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Kaposiform Hemangioendothelioma With Thrombocytopenia: Report of Two Cases

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KEYWORDS

Kaposiform Hemangioendothelioma, thrombocytopenia, Kasabach - Merritt phenomenon

Introduction

Kaposiform hemangioendothelioma is a rare vascular tumor of infancy. It is one of the three hemangioma seen in infancy with other two being infantile hemangioma and congenital hemangioma [1]. It is the least common of all three hemangiomas and intermediate malignant potential with tendency to recur locally [1]. The characteristic features of KHE include its association with Kasabach-Merritt phenomenon (coagulopathy with profound thrombocytopenia) in almost half cases and associated lymphatic malformation in almost 70% cases [1, 2]. It usually present before 2 years of age although few cases are reported in adult patients. Because of rarity multimodality imaging features of KHE are lacking. We report two neonates with Kaposiform hemangioendothelioma (KHE) associated with thrombocytopenia and describe their clinical, laboratory and imaging features.

Case 1

A 4 month old boy baby presented with gross swelling of the right thigh (Fig. 1a) for past 10 weeks. The child was normal up to 45 days. It started as a small ill-defined red patch, progressively increased in size and attained the present size. The skin was shiny and tense. The swelling was firm to hard in consistency.

Blood investigations revealed normal Prothrombin time with INR. Activated Partial thromboplastin time is mildly prolonged; patient's value is 39.5 (Normal range 25 – 34.8). Platelet count was low; 15000 / cu mm. Blood born virus screening for HIV, HBs Ag and HCV were negative.

Ultrasound showed a diffuse infiltrative enlargement of the thigh tissues involving all compartments with subcutaneous edema (Fig. 1b).

MRI showed diffuse subcutaneous fat stranding. The infiltrative lesion extended deep with involvement of the muscles of the anterior and medial compartment and relative sparing of the posterior compartment muscles. The lesion showed hyperintense signal on T2-w images and hypointense signal on T1-w images (Fig. 1c-e). The femur showed normal signal without any destruction or remodelling.

Provisional diagnosis of Kaposiform hemangioendothelioma was made. The patient was started on steroids. Patient came for follow up after a period of 3 months. Platelet counts significantly improved from 15,000/ cu mm to 55,000/ cu mm. There was also mild reduction in the size of the lesion. Patient was reviewed after 6 months. There is significant reduction in the mass. The redness over the skin also reduced. We planned to continue steroids.

Case 2

A 4 months old boy baby presented with gross swelling of the left thigh (Fig. 2a) for past 3 months. The child was normal up to 1 month. It started as a small ill-defined red patch, progressively increased in size and grown to attain the present size. The skin was shiny and stretched. The swelling was firm to hard in consistency.

Blood investigations revealed Normal Prothrombin time with INR and activated Partial thromboplastin time. Platelet count is low; 8000 / cu mm. Blood born virus screening for HIV, HBs Ag and HCV are negative.

Plain radiograph of the lower limbs showed gross swelling of the left lower limb. The bones were normal. MRI showed diffusely infiltrative lesion involving the entire left thigh with subcutaneous stranding. The lesion was iso-intense on T1, heterogeneously hyperintense on T2-w images. The underlying femur was normal; no cortical breech or bone marrow changes (Fig. 2d-f).

Differential diagnoses of a vascular lesion in an infant include kaposiform hemangioendothelioma, tufted angiomas, infantile myofibromatoses, sarcomas, and metastatic neuroblastomas

Ultrasound guided biopsy of the left thigh lesion was performed. Histopathologic features included Tumour composed of small lobules of spindle shaped cells with bland nuclei interspersed with capillary sized vascular channels – H&E 400x suggestive of kaposiform hemangioendothelioma (Fig. 2g)

The patient was started on steroids and being followed up.

Discussion

Kaposiform hemangioendothelioma is a rare vascular tumour of infancy and childhood [1, 2]. It was confused commonly with hemangioma. The disease course and the treatment are entirely different. It was first described by Zukerberg et al [1] in 1993 as an aggressive vascular neoplasm of childhood. There is no sex predilection. It is commonly associated with Kasabach - Merritt phenomenon (KMP) [4]. The other vascular tumor described with KMP is tufted angioma. In 1997, Sarkar M, et al proposed that thrombocytopenic coagulopathy (Kasabach-Merritt phenomenon) is associated with Kaposiform hemangioendothelioma and not with common infantile hemangioma. It can be seen in up to 50% of KHE [1]. Some vascular tumors are associated with severe thrombocytopenia, consumptive coagulopathy, and microangiopathic anemia. This was referred to as Kasabach-Merritt syndrome (KMS), which was later changed to KMP. This was first described by Kasabach and Merritt in 1940. Other vascular lesion that can show KMS include tufted angioma.

The common locations involved by KHE are chest wall and upper limb, lower limb, retroperitoneum and face. Children with KHE usually presents with low platelet count which is unresponsive to platelet transfusion. This is due to intralesional entrapment of platelets. This is supported by absence of hemolysis on peripheral blood smears. Children with Kasabach-Merritt thrombocytopenia are at risk for intracranial, pleural-pulmonic, intraperitoneal, or GI haemorrhage with an associated mortality of 20% to 30% [5].

KHE usually presents as large infiltrative tumors. Plain radiographs show diffuse enlargement of the extremities and if present bone remodelling and destruction. Ultrasonography and Doppler show large ill-defined lesions with diffuse edema involving all the compartments. On MRI, these lesions are hypointense on T1 and heterogeneously hyperintense on T2-w images. Associated findings would include cutaneous thickening, subcutaneous stranding and edema [6]. MRI helps to assess the deeper extent, involvement of the compartment and bones by the lesion. Differential diagnoses such infiltrative lesion in an infant include kaposiform hemangioendothelioma, tufted angiomas, infantile myofibromatoses and fibrosarcomas.

