Kaposiform lymphangiomatosis.

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Introduction

Kaposiform hemangioendothelioma (KHE) is a rare, locally aggressive or borderline vascular tumor [1] that typically affects infants and presents as a purpuric cutaneous lesion that may appear in various locations [2,3]. Histologically, KHE forms ill-defined confluent nodules of spindled endothelial cells arranged in short fascicles with numerous slitlike vascular spaces. Mitoses are infrequent and cytologic atypia is absent. Tumor cells consistently express vascular and lymphatic endothelial markers and are negative for glucose transporter-1 (GLUT-1) and human herpesvirus-8 (HHV-8) [4,5].

KHE may be associated with life-threatening coagulation disorders, such as the Kasabach-Merritt phenomenon (KMP), which is a serious complication characterized by hemolytic anemia, thrombocytopenia, and coagulation abnormalities [2]. KMP is exclusively secondary to KHE or tufted angiomas [2] but KHE can occur without KMP in up to 43% of patients [6].

Clinical and laboratory findings may suggest KHE, but imaging plays a crucial role in the assessment and early diagnosis of patients with KHE, especially those without KMP [7]. Ultrasound is the first-line imaging modality for evaluating superficial soft tissue masses in pediatric patients, but it is unable to provide a specific diagnosis or assess the exact extent of the tumor, particularly its depth [2]. Magnetic resonance imaging (MRI) is the best imaging technique for assessing disease extent and is the preferred imaging modality for evaluating these tumors [7,8]. In addition, imaging is important for follow-up and monitoring therapeutic response [3]. Treatment of KHE is complex and requires specialized management within an interdisciplinary setting [8].

Case report

A 4-month-old patient presented with an enlarging mass on the right thigh with a vinous cutaneous colouration (Fig. 1), which was initially clinically diagnosed as an infantile hemangioma. The mass was present at birth and had grown rapidly over the following months. The patient's history included abnormal bleeding associated with vaccine inoculation. On physical examination, there was a large, vinous, cutaneous mass on the right thigh that was warm and tender, as well as ipsilateral leg and thigh edema and bilateral leg petechie. Laboratory tests revealed anemia, thrombocytopenia, and hypofibrinogenemia.

MRI of the right thigh was performed and revealed a large, infiltrative, soft-tissue lesion with poorly defined margins. The central portion of the mass appeared isointense on T2-weighted images (T2WI) and T1-weighted images (T1WI), with a more infiltrative peripheral portion showing significant hyperintensity on T2WI and T1WI. The lesion also displayed diffuse, heterogeneous enhancement after contrast administration (Fig. 2). It measured about 7 × 7 × 9 cm (maximum transverse, anteroposterior, and longitudinal measurements) and involved all muscle compartments of the thigh, with a focus on the medial compartment (Figs. 2 and 3). It extended anteriorly and medially into the subcutaneous tissue, along with diffuse, enhancing cutaneous thickening of the medial thigh. There was also diffuse soft tissue edema with evidence of reticular stranding in the subcutaneous fat, mostly perpendicular to the skin surface, which displayed avid enhancement after gadolinium administration (Fig. 2). Dilated femoral and external iliac veins were also noted. The arteries of the thigh and the femur showed no significant changes. Subcutaneous edema of the leg was also present. These MRI findings were consistent with a locally invasive vascular tumor of the thigh, specifically KHE.

Percutaneous biopsy for histopathological characterization was recommended and later performed, revealing morphological and immunohistochemical findings that were compatible with KHE (Fig. 4).

