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Kaposiform hemangioendothelioma: A case report of a diagnostic challenge of a kaposiform hemangioendothelioma of the proximal tibia with an atypical presentation

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Abstract
Kaposiform hemangioendothelioma (KHE) is a rare, benign but locally aggressive vascular tumor primarily diagnosed in infancy and early childhood. It often presents with Kasabach-Merritt phenomenon (KMP), a life-threatening coagulopathy. However, atypical presentations, especially involving deep tissues or bone without skin manifestations, are rarely documented. We report the case of an 11-year-old Nigerian boy with a painful, progressively enlarging swelling over the proximal tibia, initially misdiagnosed as chronic osteomyelitis. Imaging and histopathological evaluation, including immunohistochemistry, confirmed KHE with no evidence of KMP. MRI findings revealed involvement of the gastrocnemius muscle and proximal tibia. The patient was managed successfully with intravenous vincristine and oral prednisolone, resulting in significant clinical improvement within weeks. This case emphasizes the diagnostic challenges posed by atypical KHE presentations, especially in resource-limited settings, and the critical role of early suspicion, thorough histopathology, and prompt treatment in improving outcomes.

Keywords: Kaposiform Hemangioendothelioma, kasabach-merritt phenomenon diagnostic challenge, atypical fin dings

Introduction
Kaposiform hemangioendothelioma (KHE) is a rare, benign, rapidly growing, vascular neoplasm diagnosed mostly during infancy or early childhood [1, 2, 3]. It occurs in about 0.071 per 100,000 children and has a slight male preponderance [4, 5]. Its association with Kasabach-Merritt phenomenon with its life-threatening coagulopathy occurs in about 70% of kaposiform Hemangioendotheliomas [6, 7, 8, 9, 10, 11, 12]. Previously, KHE was shown to have an equal sex predilection. However, a slight male predominance has been indicated by two large retrospective studies [7, 8]. The etiology of this disease remains unknown, and in most cases, KHE occurs insidiously. In rare scenarios, signs and symptoms of KHE can worsen with trauma or infections and there is also evidence that aggravation or manipulation of the tumor either from surgery or trauma can incite KMP [4].
The tumor though benign, has this potential of invading surrounding tissues, bones and joints are not spared [1, 13, 14]. Because of its rarity and complex manner of presentation in some cases, misdiagnosis and delay in diagnosis is common [9, 15]. Pain, swelling and limitation of activities are common symptoms. Relevant studies have shown that approximately 70% of KHE patients have a decreased range of motion within two years after the tumor is evident. The destructive growth pattern of the tumor involving bone and the development of fibrosis around the muscles and joints can cause functional limitations, all of which may affect the ability of patients to perform daily activities. They are mainly located in the soft tissue surface and visible in the muscle layer and adipose tissues even though they can also occur in viscera. Occurrence in long bones without cutaneous changes has been reported even though only few of such cases of were reported [2, 17, 18]. Lesions can be strikingly large and mimic malignant tumors. They are usually dark red with high skin tension and do not recede with age [15, 16]. Most studies on KHE with bone destruction are case reports and very limited large-sample study of KHE involving bone destruction have been conducted.

Various attempts at treatments have been instituted such as the use of vincristine sirolimus, propranalol, corticoids, and so on, yet no consensus single regimen has been recommended yet [7]. We aim to alert clinicians to look out for KHE when evaluating tumors at specific sites, which helps at arriving at an early diagnosis and institution of appropriate early clinic therapy. Only 10% of KHE patients showed deep lesions without skin involvement, while primary bone lesions were rarer, and most of the lesions were single lesions [15]. There may be atypical cases that do not present with Kasabach-Merritt phenomenon and do have the typical imaging findings. Histological features, supportive immunohistochemistry, some radiological findings and the clinical presentations are together helpful to arriving at the diagnosis of KHE and differentiating it from other vascular lesions. Early diagnosis is crucial to the institution of treatment which usually results in in improved outcome [19].

