Clinical Article

# Oronasopharyngeal Chordomas

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**Running Title:** Oronasopharyngeal Chordomas

**Abstract**

Background: Chordomas are rare tumors derived from notochordal remnants. The authors report on a series of three cases of primary familial oronasopharyngeal chordomas treated at our institution.

Methods: A retrospective chart review was completed of the three cases of primary familial oronasopharyngeal chordoma treated at the University of Utah.

Findings: All three patients (100%) were neurologically intact and presented with nasal obstruction. The patients ranged in age from 5 to 65 years and were first-degree relatives. None of the patients had bony erosion of the skull base on imaging, and all of the patients' tumors connected with the skull base via a tract. All three patients were treated with a wide excision combined with drilling of the involved skull base. They all tolerated the procedure without any complications and remain tumor free with a follow up of 12 months to 4.5 years.

Conclusion: Primary oronasopharyngeal chordomas are rare tumors that may present without bony erosion of the skull base. A wide excision with drilling of the involved bony structures may offer an oncologic cure.



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## Introduction

Chordomas are rare tumors of notochordal origin that usually affect the midline bony craniospinal axis. The sacrococcygeal region is the most commonly affected (50%), followed by the craniocervical (35%) and nonsacral spinal (15%) regions [39,46]. In the craniocervical region, chordomas often occur in the clival and basisphenoid regions, and most often present with headaches and neurological dysfunction secondary to neural compression [27,45]. Their complete surgical excision is infrequently accomplished [9], resulting in progression-free survival rates of 50% at 5 years [8].

"Ectopic" chordomas are rare variants that arise in atypical places, mostly in the head and neck, and are thought to arise from ectopic rests of notochordal elements at the cranial end of the neuraxis [17,46]. Reported sites of ectopic chordoma occurrence include the nasopharynx, nasal septum, paranasal sinuses, and orbit [3,5,6,13,14,17,21,30-33,37,40,43]. Presenting symptoms of chordomas in ectopic locations are related to the tumor's location and can include nasal blockage, proptosis, epistaxis, or headache rather than neurologic deficit. A few reported ectopic chordomas have occurred in patients with a familial history of chordoma, which is rare [13,21,43]. Since the advent of computed tomography (CT) and magnetic resonance (MR) imaging, cases of ectopic chordomas have been reported that originate from the clivus, sphenoid, or ethmoid bone. Their clinical course also seems to be more favorable.

We present three cases of familial primary oronasopharyngeal chordomas. These cases are unique from previously described ectopic chordomas because of their complete lack


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of bony erosion of the clivus or sphenoid. We discuss the clinical and radiologic characteristics of these tumors, as well as their implications for treatment. We also include a discussion of the theory of embryological development of chordoma with minimal bone involvement, with the goal of education of the clinical neurosurgeon to the consideration of chordoma presenting in such a manner.

**Methods**

A retrospective computer database search was completed to identify all patients with the pathologic diagnosis of oronasopharyngeal chordoma who were treated at the University of Utah between 1995 and 2007.

Each patient's hospital and clinic charts were reviewed. Patient demographic information, clinical presentation, imaging findings, pathology, treatment modalities used, and clinical outcomes were analyzed.

**Results**

A total of three cases of ectopic chordomas with involvement of the oronasopharynx were identified. The patients ranged in age from 5 to 65 years (Table 1). In all cases, the patients had nasopharyngeal involvement from ventral extension of the clivus or from the sphenoid rostrum. Bony erosion was absent in all cases (100%) (Figs. 1 and 2). Different degrees of scalloping of the outer table of the clivus were observed (Figs. 1 and 2). A sinus tract was present in those cases, connecting the mass to its bony attachment (Fig. 1).


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The cases were all first-degree relatives (two female siblings and their mother). All patients presented clinically because of long-standing nasal obstruction. A biopsy of the respective lesions was performed, given the atypical presentation, confirming a diagnosis of chordoma. For each patient, a second-stage operation was then performed to resect the lesion and drill off any areas of bony involvement. A transoral transpalatal approach was most commonly used (Table 1). When necessary, as in Case 1, an endonasal approach was used as a supplement to resect the sphenoid rostrum and portion of the perpendicular plate of the ethmoid bone with tumor involvement.

In each case, the mass was sharply dissected from surrounding nasopharyngeal mucosa and resected en bloc. The sinus tract was resected and followed to the affected bony areas, which were drilled using a diamond burr. Drilling of the outer table and diploe was accomplished. The inner table was respected to minimize the risk of a cerebrospinal fluid (CSF) leak. The patients underwent a gross total resection and were not offered adjuvant therapy.

