Original Research Paper

Volume-8 | Issue-10 | October-2018 | PRINT ISSN No 2249-555X

Radiology

MULTIMODALITY IMAGING EVALUATION WITH RADIOLOGICAL-PATHOLOGICAL CORRELATION OF AN INFREQUENTLY ENCOUNTERED OSSEOUS TUMOR; EPITHELIOID HEMANGIOENDOTHELIOMA

Ali Albweady Department of Radiology, Qassim University, Qassim Region, Saudi Arabia.

ABSTRACT Epithelioid hemangioendothelioma (EHE) has been described as a rare vascular bone lesion with histological features between hemangioma and high-grade angiosarcoma. Typically, the soft tissue is involved and, less frequently, the skin, bone, liver and lung are involved. The tumor cells are positive for vimentin, CD31and CD34, factor VIII related antigen, ERG, and FLI1on immunohistochemistry. Radiological features are not specific; it may appear as an osteolytic lesion. It can present as a multifocal disease in 40% of cases. Here, I'm describing the case of a 35-year-old female patient presented with vague right thigh pain.

KEYWORDS : Epithelioid Hemangioendothelioma, Bone Tumor, Albweady, Qassim University, Radiology, Musculoskeletal, Oncology,pathology

1. INTRODUCTION:

Epithelioid hemangioendothelioma (EHE) is an uncommon, well-differentiated endothelial tumor with a broad spectrum of behavior forming less than 1% of primary bone tumors. [1] The term hemangi oendothelioma is used for entity of vascular tumors with intermediate malignancy showing a histopathological feature between that of a hemangioma and angiosarcoma. [2] EHE is most commonly found in soft tissues but can also be found in skeletal tissues such as skull, spine, pelvis, femur, and tibia. [3] EHE growing in the bone is usually multifocal or multicentric in contrast to soft tissue tumors [4,5]. These tumors can be discovered as an incidental finding as they may have asymptomatic course. However, local pain is the most common presentation of EHE [6] or certain isolated tumors, curative resection with negative margins can achieve cure and long term local control. Role of chemotherapy and adjuvant radiation therapy remains unclear.

2. Case Report:

2.1. Clinical History:

A 35-year-old female presented with chronic vague right thigh pain with no definite history of major trauma. The pain was intermittent, but persistent. The patient came with a non-enhance CT, contrast-enhanced MRI and bone scan studies done outside our hospital and showed a fairly well-defined intramedullary lesion with extension to the cortex, but no aggressive features could be seen (Fig1,2,3).

Initial investigation in our hospital with a plain film of the right thigh was done which showed benign-looking cortically based lesion (Fig.4). Moreover, taking into consideration the provided outside studies and our plain film, recommendation of a biopsy was given accordingly.

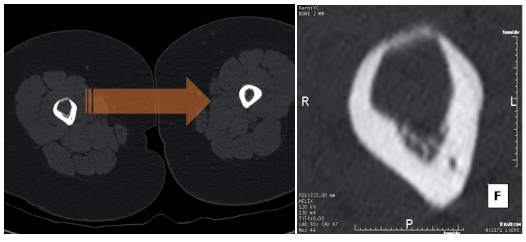

FIG.1: Axial CT and spot magnified image showing a mid-femoral diaphyseal expansile lytic lesion noted anteriorly. It measures 1.5 x 1.7 x 1.7 cm in AP, transverse and craniocaudal dimensions respectively. No internal matrix of calcification is demonstrated. The zone of transition with the adjacent cortex cannot be discerned. The anterior aspect of the cortex is markedly thinned with no evidence of cortical breakthrough. There's a solid adjacent periosteal reaction noted. No pathological fractures are demonstrated.

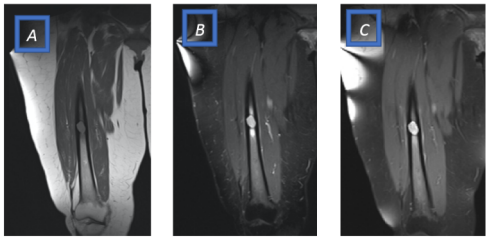

FIG.2: MRI coronal T1WI(A), T2WI(B) and T1 post-contrast(C) images of the right thigh exhibiting a well-defined intramedullary lesion with extension to the anterior cortex noted at the mid shaft of the right femur. There is thin periosteal reaction and enhancement as well as intense perilesional edema and enhancement. The signal of the lesion is mildly hyperintense on T1WI and intermediate high T2WI.

