

Case 9298

Epithelioid hemangioendothelioma of the femur

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Section: Musculoskeletal System

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Patient: 27 year(s), male

Clinical History

A 27-year-old man presented at the orthopedic department with a 3-year history of severe limitation on his locomotor capacity and persistent pain at the anteromedial side of the right thigh. These symptoms had been diagnosed and treated as a psychosomatic disorder because of his known mental illness.

Imaging Findings

Digital radiography revealed the presence of a large endosteal bone neoformation. The patient underwent radiological investigations such as CT that confirmed the presence of a gross neoplastic lesion involving the middle third of the right femur proximal diaphysis, expanding the cortical bone that appears discontinuous in several places, associated with periosteal reaction, most evident at the front side. This lesion measured approximately 177x73x69 mm and showed an inhomogeneous enhancement after contrast injection. The angio-CT documented bilateral patency of leg arteries. Subsequently T1, T2-weighted MRI and fat-suppressing sequences were performed for further characterization of the lesion, which confirmed the morpho-structural alteration of the proximal right femoral diaphysis caused by the lesion apparently constituted of a productive hypocellular tissue. At PET scan the neoplasm demonstrated a high captation of FDG. At last bone biopsy was performed for histological characterization.

Discussion

Vascular bone tumours represent less than 1% of all bone tumours and their classification seems to be still evolving [1]. It was first described by Weiss and Enzinger in 1982 and often mistaken for a

high grade malignant tumour. Considering their histological appearance and their biological behaviour these tumours are classified as intermediate between benign haemangiomas and malignant angiosarcomas and as proposed by Wanger and Wald they constitute their own subgroup between low grade and high grade bone vascular malignancies [2, 3]. So the term epithelioid haemangioendothelioma was suggested to be used to designate these biologically borderline" neoplasms. EHE occurs in the calvarium, spine, femur, tibia and feet of adults during the second or third decade. Chromosome analysis and molecular cytogenetic investigations showed that EHE is characterised by complex genetic rearrangements of 11q and 12q [4].

Usually, epithelioid haemangioendotheliomas present with pain and swelling and if present in the spine, they can cause radicular symptoms or paraplegia. Radiographically the majority of EHE show lytic bone destruction with associated alteration of matrix mineralisation, endosteal erosion and cortical thinning [5]. The contrast enhancement is easily evidenced by the Computed Tomography which can be applied for further characterisation of the neoplasm such as local and systemic extension and for vascular supply assessment of the tumour. At MRI the EHE usually presents with intermediate signal intensity on T1-weighted images, high signal on T2-weighted and homogeneous contrast enhancement without the detection of the typically serpentine vascular structures that would help in the differential diagnosis. FDG PET scanning in EHE, shows bone marrow involvement and determines the extent of the disease [6]. Although, the information gained through the imaging techniques are useful to identify and to stage the lesion, they are not entirely specific and the confirmation of the diagnosis is made possible only by biopsy and pathologic examination. The epithelioid histological subtype of haemangioendothelioma has epithelial-like cells lining the vascular channels that are large and cuboidal and contain abundant eosinophilic cytoplasm. Immunohistochemical study is helpful in confirming the diagnosis by identifying the markers for vascular endothelial cell. The clinical course and prognosis depend on uni or multifocality, histological differentiation and cytologic atypia. The overall survival is 89% in unifocal disease and 50% in multifocal involvement [7]. Surgical excision is considered the best treatment in addition or not with post-operative chemotherapy or previous embolisation of the lesion [8].