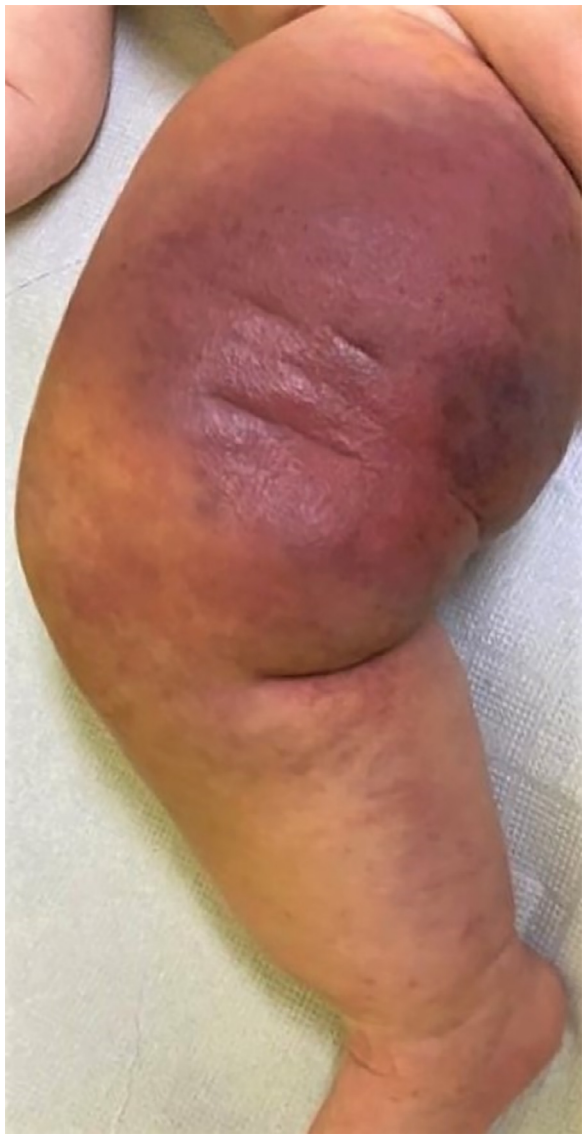

Fig. 1 – Photograph of the thigh lesion. There is a large mass with vinous cutaneous colouration on the right thigh, accompanied by leg and thigh edema and leg petechie.

