CASE REPORT

# KAPOSIFORM HEMANGIOENDOTHELIOMA OF BONE WITH NO CUTANEOUS LESION IN CHILDREN - A CASE REPORT

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## Introduction

Kaposiform hemangioendothelioma (KHE) is a rare vascular aggressive tumor with an onset during infancy or early childhood. It's diagnosis is often delayed, especially in the absence of associated thrombocytopenia. According to Boutinetal., epithelioid hemangio endothelioma of the bone, producing anosteolytic lesion sinvolving the cortex and cancellous bone of extremity, is a rare malignant vascular tumor.[1] Though a few case reports have reported the imaging findings of KHE, but no particular local data has been documented to the best of our knowledge inourregion.

## Case Presentation

A 2 years and 2 months old male patient presented with pain in the right shoulder. On clinical examination, his right upper limb was hung by the side with inward rotation and limited motion. There was no skin lesion or swelling. There were no bruising or cutaneous changes. Clinically, he was diagnosed with Erb's palsy. Before presenting tous, he had a biopsy outside our hospital for lytic lesion in the right proximal humerus which was reported to be fibro-osseous tissue with florid vascular proliferation. He was prescribed to get an MRI shoulder and humerus of the affected arm.

## Imaging Findings

Complementary radiograph of the right shoulder anterior posterior view showed soft tissue increased density. Lytic destruction of the outer cortex of the proximal diaphysis of the right humerus was seen. Irregular area of sclerosis was seen in the adjacent medullary cavity with a broad zone of transition. It measured 3 cm in craniocaudal (CC) dimension. Surrounding soft tissue swelling was seen. No definite soft tissue mass on radiograph (Fig.1).

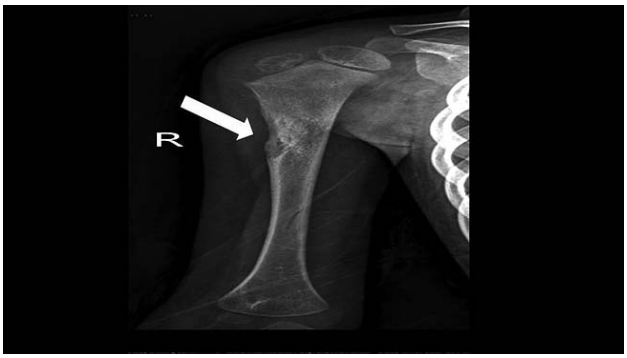


**Figure 1:** Radiograph right shoulder anterior posterior view showing lytic destruction of the outer cortex of the proximal diaphysis of the right humerus. An irregular areas of sclerosis in the adjacent medullary cavity with broad zone of the transition is seen.

Magnetic resonance (MR) imaging of the right humerus showed an ill-defined infiltrative abnormal T2-weighted and STIR hyperintense lesion arising from the proximal meta-diaphyseal region of the right humerus (Fig.2), T1 predominantly low with post-contrast homogenous enhancement was noted. There was permeative destruction of the anterio-lateral cortex of the proximal humerus with associated periosteal reaction and enhanced soft tissue components. The periosteal reaction was a butting the under surface of the deltoid muscle (Fig.3 and 4). There was no encasement of regional neurovascular structures. Multiple enlarged

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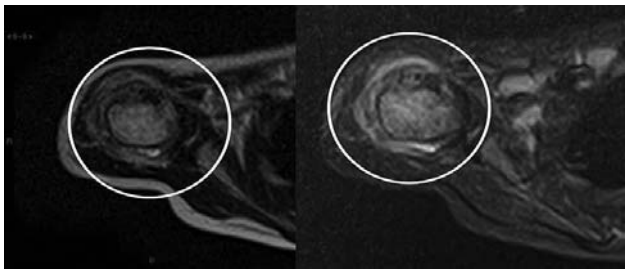


**Figure 2:** MRI shoulder axial views of T2 weighted and STIR sequences showing an ill-defined infiltrative abnormal T2-weighted and STIR hyperintense lesion of the right humerus.