Final Diagnosis

Epithelioid haemangioendothelioma

Differential Diagnosis List

Haemangio-epithelioma , Haemangioma, Angiosarcoma, Chordoma, Chondrosarcoma, Adamantinoma of Long Bones

Figures

Figure 1 Digital radiography

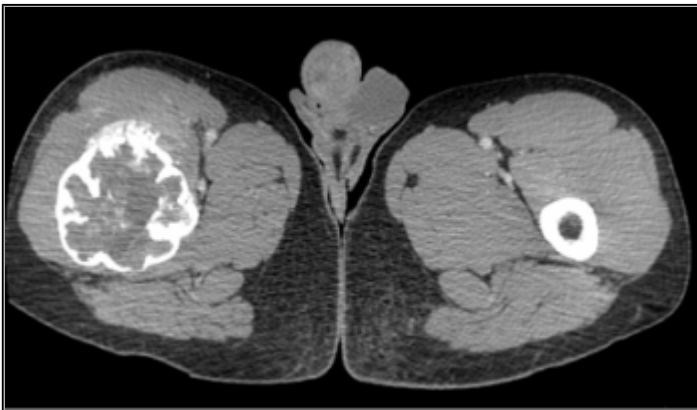




Digital radiography shows the presence of an extended endosteal bone neoformation in the context of a proximal diaphysis morphostructural alteration (cranio-caudal extension of 17 cm and maximum transverse diameters of 73 x 73 mm)

Area of Interest: Bones; Musculoskeletal bone; Musculoskeletal system;
Imaging Technique: Digital radiography;

Figure 2 CT scan



Axial CT scan reconstruction shows the presence of a gross expansive lesion involving the right femur proximal diaphysis, which appears to swell the cortical bone with discontinuities and periosteal reaction.

Area of Interest: Bones; Extremities; Musculoskeletal bone;



Coronal CT scan reconstruction shows the presence of a gross inhomogeneous expansive solid lesion with calcifications involving the right femur proximal diaphysis with the cortical bone swelling and thick periosteal reaction.

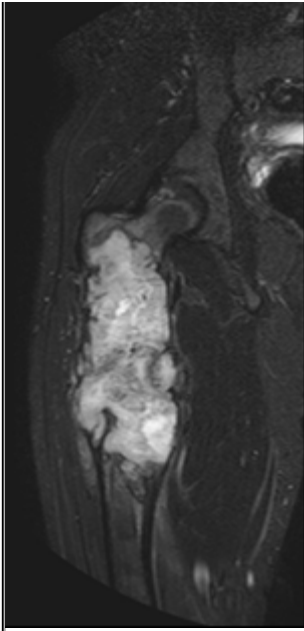
Area of Interest: Bones; Extremities; Musculoskeletal bone;
Imaging Technique: CT;



MPVR CT reconstruction shows the presence of a gross expansive lesion involving the right femur proximal diaphysis.

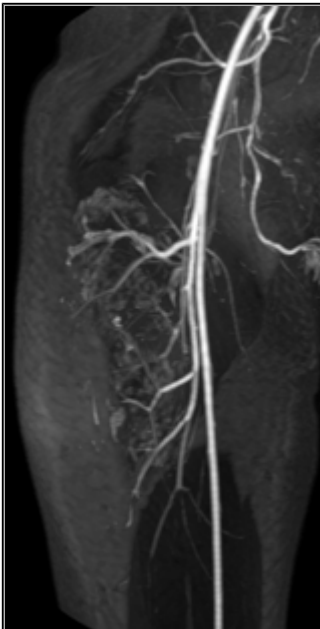
Area of Interest: Bones; Extremities; Musculoskeletal bone;

Figure 3 MR images



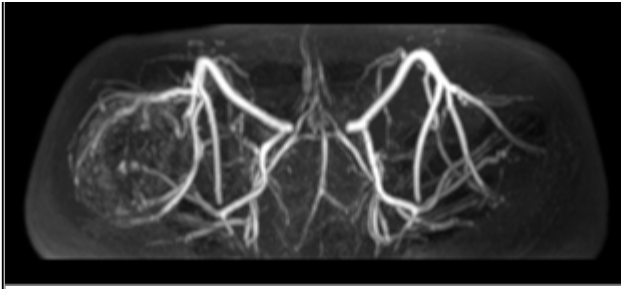
T1-weighted MR image with fat suppression after Gadolinium i.v. injection demonstrates a lesion composed of juxtaposed hypocellular tissue, with mild-high contrast enhancement.

