CHORDOMA

A chordoma is a rare slow growing malignant neoplasm. It is thought to arise from cellular remnants of the notochord. Pathologically the tumor cells demonstrate similar immunohistochemical staining patterns as notochord cells. They also lie along the neuraxis. The tumors are typically found in the clivus and sacrococcygeal regions, where notochordal cells are preferentially left behind during normal regression of the notochord in the fetus. The most common sites include: sacrum 50%, clivus 35%, vertebral bodies 15%. Chordomas, however, can arise from bone in the skull base and anywhere along the spine.

Annual incidence in the United States is 1 in one million or 300 new patients a year. There is no race predilection. Male to female ration is 2:1. Clival chordomas are considered benign tumors; but because of their critical location, local invasion, recurrence, and occasional metastatic spread, their prognosis is similar to that of malignant tumors. The prognosis of a chordoma in one study was 46% survival at 10-years. Chondroid chordomas are slower growing and have a better survival rate.

There are three histological variants of chordoma: classic chordoma, chondroid and dedifferentiated. The chondroid variant histologically shows features of both chordoma and chondrosarcoma. Differentiating chordomas from chondrosarcomas using both radiologic and histologic criteria can be difficult. Immunohistochemical studies using cytokeratin antibodies and epithelial membrane antigen (negative in chondrosarcomas, positive in chordomas) can make the distinction.

The classic form of chordoma histologically shows a lobulated tumor composed of groups of cells separated by fibrous septa.

Presenting symptoms of this lesion vary, but include headache and cranial nerve deficits. The cranial nerve most commonly involved is the CN VI abducens. Other signs include dysphagia, facial pain, facial paresis, visual loss, hearing loss, and ataxia.

If the tumor is within the sacrum there is an insidious onset of presenting symptoms. A considerable amount of time can elapse between onset of symptoms and diagnosis. The most frequent symptom in these cases is low back pain without any characteristic features. More advanced disease in this region can present with constipation, radicular leg pain and urinary complaints.
Radiographically, this lesion is characterized as a midline destructive bony lesion with predilection for the sphenoid-occipital synchondrosis. (The spheno-occipital synchondrosis is occasionally seen as a horizontal line in the midclivus, midway between the sella and the basion, or tip of the clivus, on sagittal images.)

MR images show a predominantly low signal lesion on non-contrast T1 weighted images often with mass effect on the pons. Diffusion weighted images demonstrate no restricted diffusion. Coronal FLAIR and axial T2 weighted images show these lesions to be predominantly high signal. Post-gadolinium T1 weighted images demonstrate enhancement.

MR and CT have complementary roles in the evaluation of chordoma. CT is needed to assess the degree of bone involvement or destruction and to detect patterns of calcification within the lesion. MRI provides excellent 3-dimensional analysis of the posterior fossa (especially the brainstem), sella turcica, cavernous sinuses, and middle cranial fossa.

Metastatic spread of chordomas is observed in 7-14% of patients. In these cases spread to lymph node, pulmonary, bone, cerebral, or abdominal viscera is seen predominantly with massive tumors. In true malignant forms of chordomas there occasionally are areas of typical chordoma, as well as undifferentiated areas. These undifferentiated areas are most often suggestive of fibrosarcoma. The prognosis in these patients is very poor.

The treatment for chordoma is aggressive surgical resection followed by radiation therapy. The proximity to vital neurologic structures limits the radiation dose that can be used. Therefore highly focused radiation, such as proton therapy and carbon ion therapy are used over conventional x-ray radiation.

Differential diagnosis for skull base lesions also includes metastasis, myeloma, plasmacytoma, fibrous dysplasia and Paget’s disease.


“Primary Malignant Bone Tumors: Tumors of Bones and Joints: Merck Manual Professional”.

Images of Chordoma.