

Solid Tumour Section

Mini Review

Bone: Angiosarcoma

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Identity

Alias

Primary malignant vascular tumours of bone

Note

Variable proliferation of tumour cells which show an endothelial phenotype.

Classification

Note

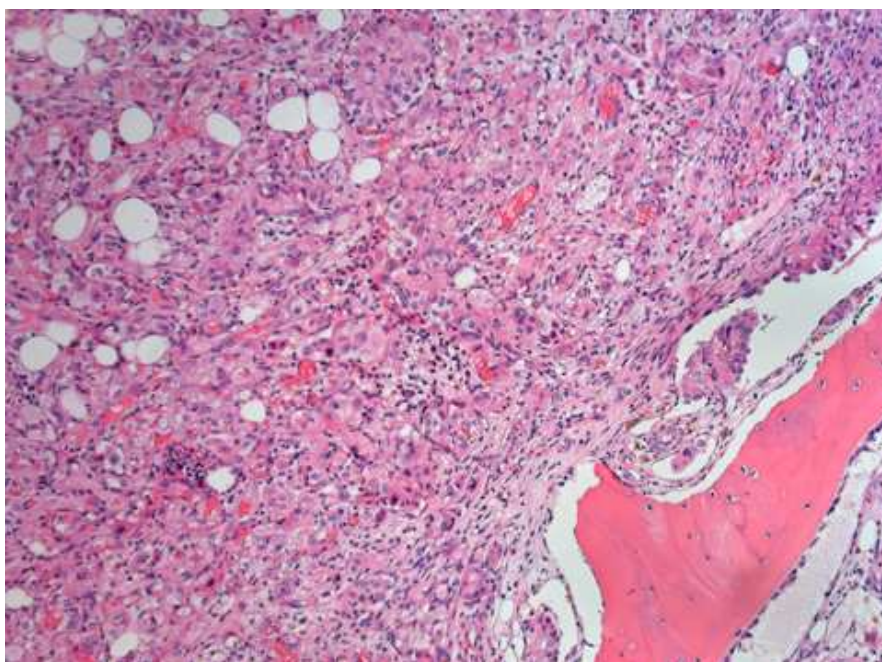
Angiosarcoma is the most accepted term for high-grade malignant vascular tumors of bone, as recognized in the 2002 World Health Organization (WHO) classification. However, the classification

of low to intermediate grade vascular tumors of bone, in particular of haemangioendotheliomas, is extremely difficult due to the lack of uniform terminology and accepted histological criteria. Many classification systems have been proposed, but none of them have been generally accepted so far.

Classification

Today, there is no generally accepted classification scheme. However, the 2002 WHO classification subdivide angiosarcoma in:

well differentiated angiosarcomas,
poorly differentiated angiosarcomas and
epithelioid haemangioendotheliomas.



Microscopy - PMVTB: epithelioid morphology - HE (10x).

Clinics and pathology

Disease

Primary malignant vascular tumours of bone

Etiology

Although there is in a very small percentage an association between angiosarcoma and radiation, the etiology of the majority of angiosarcomas remains unknown.

Epidemiology

Primary malignant vascular tumours of bone (PMVTB) are extremely rare and represent less than 1% of primary malignant bone tumours.

Clinics

Angiosarcomas of bone arise in adults and have a wide, nearly equal age distribution from the second till eighth decade. In contrast to epithelioid haemangioendotheliomas have a tendency to occur in young adults (second - third decade). It seems that males are slightly more affected than females.

Angiosarcomas of bone have a wide skeletal distribution; however, they have a tendency to occur in the short and long tubular bones of the extremities of which the femur, tibia and humerus are most often affected. In contrast to the soft tissue counterpart, one third of the tumours are multifocal, involving two or more distant bones.

In general, malignant vascular tumours of bone present as a painful mass. Depending on the size and localization of the tumour, neurological deficit or other symptoms can occur.

Pathology

Similar to the soft tissue counterpart (Angiosarcoma of the soft tissue), malignant vascular tumours of bone have variable histological features, varying from well differentiated to poor differentiated lesions. To a certain extent, the formation of vascular channels is one of the most common referred hallmarks of this tumour. These lesions show a variable amount of mitotic features, atypical mitotic figures, necrosis and cytonuclear atypia. Cells can have a spindle cell or an epithelioid morphology. Epithelioid haemangioendothelioma is considered a separate entity with distinct histomorphological features and clinically a relatively favorable prognosis. These tumours consist of strands or cords of epithelioid cells with abundant eosinophilic cytoplasm and the presence of intracytoplasmic vacuoles. Small vascular channels can be seen and the myxoid or hyalinised stromal component is most characteristic for this type of lesion.

Treatment

There is no general rule for treatment. Treatment depends upon multiple factors, such as age, size, location of the tumour, and the extent of disease.

Therapeutic options are surgical intervention (wide en bloc resection or amputation) and radiotherapy. Chemotherapy is used, although its usefulness is not well documented.

Prognosis

Survival rates of malignant vascular tumours of bone are unknown. It is accepted that histologically well differentiated tumours have a better prognosis than poorly differentiated tumours. It is suggested that multifocal lesions also have a better prognosis, most likely due to the fact that these lesions are more often well differentiated.

Cytogenetics

Note: No underlying genetic disorders or aberrations are reported in malignant vascular tumours of bone so far. In literature two independent cases of epithelioid haemangioendothelioma of bone were analyzed and revealed in one case a reciprocal translocation of chromosome 10 and 14 and in the other case complex rearrangements involving deletions and gains of 11q and 12q. No karyotypes are reported for malignant vascular tumours of bone so far.

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