

CASE REPORT

Epithelioid hemangioma of distal femoral epiphysis in a patient with congenital talipes equinovarus

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Abstract

Background: Epithelioid hemangioma (EH) is a rare benign vascular lesion of soft tissue and bone, characterized by endothelial cells with epithelioid or histiocytoid appearance. Though tubular bones, flat bones, vertebra and short bones are common sites for this lesion, the epiphyseal involvement is extremely rare. We present an unusual case of EH of the distal femur in a young boy. **Case report:** A 12-year-old boy who had congenital talipes equinovarus of the right foot presented with progressively increasing pain in the right lower thigh for six months. Physical examination revealed muscular atrophy of the right lower limb and a moderately tender swelling in the medial aspect of the right knee without restriction of knee movement. An X-ray revealed an osteolytic lesion, which appeared iso- and hypointense on T1W and hyperintense on T2W MRI images in the distal epiphysis and adjacent metaphysis of the right femur. A radiological diagnosis of chondroblastoma was entertained. The patient was treated with curettage and bone grafting. Histopathology showed a tumor composed of thin-walled arteriolar capillaries lined by large, polyhedral epithelioid endothelial cells with vesicular nuclei, finely distributed nuclear chromatin, and moderate amount of eosinophilic cytoplasm. The endothelial cells were strongly immunopositive for CD34. Mitotic activity was low and the Ki-67 proliferative rate was <2%. A diagnosis of EH was made. EH is a benign lesion and it should be differentiated from its histologically similar malignant counterparts such as epithelioid hemangioendothelioma and epithelioid angiosarcoma as the lesion can be successfully treated with curettage or resection.

Keywords: epithelioid hemangioma, congenital talipes equinovarus, chondroblastoma, femoral epiphysis

INTRODUCTION

Epithelioid haemangioma (EH) is an uncommon benign vascular tumour, which was first described by Wells & Whimster in 1969.¹ It is usually found in the dermis, subcutis, lymph nodes, and rarely in visceral organs.²⁻⁴ The tumor typically occurs during early adulthood and is more common in women.⁵ Osseous epithelioid hemangiomas are rare and may occur in any bone.⁶ Rarely it can be multifocal involving multiple bones and extraskeletal tissues.⁶ Some authors believe that osseous EH is a variant of hemangioendothelioma, which has malignant potential.⁷ However, clinical and radiological features, and follow-up studies suggest that it is a benign lesion.⁶ The common sites of osseous EH are long and short tubular

bones, flat bones, vertebra and small bones of hands.⁴ The epiphysis, however, is an unusual site for this lesion. We present a case of EH of the distal femur in a young boy with congenital talipes equinovarus.

CASE REPORT

A 12-year-old boy presented with a history of progressively increasing pain in the right lower thigh for six months. There was no history of trauma, fever, cough, night sweat or weight loss. The patient had congenital talipes equinovarus of the right foot, for which he was being treated with gentle manipulation and casting of the foot at weekly intervals (Ponseti's method) in the outpatient department. Physical examination

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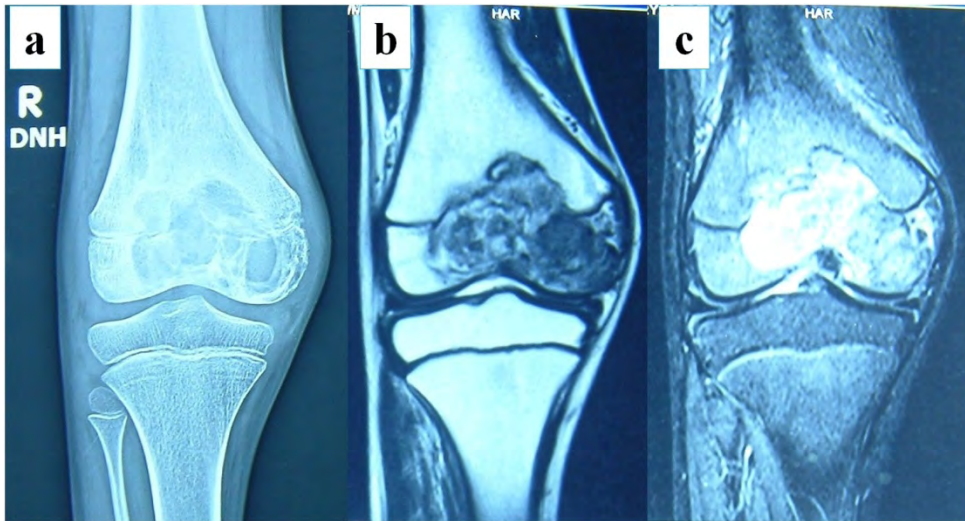


