Extraosseous Chordoma of the Nasopharynx


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Intracranial chordomas are relatively rare locally aggressive tumors thought to originate along the course of the embryonic remnant of the notochord.1 The most common location for chordoma is in the sacrum; however, the skull base/clivus region is the second most common location along the course of the notochord.2 Rarely, skull base chordomas may occur in an extraskeletal location.2 When this occurs, chordomas may mimic other lesions of the nasopharynx. These extraskeletal lesions are atypical relative to the usual presentation of clival chordoma due to their nasopharyngeal location. It is important to consider extraskeletal chordoma in the differential diagnosis of tumors in the nasopharynx because it requires a very different treatment plan and carries its own unique prognosis. In this retrospective review, we analyze the clinical and radiologic data of 5 patients with extraskeletal chordoma in an effort to improve the preoperative diagnosis of this rare tumor. In addition, we review the regional notochordal embryology to explain this variant tumor location.

Materials and Methods

Institutional review board approval was obtained. We retrospectively reviewed all imaging studies of patients who had pathologically proved extraskeletal chordomas that were treated or reviewed at our institution during the last 10 years, 1997–2007.

Each patient underwent both CT and MR imaging. CT was performed in 3-mm sections. Reformatted images were also available in 3 axial, sagittal, and coronal postcontrast images with fat saturation. All MR imaging included axial, sagittal, and coronal T1 sequences; axial and coronal T2 with fat saturation; and axial, sagittal, and coronal postcontrast images with fat saturation. All imaging studies were reviewed by 2 senior neuroradiologists.

Clinical information was compiled via a retrospective chart review. Clinical data included demographic information, presenting features, treatment, and follow-up.

Results

Five patients, 8–65 years of age (mean, 42.8 years), had pathologically proved extraskeletal chordomas. The male–female ratio was 1:1.5. Three of 5 patients presented with a sensation of a mass in the nasopharynx. The remaining 2 patients presented with a history of nasal obstruction. All patients underwent surgical resection. One of these patients received adjuvant proton beam therapy. Recurrence occurred intracranially along the clivus sinus tract in 1 of the 5 patients. The conditions of the other 4 are currently stable, without recurrence.

Four of 5 chordomas were midline with superior extension into the sphenoid sinus and anterior extension into the nasopharynx. One of the 5 lesions demonstrated preferential extension laterally into the left parapharyngeal space and left maxillary sinus.

CT of the extraskeletal chordomas showed a lobular soft-tissue mass centered in the nasopharynx with scalloping of the adjacent anterior margin of the clivus in all cases (Fig 1). All (5/5) lesions show lytic changes with a subtle sclerotic margin. Five of 5 lesions were heterogeneous but hypointense relative to adjacent muscle. Two of the 5 lesions demonstrated dystrophic calcifications within the tumor matrix (Fig 2). In 3 of 5 cases, a well-defined tract was seen extending into the
midline clivus, thought to represent the medial basal canal (Fig 2). A narrow zone of transition with a sclerotic margin was seen around the tract in all cases.

On MR imaging, the lesions were predominantly T1 hypointense relative to muscle. Scattered throughout the tumors were areas of mildly hyperintense T1 signal intensity (Fig 1). Following contrast administration, there was heterogeneous enhancement in all 5 cases. Four of 5 lesions demonstrated heterogeneous avid enhancement. Enhancement was seen predominantly in the solid portions of the tumor and along internal septations. One of 5 lesions showed peripheral enhancement.

On T2-weighted sequences, 5 of 5 cases demonstrated heterogeneous hyperintense signal intensity. Scattered areas of very high T2 signal intensity were noted, but all lesions were predominantly mildly hyperintense relative to muscle and less hyperintense compared with CSF. The internal septations on T2-weighted imaging were all uniformly hypointense (Fig 3). Three of the 5 lesions demonstrated a sinus tract extending from the predominant mass lesion posteriorly into the midline clivus (Fig 4). The sinus tract, thought to represent the medial basal canal, was T2 hyperintense in all cases. This sinus tract did not demonstrate enhancement on postcontrast sequences.