All of the patients had favorable outcomes. No cranial nerve palsies or neurologic deficits were encountered. No infections or CSF leaks were reported.. In patient follow-ups of 12 months to 4.5 years, no recurrences have been noted.

**Discussion**

Chordomas affecting the oronasopharynx have been reported with sporadic frequency in the literature, with the first report originating in 1909 [23]. For cases reported prior to the


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advent of CT and MR imaging, it is difficult to discern whether the chordomas presenting with nasopharyngeal symptoms were truly primary nasopharyngeal tumors or simply clival or high cervical lesions that grew ventrally to secondarily involve these structures [2,33,40]. Certainly, in some series in which patients presented with a pharyngeal mass on clinical examination in association with a neurologic deficits, the latter would be the more likely scenario [2,6]. More recently, advanced imaging techniques have allowed the clear anatomic delineation of these rare lesions, which appear to have a unique clinical presentation and more benign clinical course [32,33].

*Clinical Presentation of Oronasopharyngeal Chordomas*

Chordomas present with symptoms secondary to local mass effect. Typical cranial chordomas most commonly present with headache, cranial nerve deficits, and visual disturbances, and more rarely with signs of brainstem compression [27,45]. Patients with chordomas of the oronasopharynx also report symptoms secondary to local mass effect, namely symptoms of dysphagia, dysphonia, dyspnea, nasal obstruction, a sensation of mass, or hearing difficulties [32]. Because the tumors are submucosal and slow growing, they rarely present with epistaxis or mucosal ulceration, unlike other lesions that occur with greater frequency in the area [32]. In our series, all patients presented with nasal obstruction that failed to improve with conservative therapy. No neurologic deficits were present in the patients in our series, as the inner table of the affected bones of the skull base was noted to be intact at the time of surgery.

*Embryology*


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Chordomas are rare tumors believed to arise from notochordal remnants. Their typical location, along the midline of the craniospinal access, corresponds with the anatomic location of the notochordal vestiges [18,36,46]. The notochord's cephalad trajectory is known to pass through the body of the odontoid process, after which it lies on the dorsal surface of the occipital plate. The notochord then pierces the occipital plate ventrally in the vicinity of the medial basal canal and has a cephalad trajectory underlying the endoderm that will form the pharyngeal mucosa (Figs. 3 and 4). Finally, it pierces the sphenoid plate toward its cephalad end to terminate in the region of the pituitary fossa [35,36].

Ectopic chordomas arise most commonly in the head and neck region and have been reported to occur in the paranasal sinuses, orbit, septum, and oronasopharynx, where they are thought to arise from ectopic rests of notochordal tissue [3,5,6,13,14,17,21,31-33,37,40,43,46]. Such rests have been reported to occur in the nasopharyngeal submucosal tissue [19,40] and are believed to arise during embryogenesis either through the normal branching and ramification of the notochord anterior to the clivus [31] or through a process by which small derivatives of disintegrated notochord are moved by the developing craniofacial structures [5,46]. Notochordal remnants have been reported with increasing frequency in the form of ecchordosis physaliphora, benign notochordal cell tumors (BNCT), and giant notochordal rests.

*Differential Diagnosis and Radiographic Features*


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Ecchordosis physaliphora has been reported to occur along the posterior wall of the clivus intradurally and is generally thought to represent an incidental finding. Ecchordoses are hypointense on T1-weighted MR imaging and hyperintense on T2-weighted imaging but their lack of enhancement distinguishes them from a chordoma [26]. Symptomatic cases have been reported rarely, secondary to local mass effect [24,34], subarachnoid hemorrhage [16,42], or the development of a CSF leak [25]. They are generally not thought to be precursors to the development of a chordoma but their ability to enlarge has been questioned.