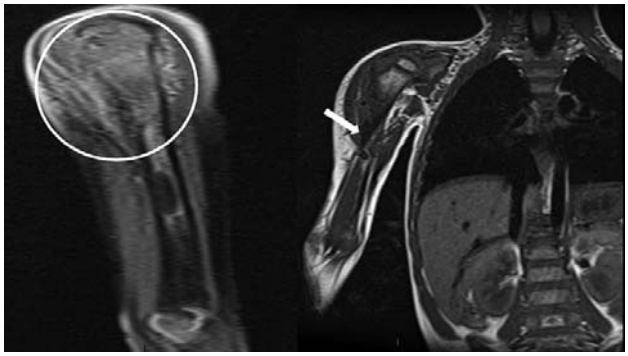


**Figure 3:** PD and T1 predominantly low lesion with permeative destruction of the anterio-lateral cortex of the proximal humerus with associated periosteal reaction.

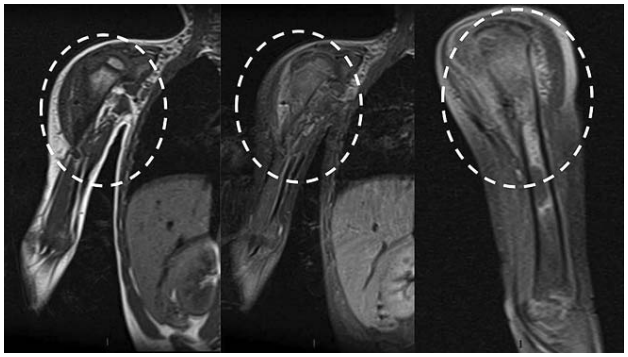
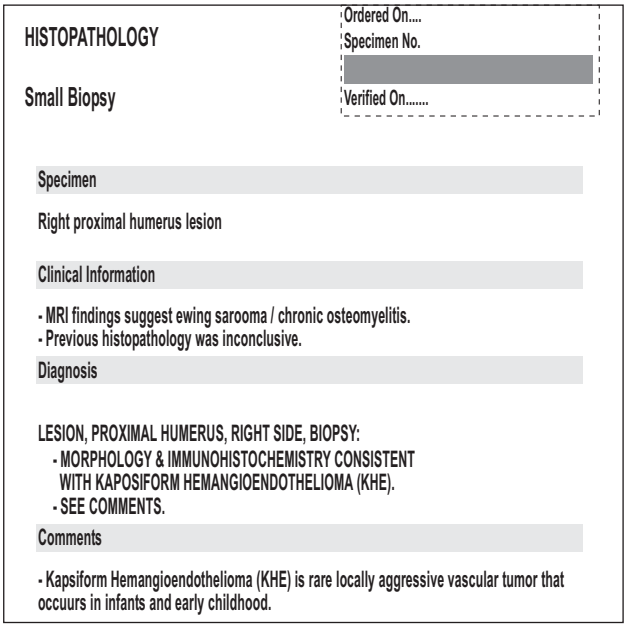


**Figure 4:** T1 pre- and postcontrast coronal and T1 contrast enhanced fat sat images showing homogenous enhancement of the lesion.

enhancing nodes in the right axilla with the largest measuring 11 mm in the short axis were seen. There was mild soft-tissue edema with minimal joint effusion in the glenohumeral joint. Edematous bone reaction surrounding the lesions was also seen. Possible differentials included malignant neoplastic osseous lesions such as Ewing sarcoma and osteo-myelitis. Tissue sampling was advised again.

Following our recommendation, an open biopsy was performed and the specimen was submitted for pathologic evaluation which revealed this epithelioid hemangioendothelioma (EH) (Fig.5).



HISTOPATHOLOGY

Ordered On....
Specimen No.
Verified On.....

Small Biopsy

Specimen

Right proximal humerus lesion

Clinical Information

- MRI findings suggest ewing sarooma / chronic osteomyelitis.
- Previous histopathology was inconclusive.

Diagnosis

LESION, PROXIMAL HUMERUS, RIGHT SIDE, BIOPSY:
- MORPHOLOGY & IMMUNOHISTOCHEMISTRY CONSISTENT WITH KAPOSIFORM HEMANGIOENDOTHELIOMA (KHE).
- SEE COMMENTS.

Comments

- Kapsiform Hemangioendothelioma (KHE) is rare locally aggressive vascular tumor that occuurs in infants and early childhood.

