Chordoma

Definition
Chordoma is a slow growing, locally invasive neoplasm that is derived from remnants of the primitive notochord.

Incidence
4.14% of malignant tumours in Mayo files
8% in NSW
Twice as common in males (2:1)
Affects older patients. It is distinctly uncommon in patients younger than 30.
Sacrococcygeal chordomas tend to occur in an older population than cranial chordomas.

Site
There is a predilection for the ends of the spinal column.
Most lesions are found in the sacrococcygeal region or the base of the skull near the site of the spheno-occipital synchrondrosis. Sacrum 50%, base of skull 40%.
Younger patients tend to get their tumours near the skull, older patients near the sacrum.
The tumour is localised to the midline.
It often involves more than one contiguous vertebral body.
It may also involve the cervical vertebrae.
The body of the vertebra is typically involved.

Symptoms
Pain
Constipation can occur with distal spinal tumours due to pressure on or destruction of nerves. The tumour usually expands anterior to the sacrum.

Haematogenous metastases develop in up to 40%, and occur late in the course of the disease.

Physical findings
Almost all sacrococcygeal chordomas have a presacral extension that is palpable on rectal examination. The mass is firm and fixed to the sacrum.
Chordomas at the base of the brain can cause signs referable to any of the cranial nerves or involvement of the pituitary gland.

Radiology
Plain XR features are expansion, destruction and frequently calcification.

CT and MRI demonstrate massive expansion anteriorly into the pelvis in many cases. Hyperintense on T2 signals.

Chordoma has similar T2 appearance to nucleus pulposus (intervertebral discs)

Pathology
Composed of highly vacuolated, soap bubbly (physaliferous) cells and mucoid intercellular material. Frequent fibrous septae lead to a lobulated appearance.

Treatment
Surgical excision or radiotherapy. All the sacral roots on one side can be removed and the patient will still preserve continence. Need to preserve both S2 nerve roots or one S2 and one S3 nerve root.

Prognosis
Cure is rare, but patients typically survive 10-15 years after diagnosis.