The existence of benign notochordal cell tumors (BNCTs) has also been reported with increasing frequency [44,48-51]. BNCTs appear as large adipocyte-like cells that exhibit only minimal nuclear atypia and lack lobulation and a myxoid stroma [12]. They are visible radiologically and are thought to represent embryologic rests of the notochord [47]. Their incidence may be as high as 20%, and they are most commonly found in the clivus and sacrococcygeal region [49]. It has been suggested, based on the coexistence of a chordoma and BNCT in some cases in the sacrum, that they may be precursors to the development of a chordoma [50,51]. They have been reported involving vertebrae caudal to C3 [47]. A more extreme variant of BNCTs has been described as giant notochordal rests, which are visible radiologically and have been identified in some patients who presented with pain along their spine [7,11,22]. They have also been referred to as giant notochordal hamartomas. They are distinguished from chordomas in pathological analysis by a lack of extracellular mucoid matrix as well as an absence of mitoses and a preservation of the bony trabecular framework. The most cephalad case reported involves


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the cervical spine at C5 [29]. Some cases have been resected surgically after biopsy that was initially compatible with a diagnosis of chordoma [7,11,22], while other cases have been followed radiologically without any changes for 10 years [29].

A continuum seems to exist between the occasional presence of notochordal remnants along its embryologic pathways and the development of certain chordomas, explaining the existence of oronasopharyngeal chordomas. All described ectopic chordomas to date have been associated with extensive bony destruction when documented with CT imaging (Table 2). Our cases may represent early stages of the development of a more benign form of chordoma along its embryologic pathway without bony destruction as has been observed in the spine by Yamaguchi et al. [50]

Preoperative diagnosis of oronasopharyngeal chordomas is difficult. Clues to accurate imaging diagnosis are given by stereotypical signal characteristics, specifically hypointensity on T1 and hyperintensity on T2 sequences with moderate enhancement [38,41,45]. CT imaging is useful to assess for lytic bony destruction. The combination of these imaging characteristics lends strongly to chordoma in the differential diagnosis when the lesion is present in a typical location (e.g., clivus). When present ectopically, the lesion must be differentiated from more common nasopharyngeal tumors, including non-Hodgkin lymphoma and nasopharyngeal carcinoma. A clue to the diagnosis of chordoma may be obtained by careful examination for a T2 sinus tract leading to the clivus, a feature noted in our patients. Scalloping of the ventral surface of the clivus was also present in the cases devoid of bony erosion. The ventral cortical surface of the clivus


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was not disrupted except in the small area where the sinus tract originated. The identification of the sinus tract also has clinical implication, and its resection is imperative to achieve complete resection and disease control. Previous published cases of nasopharyngeal chordomas in the CT era had extensive bony erosion of the clivus, sphenoid rostrum, or perpendicular plate of the ethmoid (Table 2).

Familial chordomas have been reported in the literature but are rare. A total of 13 cases have been reported, 5 of which presented as nasopharyngeal lesions [10,13,15,21,28,43]. The genetic link in these cases remains to be understood. Abnormalities at different loci have been described in familial chordomas [4,20,28,52].

*Treatment*

Treatment of cranial chordomas is difficult because of their anatomic location. Gross total resection offers the best chance of cure; however, this is rarely accomplished with cranial lesions because of complex regional anatomy and local tumor invasion [8]. Ectopic chordomas of the paranasal sinuses and oronasopharynx appear to warrant special consideration. The location of these tumors makes complete resection a feasible goal. In our series, transoral and endonasal approaches were used; these approaches provided excellent visualization of the tumor and allowed for complete resection in every case. The fact that the bulk of the tumor was present in the oro- or nasopharynx with minimal bony involvement greatly facilitated an *en bloc* removal of the tumors. Emphasis was also placed on resecting the sinus tract into the clivus with drilling of the clivus performed to its inner table over the underlying area of retropharyngeal invasion (Fig. 2).


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We did not choose to radiate any patients in this series because complete resection was obtained, with a margin of normal nasopharyngeal mucosa and bone. Although seeding of the surgical tract has been reported and the risk of such has led to the recommendation of irradiating the surgical route [1], the transoral and transpalatal approach offers direct access, which minimizes this risk. Although our follow-up period for these patients is not long, we have not seen recurrences in cases in which gross total resection was accomplished, similar to previously published series (Table 2).

**Conclusions**

Primary nasopharyngeal chordomas are unusual lesions whose origin can be understood in the context of the embryological development of notochord and craniofacial structures. The location of these lesions endows the surgeon with a better opportunity for complete resection, thus giving the patient a better chance of cure. An unusual preponderance for familial chordomas to present as nasopharyngeal masses hints at a possible genetic uniqueness of these lesions. Ectopic chordomas reported to this date have an extensive degree of bony erosion on imaging. Our series is unique because of the minimal bony involvement in these cases that was limited to the presence of a sinus tract to the clivus and some scalloping of the ventral cortical surface of the clivus. Radiological recognition of these signs is important to alert the clinician to the possibility of these masses being a chordoma. Treatment of these lesions can be limited to surgical resection, but it is recommended to resect the sinus tract as well as any bony abnormalities to prevent recurrences.


