

Chordoma

by Lester D. R. Thompson, MD

Figure 1. The classic appearance of a chordoma includes the presence of epithelioid cells in cords that are suspended in a myxoid-mucous stroma. Vacuolated cells are common.

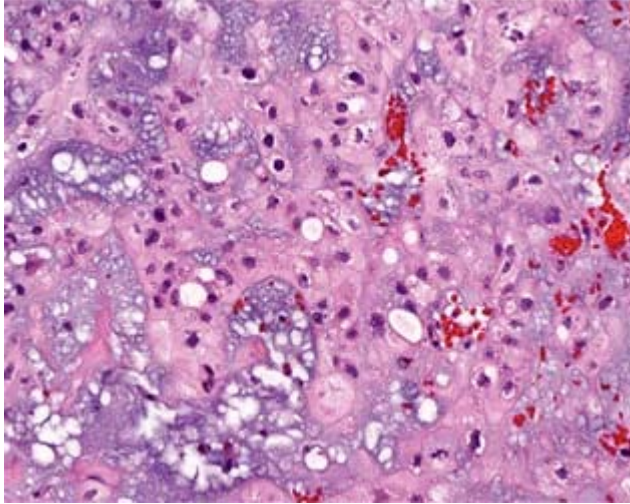
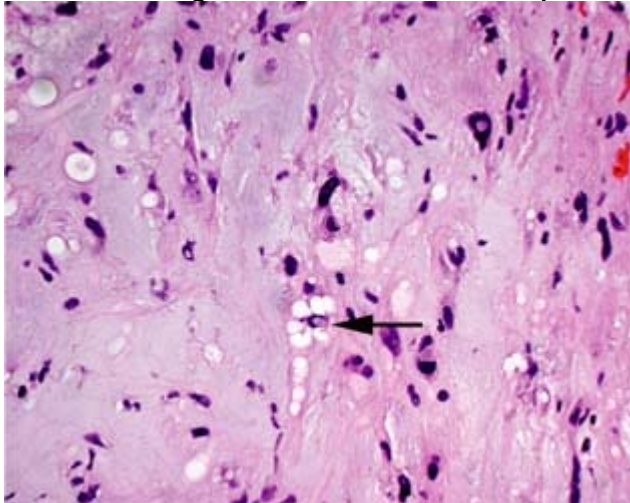


Figure 2. Physaliphorous cells are large cells with multiple vacuoles (arrow) in the cytoplasm. Note the background cells with nuclear pleomorphism.



Chordomas are low- to intermediate-grade malignant tumors that recapitulate the notochord. They are divided into three broad categories: sacrococcygeal (60% of cases), sphenoccipital (25%), and vertebral (15%). About 10% of all tumors are cervical. Vertebral or neck chordomas typically develop in the fifth and sixth decades of life; they have no predilection for either sex. Nerve impingement, progressive pain, and headaches are common. When a chordoma arises within the parapharyngeal space, the mass may be detected clinically. Radiographically, chordomas are usually solitary, lytic lesions; they are associated with matrix calcification in as many as 70% of cases.

Chordomas present as expansive, lobulated lesions with a slippery, mucoid to myxoid cut surface. The tumors usually expand into the adjacent tissues. Their size ranges from 1 to 10 cm.

Three types of chordoma can be identified histologically: classic, chondroid, and dedifferentiated. The classic microscopic appearance of a chordoma is a lobulated growth of cords and islands of polygonal tumor cells suspended in a myxoid-mucous background (figure 1). The epithelioid cells are slightly elongated, with associated large mucus-containing physaliphorous cells (figure 2). The nuclei are round and uniform, although some exhibit considerable pleomorphism (figure 3).

About 5% of chordomas contain islands of hyaline-type chondroid or cartilaginous tissue-hence the term chondroid chordoma. In fewer than 5% of chordomas, there is an association with a high-grade sarcoma (often after radiation therapy), and this is where the term dedifferentiated is applied.

The neoplastic cells are usually immunoreactive with vimentin, keratin, epithelial membrane antigen, and S-100 protein. Many times, fine-needle aspiration is performed for a "neck mass," and the smears can be misinterpreted to represent a mucinous tumor or mucoepidermoid carcinoma (figure 4). While cytogenetic abnormalities can be seen, about 70% of patients have a normal karyotype. However, when there is a genetic abnormality (frequently involving complex cytogenetic alterations of chromosomes 3, 4, 12, 13, 14, and 21), there are higher rates of recurrence and disease progression and a lower rate of survival. Chordomas frequently present within the pharynx, where the histologic distinction from a mucinous carcinoma, salivary gland tumor, or chondrosarcoma is necessary.