Case Report

This is a case of an 11yr old boy who presented to The Potter's Specialist Hospital in Jos Plateau State of Nigeria with a 5 months history of a progressively worsening pain on the proximal tibia. The pain worsens with activity and is relieved by rest and the use of analgesics. Pain became so severe that, it stops him from bearing weight on the same limb. About a month from the onset of the pain patient started noticing gradually progressive, diffuse proximal tibia swelling just around the site of pain which progressively enlarged up to 3 times the size from the time it was initially noticed. Two months prior to presentation he started having some limitation of movement around his knee. He offered a history of trauma to the affected part while playing in school but no history of pain or swelling at any other part of the body. There is no history suggestive of tuberculosis or malignancy and there is no previous history of similar lesion in him or other members of his family. He had presented to several other peripheral secondary health facilities but ended up with the Traditional Bone Setter because it was thought to be a fracture or possible soft tissue trauma but to no avail, as such decided to present to this facility.

At presentation he was found to be a young boy who walked in with bilateral axillary crutches, non-weight bearing on the affected limb with the affected limb in an attitude of 30-degree flexion. There is no wasting and no palor, and no raise body temperature. There was an obvious diffuse swelling on the proximal tibia approaching the knee. The proximal tibia was thrice the size of the contra-lateral limb and with a pointing sign on the lateral aspect of the area. The swelling had some differential warmth and was quite tender. Movements around the knee were limited due to pain both passively and actively. Distal neurovascular statuses were intact. The X-ray of the proximal tibia was suggestive of a chronic osteomyelitis. The impression of chronic osteomyelitis was then entertained to rule out osteosarcoma. X ray of the proximal tibia suggested a chronic osteomyelitis. He had an attempted sequestrectomy but during the surgery, tissues gotten were suspicious of a probable tumor (a pile of grey white and grey brown tissues were noted) and as such all curetted substances were sent for histology. The first histology report was that of an osteonecrosis, seemingly aligning with our impression but a hold on the result was declared by the most Senior Pathologist who had to read the slide again reported that the histologic sections show an intermediate grade locally aggressive vasoformative neoplasm composed of fragments of fibro-osseous tissue with infiltrating spindle to epitheloid cells disposed in short fascicles and in whorls and forming glomeruloid structures in areas. The cells formed slit-like spaces and vascular channels containing red blood cells. The tumour cells have round to oval vesicular mild to moderately pleomorphic nuclei and moderate eosinophilic cytoplasm. There were areas that showed deposition of collagen bundles within the stroma. Mitotic figures and areas of necrosis
were scanty. There are areas of bone involvement and destruction seen and lo' the overall features described were those of a Kaposiform Hemangioendothelioma (Fig 1a, b & c). Immunohistochemistry tests were then requested to confirm this diagnosis immediately. The immunohistochemistry results showed that the tumour cells were focally positive for CD31 and diffusely positive for CD34 (Fig 1d, e & f) which confirmed the vascular origin of this tumour and with the kaposiform histomorphology, the diagnosis of KHE was confirmed.

To ascertain the presence of Kasabach-Merritt phenomenon clotting profile was done.

MRI was done to buttress the diagnosis and to ascertain extent of the disease.

Following the localizers, multi-planar images were acquired in T1W; T2W AND STIR sequences using a 0.35 Tesla MRI scanner and the findings were as below:

1. An ill-defined heterogeneous lesion is seen in the upper half of the left leg within the proximal half of the medial head of the gastronemius muscle. It is mildly hyper-intense on T1 and brightly intense on T2. It is avidly enhancing on T1 post contrast.
2. A similar lesion is seen within the tibia extending from the epiphysis through the physis to the proximal third of the tibia with poor outline and cortical destruction.
3. The tibio-femoral joint compartment was preserved.
4. No evidence of joint effusion.
5. The adjacent fibular is normal in outline and signal intensity.

The conclusion of the MRI findings were also those of a Kaposiform Hemangioendothelioma of lower limb soft tissue with proximal tibia involvement.

