

CONGENITAL CUTANEOUS KAPOSIFORM HAEMANGIOENDOTHELIOMA - A CASE REPORT

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ABSTRACT

Kaposiform haemangioendothelioma is a rare, locally aggressive vascular lesion of infancy and childhood, often associated with Kasabach- Merritt syndrome and lymphangiomatosis. Despite the rarity of this lesion, the morphological and clinical appearances distinguish it from other mimics. As the congenital form of haemangioendothelioma is very rare, it precluded the reporting of this case. Our case is a male infant born with a large violaceous patch over the right loin extending to the back measuring about 12x8cm. No similar or deep lesions were found elsewhere. Incisional biopsy done, revealed the diagnosis of Kaposiform haemangioendothelioma. The tumor was immunoreactive to CD31 and CD34. A late onset of thrombocytopenia or consumption coagulopathy that is known as Kasabach- Merritt syndrome was also noticed.

Keywords: Kaposiform haemangioendothelioma(KHE), Kasabach- Merritt syndrome(KMS), Thrombocytopenia.

INTRODUCTION

Haemangiomas are the most common vascular tumors of infancy and childhood accounting for 1.1% to 2.6%.^(1,2) Kaposiform haemangioendothelioma is a very rare, aggressive vascular lesion, less than 100 cases having been reported so far in the literature.⁽³⁾ It may appear as a superficial or deep lesions, single or multiple and with or without associated complications.^(1,4,5)

In 1940s, Kasabach and Merritt described giant haemangiomas are associated with thrombocytopenia or consumption coagulopathy.⁽⁴⁾ Later on, studies showed an association between spindle cell haemangioma and consumption coagulopathy (Kasabach-Merritt syndrome). In 1993, Zukerberg et al termed it as Kaposiform haemangioendothelioma, considering the spindle cell areas and capillary haemangioma like areas.⁽¹⁾

^{4, 5)} He considers it as intermediate type of malignancy. Histologically, striking spindle cells areas, resembling Kaposi sarcoma along with capillary haemangioma like areas signals the diagnosis of Kaposiform haemangioendothelioma.^(1,3) Larger lesions especially if above 8 cms, are commonly associated with Kasabach-Merritt syndrome.⁽⁶⁾

CASE REPORT

A male child born to non consanguineous parents, by an uncomplicated vaginal delivery, showed a large violaceous patch over the right flank and loin measuring 12.5 X 9.8cm (Fig.1). The baby was then referred to the department of paediatric surgery with the clinical diagnosis of haemangioma of infancy. His base line blood examination including platelet count was normal. CT scan was done to rule out deeper lesions. The early age at presentation prompted the surgeon to perform a biopsy. So, incisional biopsy was done on 22nd day post delivery under general anaesthesia.

The biopsy specimen was received as multiple grey brown soft tissue bits in aggregates measuring 0.9x0.6x0.3cm. All the tissues were embedded and paraffin blocked. Routine H&E study revealed a nodular growth pattern composed of crescent like vessels with distinct glomeruloid appearance of the epithelioid endothelial cells. Capillary haemangioma like areas with prominent spindle cells without inflammatory cells were seen. There was minimal mitosis (less than 2/10 Hpf) and mild atypia. No nuclear pleomorphism, hyperchromasia or necrosis was noted (Fig.2). With these findings, a provisional diagnosis of Kaposiform haemangioendothelioma was made. However, what we see may not be what exists, our histopathological report was confirmed by immunohistochemistry which revealed a strong membranous positivity for CD31 and CD34 (Fig.3), thereby proving the vascular nature of the lesion.

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Initially the patient's platelet count was 90×10^9 /dl; a week later the count was dropped to 11×10^9 /dl. Bleeding time, clotting time and aPTT checked were within normal limits, suggesting Kaposiform haemangioendothelioma with a late onset of Kasabach-Merritt syndrome. The baby's immune status was normal as evidenced by normal CD4+ count and negative HIV test by ELISA.

Owing to the rarity of Kaposiform haemangioendothelioma, no standard treatment regime has been developed so far.^(5, 6) Whenever feasible, excision is the most successful treatment, as per literature⁽⁵⁾ Treatment should be tailored according to the site of presentation and associated complications.^(5, 6) In this patient, the lesion was wide and the baby's age was only 6 months which warranted a conservative approach. Multiple platelet transfusions along with minimal doses of corticosteroids were given to ameliorate his thrombocytopenia. With the above supportive measures, the patient's platelet count improved to 1, 80,000 cells/cu.mm. The mother was reassured and the infant was placed on regular follow up.

