Bone: Chondroblastoma

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Published in Atlas Database: November 2017

Online updated version : http://AtlasGeneticsOncology.org/Tumors/ChondroblastomaID5148.html
DOI: 10.4267/2042/68973

This article is an update of : Romeo S, Hogendoorn PCW. Bone: Chondroblastoma. Atlas Genet Cytogenet Oncol Haematol 2003;7(3)

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Abstract

Review on Chondroblastoma, with data on clinics, and the genes involved.

Keywords
Chondroblastoma; H3F3B

Identity

Note

Chondroblastoma is a locally aggressive, rarely metastasizing bone tumour typically affecting the epiphyses of long bones from individuals with an immature skeleton.

Radiology of Chondroblastoma.

Phylum
Bones:Cartilage tumors:Chondroblastoma

Figures 1, 2 and 3: Typical radiological findings of a lytic eccentric lesion affecting the epiphysis of the humerus (1 RX, 2 NMR and 3 bone scan)
**figure 4:** The cellular areas are made up of polygonal cells with scattered multinucleated giants cells (Haematoxylin-Eosin stain).

**figure 5:** The polygonal cells are positive for S-100 immunostain.

**figure 6:** The polygonal lesional cells are focally positive for DOG1
Epidemiology
Chondroblastoma is a rare neoplasm accounting for less than 1% of all bone tumors. Age of occurrence is usually between 10 and 25 years with a male predominance. Older age of presentation for skull lesions is reported.

Clinics
Usual symptom at presentation is mild localized pain. Radiologically it occurs more often as an eccentric lytic lesion, with sclerotic borders, involving epiphyses of the long bones.

Pathology
The tumour is composed of cellular and matrix rich areas. Cellular areas are made up of so called "chondroblasts": round, or polygonal cells, with an oval to round nucleus and with well-defined eosinophilic cytoplasm. Mainly in non-decalcified sections the chondroblasts appear focally delimited by a thin calcification rim, so called "chicken wire". Matrix rich areas are composed of different types of matrix: chondroid, osteoid, fibrous and rarely mature hyaline cartilage. Mitoses, always typical, can be quite frequent, especially in the cellular areas.

Immunohistochemical stainings show reactivity of the neoplastic cells for S-100, DOG1 (focal) and Vimentin; although several other antigens are reported to be expressed (i.e. Smooth muscle actin and Cytokeratin). Multinucleated giant cells, especially at the periphery of matrix-rich areas, are almost always found. An associated aneurysmal bone cyst occurs in about 1/3 of the cases. Histological features of chondroblastoma.

Treatment
Simple curettage is the standard treatment. 

Evolution
Rate of recurrence is between 14-18% mainly occurring within 2 years, and showing a higher occurrence rate in case of temporal bone location. Rare lung metastases in benign chondroblastomas, are documented. However they are not progressive, and therefore simple observation is sufficient, if necessary followed by simple surgical resection.

Prognosis
The prognosis is good. Rare and doubtful malignant progression are described, but no universal criteria for this event are currently available, and several authors consider this as cases of a misdiagnosis.

Genetics
Recently a specific driver mutation in the histone 3 gene H3F3B (K36M), was identified in approximately 95% of chondroblastomas. H3F3B mutation detection can be used as a diagnostic tool for the distinction of chondroblastoma from other giant cell-containing tumors. The mutation can be detected using a mutation specific antibody for the K36M mutation.

Cytogenetics
DNA flow cytometry studies show chondroblastoma mainly to be a low proliferative diploid neoplasm, however aneuploid near-diploid populations have been reported.

Karyotypic results of 7 cases are available in the literature. No specific cytogenic abnormalities neither specific type of aberrations are reported sofar. However some chromosomes seem to be more often involved: 3 cases for chromosome 5, 2 for chromosome 8, 2 for chromosome 11 and 2 for chromosome 17.

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