Figure 3. This chordoma features a pseudoglandular appearance with hyalinization of the stroma. The epithelioid cells show nuclear pleomorphism and intranuclear cytoplasmic inclusions.

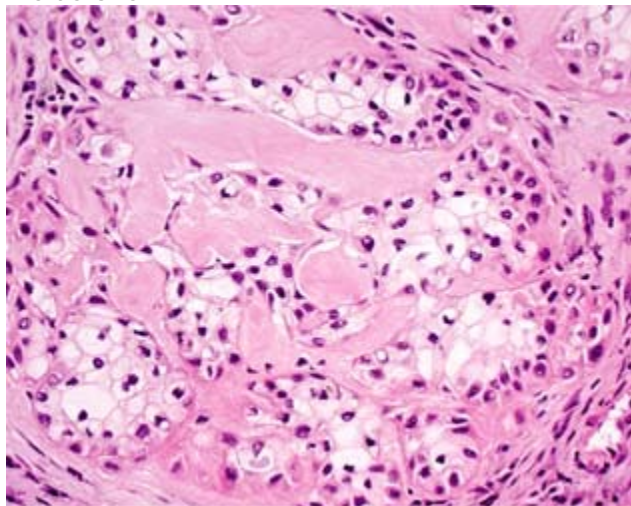
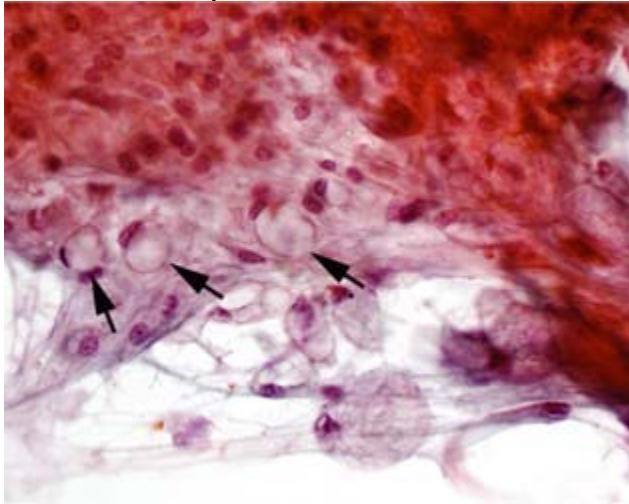


Figure 4. This smear of a fine-needle aspirate shows a number of cells with large vacuoles (arrows) in the cytoplasm. These physaliphorous cells can be misinterpreted as mucocytes within a mucoepidermoid carcinoma.



Chordomas are low- to intermediate-grade tumors that rarely develop distant metastases. However, they are indolent tumors associated with a 5-year survival rate of 65%; nearly 60% of patients ultimately die of this tumor. The prognosis for patients with the chondroid variant may be better. Radical, complete surgical removal of a chordoma is associated with longer survival and delayed recurrence, but this is often difficult to achieve in the anatomic confines of the neck or pharynx. For these tumors and for unresectable tumors, adjuvant radiotherapy is often employed.

From the Department of Pathology, Woodland Hills Medical Center, Southern California Permanente Medical Group, Woodland Hills, Calif.

Suggested reading

1. Mendenhall WM, Mendenhall CM, Lewis SB ,et al. Skull base chordoma. *Head Neck* 2005; 27 (2): 159-65.
2. Mirra JM, Nelson SD, Della Rocca C, Mertens F. Chordoma. In: Fletcher CD, Unni KK, Mertens F, eds. *Pathology and Genetics of Tumours of Soft Tissue and Bone*. World Health Organization Classification of Tumours. Lyon, France: IARC Press; 2002: 316-17.
3. Yamaguchi T, Suzuki S, Ishiiwa H ,et al. Benign notochordal cell tumors: A comparative histological study of benign notochordal cell tumors, classic chordomas, and notochordal vestiges of fetal intervertebral discs. *Am J Surg Pathol* 2004; 28 (6): 756-61.

Ear Nose Throat J. 2011 January; 90(1): 16-18.