Primary chordoma of the lateral nasal wall: Case report and review

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Abstract
Chordomas are malignant, nonepithelial neoplasms derived from notochordal tissue. A primary chordoma of the nasal cavities and paranasal sinuses is extremely rare compared with clival chordomas, which often present as nasal masses after spreading anteriorly. Only a few cases of primary chordoma of the nasal cavities and paranasal sinuses have been reported in the literature. We report a case of a primary chordoma of the lateral nasal wall. Pathologic diagnosis was obtained using an intranasal endoscopic excision. Asin our patient, a primary chordoma of the nasal cavity or paranasal sinuses may present with symptoms related to mechanical obstruction secondary to the tumor mass. We summarize our case in the context of the other reported cases, and we discuss treatment options, natural history, and prognosis.

Case report
A 35-year-old woman presented with a long history of nasal obstruction. On anterior rhinoscopy, she was found to have a right-sided, large, intranasal lesion and, as an incidental finding, a right, polypoid, right-vocal-fold lesion. Computed tomography (CT) of the paranasal sinuses revealed a large soft-tissue mass filling the entire right nasal cavity. On CT (coronal bone window, figure 1), the mass appeared to be contiguous with the middle and inferior turbinates. Large areas of calcification were noted in the center of the lesion. No bone erosion was found, although there was significant remodeling of the anterior ethmoid bulla, inferior turbinate, septum, and lateral nasal wall. The left nasal cavity and nasopharynx were normal.

An endoscopic sinus surgery was performed. After topical oxymetazoline and 4% cocaine were introduced into the nasal cavity, the lesion was seen to have a mottled mucosal appearance. It completely filled the right nasal cavity. The left nasal cavity showed a septal deviation secondary to the mass effect of the lesion. The mass was infiltrated with 1% lidocaine with 1:100,000 epinephrine for further hemostatic control, and multiple samples of the lesion were removed and sent for frozen-tissue examination. After further debridement, the lesion was found to contain areas of a bony, ossified texture. The lesion clearly originated from the right medial turbinate. The remnant mass was widely excised en bloc along with the superior portion of the right middle turbinate. The right nasal cavity was remarkable only for extensive remodeling of the anterior ethmoid bulla, inferior turbinate, septum, and lateral nasal wall. The left nasal cavity and nasopharynx were normal.

The frozen-section diagnosis read, “Lesion suspicious for pleomorphic adenoma with no suspicion of malignancy.” The permanent section revealed a tumor arranged in lobules in myxoid tissue, separated by fibrous bands. Physaliphorous cells were noted (figure 2), which stained strongly for S-100, epithelial membrane antigen (EMA) and, focally, AE1-3 cytokeratin. The same cells were negative for glial fibrillary acidic protein, high-molecular-weight keratin, and muscle-specific actin and desmin. The final diagnosis was chordoma. The patient was discharged on the same day as the surgery and was disease-free 6 months later.

Discussion
Chordomas are nonepithelial tumors constituting less than 1% of all tumors of the central nervous system. They have been found in all age groups and appear to have an equal sex distribution. Chordomas are classified according to their location in the neuroaxis, with most chordomas classified as sphenoid-occipital, vertebral, or sacrococcyegeal. An estimated 50% of chordomas are sacrococcygeal, and 35% are sphenoid-occipital.

Chordomas were independently described by Virchow and Luschka in the 1850s. Virchow believed that they originated from the sphenoid-occipital synchondrosis. Muller in 1958 linked the tumor to notochordal tissue and renamed it “ecchordosis physaliphora” because of the presence of the physaliphorous cells. Notochordal
tissue vanishes as part of normal human development, the only remnant being the nucleus pulposis in the adult. Areas of ectopic notochord tissue coincide with all those areas where chordomas are found, including the clivus, the bodies of the vertebrae, and the nasopharynx, nasal cavity, and paranasal sinuses.

A few chordomas have derived from the nasal cavity and paranasal sinuses. These cases constitute a very rare subgroup. Perzin and Pushparaj note only 11 cases of chordoma of the nasal cavity, paranasal sinuses, and nasopharynx in 480,000 surgical pathology cases reviewed in their institution during 70 years. Most patients presenting with nasopharyngeal symptoms due to chordoma will have a sphenoid-occipital tumor arising from the clivus that has extended downward into the nasal cavity rather than a true ectopic chordoma deriving from the paranasal sinuses, the nasopharynx, or the nasal cavity.