FIG. 1: (a) Plain radiograph of right thigh showing an osteolytic lesion in the distal epiphysis and adjacent metaphysis of the right femur. Coronal T1W and T2W fat saturated MR images show hypointense lesion on T1-weighted image (b), and hyperintense lesion on T2-weighted image (c), involving distal end of the right femur.

revealed atrophy of muscles of the right lower limb and a moderately tender swelling in the medial aspect of the right knee. There was no restriction of knee movement. The right foot showed severe talipes equinovarus. Examination of the other systems was unremarkable.

An X-ray of the right lower thigh showed an osteolytic lesion in the distal epiphysis and adjacent metaphysis of the right femur (Figure 1a). Magnetic resonance imaging (MRI) scan revealed heterogeneous signal intensity with hypo- and isointense lesion on T1-weighted (Figure 1b) and markedly hyperintense on T2-weighted images (Figure 1c). A radiological diagnosis of chondroblastoma was offered. Chest X-ray, complete blood count and erythrocyte sedimentation rate (ESR) were within normal limits. The patient was treated for the lesion with local curettage and bone grafting.

Pathology

The entire curetted tissue was processed for microscopic examination. Hematoxylin and eosin stained sections showed a tumor composed of thin-walled arteriolar capillaries lined by large, polyhedral epithelioid endothelial cells with vesicular nuclei, finely distributed nuclear chromatin, and moderate amount of eosinophilic cytoplasm (Figures 2a). No intracytoplasmic vascular lumina were identified. Few eosinophils, lymphocytes and plasma cells were also seen. Focal haemorrhage was present but no brisk mitoses were seen. The endothelial cells were

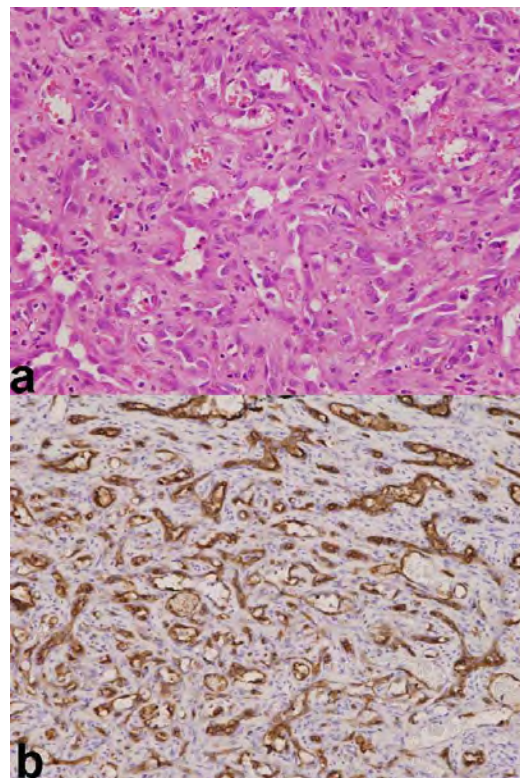


FIG. 2: Photomicrographs showing solid tumour composed of thin-walled arteriolar capillaries lined by prominent epithelioid endothelial cells (a, H&E original magnification x200). The tumour cells are immunopositive for CD34 (b, Original magnification x100).

strongly immunopositive with the antibody against CD34 (1:100) (Diagnostic BioSystems, USA) (Figure 2b). The tumor cells revealed low proliferative activity with Ki 67 labelling index <2%.

DISCUSSION

Epithelioid hemangioma, is a rare benign, slow growing vascular lesion that typically occurs in the head and distal extremities. The possible pathogenesis is (i) benign vascular neoplasm or (ii) reactive process to a preceding trauma. It occurs in mid-adult life and is slow growing, with an average time between appearance and excision of four and a half months.⁸ Osseous hemangiomas account for less than 1% of all primary bone tumors and most often involve vertebral bodies and calvarium.⁹ Peripheral tubular bones and the ribs are infrequently affected.⁹ The histopathological subtypes of osseous hemangiomas are cavernous, capillary, arteriovenous, venous and epithelioid variants.² Most of the solitary hemangiomas of bone are asymptomatic and diagnosed as an incidental finding on radiographs. Middle-aged patients are usually affected with a predilection for females.⁵ Hemangioma of the tubular bone may be periosteal, intracortical, or medullary type which is further subdivided into the diaphyseal (48%), metadiaphyseal (30%), metaphyseal (12%), metaepiphyseal (4%), epimetadiaphyseal (3%) and epiphyseal type (1%).¹⁰ Epiphyseal or juxta-articular hemangiomas are rare.¹¹ Capillary and cavernous hemangiomas are the most common in bone, whereas EH, also known as histiocytic hemangioma, and angiolymphoid hyperplasia with eosinophilia is less common. The endothelial cells of EH are referred to as 'histiocytoid' because of their lobated nuclei, decreased alkaline phosphatase and increased acid phosphatase content compared to normal endothelium.⁵