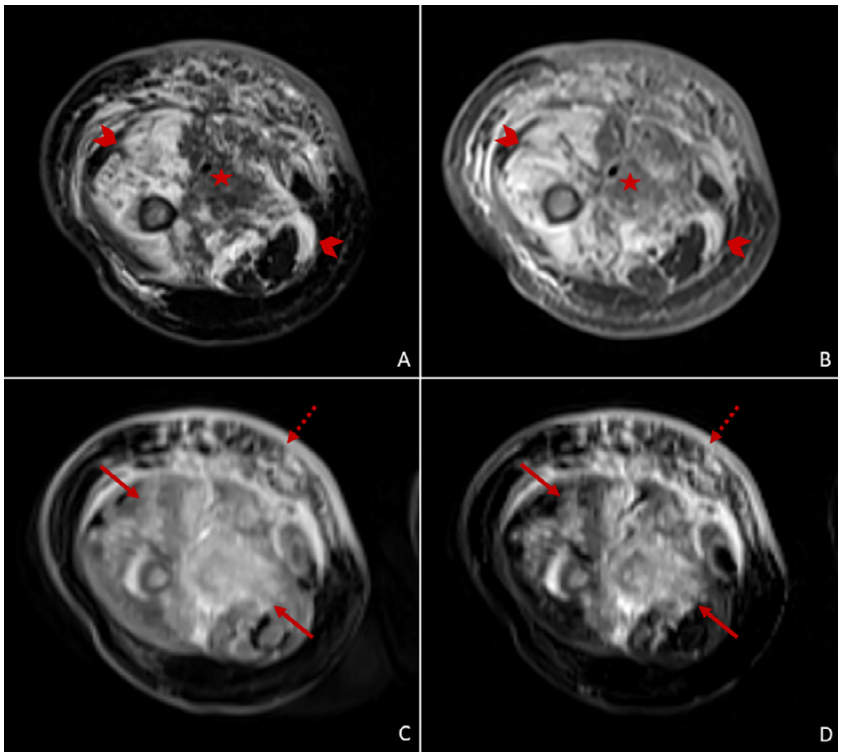

Fig. 2 – Magnetic resonance imaging of the right thigh. Axial T2-weighted image (A), axial T1-weighted image (B), axial postcontrast T1-weighted image with fat suppression (C), and axial postcontrast T1-weighted image with fat suppression subtraction image (D) show an infiltrative soft tissue lesion with poorly defined margins that involves all muscle compartments of the thigh, centred on the medial one. The central portion is isointense on the T2 and T1 weighted images (star, A and B), while the more infiltrative peripheral portion shows significant hyperintensity on the T2 and T1 weighted images (arrowheads, A and B). After contrast administration, the lesion demonstrates diffuse heterogeneous enhancement (arrows, C and D). There is also diffuse soft tissue edema with enhancing reticular stranding in the subcutaneous fat, mostly perpendicular to the skin surface, associated with enhancing cutaneous thickening (dashed arrow, C and D).

Discussion

KHE is a rare condition and prompt and accurate diagnosis can be difficult for clinicians [3]. KHE most commonly presents as a purpuric mass on the extremities and if large is associated with swelling of the affected extremity [8].

MRI is the preferred imaging modality for evaluating KHE [8]. On MRI, KHE typically presents with an infiltrative morphologic pattern that involves multiple tissue planes and has poorly defined borders, along with stranding of the adjacent subcutaneous fat and cutaneous thickening [9]. The maximum diameter of the lesion is usually more than 5 cm [9]. Heterogeneous regions of isointensity or mild hyperintensity on T2WI, heterogeneous enhancement, and destruction/remodeling of adjacent bone are also commonly seen [2]. Soft tissue edema is frequent [9], and MRI shows the associated lymphedema with characteristic septa perpendicular to the skin surface that display prominent enhancement after gadolinium administration [8]. Dilated draining veins are also often present [8]. Most of these MRI features were noted in the presented case. There were no skeletal changes, but it is known that these complications are more common in patients older than 6 months of age [3].

The differential diagnosis for KHE includes other vascular tumors such as congenital or infantile hemangioma, which can sometimes be misdiagnosed due to a similar age of presentation and are difficult to differentiate based on clinical presentation alone [2,3]. Infantile hemangiomas are the most common vascular tumors in children and typically do not require treatment [8]. KHE has a worse prognosis and requires more aggressive treatment [2]. The infiltrative nature and imaging characteristics of KHE can differentiate it from congenital and infantile hemangioma [3]. Hemangiomas typically present as well-defined lesions with marked hyperintensity on T2WI and homogeneous enhancement, usually without destruction of adjacent bone [1]. Kaposiform lymphangiomatosis (KLA) can also be included in the differential diagnosis for KHE when it involves multiple planes [10]. However, unlike KLA, KHE is usually unifocal and involves the skin [10].

KHE has a generally poor prognosis due to complications such as hemorrhage or infiltration of vital organs [5]. The KMP contributes to the poor prognosis of patients with KHE and is associated with high mortality rates (up to 30%) [2]. The frequency of KMP in patients with KHE is 42%, and age and lesion size are risk factors [3]. Our patient had a large lesion and presented with KMP.