Pathologically, chordoma is a gelatinous, multilobulated mass with areas of dystrophic calcification. Microscopic examination reveals a highly varied internal structure but includes the pathognomonic physaliphorous cells: eosinophilic cells with bubbly cytoplasm indenting the nucleus. Chordomas will stain positive with monoclonal staining for S-100 and EMA, but negative for glial fibrillary acidic protein, high-molecular-weight keratin, and muscle-specific actin and desmin. Chordomas are locally invasive but are rarely metastatic.

The presenting signs and symptoms of chordoma are related to the site of origin and the area of extension. The presenting complaint of the patient in this report was longstanding nasal obstruction caused by the mass effect of the tumor. In a review of 20 cases of chordomas involving the nasal cavity, paranasal sinuses, or nasopharynx, Perzin and Pushparaj found that the most common presenting symptom was diplopia, followed by localized head pain and nasal obstruction.

Radiographically, chordomas are associated with bony destruction and an extraosseous soft-tissue mass with intralesional destruction. Treatment of the lesion depends on the location and pattern of extension. An anterior lesion, as in the case presented here, appears to be amenable to wide local excision, with good results. Clival or sphenoid lesions are usually more advanced on presentation and are typically treated with debulking and postoperative radiation.

Perzin and Pushparaj reviewed a series of 20 chordomas as part of a larger series of nonepithelial tumors of the nasal cavity, paranasal sinuses, and nasopharynx. However, given the limited clinical data on each patient, it is impossible to state that any of the 20 had a primary chordoma of the nasal cavity or paranasal sinuses.

Shugar et al presented a case of a primary chordoma of the maxillary sinus. Their patient, a 30-year-old black woman, presented with a chief complaint of nasal stuffiness for several months’ duration and was treated with a radical maxillectomy. The patient was tumor-free 19 months postoperatively.

Berdal and Myhre reviewed six cranial chordomas involving the paranasal sinuses, nasal cavity, and palate.
these six cases, only three appeared to originate from the paranasal sinuses or nasal vault. In their case 1, a 46-year-old woman presented with left nasal stuffiness obstruction and was treated with ethmoid resection. The patient was reported to be symptom-free 19 years later, although a radiograph showed a swelling of the mucous membrane of the right frontal sinus. In their case 5, an 11-year-old schoolgirl presented with left-sided nasal obstruction of 6 months’ duration and was treated with an external ethmoidectomy. No follow-up information was available. Case 6 was that of a 54-year-old housewife who presented with a swelling over the right maxillary alveolar ridge. She was diagnosed as having a maxillary/hard palate chordoma and treated with radiation therapy and extensive wide local excision. Ten-year follow-up revealed local recurrence.

Loughran et al presented a case of a primary chordoma of the ethmoid sinus. The patient, a 42-year-old woman, presented with a chief complaint of eye discomfort and swelling of the medial canthus of the left eye. She was treated with wide local excision through a lateral rhinotomy approach. Twelve-month follow-up showed the patient to be tumor-free. The table summarizes the previously published reports of primary chordomas of the nasal cavity and paranasal sinuses.

In conclusion, a primary chordoma of the paranasal sinuses or nasal cavity is extremely rare, with only a handful of cases having been reported. Diagnosis is typically made after endoscopic biopsy. Unlike most cases of chordoma, chordomas of the nasal cavity and paranasal sinuses are amenable to wide local excision, with good results.

### Table. Primary chordomas of the nasal cavity and paranasal sinuses

<table>
<thead>
<tr>
<th>Reference</th>
<th>Age (yrs)/gender</th>
<th>Tumor location</th>
<th>Symptoms/signs</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Shugar et al¹</td>
<td>30/F</td>
<td>Maxillary sinus</td>
<td>Nasal stuffiness</td>
<td>Radical maxillectomy</td>
</tr>
<tr>
<td>Berdal &amp; Myhre²</td>
<td>46/F</td>
<td>Ethmoid sinus</td>
<td>Nasal stuffiness</td>
<td>Ethmoid resection</td>
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<tr>
<td>Berdal &amp; Myhre⁶</td>
<td>11/F</td>
<td>Ethmoid sinus</td>
<td>Nasal obstruction</td>
<td>External ethmoid resection</td>
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<tr>
<td>Berdal &amp; Myhre⁶</td>
<td>54/F</td>
<td>Maxillary sinus/hard palate</td>
<td>Swelling of alveolar ridge</td>
<td>Radiation therapy and excision</td>
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<tr>
<td>Loughran et al⁹</td>
<td>42/F</td>
<td>Ethmoid sinus</td>
<td>Eye pain and swelling</td>
<td>Wide local excision</td>
</tr>
<tr>
<td>Lynn-Macrae et al (present case)</td>
<td>35/F</td>
<td>Lateral nasal wall</td>
<td>Nasal obstruction</td>
<td>Wide local excision</td>
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### References