Content available at: https://www.ipinnovative.com/open-access-journals

IP Journal of Diagnostic Pathology and Oncology

Journal homepage: https://www.jdpo.org/

Case Report

Chondroid chordoma of the nasopharynx - A rare case report

Pallavi Padamsing Ghunawat[1,*], Madhuvantee Shashikant Sonawane[1], Shashank Ramesh Godbole[1], Anitha Padmanabhan[2], Nitin Gadgil[1]

[1]*Dept. of Pathology, Lokmanya Tilak Municipal Medical College and General Hospital, Mumbai, Maharashtra, India*
[2]*Lokmanya Tilak Municipal Medical College and General Hospital (LTMGH), Mumbai, Maharashtra, India*

ARTICLE INFO

Article history:
Received 06-04-2023
Accepted 03-06-2023
Available online 28-08-2023

Keywords:
Nasopharynx
Chordoma
Physaliphorous cells

ABSTRACT

Chordomas are slow-growing tumors that account for 1-4% of all bone tumors. They are most commonly seen in the sacrum, followed by the base of the skull and spine. Males are affected more than females. A 28-year-old female presented with nasal bleeding and bilateral nasal obstruction for 1 month. Clinical, physical, and radiological examinations revealed a mass in the nasopharynx. The radiological diagnosis was a large polypoidal adenomatous neoplastic mass. However, histopathological examination revealed a chordoma. The nasopharynx is an unusual site for chordoma presentation, and it has a nonspecific clinical and radiological appearance. Hence, it should be considered as a differential diagnosis of a nasopharyngeal mass.

1. Introduction

Chordomas are rare, locally aggressive tumors that originate from the embryonic remnant of the notochord. They account for 1-4% of all primary malignant bone tumors, with an incidence rate of 0.8 per 100,000 per year.[1] They occur twice as frequently in men compared to women, with an age range around 40-60 years.[2] The sacrococcygeal area (65%) is the most common site, followed by the spheno-occipital area (25%), and rarely the cervico-thoraco-lumbar spine (15%) is involved.[3] Skull base tumors may occur at a younger age and have been reported in children and adolescents.[2] Primary chordoma of the nasopharynx commonly presents as a nasal mass, which causes headaches, nasal obstruction with discharge and pressure symptoms, and dysphagia in those with pharyngeal spread.[2]

Radiographically, chordomas are destructive midline lesions that are readily evident on CT and MRI, but are difficult to identify on plain radiographs. Lesions arising in the skull base arise in the midline, destroy surrounding bone, and encroach on the brain stem or optic nerve. In the sacrum and coccyx, they form lobular masses that extend anteriorly toward the rectum and bladder.[4]

Grossly, chordomas are lobulated, gelatinous, soft, and contain areas of haemorrhage. Surgical excision is the treatment of choice because chordomas have poor sensitivity to radiotherapy and chemotherapy.

2. Case Report

A 28-year-old female presented with right nasal bleeding for the last month. She also complained of bilateral nasal blockage, bilateral supraorbital headache, purulent discharge, and nasal twang for the same duration.

A CT scan of the paranasal sinuses and neck revealed evidence of a well-defined soft tissue density lesion

* *Corresponding author*.
E-mail address: pallavighunawat133@gmail.com (P. P. Ghunawat).

https://doi.org/10.18231/j.jdpo.2023.037

involving the sphenoid sinus, posterior ethmoidal air cells, and nasopharyngeal region. The lesion caused expansion and erosion of the floor of the sella, roof of the sphenoid, and floor of the anterior cranial fossa. Furthermore, there was destruction of the septum encroaching on the right retro-orbital region, with indentation over the medial rectus muscle and destruction of the medial wall of the right orbit.

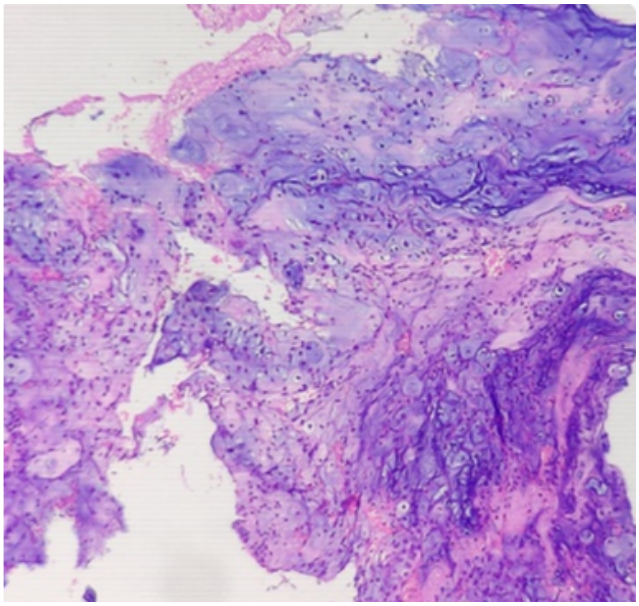

Fig. 1: Scanner view shows chondroid matrix and foci of tumor cells arranged in sheets. (H&E 40X)

