# Chordoma and Chondrosarcoma

Jamie J. Van Gompel, MD[a,*], Jeffrey R. Janus, MD[b]

**KEYWORDS**

• Chondrosarcoma • Chordoma • Outcome • Proton beam • Surgery

**KEY POINTS**

- Chordomas are locally aggressive tumors derived from notochord remnants.
- Chondrosarcomas are generally low-grade, indolent malignancies that cause morbidity through compression of neurovascular structures at the skull base.
- The primary treatment of both chordoma and chondrosarcoma is aggressive primary resection followed by adjuvant radiation therapy.
- Heavy particle therapy, such as proton beam or carbon ion, provides higher treatment doses with less toxicity and currently is most commonly recommended for chordomas and chondrosarcomas after surgical resection.
- Chordomas and chondrosarcomas create multifaceted clinical challenges and should be managed by multidisciplinary skull base teams with expertise in their treatment.

**Videos of chordoma and chondrosarcoma surgeries accompany this article at http://www.oto.theclinics.com/**

## INTRODUCTION

Chordoma and chondrosarcoma have been grouped together historically because of the midline presentation, similar radiography, and confusion in initial pathology. However, these lesions are distinct clinicopathological entities and vary significantly in their clinical outcome. Both lesions are uncommon and represent less than 1% of intracranial lesions.[1,2] Each poses a daunting challenge to effective treatment due to the perilous proximity of critical cranial nerves, the brainstem, and the skull base vasculature. To that end, both tumors have a tendency to recur locally, even after aggressive resection, especially if known residual tumor is left behind. Chordomas may metastasize

Disclosures: The authors report no conflicts of interests.
[a] Departments of Neurosurgery and Otolaryngology, Mayo Clinic, 200 First Street, Southwest, Rochester, MN 55905, USA; [b] Division of Otolaryngology Head and Neck Surgery, Department of Otolaryngology, Mayo Clinic, Rochester, MN 55905, USA
* Corresponding author.
*E-mail address:* vangompel.jamie@mayo.edu

Otolaryngol Clin N Am 48 (2015) 501–514
http://dx.doi.org/10.1016/j.otc.2015.02.009 **oto.theclinics.com**
0030-6665/15/$ – see front matter 

**Abbreviations**

Gy Gray
WHO World Health Organization

with time; however, they rarely present as such. Despite these concerns, the combination of maximal resection with minimal morbidity, followed by aggressive high-dose postoperative radiation therapy, can cure most low-grade chondrosarcomas, and provides good long-term local control and quality of life in patients with chordoma and high-grade chondrosarcoma.

Chordomas are malignant primary bone tumors that are derived from notochord remnants. They were first recognized in 1856 by Lushka and concomitantly by Virchow in 1857 at autopsy.[3,4] Originally thought to be of cartilaginous origin, Muller proposed these tumors were derived from notochord in 1958.[5] The term chordoma was later suggested after these tumors were directly observed in the nucleus pulposis, incriminating their origin.[6] They occur in the axial skeleton, where the notochord resides developmentally. Their occurrence in the axial spine can be thought of in terms of thirds, with a third arising in the skull base, spine, and sacrum 32%, 33%, and 29% of patients, respectively. It should be noted, however, that other studies support a more caudal cranial distribution of cases with less axial spine involvement.[7] Chordoma has an incidence of 8.4 cases per 10 million population in the United States.[7] They frequently recur locally despite aggressive surgical treatment. Although metastases do occur, these are uncommon and are not frequently existent at initial presentation. Ecchordosis physaliphora, which is also a remnant of the embryonic notochord, is a different entity and is found typically intradurally behind the clivus and anterior to the brainstem.[8,9] Pathologically, ecchordosis physaliphora is virtually identical to chordoma.[8,9] In the context of a patient, however, these are commonly smaller; there is complete lack of clival involvement, and they frequently follow a benign clinical course.[8,9]

Chondrosarcomas are rare cartilaginous tumors that may present as low grade (World Health Organization [WHO] 1 or 2), which are far and away more common, or high grade (WHO grade 3), as well as a mesenchymal chordoma subtype. WHO grade 3 and mesenchymal subtype tumors forbear a worse prognosis.[1,2] Only 1% of chondrosarcomas occur at the skull base.[10] The cell of origin is unknown; however, typically these are thought to arise from synchodroses of the skull base, such as the spheno-occipital suture or petroclival suture. They frequently present eccentric, but do not stray far from midline.[1,2] Sixty-six percent of skull base chondrosarcomas arise from the petroclival fissure; an additional 28% percent arise from the clivus proper, and a further 6% arise from the sphenoethmoidal complex.[11] The lattermost of these is a fusion plane, which is off midline, accounting for eccentric presentation just lateral to midline. Chondrosarcomas are typically less aggressive than chordomas, rarer, and very uncommonly metastasize.[1,2]

## PRESENTING CHARACTERISTICS

### *Chordoma*

These are the most common extradural clival tumors. The median age of presentation for skull base chordomas appears to be 60 years of age; however, there is a large range of presenting ages, including pediatric cases.[7,12–15] Chordomas produce morbidity and mortality through local growth. Often upper clival chordomas will present with sixth nerve palsies secondary to the tumor accessing the basilar venous plexus between the