

CASE REPORT

Massive Congenital Vascular Tumor of Lower Limb: Kaposiform Hemangioendothelioma

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ABSTRACT

We report a neonate with congenital Kaposiform hemangioendothelioma of left thigh and diagnostic dilemmas encountered in the management. The lesion was dealt with en-bloc resection in early infancy which was proven curative. Currently patient is doing well on follow-up.

Key words: Kaposiform hemangio-endothelioma; Congenital hemangioma; Kasabach Merritt syndrome; Lymphangiomatosis

INTRODUCTION

Kaposiform hemangioendothelioma (KHE), first described about 25 years ago, is a locally infiltrative vascular tumor, which may have an alarming presentation and is shrouded with diagnostic dilemma. It is often confused with hemangioma and mistreated. Surgical excision is usually curative.[1,2]

CASE REPORT

A 20-day-old boy presented with a progressively increasing large vascular mass of left thigh, spanning from groin to knee joint, present since birth (Fig.1A). On examination, the swelling was large (12 x 10 cm) erythematous, and mildly hot (non-infective) on the anterior aspect of left thigh. The baby was severely anemic and thrombocytopenic on CBC (Hemoglobin 7.2g/dl, platelet count 20,000/cmm). X-ray left thigh showed a soft mass and no bony erosion. Ultrasound doppler study showed a large heterogenous mass lesion in the left thigh, with marked internal vascularity (both arterial and venous waveforms) without any cystic components, or calcification. MRI revealed a nodular longitudinal-oriented area of altered signal in-

tensity on medial, anterior and lateral thigh. The lesion appeared hypo-intense on T₁-weighted and hyper-intense on T₂ weighted images, involving muscles in medial and anterior compartments of thigh. There were hyper-intense speckled areas on T₁ weighted images likely representing blood degradation products in lesion and associated thickening of surrounding soft tissue in thigh (Fig.1B). A differential diagnosis of arterio-venous malformation, or hemangioma was reported on MRI. Hemangiomas of other sites were ruled out.

Keeping hemangioma in mind, the neonate was initially started on propranolol and later oral prednisolone was also added, but in vain. The lesion also did not respond to intralesional bleomycin. He underwent USG guided FNAC, after one month of treatment, which showed few clusters of spindle and endothelial cells with immature erythroblastic cells suggestive of hemangioma. He underwent excision of the vascular tumor with split skin grafting at 4 months of age; the skin graft was harvested from the excised tumor surface (Fig.1C). The histopathology was reported as Kaposiform hemangioendothelioma(Fig.1D). Immunohistochemistry (IHC) was positive for CD31, CD34. Postoperative recov-

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ery was uneventful. The boy had no recurrence on one-year follow-up.



Figure 1: A) Clinical Picture at presentation. B) MRI images. C) Post excision split skin grafting. D) Histopathology picture: glomeruloid area (←) in Kaposiform hemangioendothelioma.

DISCUSSION

KHE is a rare locally infiltrative vascular neoplasm seen usually in early childhood that has little tendency to involute spontaneously.[3] It may present with large erythematous or ecchymotic mass which simulated lesions of child abuse in one case. [4] It usually affects skin, with infiltration to deeper tissues. It commonly involves trunk, extremities and retroperitoneum but may involve other rare sites. Similarly in our case it involved thigh. It may be accompanied by lymphangiomatosis and Kasabach-Merritt syndrome. [3,4] Our case also had presentation with KMS with thrombocytopenia but no features of congestive heart failure.

KHE may be seen at birth; cutaneous KHE usually manifests later when compared to infantile hemangioma.[4] Pre-operative diagnostic biopsy is advised in newly-onset proliferative vascular mass in children older than 3 months of age.[5]

Investigative modalities include doppler ultrasonography and MRI. On doppler study, KHE shows an ill-defined heterogeneous lesion with high internal vascularity and high systolic flow. Radiologically, KHE is a highly vascular well-enhancing tumor, but no characteristic findings differentiate it from other soft-tissue tumors, particularly when the tumor does not have any locally aggressive features or identifiable large vessels. Two important differential diagnoses include hemangioma of infancy and Kaposi sarcoma. Juvenile hemangiomas show positivity to GLUT 1 on IHC, whereas KHE shows positivity with CD 31 and CD34.[3] KHE may be differentiat-

ed from capillary hemangioma with lack of well-formed spindle cell fascicles and from Kaposi sarcoma due to lack of periodic acid Schiff positive globules.[1] Other useful microscopy findings to differentiate KHE from Kaposi sarcoma are a lobular pattern, a greater tendency to form complete vascular spaces, and the presence of scattered fibrin thrombi.[5]

Treatment of KHE is decided by its size, location and expected side effects of various available therapies. Sirolimus is now been used as first line therapy or as part of multidisciplinary approach for treatment of KHE.[6]. Tumors confined to the superficial soft tissues are best treated by wide local excision of the infiltrated skin and underlying soft tissues; surgical margins should be clear.[2] The use of cyber knife for KHE of ilium has been reported in literature.[7] The treatment of associated KMS is more troublesome and may need additional multimodal therapy approach in form of steroids, interferon and cytotoxic drugs.[8] Steroids and interferon are usually favored, though no strict guidelines available owing to its rarity. Low dose radiotherapy, angio-embolisation, chemo-embolisation, and intralesional bleomycin/ ethanol has also been described recently in literature.[9-11] Treatment with propranolol, steroids, and intralesional bleomycin did not work in the index case.

In conclusion, the suspicion of KHE should be raised in rapidly proliferating vascular tumor and need of prompt biopsy depending on its biological behavior. The diagnosis of KHE is at stake of an expert histopathologist, thus highlighting need of thorough histopathological examination and immunohistochemistry.

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Authors' contribution: All the authors equally contributed in concept, design, drafting of manuscript, and approved final version of the manuscript.

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