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## Chondrosarcoma of the Nasal Septum A Case Presentation

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**Abstract:** Chondrosarcoma of the faciomaxillary area is an uncommon entity and only a few cases have been reported of this cartilaginous tumor of the nose and paranasal sinuses. A case of chondrosarcoma of the nasal septum is presented along with the result of treatment. Total removal of the tumor was achieved by a maxillary swing approach. Histological examination of the excised tumor showed that the lesion was a well-differentiated chondrosarcoma.

**Introduction:** Chondrosarcomas are rare tumors and approximately 5% to 10% are located in the head and neck region.<sup>1</sup> The nasal septum is a particularly rare site of origin of this tumor.<sup>2</sup> Chondrosarcomas arise in tissues known to be formed of cartilage. It is a slow-growing tumor, occurring mostly in middle-aged men. Primary chondrosarcoma of the nasal and paranasal sinus region, including the nasal septum, rarely extends into the cranial or intracranial areas unless there is recurrence.<sup>3</sup> The widest possible exposure, to allow wide surgical excision, is considered the treatment of choice.<sup>4</sup>

**Case Report:** A 40 year old female, presented with a history of a mass in the nasal cavity and bulging of the hard palate for 1 year duration. The mass was very slow-growing and painless. The patient reported one episode of nasal bleeding. During the previous year, the growth had completely blocked the nose resulting in bilateral nasal obstruction, forcing the patient to breath from her mouth. There was no history of loss of vision, fever, loss of appetite or weight loss. There was no history of tuberculosis or diabetes mellitus. Anterior rhinoscopy revealed the presence of a firm, non-tender, fleshy intranasal mass occupying

both nasal cavities. On examination, a probe could not be passed along the nasal septum. There was no bleeding. Posterior rhinoscopy revealed a midline mass in the nasopharynx. Intraorally, there was a hard, nontender bulge over the hard palate which was covered by intact mucosa. There were no palpable lymph nodes. Her general and systemic examinations were normal. Both eyes were normal. The patient had no symptoms of increased intracranial pressure and neurological examination was normal.

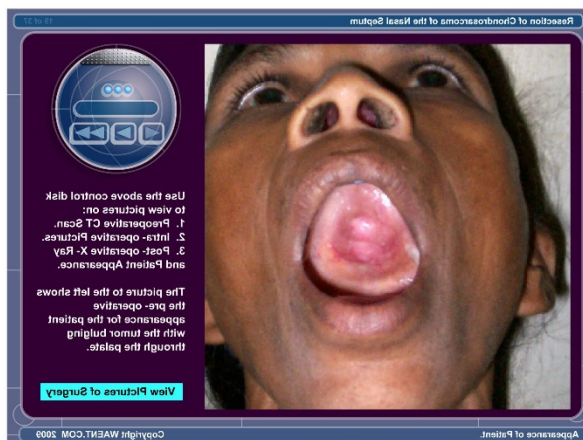
The CT scan of the patient's paranasal sinuses showed a well-circumscribed midline, destructive mass with multiple specks of calcific foci. The mass occupied the nasal septum and both of the nasal cavities. It involved both ethmoid sinuses, and partially both sphenoid sinuses with destruction of their walls. In addition, it involved the right lamina papyracea, both cribriform plates and part of the crista galli. The mass was also encroaching upon both maxillary sinuses, minimally encroaching upon the right orbit, displacing medial and inferior recti muscle laterally. The mass extended posteriorly through choana and bulging on the left side through the hard and soft palatal junction.

A wedge biopsy of the nasal mass was taken. Histopathology was suggestive of chondrosarcoma.

The tumor was excised under general anesthesia by a maxillary swing approach. A firm, gritty white tumor was attached to the nasal septum and extended into both nasal cavities filling the nasopharynx. The tumor also compressed the medial wall of the right maxilla and extended onto the hard palate. A part of the tumor was adherent to the base of the anterior cranial fossa. Complete removal of the tumor was accomplished.

