

# Solid Tumour Section

## Mini Review

## Bone: Chordoma

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### Identity

#### Note

Chordoma is a malignant tumour derived from remnants of the fetal notochord; it occurs along the spinal axis, predominantly in the sphenoccipital (35%), vertebral (15%) and sacrococcygeal (50%) regions.

### Clinics and pathology

#### Etiology

Although most chordomas are sporadics, five families with chordoma occurrence have been reported, two of them displaying an autosomal dominant transmission with incomplete penetrance (MIM. \*215400); preliminary linkage data in a three generation family suggest that the disease locus might be assigned to chromosomes 1,17 or 19.

#### Epidemiology

Chordomas accounts for 1-4% of all primary bone tumours; the sacrococcygeal lesions are more common in the fifth decade of life, whereas the sphenoccipital tumours occur predominantly in children.

#### Clinics

Chordoma is a slowly-growing tumour, characterized by local destruction of bone and rarely distant metastatic spread.

The differential diagnosis includes renal tumours, chondrosarcomas and myxo-papillary ependymoma.

### Pathology

Microscopically, it resembles normal fetal notochord in its different stages of development; it is composed of extremely large cells (know as physaliferous) and other small tumour cells; areas of cartilage and bone may be present.

### Cytogenetics

#### Cytogenetics Morphological

In eight sporadic sacral chordomas with abnormal karyotype, a tumour-specific rearrangement has not been identified; the karyotypes, characterized by a hypo-or near-diploid chromosome number, showed complex rearrangements affecting several chromosomes, among them chromosome 1 was frequently involved in losses and translocations with different partners; no FISH experiments to clarify the complex cytogenetic picture have been performed so far.

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