

## **Chordoma**

The chordoma is a very rare malignant tumor of bone that accounts for only four per cent of all malignant bone tumors. It arises from the primitive notochord of the axial skeleton and is most commonly seen in the lower portion of the sacrum, accounting for 50 per cent of the chordomas. Thirty-seven per cent arise in the spheno-occipital area and a small number occur in the cervical and lumbar spine. The more common sacral lesions are seen in an older age group between the ages of 40 and 80 years, compared to the spheno-occipital chordomas that occur in a younger adult age group. The latter carry a much worse prognosis because of the location in the base of the skull. The chordoma is clinically similar to a mucinous type of chondrosarcoma.

In the sacral area, chordomas are usually attached to the anterior portion of the distal three segments of the sacrum and grow in the retroperitoneal space, pressing up against the rectum where eventually they will cause clinical symptoms such as constipation and can be picked up on a rectal examination. Because the tumor is very slow growing, it rarely causes significant pain symptoms. The radiographic findings are not very obvious, even with large tumors that are better evaluated by soft tissue techniques, such as CT scan or, better yet, magnetic resonance imaging. Microscopically, the chordoma has a mucinous appearance similar to a low-grade myxoid chondrosarcoma, but the diagnostic feature is the "signet ring" appearance of the physaliferous cells that have a peripheral nucleus, a large cytoplasmic inclusion of physaliferous mucinous material and can look like a liposarcoma.

Treatment for the chordoma consists primarily of wide surgical resection, which can sometimes be very difficult, especially when lesions extend into the upper sacral segments where the nerve roots are an issue. Significant neurogenic bowel and bladder complications may result. Even with surgical treatment, the local recurrence rate is very high so postoperative radiation therapy is recommended. It has cut the local recurrence rate to about 30 per cent compared to 65 per cent without radiation therapy. Local recurrences can occur up to fifteen years after the original surgery. Pulmonary metastases are extremely rare and systemic chemotherapy is not indicated for this tumor.