Bone: Vascular Tumours

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Identity

Note
Benign vascular lesions of bone are relatively common and occur most frequent as an asymptomatic incidental finding in the skull or spine. Primary malignant vascular tumors of bone are rare. They represent less than 1% of primary malignant bone tumors reported by the Netherlands Committee on Bone Tumors and 0.5% of those registered at the Mayo Clinic. Clinically they are extremely aggressive and have a very poor prognosis. Survival rates are unknown, but in those cases reported patients die very soon after diagnosis due to wide spread metastasis and lack of tailored therapy.

Classification

Note
Over the years, the terminology and classification of vascular tumors of bone has been highly controversial and in literature a great variety of names has been proposed. Today, angiosarcoma is the most acceptable term for high-grade malignant vascular tumors of bone, as recognized in the 2002 World Health Organization classification. However, the classification of low-grade vascular to inter-mediate tumors of bone, in particular of haemangioendotheliomas, is extremely difficult due to the lack of uniform terminology and accepted histological criteria. Many authors have proposed different classification systems, but due to small numbers of cases, their large diversity and the lack of good correlation with clinical outcome none of them have been generally accepted so far. The large variety of histological features of vascular tumors of bone suggests that it should be regarded as a spectrum with on one side the overtly benign lesions and on the other side the frankly malignant lesions. In between there are the low and intermediate grade lesions in which numerous histomorphological diversity can be seen and for which classification is most difficult.

Classification
Today, the most accepted classification of vascular tumours of bone is the 2002 WHO classification: Haemangioma and related lesions Angiosarcoma.

Schematic representation of histological spectrum of vascular tumors of bone: accepted histological criteria to classify vascular tumors of bone are lacking.
Clinics and pathology

Disease
Haemangioma and related lesions
Note
Multiple lesions are defined as (haem) angiomatosis

Phenotype / cell stem origin
Endothelial cell.

Epidemiology
Haemangiomas are relatively common.

Clinics
In general asymptomatic.

Cytogenetics
No cytogenetic investigations reported.

Prognosis
Haemangiomas have a good prognosis and low recurrence rate.

Disease
Angiosarcoma.

Epidemiology
Extremely rare.

Clinics
In general, presents as a painful mass. Depending on the size and localization of the tumour, neurological deficit or other symptoms can occur.

Cytogenetics
No cytogenetic investigations of angiosarcoma in bone are reported.

Prognosis
It is accepted that histologically well differentiated tumours have a better prognosis than poorly differentiated tumours.

References
Fletcher CDM, Krishnan Unni K, Mertens F. Pathology and genetics of tumours of soft tissue and bone. WHO Classification of Tumours: IARC press Lyon 2002.

This article should be referenced as such: