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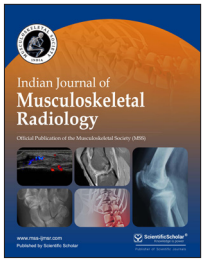

Case Report

Multicentric epithelioid hemangioendothelioma of humerus and scapula

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ABSTRACT

Epithelioid hemangioendothelioma (EH) is a rare malignant vascular tumor occurring mainly in the liver and lungs, with bones being a rare site and primarily seen in the adult population. We present a rare case of multicentric EH in the right humerus in an adolescent male, who presented with complaints of pain and swelling in the right shoulder. Radiographs and Computed tomography showed a large expansile lytic lesion in the proximal end of the right humerus with areas of cortical destruction and matrix calcification. On Magnetic resonance imaging, the lesion was predominantly isointense on T1-weighted image, hyperintense on T2-weighted, and Short Tau Inversion Recovery images with prominent flow voids within. On post-contrast sequences, the lesion showed intense heterogenous enhancement with a non-enhancing central necrotic area. Multiple smaller lytic lesions with similar imaging characteristics were also found in the mid and distal shaft of the right humerus and the right coracoid process. Differentials of Telangiectatic osteosarcoma, Giant cell tumor, brown tumors of hyperparathyroidism, and metastasis were considered. Core biopsy revealed that the lesion was an EH. Though the imaging features of EH are non-specific, it may be considered in the differential diagnosis of an expansile lytic bone lesion with no periosteal reaction, showing cortical break and soft tissue component, especially if it is multifocal and multicentric.

Keywords: Epithelioid hemangioendothelioma, Multicentric, Magnetic resonance imaging, Computed tomography, A case report

INTRODUCTION

Primary vascular tumors originate from the elements of blood vessels like endothelial cells or pericytes. Epithelioid hemangioendothelioma (EH) is a vascular tumor that can occur anywhere in the body but frequently involves the liver, lungs, and bones.[1] On radiographs and Computed Tomography (CT), EH of bone presents as a multifocal or multicentric locally aggressive expansile, lytic lesion with cortical destruction and a soft tissue component.[1,5]

On magnetic resonance imaging (MRI), the signal intensity characteristics are non-specific. The lesion appears hypo-to-intermediate signal intensity on T1-weighted images and hyperintense signal intensity on T2-weighted images and shows restriction on diffusion-weighted imaging.[5]

To the best of our knowledge, this is a rare case of multicentric EH found in proximal humerus and scapula, having an unusual feature of matrix calcifications, which posed difficulty in arriving at a definitive diagnosis, thereby leading us to consider various other bone tumors in the differential diagnosis. As a result, the final diagnosis was confirmed only on histopathology.

CASE REPORT

An 18-year-old male presented to the Orthopaedics department with complaints of pain in the right shoulder for 2 years, swelling, and restriction of motion in the right shoulder for 1 year. The pain began after an episode of trivial trauma; it was gradually progressive, non-radiating in nature and aggravated on moving the limb, and relieved at rest. There were no constitutional symptoms. On examination, the swelling was large, measuring approximately 15 cm × 7 cm present in the right shoulder, extending up to mid-arm with a smooth surface and stretch marks. Palpation revealed the swelling was immobile, firm in consistency, tender with the local rise of temperature. The range of movements of the right shoulder was significantly reduced due to pain. There was no palpable axillary lymphadenopathy.

Blood investigations revealed anemia with hemoglobin of 7.8 g/dL, elevated ALP of 245 U/L. The rest of the parameters were within normal limits.

Radiograph of the right shoulder including humerus [Figure 1] and CT [Figure 2] showed a large expansile, lytic lesion involving the proximal epiphysis, metaphysis, and diaphysis of right humerus extending till the subarticular surface with areas of thinning and destruction of overlying cortex. The matrix of the lesion showed coarse calcifications and fine internal septations. There was no periosteal reaction. Few other similar smaller lytic lesions were present along the inner cortex and medulla in the mid and distal shaft of the right humerus. CT confirmed the findings on the radiograph but also showed another lytic lesion in the right coracoid process.

MRI of the right shoulder [Figure 3] showed a well-defined large expansile lesion measuring 9.7 cm × 10.6 cm × 15.6 cm involving the head, greater and lesser tuberosities, proximal meta-diaphysis of the right humerus. The lesion showed predominantly isointense signal on T1-weighted images and hyperintense signal on T2-weighted and Short tau inversion recovery images. Few hyperintense areas on T1-weighted images, which are hypointense on T2-weighted images, are seen within the lesion, corresponding to areas of calcification. The lesion is compressing and showing loss of fat planes with deltoid, rotator cuff muscles, and triceps. The lesion is displacing the axillary neurovascular bundle medially. On post-contrast images, the lesion showed heterogeneous enhancement with a central non-enhancing area suggestive of necrosis.