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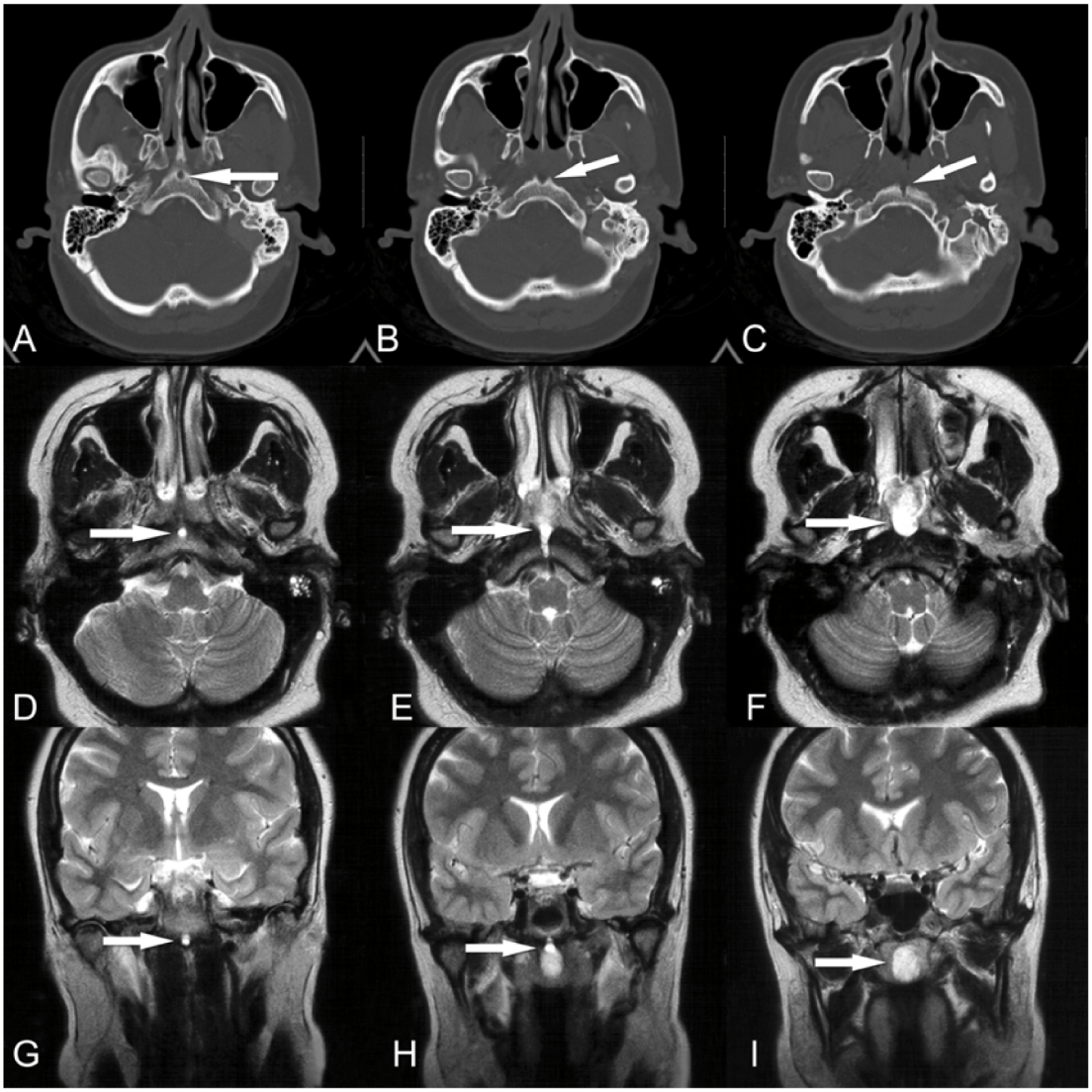


Figure 1: Case 1. Consecutive caudal to cranial sequences on noncontrast CT (a, b, c), axial (d, e, f), and coronal (g, h, i) T2-weighted MR imaging demonstrating the presence of a well-corticated canal within the clivus (arrows in a, b, c, d, g) in continuity with the chordoma (arrows in e, f, h, i).



