Chordoma

Lester D.R. Thompson, MD

Chordomas are low- to intermediate-grade malignant tumors that recapitulate the notochord. They are divided into three broad categories: sacrococcygeal (60% of cases), spheno-occipital (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...
Silence is a Science

It’s not easy treating tinnitus. Prescription medications may help, but come with unwanted side effects. Surgery is costly and applies only to a small number of patients. Arches Tinnitus Formula™ provides the benefits of clinically proven Ginkgo Biloba Extract, designed specifically for tinnitus, with zinc picolinate and odorless garlic. Our unique formulation works through the following mechanisms of action: glutamate antagonism, neuroprotection, antioxidant activity, and increased circulation to the cochlea.

Review the science. Contact us today.

For a free CD of clinical studies, physician’s booklet, and patient brochures, call 800.350.9631 or email md@archesnp.com

Arches Tinnitus Combo Pack

“I recommend the Arches Tinnitus Combination Pack, a three-month supply of Arches Tinnitus, Stress and B-12 Formulas. At a minimum, I tell my patients to take at least four bottles (three months) of Arches Tinnitus Formula™ to determine their total degree of relief.”

Michael Seidman, MD, FACS, inner ear specialist.

www.tinnitusformula.com
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.

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 chordoid 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.

**Suggested reading**