Smaller focal satellite lesions were present in the mid and distal shaft of the right humerus and in the right coracoid process, which showed similar imaging characteristics. Based on the age, clinical and imaging features, differentials of Telangiectatic osteosarcoma with skip lesions, Giant cell

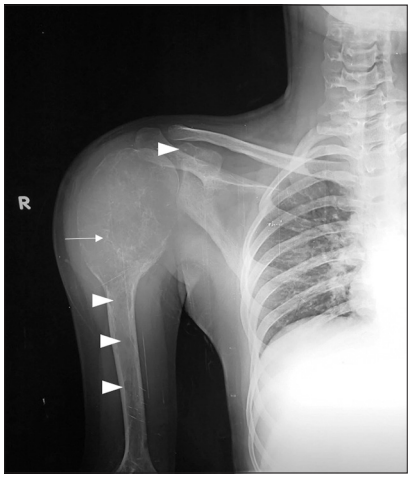

Figure 1: Multicentric Epithelioid Hemangioendothelioma in an 18-year-old man with pain and swelling in the right shoulder. Anteroposterior radiograph of right shoulder showing a large, expansile, lytic lesion involving the proximal epiphysis, metaphysis and diaphysis of right humerus with subarticular extension. Matrix calcifications are seen within the lesion (arrow). There is thinning of overlying cortex with cortical destruction at places along the medial cortex. Multiple smaller satellite lesions are also seen in the right humerus and coracoid process of scapula (arrowheads).

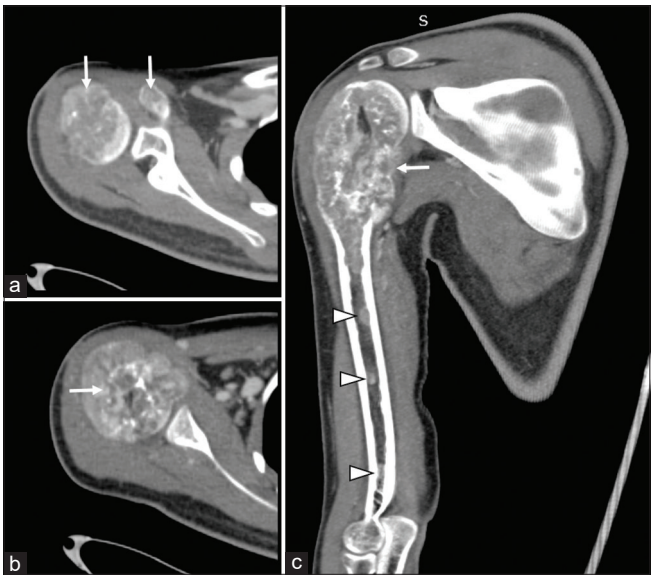

Figure 2: (a-c) Multicentric Epithelioid Hemangioendothelioma in an 18-year-old man with pain and swelling in the right shoulder. Computed tomography showing multiple lesions in right humerus and coracoid process of scapula. Axial bone window images (a) and (b) showing geographic, expansile, lobulated lytic lesion in proximal end of humerus and coracoid process (arrows in a) with central areas of ossification (arrow in b). Oblique coronal bone window image (c) showing geographic, expansile, lobulated lytic lesion in proximal end of humerus with areas of cortical break along the medial aspect (white arrow). Multiple other medullo-cortical satellite lesions in the diaphysis (arrowheads).