He was started on intravenous Vincristine 1.7mg weekly for 6 weeks and tabs Prednisolone 70mg daily. Two weeks after the start of chemotherapy, the boy resumed ambulation with significantly reduced pain and the swelling regressed appreciably well.

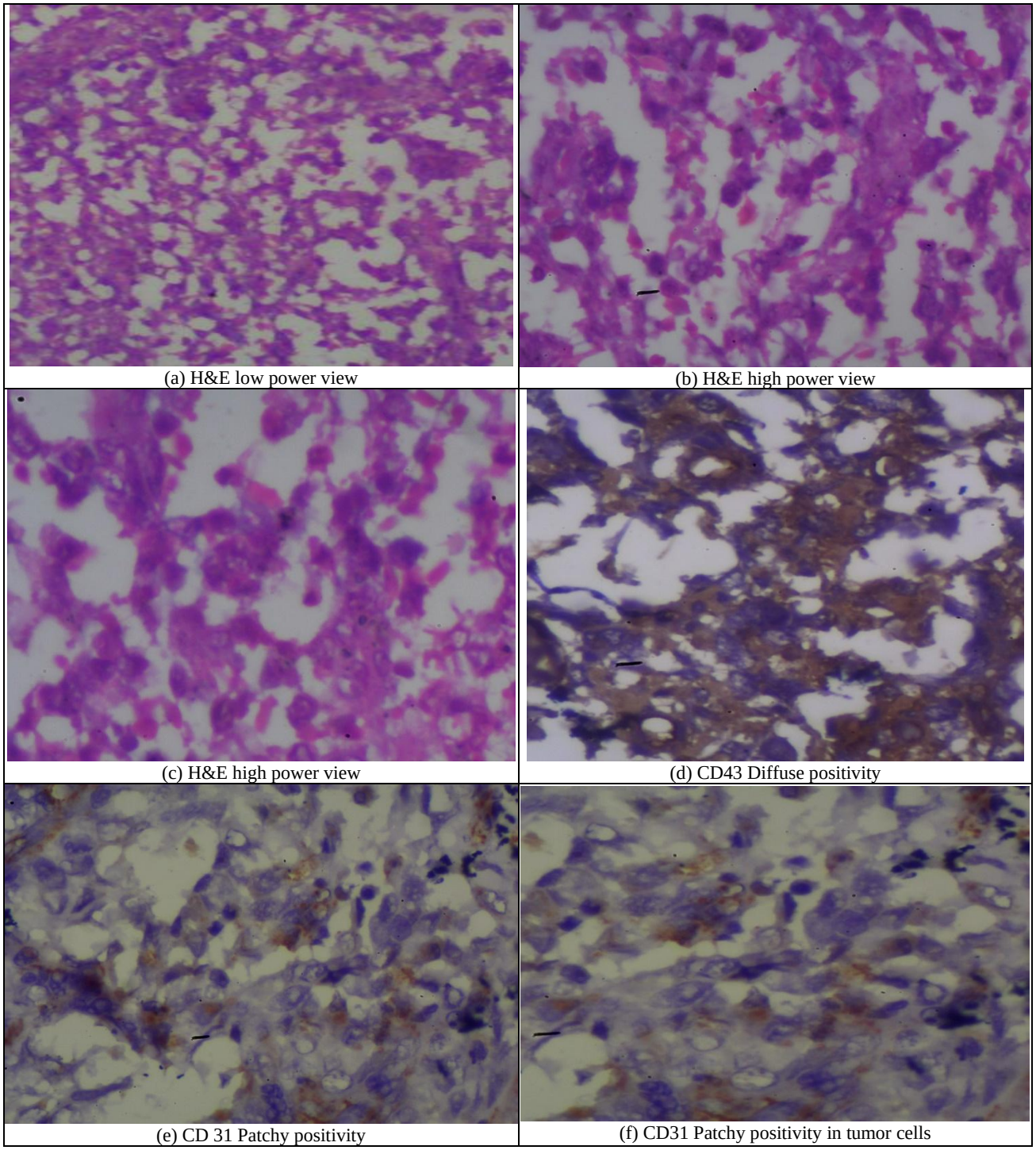

Fig 1 (a-f): Showing H&E and Immunohistochemistry-stained photomicrographs of a case of kaposiform hemangioendothelioma involving soft tissues and adjacent tibial bone in an 11 year old male

