

CHAPTER 17

Notochordal Tumours

Notochordal tumours arise from remnants of the notochord and hence occur exclusively along the midline. Tumours which occur elsewhere may resemble chordomas.

The majority of the tumours occur in the sacrum or in the clivus. Involvement of the remainder of the spine is unusual. One of the characteristic histological features of chordoma is a lobulated growth pattern.

Chondroid chordomas occur exclusively in the base of the skull and show features of both low grade chondrosarcoma and chordoma. Some studies have indicated a better prognosis for this subtype.

Chordoma

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Definition

Chordoma is a low to intermediate grade malignant tumour that recapitulates notochord.

ICD-O codes

Chordoma NOS	9370/3
Chondroid chordoma	9371/3
"Dedifferentiated" chordoma	9372/3

Epidemiology

Chordomas account for 1-4% of all primary malignant bone tumours. Chordoma most commonly presents after age 30, with the most common decade being the sixth (30% of patients). It is very rare under age 20 (1%). Male:female ratio is 1.8:1.

Sites of involvement

Axial spine (sacral 60%; sphenoccipital/nasal 25%; cervical 10%; & thoracolumbar 5%).

Clinical features / Imaging

The clinical features are related to the location and spread of the neoplasm. Being a slow-growing mass chordoma usually produces non specific symptoms for months to years before the diagnosis is made.

In the sacrococcygeal presentation pain is the most frequent symptom. It is usually referred to the lower back or tip of the spinal column. Constipation due to obstruction may develop. Almost all these neoplasms spread in the presacral area allowing physical detection by rectal examination. Nerve dysfunctions, such as anesthesia and paresthesia, are unusual and late manifestations. Those located in the sphenoccipital region are often associated with a chronic headache and symptoms due to compression of a cranial nerve. Ocular nerve involvement is the most frequent; compression and destruction of the pituitary gland may lead to endocrine disturbances; if spread is lateral a cerebellopontine angle tumour symptomatology can be evident. In case of spread inferiorly nasal obstruction, bleeding and

even a nasal mass may appear.

Chordomas arising in the cervical, thoracic and lumbar spine usually produce symptoms related to nerve roots or spinal cord compression and / or a palpable mass can be present. Characteristically cervical chordoma may clinically manifest as a parapharyngeal mass. Clinically, most patients experience progressive pain, swelling and/or neurological deficits that may ultimately be incapacitating.

Radiologically, chordomas are typically solitary, central, lytic, destructive lesions of the axial skeleton {2058} They are almost always associated with a soft tissue mass and shards of bony detritus. Intratumoural calcification may be present particularly in sacral tumours. In the sacral area they tend to displace the bowel and/or bladder {1302}. MRI studies best visualise soft tissue extension and its relationship to anatomic structures. On MRI, T-1 weighted images are hypo- or isointense {418}, while T-2 weighted images are of high signal intensity {418,1551}.

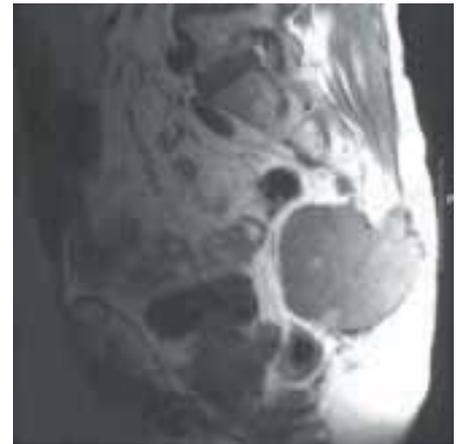


Fig. 17.01 Chordoma. T2 MRI showing a dark, lobulated, destructive mass in the sacrococcygeal region.

Macroscopy

Chordoma is a lobulated, glistening, greyish tan to bluish white, muco-gelatinous to friable, dark-red haemorrhagic tumour, generally from 5 to 15 cm. In most cases it is associated with extension beyond the contours of the bone into the surrounding soft tissues {418, 1468}.