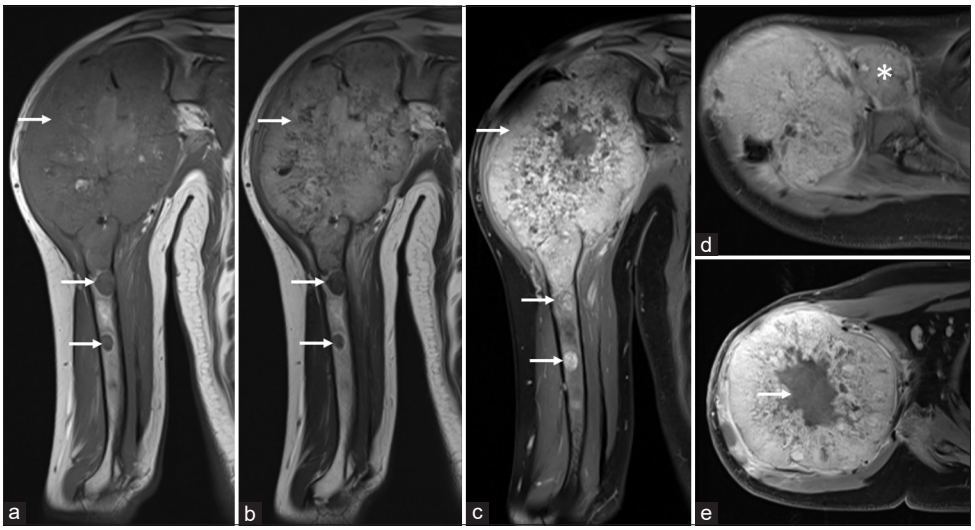

Figure 3: (a-e) Multicentric Epithelioid Hemangioendothelioma in an 18-year-old man with pain and swelling in the right shoulder. Coronal Magnetic resonance images showing a geographic, expansile, lobulated lytic lesion in proximal end of humerus with multiple smaller satellite lesions in the medulla of diaphysis of right humerus and coracoid process of scapula. The lesions appear heterogeneous, isointense signal intensity on T1 weighted (arrows in a), heterogeneous, hyperintense signal intensity on T2 weighted (arrows in b) with heterogeneous enhancement on post contrast (arrows in c) images. Axial post contrast Magnetic resonance images showing enhancing lesion in the coracoid process of scapula (asterisk in d) and central area of non-enhancement suggesting necrosis (arrow in e).

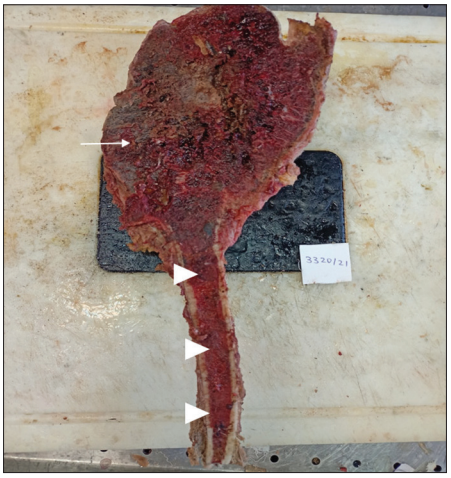

Figure 4: Multicentric Epithelioid Hemangioendothelioma in an 18-year-old man with pain and swelling in the right shoulder. Photograph of post forequarter amputation right upper limb specimen depicting multifocal lesions, largest in proximal humerus (arrow), multiple skip lesions are seen distal to it within diaphysis (arrowheads).

tumor, brown tumors of hyperparathyroidism, and metastasis were considered. Telangiectatic osteosarcoma with skip lesions was considered as the patient was an adolescent and the lesion was expansile, lytic with matrix mineralization. Giant cell tumor was considered because of expansile, lytic lesion involving epi-metaphyses and extending to subarticular location; however, matrix calcification and multicentricity are rare. Brown tumors of hyperparathyroidism often present as multiple bone lesions; however, the lesions tend to be purely lytic and often associated with decreased bone density and subperiosteal resorption. Metastasis from the kidney or thyroid present as expansile, multifocal, or multicentric lytic lesions.

The patient underwent high-resolution ultrasound of the neck, which was normal. Ultrasound-guided core needle biopsy of the lesion in the proximal end of the humerus was then performed. The histopathology revealed a vascular neoplasm, with morphology and immunohistochemistry favoring an EH.

Given the long length of the lesion in the proximal humerus, the satellite foci in the mid and distal diaphysis of the humerus and another focus in the coracoid process, forequarter amputation of the right upper limb and excision of the coracoid process were performed and the amputated limb was sent for gross [Figure 4] and histopathological examination [Figure 5].

DISCUSSION

Primary vascular tumors are rare. They originate from the elements of blood vessels like endothelial cells or pericytes. EH is one such type of vascular tumor that accounts for <1% of all vascular tumors.[1] Weiss and Enzinger coined the term EH.[2] In the recently revised classification in 2018, the ISSVA has reclassified EH as a malignant vascular tumor.[3]

The etiology of EH remains unknown; however, WW domain-containing transcription regulator 1 gene and