Area of Interest: Bones;
Imaging Technique: MR;



MRA MIP reconstruction coronal plane: vascular pedicles feeding the lesion from the right femoral artery.

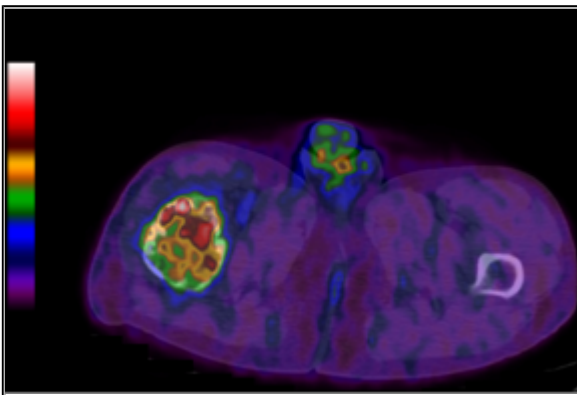
Area of Interest: Vascular;
Imaging Technique: MR-Angiography;



MRA MIP reconstruction axial plane: vascular pedicles feeding the lesion from the right femoral artery.

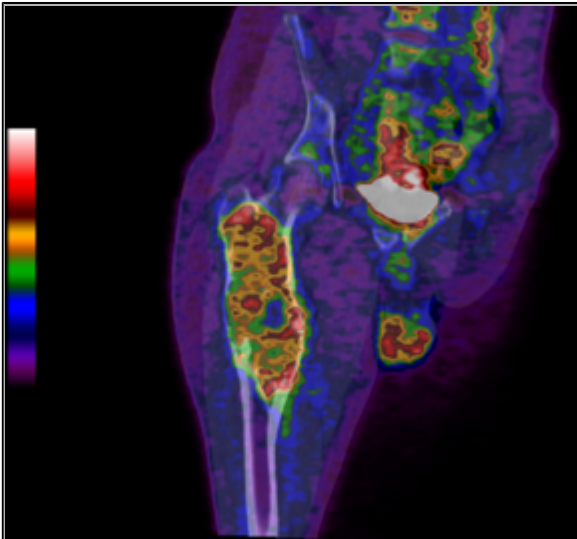
Area of Interest: Vascular;
Imaging Technique: MR-Angiography;

Figure 4 Fusion PET-CT images



Fusion PET-CT axial image shows high and heterogeneous ^{18}F -FDG uptake by the neoplastic mass (SUV max.3.5).

Area of Interest: Bones; Oncology;
Imaging Technique: PET-CT;



Fusion PET-CT coronal image shows an ^{18}F -FDG avid neoplastic mass.

Area of Interest: Bones;
Imaging Technique: PET-CT;

Figure 5 Post-surgery digital radiography



Post-surgery digital radiography documents cadaveric femur transplantation and fibula autologous graft with multiple bone screws at the diaphysis of the right femur.

Area of Interest: Bones;
Imaging Technique: Digital radiography;

Figure 6 Post-surgery DSA image



Post-surgery DSA image with superficial femoral and fibular pedicle arteries latero-lateral anastomosis.

Area of Interest: Vascular;

Figure 7 3D shaded-surface rendering CT reconstruction

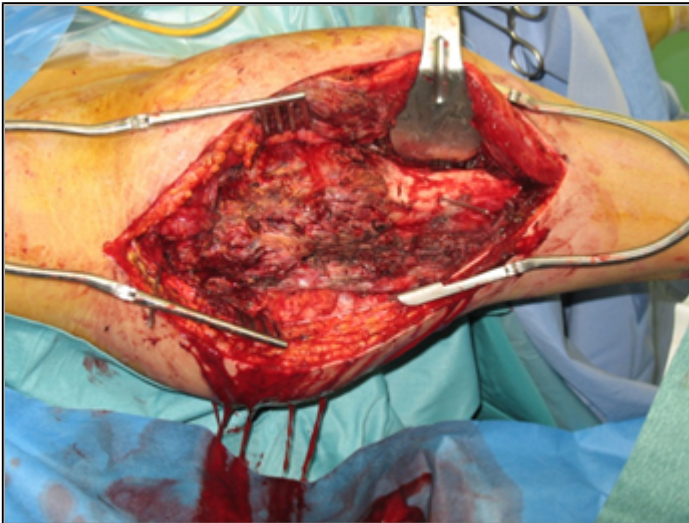




3D shaded-surface rendering CT reconstruction highlights anatomical proximity of screws (red) and grafts.