**Figure 5:** Repeat biopsy report documenting the lesion to be Kaposi hemangoendothelioma.

Following the diagnosis of epithelioid hemangio-endothelioma (EH) on the basis of findings at open biopsy, the patient's parents decided after discussion with his orthopedic surgeon, to undergo chemo-therapy. The patient had injectable vincristine for 4 weeks and reassessed. After the completion of 8 cycles of chemotherapy, there was some decrease in tumor size (Fig.6). The orthopedic surgeon decided to proceed with excision of tumor. Plastic surgery team was taken on board for graft and aesthetic reasons. Following the completion of surgery with

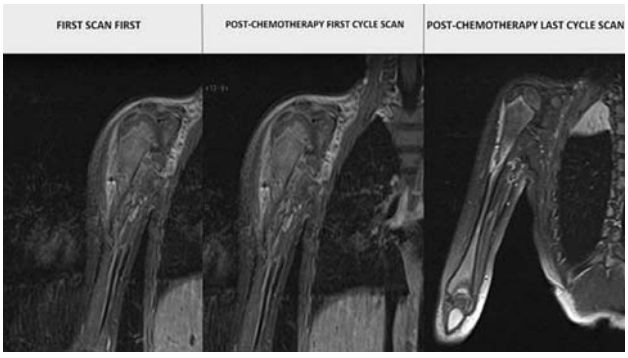



**Figure 6:** Coronal view of postcontrast T1 humerus, first image showing pre-chemotherapy image followed by then at first and last chemotherapy showing decrease in the size of the primary mass.

neoadjuvant chemotherapy, the patient's father was contacted through the given demographic number after three-month time who reported smooth recovery of the child.

## Discussion

Kaposi hemangioendothelioma is a rare endothelial vascular neoplasm, with its peak onset during infantile year.[2] This locally aggressive tumor has its annual prevalence and incidence to be 0.91 and 0.71 per 100,000 children, respectively.[3] Weiss and Enzinger described the first ever identified case as a soft tissue mass.[2] In 1993, Zukerberg and his coworkers designated KHE as a distinct entity from infantile hemangioma, due to its locally aggressive invasive growth.[4] However, the reisa paucity of literature particularly addressing the incidence of KHE. Its etiology remains unknown, as of now.[5] Combination of clinical assessment, imaging, histological, and hematological findings are essentials for making a diagnosis. Yet few studies still report that all of these might not be helpful in making a diagnosis readily in few cases.[6] Diagnosis is especially delayed if this presents as deep KHE without KMP (bone and/or joint and so on), just similar to our case.[5] Some studies support sonographic evaluation via ultrasound to be the preferred modality for small and superficial lesions, unlike the case we are presenting.[7] Ultrasound is the modality of choice forsmall and superficial lesions. However, similar to our case, magnetic resonance imaging (MRI) is considered the first-line assessment because the deep infiltrating nature of KHE may not be apparent on physical exam or on ultrasound.

Pre- and post-contrast MRI acts as a workhorse in clearly determining the disease extent and treatment response.[8] Similar to our case, MRI exhibits T1-weighted iso- and T2-weighted hyperintense with adjacent muscles, signal abnormalities with ill-defined margins, and characteristic diffuse enhancement. It also shows fat stranding in unusual sites. Changes in adjacent bones and/or joints are commonly seen. Similar to this, case, studies have reported cortical destruction, epiphyseal injury, and invasion of adjacent joints.[9] One of the studies also states that there is no specific MR pattern of this disease, especially inbones.[5] In most cases, clinical history and examination showing cutaneous purpura and severe anemia, the differential must include KHE. Differential diagnosis for EH of bone should include angiomatosis, Langerhans cell histiocytosis (LCH), angiosarcoma, infection, myeloma, metastasis, and lymphoma.[10]

## Conclusions

Kaposi hemangioendothelioma occurs in a variable location and affects mostly infant. Awareness of these features should prompt radiologists to include KHE in the differential diagnosis for pediatric mass, although the incidence of KHE is low, it can cause morbidity and mortality in children and adults. Consequently, prompt diagnosis and appropriate management are crucial to improving the long-term prognosis of patients.

**Conflict of Interest:** None

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