The aetiology of EH remains unknown. Boudousquie *et al* reported that epithelioid hemangioendothelioma which is the low grade malignant counterpart of EH has a complex translocation between chromosome 7 and 22.¹² Osseous EH should be distinguished from some histologically similar low grade malignant tumors like epithelioid hemangioendothelioma and epithelioid angiosarcoma.^{2,13} Many of the tumors previously reported as low-grade hemangioendothelial sarcoma indeed represented EH.¹³ Microscopically EH has

lobular organization of capillaries lined by large polyhedral epithelioid endothelial cells with oval to bean shaped nuclei having fine chromatin. The cytoplasm is usually abundant and deeply eosinophilic. Occasional one or more round, clear spaces containing red blood cells are seen. The endothelial cells may be plump and intruding into the lumen like a tombstone.⁶ As EH is a benign tumor, the cells have low mitotic index and absent cellular atypia.¹⁴ Prominent inflammatory infiltrate including eosinophils, lymphocyte and plasma cells may be present. Occasionally a central parent vessel can be identified in the centre of the lesion.^{2,6} Epithelioid hemangioendothelioma on the other hand is a borderline malignant tumor with less vascular differentiation and a tendency to recur and metastasize. It is more often multifocal and tends to have a more aggressive clinical course. Unlike EH, in haemangioendothelioma the endothelial cells are usually arranged in cords and nests in a distinct myxohyaline or chondroid background without prominent inflammatory component. There is nuclear atypia and the eosinophils are generally absent.¹⁵ Epithelioid angiosarcoma is a malignant tumor, which usually shows larger cell size, larger, irregular nucleoli, higher nucleo-cytoplasmic ratio, greater mitotic activity and frequent necrosis.^{2,13} It has a more aggressive clinical course than epithelioid hemangioendothelioma. Proliferation markers like Ki-67 labelling index will be higher in epithelioid hemangioendothelioma and angiosarcoma as against EH. In our case there was minimal cellular atypia with low mitotic index.

Radiologic examination classically demonstrates osteolytic lesion with or without surrounding sclerosis. MRI is helpful in evaluating osseous lesions. EH on T1-weighted MRI images shows areas of decreased signal intensity; while on T2-weighted images the vascular components demonstrate markedly hyperintense signal. Cortical expansion and cortical destruction have been described in rare cases.¹¹ T1-weighted image is based on relaxation properties of various protons thereby assessing the marrow involvement by the tumor, identifying fatty tissue, and hemorrhage. T2-weighted image is created based on decay properties of various protons in the body and is useful for detecting abnormalities like oedema, soft tissue component and skip lesions. The radiological differential diagnoses may be giant cell tumor, aneurysmal bone cyst, plasmacytoma,

osteoblastoma, infection, benign vascular lesion etc.^{11,16} The radiologic diagnosis of our case was chondroblastoma.

Nielsen *et al* reported 50 cases of osseous EH with ages ranging from 10 to 75 years with a mean of 35 year. The majority of the lesions were in long and short tubular bones (58%), and 18% of patients had more than one bone involvement.⁶ In our case there was a single lesion that involved the epiphysis, epiphyseal plate and adjacent metaphysis of the distal femur. The patient in addition had congenital talipes equinovarus, which is a poorly understood developmental disorder of the lower limb with fixation of the affected foot in adduction, supination and varus and concomitant soft tissue abnormalities. The coexistence of EH and talipes equinovarus in our case could be an incidental association. The patients with EH are treated with curettage or by local resection depending on the location and extent of the lesion.² Complications of this entity includes local recurrence though rare.⁵

EH of bone is a distinct vascular tumor, which should be distinguished from epithelioid hemangioendothelioma and epithelioid angiosarcoma as it is a benign lesion which can be successfully treated with curettage or marginal en block resection.

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