Area of Interest: Bones;
Procedure: Computer Applications-3D;

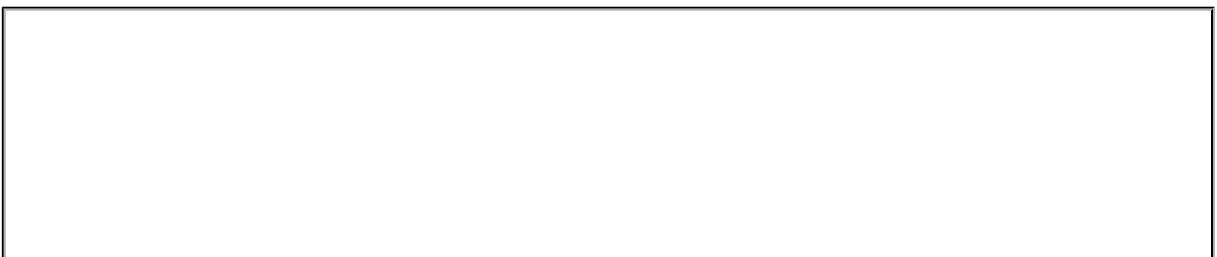
Figure 8 Lesion exposure



Lesion exposure during surgical excision.

Area of Interest: Anatomy; Bones;
Procedure: Intraoperative; Removal; Surgery;

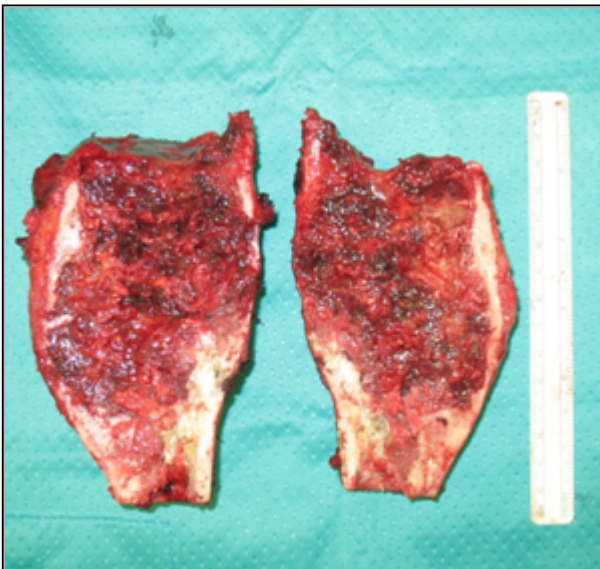
Figure 9 Gross specimen





Gross specimen cut surfaces with solid inhomogeneous hypervascularised lesion. Poorly-defined intramedullary mass, extending through cortex with moth-eaten bone destruction and aggressive periosteal reaction.

Area of Interest: Anatomy; Bones;
Procedure: Biopsy; Removal; Surgery;

