Case Report

# Chordoma of skull base presenting as nasopharyngeal mass

Sant Prakash Kataria, Ashima Batra, Gajender Singh, Sanjay Kumar, Rajeev Sen

*Department of Pathology, Pt B.D. Sharma PGIMS, Rohtak, Haryana, India*

**ABSTRACT**

While the nasopharynx is most commonly regarded by the otolaryngologist as a primary site of neoplastic involvement, it is also an avenue of spread of base-of-the-skull tumors presenting as bulging nasopharyngeal masses. Chordoma is a relatively rare tumor of the skull base and sacrum thought to originate from embryonic remnants of the notochord. Chordomas arising from the skull base/clivus are typically locally aggressive with lytic bone destruction. The optimal treatment may be photon/proton radiotherapy alone or combined with a gross total resection, when feasible. We report a case of intracranial chordoma presenting as nasopharyngeal mass.

**Key words:** Chordoma, nasopharyngeal mass, skull base

## Introduction

Intracranial chordomas are relatively rare locally aggressive tumors thought to originate along the course of the embryonic remnant of the notochord. The most common location is in the sacrum; however, the skull base/clivus region is the second most common location along the course of the notochord.[1]

Skull base chordomas usually arise in the clivus and are rarely completely resectable.[2] They may cause extensive bony erosion of the petrous apex, sphenoid sinus, and clivus and may suggest a more rapidly growing and aggressive tumor type. The extent of the tumor may be accurately determined by conventional tomography, computerized axial tomography, and arteriography.[3] Most are treated with radiotherapy (RT) because of extensive involvement making them unresectable. Because of the risk of severe late complications, the dose is often limited with conventional photon RT, and the probability of cure is low.[2]

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## Case Report

A 28 year-old female sought medical attention for pain in throat and difficulty in deglutition for past 2 months. On anterior rhinoscopy, a pale mass was seen occupying right nasal cavity. The left nasal cavity was normal. Oral cavity examination revealed a smooth bulge on right-sided soft palate and anterior pillar of right tonsil, because of which the right tonsil could not be visualized. Uvula was in the midline. Indirect laryngoscopy could not be done. On central nervous system (CNS) examination, bilateral occulomotor and right hypoglossal nerves were involved. Fibreoptic nasopharyngoscopy was done under local anesthesia. A mass was seen in superior part of nasopharynx. Other sites of nasopharynx including fossa of Rosenmuller, eustacian tube opening, and torus tubarius were unremarkable.

Computed tomography (CT) scan of head and neck [Figure 1a and b] showed that base of skull was eroded with extracranial extension of bilateral temporal lobes and occipital lobe. The mass was compressing and pushing brainstem and extending into orbit and bilateral pterygopalatine and infratemporal fossae. Biopsy of the mass was performed.

Histological examination revealed round to oval cells with central nuclei and a vacuolated cytoplasm (physaliphorous cells) [Figure 2a]. The cells stained positive with Periodic acid-Schiff (PAS) [Figure 2b] and monoclonal staining

**Address for correspondence:**
Dr. Ashima Batra, H. No. 901 Ward No. 22, Jhang Colony, Rohtak, Haryana, India. E-mail: drashimabatra@gmail.com

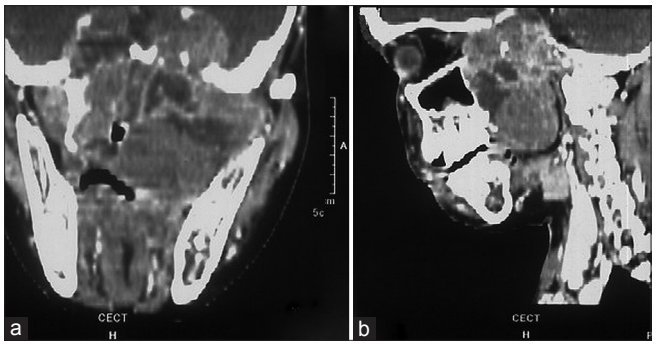


**Figure 1:** (a and b) CT scan showing erosion of base of skull with extracranial extension

for epithelial membrane antigen (EMA) [Figure 2c]. A histopathological diagnosis of chordoma was made.

The patient was advised RT but the patient did not comply and became lost to follow up.