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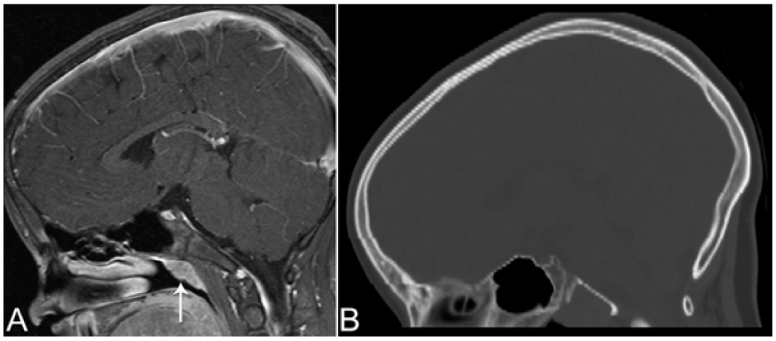


Figure 2: Case 2. (Left) Sagittal T1-weighted MR image with gadolinium enhancement demonstrating chordoma involving the nasopharynx (arrow). (Right) Postoperative sagittal noncontrast CT image showing drilling of the outer cortex and diploe of the clivus.



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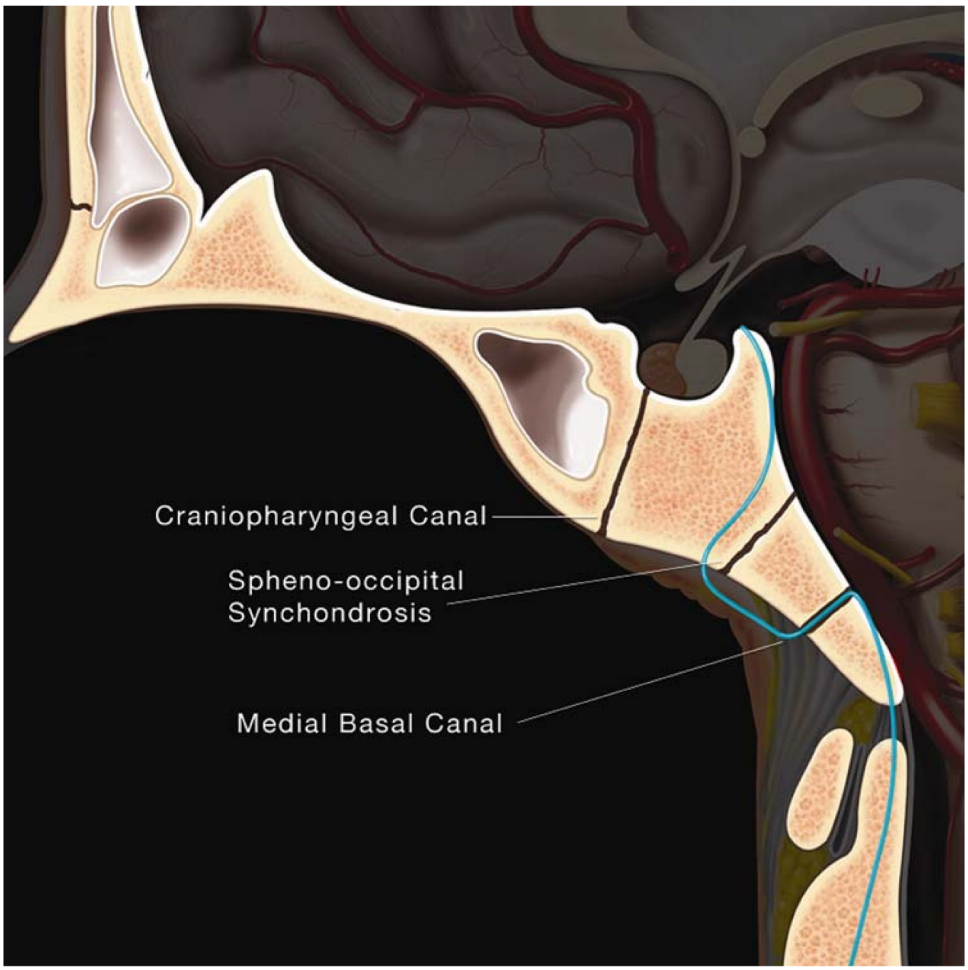


Figure 3: Sagittal illustration of the previous cephalad trajectory of the embryonic notochord in the adult (green line). (Used with permission from Amirsys®, Salt Lake City, UT.)