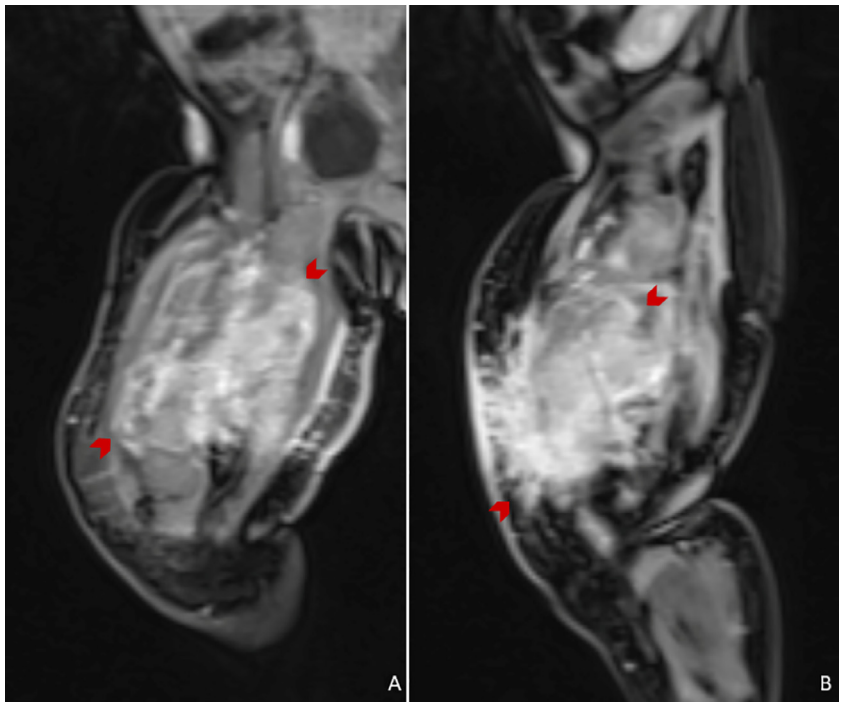

Fig. 3 – Magnetic resonance imaging of the right thigh. Coronal (A) and sagittal (B) postcontrast T1-weighted images with fat suppression demonstrate the infiltrative and locally invasive nature of the lesion. The large soft tissue mass involves all muscle compartments of the thigh and shows heterogeneous enhancement after contrast administration (arrowheads, A and B).

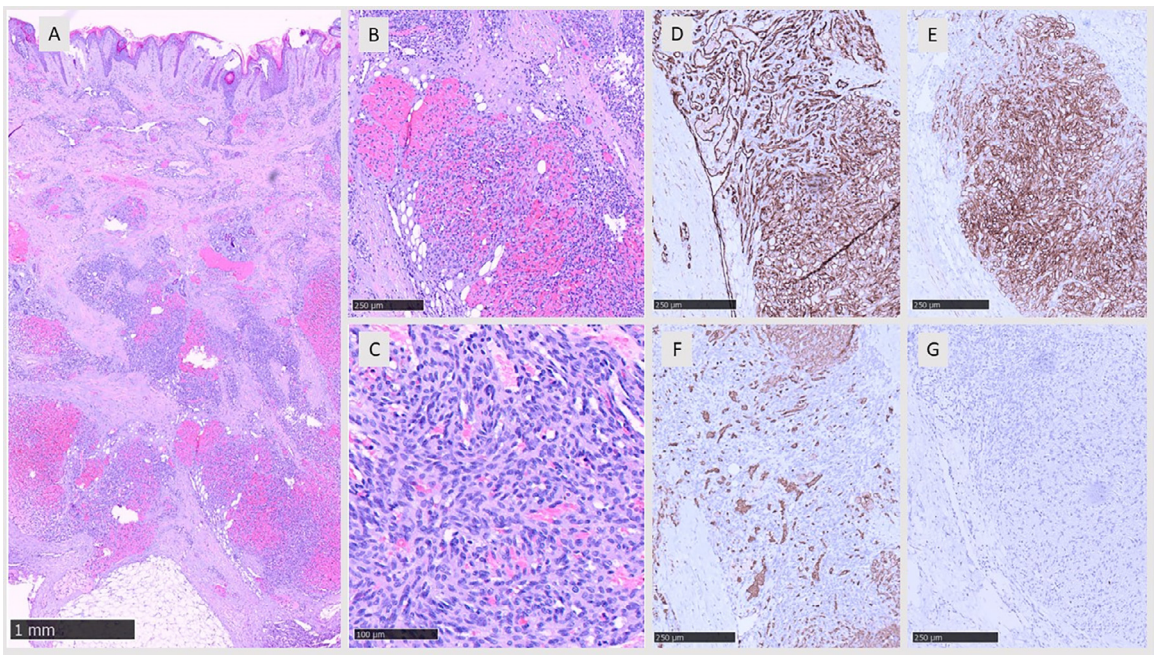

Fig. 4 – Histology of the lesion. Hematoxilin-eosin slides of the biopsy specimens (A–C) revealed an infiltrative dermal centred vascular lesion arranged in confluent nodules (A) composed of spindled endothelial cells and numerous vascular splitlike spaces (B) that lack mitotic activity and atypia (C). Immunohistochestry (D–G) shows reactivity for CD31 (D) and podoplanin (D) in the endothelial of the vascular spaces. There was no expression of GLUT-1 (E) or HHV-8 in the above-mentioned cells.