Fig1: Show a large violaceous lesion over the right flank region.



Fig2: H&E section show nodular growth pattern of capillary sized vessels. (4x) Inset shows spindled endothelial cells. (40x)

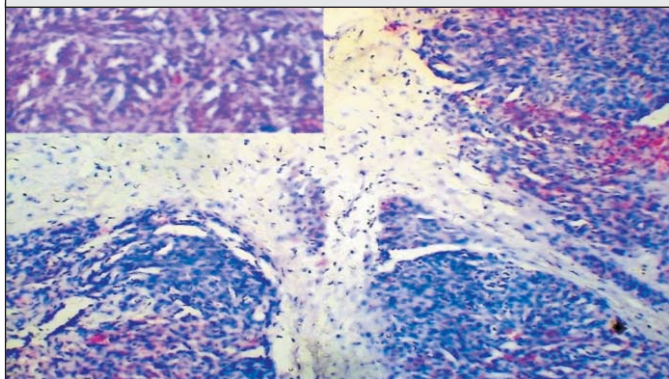
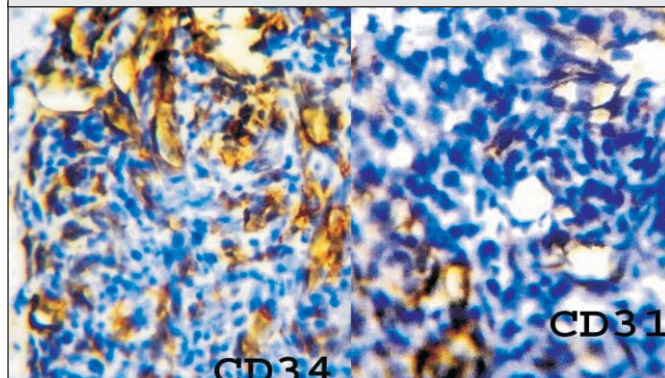


Fig3: Immunocytochemistry of CD34 & CD31 show diffuse membrane positivity. (40X)



DISCUSSION

Kaposiform haemangioendothelioma is a rare, locally aggressive vascular lesion of infancy and childhood, often accompanied by thrombocytopenia and lymphangiomatosis.^(1,2,4) Clinically, the Kaposiform haemangioendothelioma presents either as a superficial or deep lesion mainly over the extremities and trunk or in the retroperitoneum.^(3,4,6) The present case scenario the lesion was situated in the skin over the right side of the abdomen.

The vast majority of the haemangiomas are evident by the fourth week of life and rarely at birth.^(3,5) In this patient it was present at birth as a large violaceous superficial lesion which has persisted till now, (ie.6 months). An abnormal age at presentation prompted us to biopsy the lesion.

Morphologically, the lesion showed a nodular growth pattern composed of crescent like vessels with areas of spindled endothelial cells. In few vessels fragmented red blood cells were seen (Fig.2). There were no inflammatory cells, necrosis, nuclear atypia or pleomorphism.^(3,6,7) With this morphological pattern the differential diagnosis of Tufted angioma, Juvenile haemangioma and Kaposi sarcoma were considered^(3,6,7)

In Tufted angioma, the mid dermis shows, tightly packed, well-formed capillaries in a cannon ball pattern. As opposed, in this patient it revealed the presence of spindle cell areas, poor canalized vessels with fragmented red blood cells, favours the diagnosis of KHE^(3,6)

In Juvenile haemangioma the age at presentation is during the first year of life, as a flat lesion over the head and neck region, whereas in our patient it was a large violaceous patchy lesion over the flank, presenting at birth. Histologically in juvenile haemangioma, there are well formed capillary vessels without spindled areas. On the contrary, KHE has poorly canalized vessels with prominent spindled areas.

The other differential diagnosis is Cutaneous Kaposi sarcoma that occurs especially in endemic areas and other known groups of Kaposi sarcoma (Mediterranean KS, HIV related KS). It can be distinguished by the immune status as well as by morphology, by the absence of inflammatory cell infiltrates and haemangioma like areas.^(2,3,6) In this patient the immune status was well maintained which was revealed by the clinical status, negative HIV test and normal CD4+ count. One of the most remarkable features is the strong association of KHE with KMP.^(1,4,5)

To conclude, we present this case, as less than 100 cases have been reported, so far in literature. Though the term was coined as early as 1940, there is no standard treatment regimen has been hypothesized so far. The clinical assessment of KHE with KMS has to be improved in order to standardize the treatment regimen. More data's regarding treatment and follow up have to be recorded and reviewed so as to enable the prognostication of this lesion.

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