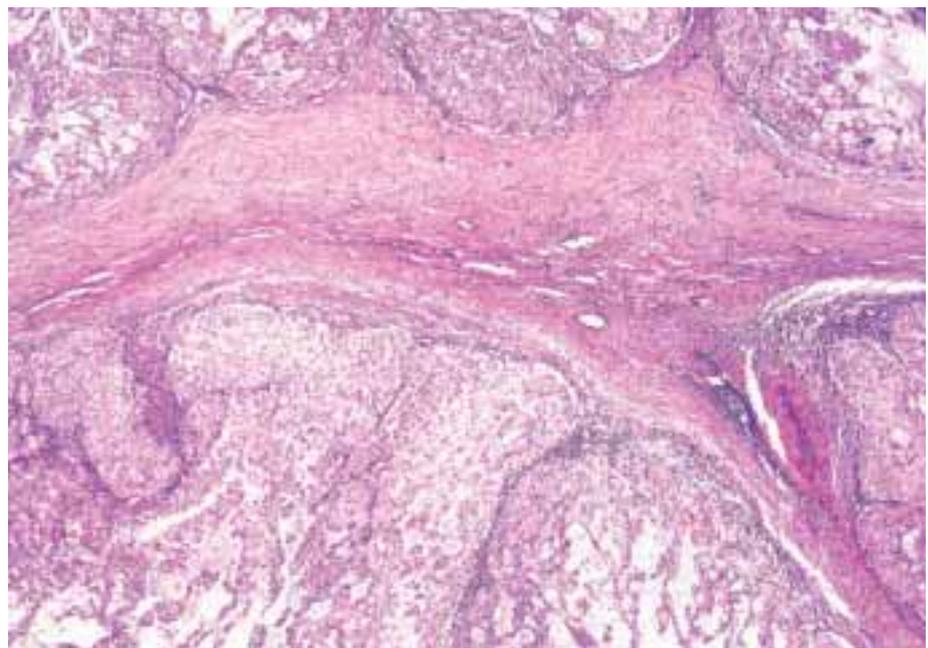


Fig. 17.02 Chordoma. Bands of fibrosis and lobularity typify this neoplasm on low power.

Histopathology

Chordomas are lobulated tumors, with individual lobules being separated by fibrous bands. The tumour cells are arranged in sheets, cords or float singly within an abundant myxoid stroma. They typically have an abundant pale vacuolated cytoplasm (the classic "physaliphorous cells"). They show mild to moderate nuclear atypia. There may be considerable variability in the appearance of the tumour from area to area. Mitoses are infrequent [1468]. In the chondroid variants, there are areas that may mimic hyaline or myxoid cartilage [925]. Chordoma associated with a high grade sarcoma is called a "dedifferentiated" chordoma [1398] or sarcomatoid chordoma [1506]. They account for less than 5% of all chordomas.

Immunophenotype

Chordomas are reactive with antibodies against S100 protein, pan-keratin, low molecular cytokeratins and Epithelial Membrane Antigen (EMA).

Genetics

Clonal chromosome aberrations have been detected in 16 cases [1477, 2082]. Nine of them had a hypodiploid stemline, with a chromosome number ranging from 33 to 44. Frequent numerical changes include loss of chromosomes 3, 4, 10, and 13, and the most commonly (half of the cases) deleted segments are 1p31-pter, 3p21-pter, 3q21-qter, 9p24-pter, and 17q11-qter. These results are in agreement with data obtained by comparative genomic hybridisation (CGH) [1880]. By CGH, also gains of chromosome arms 5q and 7q and chromosome 20 are frequently seen. The possibility of a tumour suppressor locus of significance for chordoma development at distal 1p is further strengthened by the finding of loss of heterozygosity at band 1p36 in sporadic as well as familial chordomas [1465].

Prognostic factors

Prognosis has improved considerably with modern surgical techniques of resection especially with tumours of the sacrum [1051,2027] and even of mobile spine [210]. The chondroid variant has been reported to be associated with a better prognosis [925] although this