Timely diagnosis of KHE is essential. KHE should be considered in the differential diagnosis of a superficial soft tissue mass in infants or children when it exhibits multicompartment involvement with poorly defined margins, intense heterogeneous enhancement and adjacent fat stranding in unusual locations, especially if the mass presents with rapid growth, purpuric discoloration and is associated with KMP [2]. Although MRI findings are not specific for the diagnosis of KHE, awareness of these features should prompt radiologists to include KHE in the differential diagnosis for pediatric soft tissue vascular tumors.

Conclusion

The differential diagnosis of a soft tissue mass in infants can be very challenging, and histopathologic characterization is necessary. However, imaging plays a crucial role in the diagnostic workup, particularly MRI, as it can narrow the differential diagnosis by providing decisive information such as the anatomical extent of the tumor.

Patient consent

Written informed legal representative consent for publication has been obtained.

REFERENCES

[1] International Society for the Study of Vascular Anomalies ISSVA classification of vascular anomalies. issva.org/classification; 2018 [accessed 15.09.23].

[2] Ryu YJ, Choi YH, Cheon JE, Kim WS, Kim IO, Park JE, et al. Imaging findings of Kaposiform hemangioendothelioma in children. Eur J Radiol 2017;86:198–205. doi:10.1016/j.ejrad.2016.11.015.

[3] Peng S, Xia C, Yang K, Chen S, Ji Y. Kaposiform haemangioendothelioma: magnetic resonance imaging features in 64 cases. BMC Pediatr 2021;21(1):107. doi:10.1186/s12887-021-02573-8.

[4] Putra J, Gupta A. Kaposiform hemangioendothelioma: a review with emphasis on histological differential diagnosis. Pathology 2017;49(4):356–62. doi:10.1016/j.pathol.2017.03.001.

[5] Hu PA, Zhou ZR. Clinical and imaging features of Kaposiform hemangioendothelioma. Br J Radiol 2018;91(1086):20170798. doi:10.1259/bjr.20170798.

[6] Croteau SE, Liang MG, Kozakewich HP, Alomari AI, Fishman SJ, Mulliken JB, et al. Kaposiform hemangioendothelioma: atypical features and risks of Kasabach-Merritt phenomenon in 107 referrals. J Pediatr 2013;162(1):142–7. doi:10.1016/j.jpeds.2012.06.044.

[7] Drolet BA, Trenor CC 3rd, Brandao LR, Chiu YE, Chun RH, Dasgupta R, et al. Consensus-derived practice standards plan for complicated Kaposiform hemangioendothelioma. J Pediatr 2013;163(1):285–91. doi:10.1016/j.jpeds.2013.03.080.

[8] Wildgruber M, Sadick M, Muller-Wille R, Wohlgemuth WA. Vascular tumors in infants and adolescents. Insights Imaging 2019;10(1):30. doi:10.1186/s13244-019-0718-6.

[9] Calvo-Garcia MA, Kline-Fath BM, Adams DM, Gupta A, Koch BL, Lim FY, et al. Imaging evaluation of fetal vascular anomalies. Pediatr Radiol 2015;45(8):1218–29. doi:10.1007/s00247-014-3248-x.

[10] Goyal P, Alomari AI, Kozakewich HP, Trenor CC 3rd, Perez-Atayde AR, Fishman SJ, et al. Imaging features of kaposiform lymphangiomatosis. Pediatr Radiol 2016;46(9):1282–90. doi:10.1007/s00247-016-3611-1.