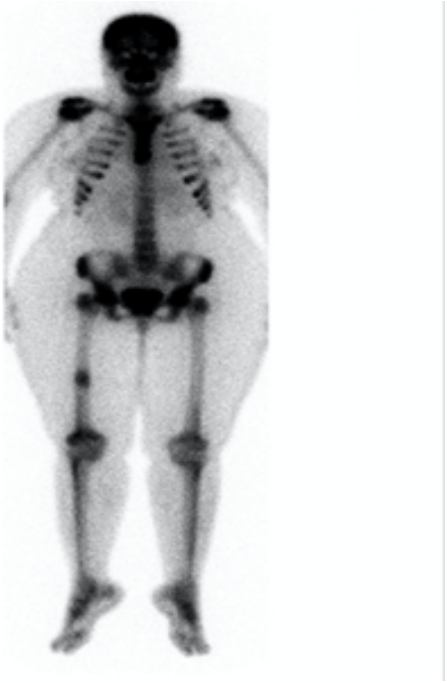

FIG.3:
Whole body bone scan showing intense focus of uptake (blue arrow), at the corresponding right femoral mid-shaft lesion.

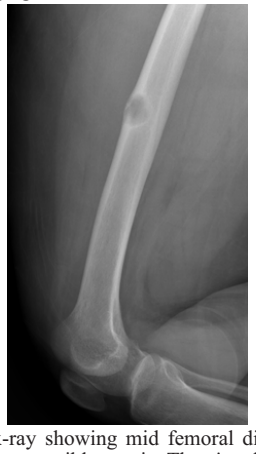

FIG.4: Plain film x-ray showing mid femoral diaphyseal expansile lytic lesion with no perceptible matrix. There's a thin narrow sclerotic zone of transition (red arrow).

2.2. Pathology Reports:

The obtained biopsy was further investigated utilizing H&E (Fig.5) staining and CD31 immunostaining (Fig.6) that revealed endothelial cells with focal vascular hyperplasia.

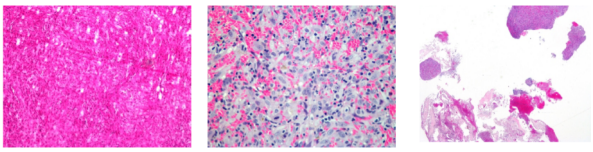

FIG.5:
H&E slides showing numerous small vessels, plump endothelial cells, occasional mitoses and one shows bony fragments.

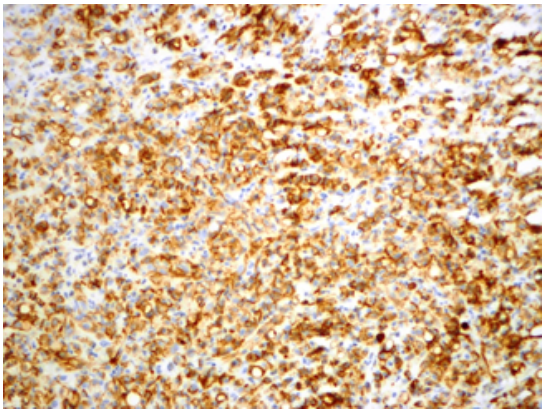

FIG.6:
Positive immunostain Cd31.

2.3. Follow up study, post biopsy:

A post biopsy follow-up MRI was obtained as follow (Fig.7).