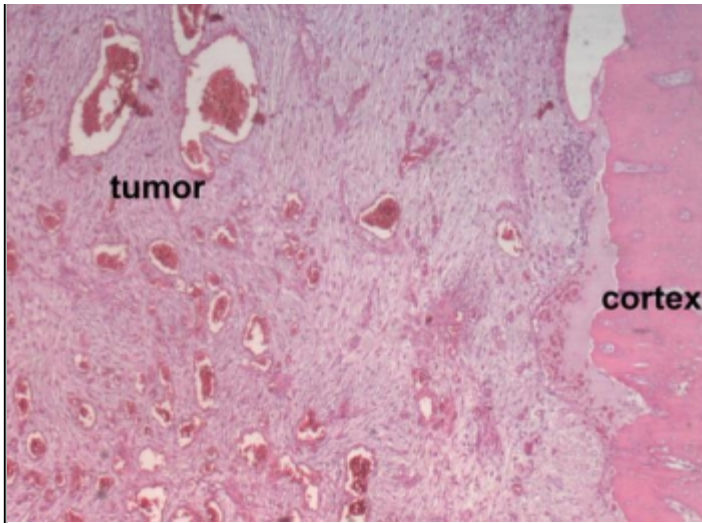


Gross specimen cut surfaces with solid inhomogeneous hypervascularised lesion. Poorly-defined intramedullary mass, extending through cortex with moth-eaten bone destruction and aggressive periosteal reaction.

Area of Interest: Anatomy; Bones;
Procedure: Biopsy; Removal; Surgery;

Figure 10 Hematoxil-Eosin histopathology

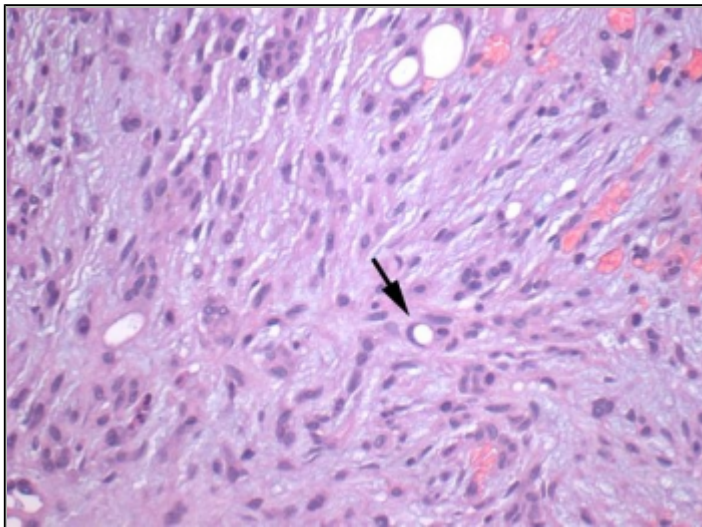




Lining cells of gland-like structure show irregular vesicular or optically clear nuclei with prominent basophilic nucleoli and occasional vacuolated cytoplasm. Epithelioid haemangioma morphology characterised by epithelioid endothelial cells.

Area of Interest: Musculoskeletal soft tissue;

Figure 11 Hematoxylin-Eosin histopathology

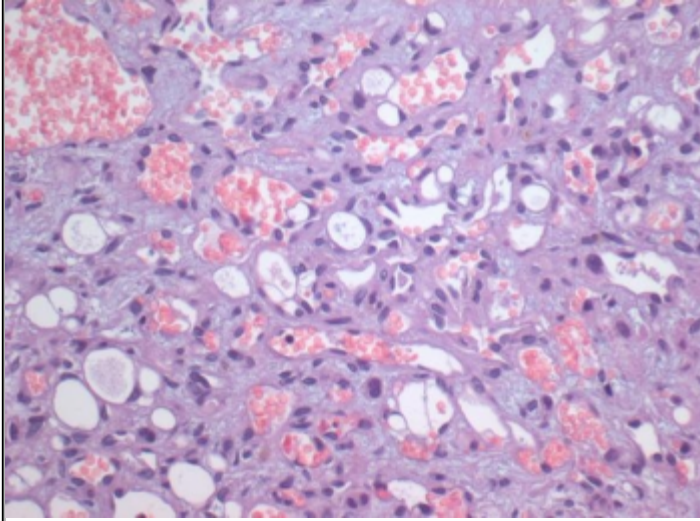


The epithelioid histological subtype of haemangioma has epithelial-like cells lining the vascular channels that are large and cuboidal and contain abundant eosinophilic cytoplasm.

