Unusual presentation of cervical chordoma with long-term survival

Case report

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A 33-year-old woman presented with upper respiratory obstruction due to a posterior pharyngeal mass, which was subtotally resected. The mass was a chordoma arising from the C-2 vertebral body. The patient then remained asymptomatic for 15 years before presenting with signs of cord compression. A chordoma, extending from C-1 to C-4, was again subtotally resected. The unusual biological behavior of this neoplasm is discussed, and a brief review of the pertinent literature is presented.

KEY WORDS • cervical spine chordoma • posterior pharyngeal mass • respiratory obstruction • cervical spine

CHORDOMA of the cervical spine is a relatively rare neoplasm. It usually presents with pain as the initial manifestation and often carries a poor prognosis. We present a case in which the patient's first symptom was that of upper respiratory obstruction. The patient has survived 16 years to date.

Case Report

First Admission. This 33-year-old Caucasian woman presented in 1966 with a 2-day history of non-productive cough and orthopnea. She felt as though there was "something" in her throat. Examination revealed a 5 x 3-cm mass in the posterior pharyngeal wall, which lay to the right of midline and extended from the soft palate to the epiglottis. Although no injection or fluctuance was noted and the patient was afebrile, the initial impression was that this represented an abscess. Needle aspiration was unsuccessful, and the pharyngeal mucosa was incised. A soft gray lobulated neoplasm was encountered, which resembled a mixed salivary gland tumor on frozen section. The tumor was debulked down to the spine, as total excision was thought to be impossible. The permanent pathological specimen was consistent with the diagnosis of chordoma (Fig. 1). No additional treatment was given. The patient recovered uneventfully. Plain x-ray films and tomograms taken postoperatively revealed no intracranial involvement, and the only abnormality in the spine was enlargement of the right C-2 intervertebral foramen.

Second Admission. The patient remained asymptomatic for approximately 16 years. She then began to experience paresthesiae in her hands and difficulty with fine coordinated movements which progressed over 1 year to the point where these problems became disabling. Examination revealed no evidence of a posterior pharyngeal mass. Deep-tendon reflexes were hyperactive, and there was impairment of rapid and skilled hand movements. A metrizamide computerized tomography scan revealed an epidural mass on the right side of the spine which had displaced the cord and extended from C-1 to C-4 (Fig. 2). Bone erosion could be seen, primarily involving C-2 on the right side. An en bloc laminectomy was performed from C-1 through C-4 using a high-speed air drill. Under the operating microscope, all accessible tumor in the right lateral gutter was excised. The gross and subsequent microscopic appearance was consistent with chordoma, unchanged from the original specimen obtained 15 years before. The patient again made an uneventful recovery, with rapid resolution of the signs and symptoms of cord compression. Postopera-
Cervical spine chordoma

Fig. 1. Photomicrograph of the pathological specimen showing the characteristics of a chordoma. H & E, × 300.

Discussion

Chordoma is a relatively uncommon neoplasm. The incidence has been estimated to be 0.51 cases per million. The most common location is the sacrococcygeal region, followed by the clivus. These two locations account for approximately 90% of chordomas, with the remainder occurring in the spine in decreasing frequency from the cervical to lumbar region. There is a male preponderance of up to 2:1 in some series.

Chordomas are thought to arise from neoplastic transformation of ectopic rests of notochord. These rests, termed "ecchordosis physaliphora," can be found in approximately 2% of autopsies. It is of interest that the only "normal" remnant of the fetal notochord is the nucleus pulposus of the intervertebral disc. Although chordomas have been induced in the experimental animal by opening the disc and exposing the nucleus, there has been no documented case in man of chordoma in continuity with or arising from the disc.

That a chordoma can present in the pharynx can be explained on the basis of the embryology of the notochord. This structure consists of a column of cells ventral to the neural tube which appears in the 4th week of embryonic life and regresses by the 7th week. The spine forms as a result of condensation of mesoderm around the notochord. At approximately the 5th week, the rostral end of the notochord is in apposition or contact with the endoderm destined to become the pharyngeal epithelium. Notochordal tissue at times has been seen to extend laterally into the parapharyngeal spaces and even as far anteriorly as the site of the future paranasal sinuses.

The number of reported cases of chordoma presenting with symptoms of upper airway obstruction has been small, and the majority of tumors originated adjacent to the clivus with extension into the sphenoid sinus. An even smaller number seemed to have arisen in the maxillary sinus. The majority of reported cases of chordoma originating in the cervical spine have presented initially with pain, occasionally with dysphagia, and rarely with respiratory difficulty. The usual radiological findings in chordomas of the spine are destructive or lytic lesions with occasional sclerotic changes. Epidural extension is common by the time the tumor is clinically manifest. In contradistinction to metastatic epidural neoplasms, chordomas tend to lie anterolateral to the cord rather than dorsal to it, and have been known to invade the dura, which is rare with metastases.

Surgery, with radical excision if possible, is considered the treatment of choice for chordomas. Unfortunately, total resection is rarely possible due to the location and extent of the disease and its proximity to vital structures. Repeated operations for local recurrences are worthwhile in some patients, and fusion may be required in the case of spinal chordomas for late instability or for vertebral body resection associated with an anterior surgical approach.

Although chordomas are generally believed to be relatively radioresistant, radiation therapy is commonly recommended in the literature as an adjunct to incomplete surgical excision. Suggested dosages are in the range of 6000 to 8000 rads. Objective evidence of tumor regression is rarely seen, and therefore the effect of radiation on these slow-growing
neoplasms is difficult to assess other than by the diminution of pain. Had our patient been irradiated after incomplete resection 16 years ago (which she was not), it would have been tempting to attribute the long interval before recurrence to the radiation.

The prognosis for chordomas in general is poor, due to the difficulty of surgical excision, their relative radioresistance, and their biological behavior which is one of relentless progression. The average survival period in these patients is approximately 5 years. Metastases do not seem to directly affect survival in that most deaths are said to occur as a result of complications related to neurological impairment. Metastases have been noted in lymph nodes, lungs, liver, bone, and virtually every other organ system in the body. A pathologically distinct subgroup of chordomas, known as the chondroid type, occurs in the sphen-occipital area and seems to be associated with a better prognosis.

We have presented a case in which a chordoma of the upper cervical spine presented in the pharynx with respiratory obstruction. The tumor was subtotally resected and no other therapy given. Recurrence occurred 16 years later in the form of epidural spinal cord compression. Subtotal resection was again accomplished via laminectomy. The anterior approach was rejected since that would have been a more formidable procedure necessitating spinal fusion. Radiation therapy has been withheld to date in light of its questionable efficacy in patients with chordomas in general, and in particular in tumors such as the one in this case which seems thus far to have a very slow-growing biological behavior. We also considered that the risks of irradiating a neoplasm in close proximity to the spinal cord in a young patient outweighed the potential benefits, at least at this time.

A recent report in the literature suggests that heavy-particle or proton-beam irradiation may be efficacious in treating chordomas. This modality allows a higher dose to be delivered accurately to the tumor with less radiation to adjacent or surrounding structures than can be achieved with standard photon therapy. Although the results reported by Suit, et al., are encouraging, there were only two cases of cervical chordoma in their series, and the follow-up period was only 22 and 52 months, albeit with no evidence of disease. We anxiously await further reports on proton therapy for patients with chordomas.

References


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