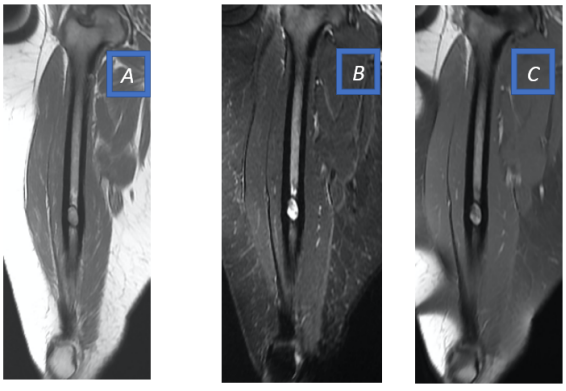

FIG.7:
MRI coronal T1WI(A), STIR(B), and Post-contrast (C) images show again the known intramedullary lesion at the mid femoral diaphysis. The lesion is stable in size in all three dimensions. The lesion exhibits iso signal intensity on T1 and intermediate to high signal intensity on T2 with heterogeneous enhancement after contrast administration. The previously noted periosteal enhancement and edema along the biopsy tract has completely resolved.

3.DISCUSSION:

EHE was described for the first time in 1982 by Weiss and Enzinger as a subgroup of the hemangioendotheliomas having an important endothelial or histiocytic component and they described EHE for soft tissue neoplasm. [7] EHE of the bone is rare and a review of literature reveals a slight male predominance. [8] Since EHE has a higher incidence of multicentricity compared to conventional angiosarcoma of bone, a complete bone scan is needed to rule this out.

Local pain, swelling, and pathological fracture are the common presenting clinical scenarios. Although the radiographic appearance of the EHE is variable, it usually presents as an expansive, osteolytic, and poorly demarcated lesion as seen in our case here. [9] Cortical disruption and extension into soft tissues can be present with joint invasion. [10]

The histopathological examination of EHE shows strands or solid nests of epithelioid endothelial cells usually growing centrifugally around a central vein in a stroma varying from myxoid to hyalinized. The cells form primitive vascular channels, occasionally containing erythrocytes. The epithelioid endothelial cells typically have abundant eosinophilic cytoplasm with vacuoles. The nucleus is usually oval shaped and concentrically placed [11,5]. The EHE cells show immunoreactivity for CD31, CD34, factor VIII, and vimentin. [12] Positive reaction for cytokeratin and epithelial membrane antigen (EMA) have been noted in one- fourth of the cases but the expression is weak and focal. [11,5]

The differential diagnosis for EHE of the bone includes osteomyelitis and various tumors such as skeletal angiomatosis, Langerhans cell histiocytosis, myeloma, lymphoma, angiosarcoma, and metastasis. A definitive diagnosis based on radiological findings alone is often difficult.[12] Histopathology remains the mainstay for the diagnosis.

The ideal treatment of EHE of the bone is wide resection to reduce local recurrence. Radiation therapy increases the risk of radiation sarcoma and should be reserved for inoperable cases.[13] Nevertheless, the role of radiotherapy remains controversial. In the series of Kleer et al. [5] four out of 10 patients who underwent radiotherapy for EHE had a favorable evolution. However, other studies conclude that due to risk of radiation-induced sarcomas, radiation therapy should be reserved to those cases not amenable to wide surgery or when lesions are seated locations difficult to treat.[13] The role of adjuvant chemotherapy, on the other hand, in the treatment of EHE, has not been established so far. [11]

4.CONCLUSION:

Primary vascular tumors of the bone are very rare. EHEs are a select subgroup of vascular tumors that do not lend itself well to characterization both clinically and histologically. Immunohistoche mistry is a useful adjunct in diagnosing these tumors. The behavior of these tumors is intermediate; hence, it is important not to misdiagnose these as angiosarcomas. Variously known as low degree anaplastic angiosarcoma, cellular hemangioma, histiocytoid hemangioma, or hemangioendothelioma, are locally aggressive and have high rates of recurrence, thereby necessitating aggressive local treatment which is mostly surgical. Although wide surgical excision is usually required, treatment plans should depend on extension and location of the disease.

Conflict of Interests

The authors declare that there is no conflict of interests regarding the publication of this paper.

ACKNOWLEDGMENTS

The authors thank the Consultant Anatomic Pathologist: Muhammad Amin ur Rahman, MD, FRCPath (UK)., for the pathological and histological slides preparation, as well as the Department of Pathology at King Fahad Medical City, Riyadh-Saudi Arabia.

Financial Support
Nil.

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