Area of Interest: Musculoskeletal soft tissue;

Figure 12 Hematoxylin-Eosin histopathology

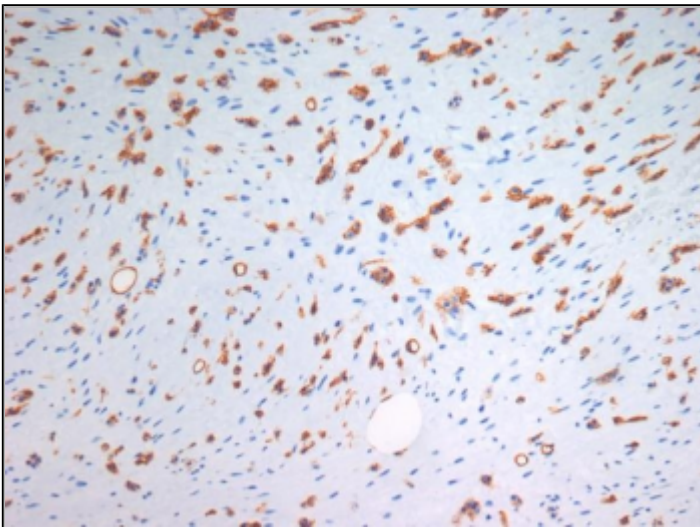




The epithelioid histological subtype of haemangioma has epithelial-like cells lining the vascular channels that are large and cuboidal and contain abundant eosinophilic cytoplasm.

Area of Interest: Musculoskeletal soft tissue;

Figure 13 CD34 immunopathology

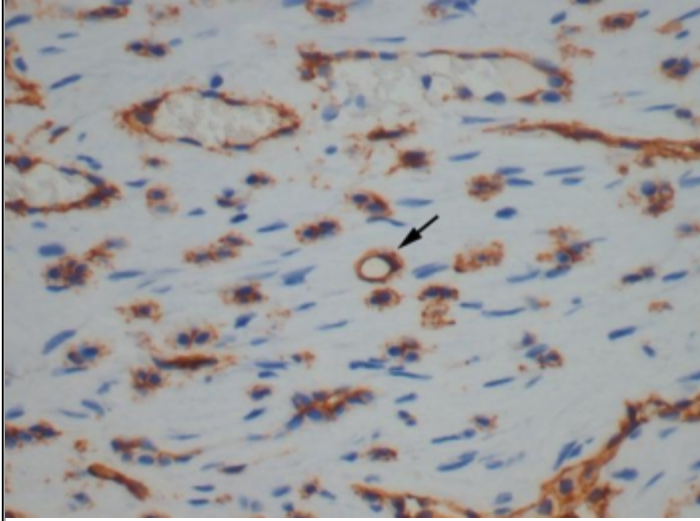


Immunohistochemical study is helpful in confirming the diagnosis by identifying the markers for vascular endothelial cell (CD34+).

Area of Interest: Musculoskeletal soft tissue;

Figure 14 CD34





Immunohistochemical study is helpful in confirming the diagnosis by identifying the markers for vascular endothelial cell (CD34+).

Area of Interest: Musculoskeletal soft tissue;

MeSH

Hemangioendothelioma, Epithelioid [C04.557.645.375.370.380]

A tumor of medium-to-large veins, composed of plump-to-spindled endothelial cells that bulge into vascular spaces in a tombstone-like fashion. These tumors are thought to have "borderline" aggression, where one-third develop local recurrences, but only rarely metastasize. It is unclear whether the epithelioid hemangioendothelioma is truly neoplastic or an exuberant tissue reaction, nor is it clear if this is equivalent to Kimura's disease (see ANGIOLYMPHOID HYPERPLASIA WITH EOSINOPHILIA). (Segen, Dictionary of Modern Medicine, 1992)

Anastomosis, Surgical [E04.035]

Surgical union or shunt between ducts, tubes or vessels. It may be end-to-end, end-to-side, side-to-end, or side-to-side.

Magnetic Resonance Angiography [E01.370.350.500.500]

Non-invasive method of vascular imaging and determination of internal anatomy without injection of contrast media or radiation exposure. The technique is used especially in CEREBRAL ANGIOGRAPHY as well as for studies of other vascular structures.

References

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Citation

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