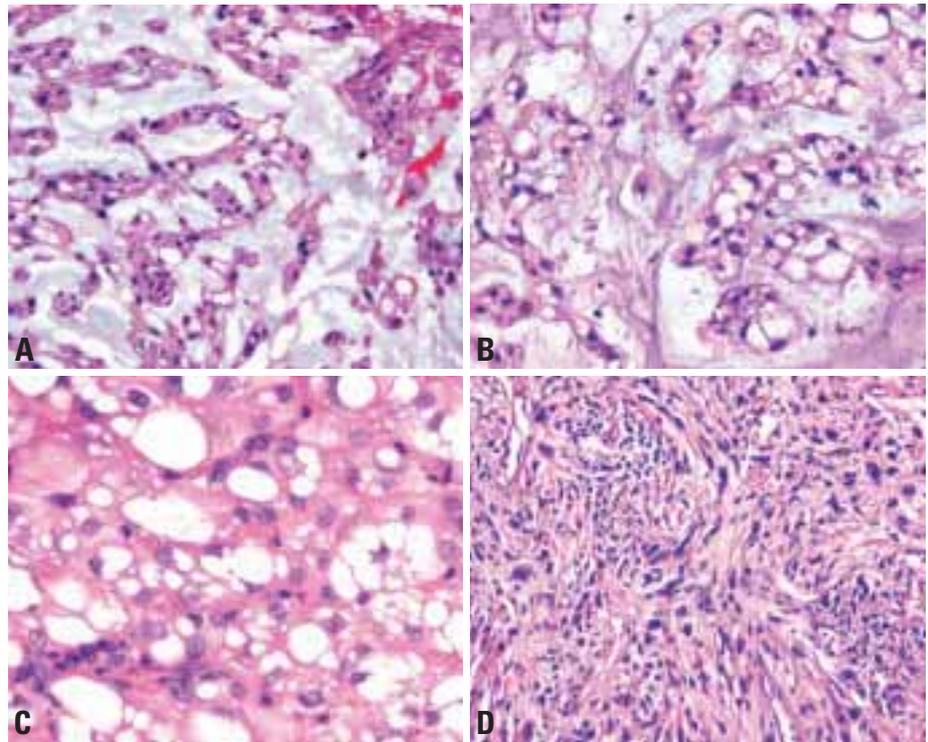


Fig. 17.03 Chordoma. **A,B** Chords of tumour cells in a myxoid background. Note occasional cells displaying a bubbly cytoplasm. **C** Some of the classic physaliphorous cells contain multiple intracytoplasmic bubbles that may cause nuclear indentations similar to those seen in lipoblasts. **D** Sarcomatoid, or "dedifferentiated" chordoma displaying prominent storiform architecture. Note the large, pleomorphic nuclei and the rather solid arrangement of cells without a prominent myxoid background.

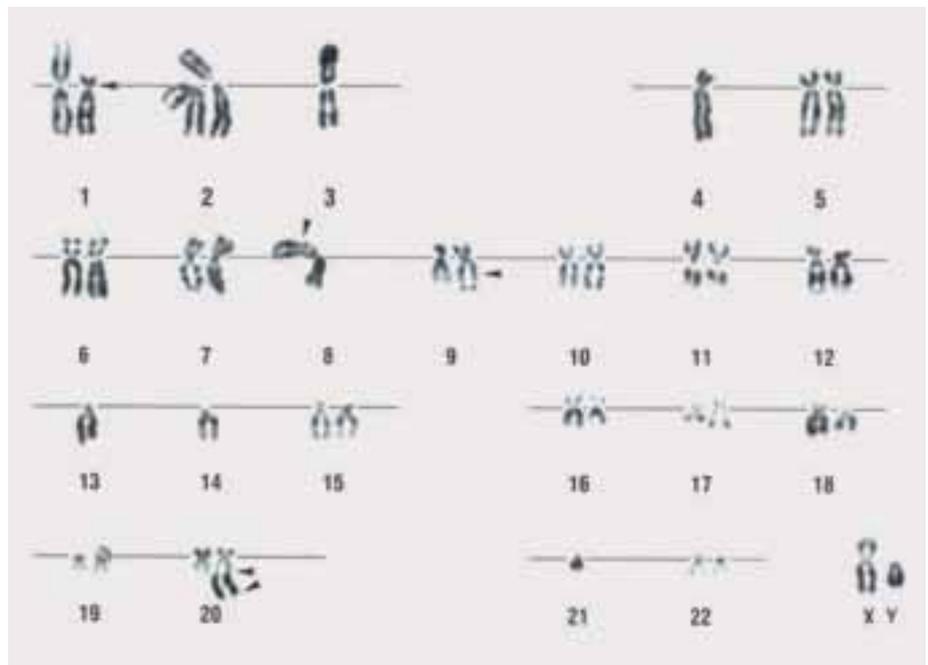


Fig. 17.04 Chordoma with complex karyotype, including the characteristic loss of chromosomes 3, 4 and 13. Arrowheads indicate breakpoints in structural rearrangements.

experience is not universal. Metastases to lung, bone, soft tissue, lymph node and skin occur, and are more frequent in patients with advanced disease.