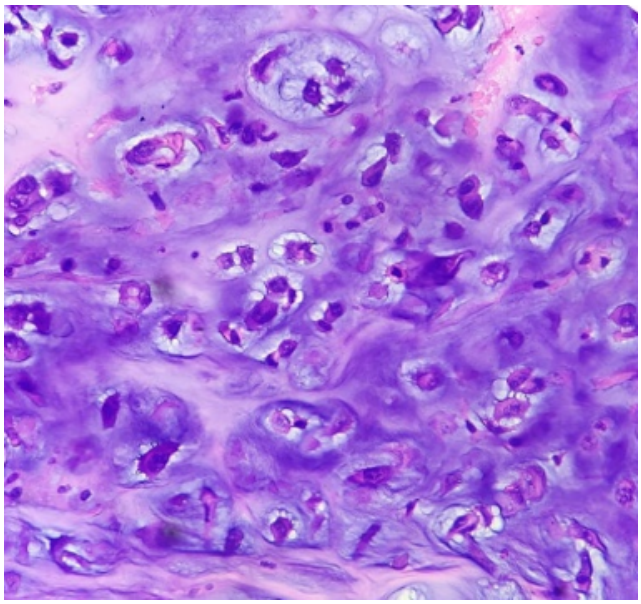

Fig. 2: High power view shows physaliphorous cells showing abundant eosinophilic vacuolated cytoplasm and moderate pleomorphic nuclei. (H&E 400X).

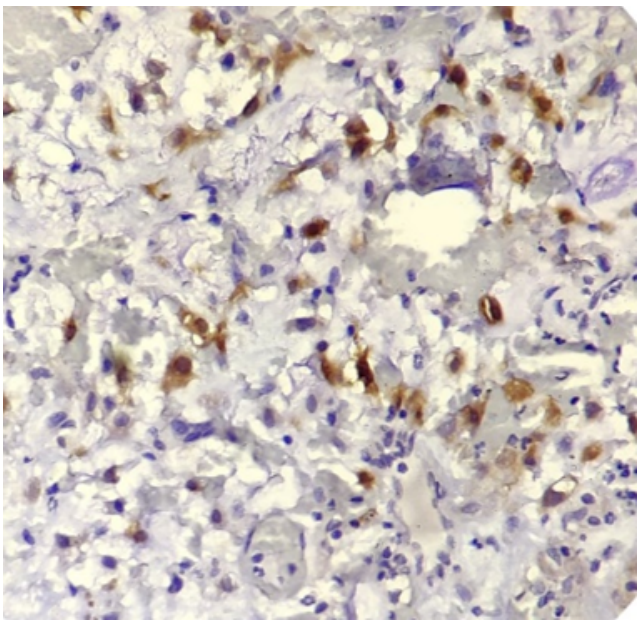

Fig. 3: Pan cytokeratin.

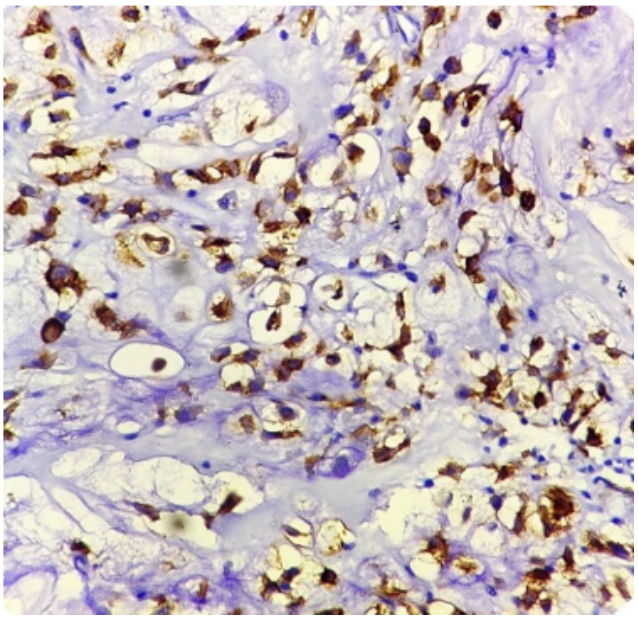

Fig. 4: S100 shows nuclear positivity.