## Discussion

Chordoma is a rare, low-grade, primary malignant bone tumor arising from primitive notochord remnants of the axial skeleton. It accounts for 1-4% of all primary skeletal tumors and its incidence rate is inferior to 0.1 per 100,000 inhabitants per year.[4] They arise from the sacrum in approximately 50-60% of cases, from the skull base region (spheno-occipital/nasal) in approximately 25-35% of cases, from the cervical vertebrae in approximately 10% of cases, and from the thoracolumbar vertebrae in approximately 5% of cases. The median age at presentation is around 60 years; however, presentation with skull base tumors may occur at a younger age and has been reported in children and adolescents.[5] Intracranial chordomas have a male (2:1) predilection.[6]

The clinical features of chordoma will depend on the site and line of spread of the tumor. In the cranial case, which arises in the region of the basi-sphenoid, the growth spreads intracranially at an early stage and death eventually results from compression of vital structures. Apart from invading the cranial cavity, the growth sometimes projects into the nasopharynx as well when symptoms of obstruction will be produced. The prominent complaints will therefore consist of headache and cranial nerve palsies due to intracranial spread, nasal obstruction with discharge, and dysphagia in those with pharyngeal spread. Epistaxis as in nasopharyngeal carcinoma is not uncommon.[7]

Chordomas are usually relatively slow-growing, low-grade malignancies.[5] They have been considered of low

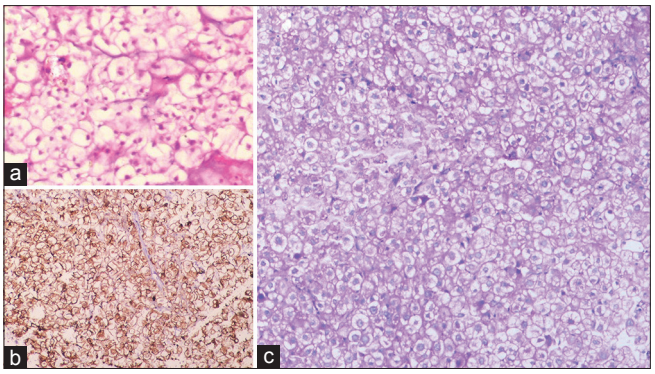


**Figure 2:** Photomicrograph showing round to oval cells with central nuclei and vacuolated cytoplasm (physaliphorous cells) (a) H and E (×40). These cells stained positive with (b) PAS (×40) and (c) monoclonal staining for EMA (×100)

metastatic potential; however, distant metastasis to lung, bone, soft tissue, lymph node, liver, and skin has been reported in up to 43% of patients.[8] However, metastatic sites usually occur late in the course of the disease.[4]

Both CT and magnetic resonance (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.[1]

Histologically, they display lobules and vacuolated (physaliphorous), moderately atypical, neoplastic cells across a myxoid stroma separated by fibrous bands.[9]

Adequate wide surgery still remains the cornerstone of chordoma treatment even though safe margins are often hard to obtain because of its anatomical sites of origin.[8] Surgery for skull base and upper cervical spine tumors poses a risk of damage to normal structures, including the spinal cord and cranial nerves. These same normal tissue structures also impose a limitation on the external-beam RT dose, which adds additional difficulty. The advent of advanced imaging, planning, and delivery of photon RT over the past one to two decades has provided opportunities for delivering high doses of radiation safely to patients with skull base and cervical spine tumors.[4] Proton RT alone or combined with photon RT (proton/photon RT) offers the advantage of improved dose distribution and the ability to treat the tumor to a higher dose without exceeding normal tissue tolerance.[2] Sensitivity to chemotherapy is very low and generally reported in the small subgroup of patients with high-grade dedifferentiated chordomas with agents active in high-grade sarcomas.[10]

The best results in the treatment of chordomas of the skull base are reported when using surgery and adjuvant high-dose proton RT.[4]

## Conclusion

Chordoma of skull base is a rare entity. It can sometimes present as nasopharyngeal mass which can be mistaken for primary nasopharyngeal mass. To minimize delay in diagnosis, nasopharyngeal extension of an intracranial chordoma should be considered in the differential diagnosis of any mass forming lesion in nasopharynx.

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