Histopathology of the excised tumor revealed lobules of chondromatous tissue and at places was lined by nasal mucosa. Few lobules were relatively acellular and resembled mature cartilage, while others showed marked increase in cellularity with large chondrocytes having enlarged multinucleated and binucleated cells. The stroma consisted of collagen strands having congested blood vessels. Histology of the specimen was suggestive of well-differentiated chondrosarcoma.

The postoperative period was uneventful. Her ocular movements were normal. Vision and olfactory function were the same as the preoperative status. No adjuvant treatment was given. There was no evidence of local recurrence during the one year followup.



#### View Flash Animation of Resection:

<http://www.waent.org/archives/2009/vol2-2/20090729-chondrosarcoma/chondrosarcoma.htm>

**Discussion:** Chondrosarcomas constitute an uncommon group of cartilaginous tumors. They comprise only 10% to 20% of all malignant primary bone tumors, of which only 10% arise from the head and neck areas.<sup>5</sup> In the face, they commonly involve the nasal cavity, paranasal sinuses, nasopharynx, larynx, and mandible.<sup>3</sup> Chondrosarcomas constitute only 4% of nonepithelial tumors of the nasal cavity, paranasal sinuses and nasopharynx. Hence, this malignancy is

considered as one of the rarest and the origin of chondrosarcomas from the nasal septum is an extremely rare occurrence.<sup>3</sup> The usual age of presentation varies between 40 and 50 years,<sup>3</sup> with higher incidence

among males. Hence, this case of a female presenting with a chondrosarcoma of the nasal septum is an unusual one.

Chondrosarcomas arises from primitive mesenchymal stem cells or may arise in tissues known to be formed of cartilage from nests that remain after ossification.<sup>2</sup> Due to their slow growing nature, chondrosarcomas tend to be large at presentation.

Symptomatology varies according to the site and size of the tumor. Usually, nasal obstruction, nasal discharge, facial asymmetry, headache, restricted ocular movements, diplopia, proptosis, facial pain and ear fullness are presenting symptoms. Recurrence of the lesion with intracranial involvement is characterized by loss of vision, proptosis and multiple cranial nerve involvement.<sup>3</sup> In this case, the patient didn't have any other symptoms than nasal obstruction and one episode of epistaxis.

Radiological imaging is essential before attempting surgical intervention. Computer Tomographic (CT) scan in coronal and saggital plain is the most informative investigation in evaluating the extent and exact location of the tumor.<sup>1</sup> Bony destruction and calcification with an isodense or hypodense nonenhancing mass are common findings on CT scan. Magnetic Resonance Imaging, if available, is equally helpful. Biopsy of the lesion is the only way to confirm the diagnosis.

Although the histological appearances of this tumor was generally characteristic of a chondrosarcoma, a differential diagnosis of a chondroma, chordoma, and chondromyxoid fibroma should be considered.

Since it is described as a radio-resistant tumor due to its prolonged response time to radiation, the standard treatment of choice is wide radical surgical resection.<sup>6</sup> These tumors are not routinely offered post-operative adjuvant radiation therapy. Adjuvant radiation therapy or chemotherapy may have to be utilized for residual, recurrent disease or for palliation.<sup>6</sup> In the patient documented here, wide surgical excision with en bloc tumor removal was performed. Radiotherapy or chemotherapy was not given postoperatively.

The prognosis depends upon the location and extent of the lesion, adequacy of treatment and degree of differentiation. Patients with incomplete resections requiring further radiotherapy or chemotherapy have a bad prognosis. Generally, the posterior nasal cavity and nasopharynx are the sites with poor prognosis because of late presentation.<sup>6</sup> Chondrosarcomas located in the head and neck have a worse prognosis than those arising in other regions.<sup>7</sup>

**Conclusion:** Chondrosarcoma of nasal septum is an extremely rare occurrence especially in a middle aged female patient. Biopsy is important to obtain a diagnosis and a CT is required to know the exact extent of the tumor. Being a slow growing radioresistant tumor, radical surgical excision provides a good chance of cure. The maxillary swing approach provides excellent exposure of the nasal cavities, nasopharynx and the pterygoid region. Periodic follow up is necessary to monitor for recurrences.

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