The features that help to differentiate hemangioma form KHE include association of KHE with Kasabach-Merrit phenomenon on laboratory findings, diffuse infiltrative nature of the lesion involving all tissues on imaging, associated lymphatic malformation on pathology and positivity of certain factors on immunohistochemical staining especially antibody D-240 [1, 2].

There is no best single treatment available for KHE. As this is a rare entity, there is no consensus on the treatment. Various treatment modalities used to control or treat KHE include medical, surgical and endovascular. Surgery can be used if the lesion is focal, well circumscribed and away from neurovascular bundle. Most of the KHE are diffuse and involve neurovascular bundles as they are infiltrative and locally aggressive. Surgery cannot be done for large infiltrative lesions.

Medical treatment would include steroids, interferon alfa and combination chemotherapy with Vincristine, cyclophosphamide and actinomycin D [7]. Most commonly used single chemotherapeutic agent is Vincristine.

Embolization of the feeder arteries is an alternative option. This can be used as an adjunct to medical treatment. There are case reports supporting use of embolization and vincristine for treating KHE [8].

Conclusion

KHE is rare vascular tumour. Clinical findings, laboratory parameters and imaging findings can suggest the diagnosis. But biopsy is usually required to confirm the diagnosis. Apart from suggesting the diagnosis, imaging helps to evaluate extent and involvement of various structures by the lesion. Aggressive therapy should be done to achieve control of the tumour. Thrombocytopenia and local complications can lead to death unless primary tumour and thrombocytopenia are treated.

Figure Legends

Figure 1a. Right thigh is grossly swollen. The skin was shiny

Figure 1b. Ultrasound of the right thigh shows subcutaneous stranding and thickening with enlarged muscles.

Figure 1c, d and e. MRI of the right thigh showed swelling of the muscles of the thigh and subcutaneous stranding.

Figure 1f. Follow up photograph shows significant reduction in the size of the tumor.

Figure 2a. Gross swelling of the left thigh. The skin was shiny.

Figure 2b. Plain radiograph of lower limbs show gross swelling of the left thigh and knee.

Figure 2c. Ultrasound of the left thigh shows subcutaneous stranding and thickening with enlarged muscles.

Figure 2d. T1 W axial section at the mid-thigh showed swelling of the muscles of the thigh and subcutaneous stranding.

Figure 2e and f. MRI - T2 STIR sequence coronal and sagittal views also confirms the swollen left thigh and subcutaneous stranding.

Figure 2g Histopathology slide - Tumour composed of small lobules of spindle shaped cells with bland nuclei interspersed with capillary sized vascular channels – H&E 400x. suggestive of kaposiform hemangioendothelioma.

References

1. Navarro O, Laffan EE, Ngan BY (2009) Pediatric Soft-Tissue Tumors and Pseudo-tumors: MR Imaging Features with Pathologic Correlation Part 1. Imaging Approach, Pseudotumors, Vascular Lesions, and Adipocytic Tumors. Radiographics 29:887-906.
2. Rekhi B, Sethi S, Kulkarni SS, Jambhekar NA (2011) Kaposiform hemangioendothelioma in tonsil of a child associated with cervical lymphnagioma: a rare case report. World J Surg Oncol 9-57.
3. Zukerberg LR, Nickoloff BJ, Weiss SW. Kaposiform hemangioendothelioma of infancy and childhood. An aggressive neoplasm associated with Kasabach-Merritt syndrome and lymphangiomatosis. Am J Surg Pathol. 1993 Apr;17(4):321-8. PubMed PMID: 8494101.
4. Sarkar M, Mulliken JB, Kozakewich HP, et al. Thrombocytopenic coagulopathy (Kasabach-Merritt phenomenon) is associated with Kaposiform hemangioendothelioma and not with common infantile hemangioma. Plast Reconstr Surg 1997; 100:1377–86.
5. Fevurly RD, Fishman SJ. Vascular anomalies in pediatrics. Surg Clin North Am. 2012 Jun;92(3):769-800, x. doi: 10.1016/j.suc.2012.03.016. Epub 2012 Apr 26. Review. PubMed PMID: 22595720.
6. Chen YJ, Wang CK, Tien YC, Hsieh TJ. MRI of multifocal kaposiform haemangioendothelioma without Kasabach-Merritt phenomenon. Br J Radiol. 2009 Mar;82(975):e51-4. doi: 10.1259/bjr/16482217. PubMed PMID: 19211904.
7. Mukerji SS, Osborn AJ, Roberts J, Valdez TA. Kaposiform hemangioendothelioma (with Kasabach Merritt syndrome) of the head and neck: case report and review of the literature. Int J Pediatr Otorhinolaryngol. 2009 Oct;73(10):1474-6. doi: 10.1016/j.ijporl.2009.06.019. Epub 2009 Jul 29. Review. PubMed PMID: 19643504.
8. Garcia-Monaco R, Giachetti A, Peralta O, Napoli N, Lobos P, Gioseffi L, Mariani G. Kaposiform hemangioendothelioma with Kasabach-Merritt phenomenon: successful treatment with embolization and vincristine in two newborns. J Vasc Interv Radiol. 2012 Mar;23(3):417-22. doi: 10.1016/j.jvir.2011.12.007. PubMed PMID: 22365299.