a) Low power view of H&E stained slide of Kaposiform hemangioendothelioma showing multiple small calibre vascular spaces lined by minimally pleomorphic plump endothelial cells containing red blood cells
b) Figures b and c show high power view of the cells and vascular spaces described above
c) Figure d shows diffuse cytoplasmic/membranous positivity of the tumour cells
d) Figures e and f show patchy cytoplasmic/membranous positivity to CD31

Discussion

Kaposiform Hemangioendothelioma (KHE) is a rare, benign, rapidly growing, vascular neoplasm diagnosed mostly during infancy or early childhood [1, 2, 3]. This case is that of an 11yr old boy. Even though he is in his late childhood, reporting such late presentations is not uncommon [1, 2, 5]. The index case is a male child. Some studies noticed some male predominance even though most studies really did not find any of such [4, 8]. This is probably due to the rarity of the disease and as such, large scale studies have not been done to elucidate significantly on gender variability. Pain and swelling were noted at the proximal tibia for about 5 months before presentation.

Proximal tibia involvement may not be commonly reported but affectation of long bones has been reported a number of times [2, 7, 18]. The stiffness that this patient noticed two months after the onset of the other symptoms is not a rare finding especially

when peri-atricular areas are the common sites of occurrence of the tumor. Preceding history of trauma offered could be true because there are some associations noted between trauma and the onset of KHE even though sometimes parent's insistence of finding the cause of this disease of unknown etiology may make the child imagine some vague trauma as the cause.

The patient's condition was misdiagnosed several times at other health facilities as a case of trauma. Traditional Bone Setters also made attempts of treatment as a trauma to no avail. This boy's tumor came devoid of the conventional clinical features that are classical for Kaposiform Hemangioendothelioma. No skin changes at all apart from the tenderness and swelling the child noticed. Jie Ma and colleagues in there article 'Primary kaposiform hemangioendothelioma of long bone" reported two cases of long bone tumors without cutaneous changes [2]. This suggests the probable atypical origin of the disease is the long bone. Even at presentation our first impression was that of a chronic osteomyelitis but it was debunked by the intra-operative findings and confirmed by the histology report. Most of the studies have this disease in its earlier stages misdiagnosed commonly as cellulitis, chronic osteomyelitis, Ewings tumor, osteo-sarcoma and fractures, to mention but a few. Being able to make early Diagnosis of the disease at rarer sites helps the clinician to institute early treatment which usually give a more favorable outcome [18]. [xviii] Kasabach Merritt Phenomenon was not present in this index case. KMP is usually found in about 70% of cases of KHE. It is more common in those with typical than those with atypical presentations and more common when the disease occurs in early childhood [18]. Relevant clinical history with Histology, immune-histochemistry and MRI findings are helpful in arriving at the diagnosis as in this case, even though atypical presenting features were found almost all along the diagnostic workup.

Although no consensus on the effectiveness of the different tried regimens was arrived at, but Vincristine and prednisolone have been tried by in a number of reported cases, some to no avail but in this case we found that there was good response in 4weeks. Pain, swelling and the limitation of movements were significantly reduced and the patient was able to walk unaided and without a limp.

Conclusion

Kaposiform Hemangioendothelioma can be confusing with quite a number of pathologies making early diagnosis an uphill task and delay is catastrophic. It can present atypically, arising from deeper tissues with little or no cutaneous or subcutaneous finding, as high index of suspicion is the key to early diagnosis in such scenarios. Early intervention can quite rewarding and significantly restores motility, hence reducing morbidity significantly as in this index case.

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