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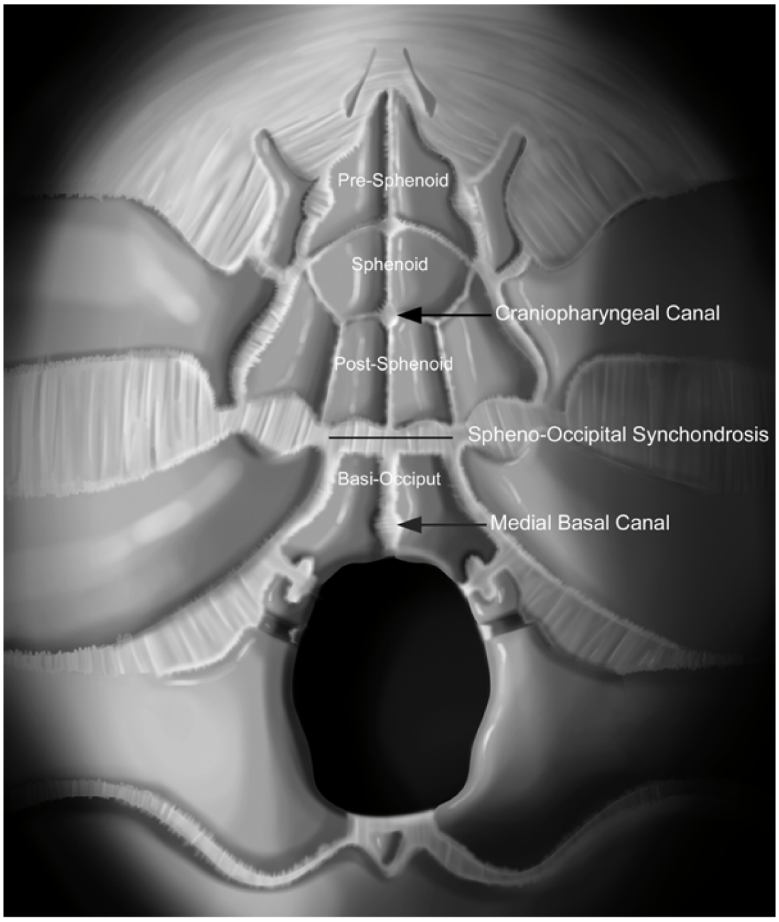


Figure 4: Illustration of the inferior surface of the skull base during development with its ossification centers depicting the relationship of the medial basal canal. (Used with permission from Amirsys®, Salt Lake City, UT.)



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Table 1. Patient characteristics of our series of ectopic chordomas

| Patient | Age (yr) | Sex | Presenting symptom | Chordoma location | Bony involvement | Tract | Treatment | Recurrence | Follow-up |
|---|---|---|---|---|---|---|---|---|---|
| 1 | 32 | F | Nasal obstruction | nasopharynx | scalloping of clivus | yes | transnasal and transoral transpalatal resection | no | 4.5 yr |
| 2 | 8 | F | Nasal obstruction | nasopharynx | scalloping of clivus | yes | transoral transpalatal resection | no | 14 mo |
| 3 | 5 | F | Nasal obstruction | nasopharynx | scalloping of clivus | yes | transoral transpalatal resection | no | 12 mo |

M, male; F, female; NA, not available.

Table 2. Ectopic nasopharyngeal chordomas reported in the literature during the CT and MRI era

| Study | N | Symptoms | CT era | Location | Treatment | Bony erosion | Pathology |
|---|---|---|---|---|---|---|---|
| Shugar et al. 1980 | 1 | nasal stuffiness | yes | maxillary sinus | resection | yes | chordoma |
| Loughran et al. 2000 | 1 | eye pain | yes | ethmoid sinus | resection | yes | chordoma |
| Scartozzi et al. 2003 | 1 | nasal obstruction | yes | nasopharynx, maxillary sinus | resection | yes | chondroid chordoma |
| Lynn-Macrae et al. 2005 | 1 | nasal obstruction | yes | lateral nasal wall | resection | yes | chordoma |
| Kitai et al. 2005 | 1 | epistaxis | yes | sphenoid sinus | resection | yes | chordoma |
| Gupta et al. 2007 | 1 | nasal obstruction | yes | nasopharynx | resection | yes | chordoma |


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ERRATUM

Two of the patients in this paper were included in a recent publication in the radiological literature on this subject: Nguyen et al. (2009) Extraosseous chordoma of the nasopharynx. AJNR Am J Neuroradiol. doi:10.3174/ajnr.A1446.



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