Discussion
Chordomas account for 1% of all intracranial tumors.1-4 Intracranial chordomas generally occur in the vicinity of the clivus, often in the region of the sphenoid-occipital synchondrosis. There have been case reports of extraosseous chordomas in the literature, but none have been reported in the nasopharynx, as defined in our series.2,3 It is important to consider
canal. The cranial extent of this tract is situated between the
medial basal canal is that of the craniopharyngeal
tract as described by Virchow. The notochord is a primitive cell
ponent along the course of the medial basal canal. Therefore this
anatomic distinction has significant differential diagnosis implications.

**Imaging Characteristics**

Both CT and MR imaging are used in the evaluation of chordoma. CT is ideal for evaluating the bony involvement, whereas MR imaging is useful in evaluating the surrounding soft tissues and extension into adjacent structures. MR imaging is considered the gold standard in pretreatment and post-treatment evaluation of chordomas.

On CT, the typical appearance for an extraosseous chordoma is a lobular hypoattenuated soft-tissue mass with areas of dystrophic calcification and lytic changes of affected osseous structures. Scattered areas of hyperattenuation are consistent with descriptions in the literature of blood products and intratumoral hemorrhage in typical chordomas.

The interesting feature in our series is a sinus tract along the expected course of the notochordal remnant. The associated midline bony tract is an important clue to the notochordal origin of this extraosseous chordoma because other nasopharyngeal malignancies may destroy clival bone but do not demonstrate this midline tract. One of 5 cases we present is a Tornwaldt cyst mimic. In this specific example, the subtle sinus tract into the clivus helped distinguish this lesion from a Tornwaldt cyst.

With MR imaging, the features of the extraosseous chordomas are similar to typical skull base chordomas. The lesions were predominantly hyperintense compared with muscle on T2 sequences. The lesions were heterogeneous and demonstrated intratumoral septations. These findings suggest that though extraosseous chordomas can have atypical locations, the tumors will continue to demonstrate typical MR imaging characteristics.

**Treatment**

Improving outcomes are being obtained with surgical resection of intracranial chordomas. The main complication to treatment of chordomas remains local recurrence. In the case of an extraosseous chordoma, the removal of chordoma along the clival bony sinus tract along with the extraosseous tumor may be important to limiting the early recurrence of this lesion. MR imaging remains the standard for evaluating postsurgical patients and surveillance for recurrence. The most commonly cited finding of recurrence is hyperintensity on T2-weighted sequences rather than morphologic changes. Contrast-enhanced imaging further aids in detecting areas of recurrence along the surgical margins. Distant metastasis is very rare in chordomas.

**Differential Diagnosis**

Extraosseous chordoma is primarily a lesion of the nasopharyngeal soft tissues. For the purposes of our discussion, due to the location, the differential diagnosis primarily includes nasopharyngeal tumors. The differential diagnosis list is signifi-

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Fig 3. A 53-year-old man presented with dryness of mouth and difficulty breathing. Axial T2-weighted MR image with fat saturation shows a locally invasive heterogeneously hyperintense mass with internal septations (black arrow) centered in the nasopharynx, with invasion of the right masticator space, nasal cavity, and maxillary sinus. Note involvement of the anterior clivus (curved arrow), which may help preoperatively to diagnose this lesion as an extraosseous nasopharyngeal chordoma.
cantly different from the standard differential considerations for a classic chordoma. The differential for this variant lesion includes nasopharyngeal carcinoma and non-Hodgkin lymphoma. Nasopharyngeal carcinoma differs from chordoma in that there is a demographic predilection among Chinese and North Africans. Non-Hodgkin lymphoma of the nasopharynx is a relatively uncommon tumor. The age distribution for non-Hodgkin lymphoma of the head and neck is in the sixth decade. Combined chemotherapy and radiation therapy remain the primary treatment modalities for both nasopharyngeal carcinoma and lymphoma. Interestingly, as seen in our small series, a small extraosseous chordoma may mimic a Tornwaldt cyst, a benign notochordal remnant entity.

Conclusions

Extraosseous chordoma of the nasopharynx is a rare but important tumor to be considered in the differential diagnosis of nasopharyngeal masses. When a midline nasopharyngeal mass is found with an associated clival sinus tract, extraosseous chordoma moves to the top of the differential diagnosis list. Complete removal of the soft-tissue tumor and the clival sinus tract is the treatment of choice in such cases.

References