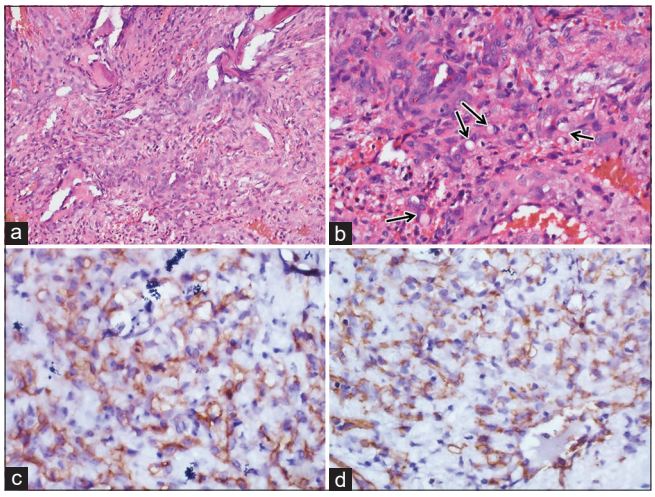

Figure 5: (a-d) Multicentric Epithelioid Hemangioendothelioma in an 18-year-old man with pain and swelling in the right shoulder. Hematoxylin and Eosin stained (a) and (b) section shows bony trabeculae entrapped and surrounded by cellular lesion comprised of plump spindle to epithelioid cells with round to oval vesicular nuclei and eosinophilic cytoplasm along with some congested capillaries. On higher magnification some of the cells having round cytoplasmic vacuoles representing intracytoplasmic lumina pushing the nucleus to periphery can be seen (black arrows) (a ×200; b ×400). Immunohistochemistry with (a) CD34 and (b) CD31 shows positivity within the tumor cells (HRP-Polymer; c ×400, d ×400).

Calmodulin-binding transcription activator 1 gene are known to be associated with EH through chromosomal translocation.[2]

EH is known to occur commonly in the 2nd to 3rd decades of life. It has a male predilection with a male-to-female ratio of 2:1.[4] It is commonly seen in the liver and lungs, with bones being the third most common site.[2] The long bones of the lower extremity are the most common site.[2,4] In the long bones, EH presents as multiple (multifocal) or can involve multiple bones (multicentric).[4]

The clinical presentation is nonspecific, most patients present with local pain and swelling of the affected site.[2,4] In addition, EH of bone can rarely be associated with paraneoplastic manifestations such as hemolytic anemia and consumptive coagulopathy.[4]

On gross anatomy, the tumor appears as a well-defined mass with irregular borders and bright red appearance. On microscopic examination, the tumor mass consists of solid nests and cords of round, polygonal and spindle-shaped cells with eosinophilic cytoplasm. The most characteristic feature is the presence of intracytoplasmic vacuoles, indicating primitive vascular channels.[4]

On radiographs and CT, EH of bone can present as multifocal lesions in a single bone or multicentric lesions involving multiple bones. Lesions are locally aggressive, expansile, lytic with cortical destruction and a soft tissue component. Matrix mineralization and periosteal reaction are usually absent.[1,5] The presence of matrix calcification is a very unusual appearance and has been described only in very few cases.[6]

On MRI, it appears as a hypointense lesion on T1-weighted images and iso to hyperintense on T2-weighted images with homogenous enhancement on post-contrast images.[4] The lesion shows restriction on diffusion-weighted imaging.[7] The lesions show increased uptake on 99^{m}Tc Sestamibi bone scintigraphy and 18 F-FDG Positron emission tomography-CT.[2,7] Nuclear scintigraphy is useful to detect the multifocal and multicentric nature of the disease.[1]

However, the imaging features of EH are somewhat non-specific and the radiological differential diagnoses depend on the age of the patient. In young patients with the multifocal or multicentric disease, the differential diagnosis would include Brown tumors of hyperparathyroidism, lytic variant osteosarcoma with skip lesions, and metastasis. In adults, differentials such as brown tumors of hyperparathyroidism, multicentric giant cell tumor, metastasis, multiple myeloma, and angiosarcoma may be considered.[1,4]

Perhaps the most unusual imaging feature in our case was the presence of matrix calcifications, which made us think of other differentials which included the Telangiectatic variant of osteosarcoma and skeletal metastasis.

There is no standard treatment option for EH and therapy must be individualized. En bloc resection is done for solitary bone lesions and amputation for multicentric disease. Radiofrequency ablation can be used before surgery to decrease intraoperative bleeding and reduce the extent of resection.[2] Radiation therapy can be used for unresectable and metastatic diseases.[4] The role of chemotherapy is not yet established. The prognosis of EH varies individually, solitary lesions have a better prognosis, while visceral involvement and multifocal disease have a poor prognosis.[4]

CONCLUSION

EH is a rare, malignant vascular tumor that frequently involves the liver and lungs, with bones being a rare site. The clinical and radiological features are nonspecific and histopathological evaluation may be needed for definitive diagnosis. However, EH may be considered in the differential diagnosis of slow-growing, multifocal, or multicentric lytic tumor in long bones which does not show any matrix calcification or periosteal reaction.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

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