A biopsy was taken from the right nasal mass and sent to the surgical pathology section. The biopsy was processed, and a hematoxylin and eosin stain was performed on it, revealing small foci of a tumor composed of sheets of physaliphorous cells with moderate pleomorphic nuclei. Additionally, a few mitotic activities were observed. The biopsy also showed extensive extracellular chondromyxoid material with haemorrhage and a dense mixed inflammatory infiltrate. Immunohistochemistry performed on the biopsy showed positive reactivity to Pan-CK, S100, and diffuse

positive epithelial membrane antigen (EMA). Based on the hematoxylin and eosin slides and the results of immunohistochemistry, the diagnosis of chondroid chordoma was confirmed.

3. Discussion

Chordoma is a rare nonepithelial malignant bone tumor primarily involving both ends of the axial skeleton, and it arises from remnants of the notochord. Primary nasopharyngeal chordoma arises in extraosseous nasopharyngeal soft tissue. It commonly occurs in the young adult and adolescent age groups. It affects males more commonly than females, with a male-to-female ratio of 2:1.[5] The sacrococcygeal area is the most common site, followed by the spheno-occipital area, and rarely the cervico-thoraco-lumbar spine is involved.[3] The most common clinical presentation is nasal obstruction. It also presents with other symptoms such as hearing impairment, headache, difficulty in swallowing, dryness of the mouth, nasal bleeding, diplopia, nasal speech, and difficulty in breathing. On gross examination, chordomas are lobulated, gelatinous, soft, and contain areas of haemorrhage.

Chordomas are classified into three different subtypes: conventional or classic chordomas, chondroid chordomas, and dedifferentiated types. Classic chordomas show numerous physaliphorous cells arranged in cords and lobules with a myxoid stroma separated by fibrous tissue. Chondroid variants show chondroid differentiation, and dedifferentiated variants show a biphasic tumor with two components: the first component being a conventional chordoma and the second component being a high-grade sarcomatous component. A few cases show a poorly differentiated type.[6–12]

The patient in the present case report had a chondroid type chordoma. Microscopically, it showed foci of tumor cells arranged in sheets composed of physaliphorous cells characterized by large intracytoplasmic vacuoles and moderate pleomorphic nuclei embedded in chondromyxoid stroma (Figures 1 and 2). The tumor cells stained strongly positive for cytokeratin, S100, and EMA (Figures 3 and 4).

Chondrosarcoma would be an important differential diagnosis of chondroid chordoma. Cytokeratin will be positive for chondroid chordoma and negative for chondrosarcoma.[3]

Local recurrence with an incidence of 50-68% is common due to the aggressive nature of this tumor, and systemic metastasis to the lung, bone, liver, and lymph nodes are found in 17.8-43% of patients with chordoma.[1]

Adverse prognostic factors include subtotal resection, dedifferentiated histological subtype, age >40 years, history of prior treatment, tumor size ≥4cm, gross tumor volume >25cm^3, pharyngeal involvement, optic nerve and brain stem compression, none or low-dose radiation therapy, and female sex.[1]

Surgery is the primary modality to achieve the best long-term control.

4. Conclusion

In conclusion, chondroid chordoma of the nasopharynx is a rare condition that poses challenges in clinical diagnosis, often resulting in misdiagnosis. Due to its rarity and nonspecific symptoms, it is important to rely on histopathological examination to establish an accurate diagnosis. This examination helps differentiate chondroid chordoma from other similar conditions and enables appropriate management planning. Surgical intervention remains the primary treatment modality for achieving optimal long-term control. Awareness of this rare entity and its characteristic features is crucial for timely diagnosis and appropriate management of chondroid chordoma of the nasopharynx.

5. Authors' Contribution

All authors contributed equally to the manuscript.

6. Conflict of Interests

The authors have no conflict of interest to disclose.

7. Source of Funding

None.

References

1. Yeh CY. Radiotherapy of extraosseous nasopharyngeal chordoma: A case report and literature review. *Mol Clin Oncol*. 2021;14(4):84. doi:10.3892/mco.2021.2246.
2. Dwianingsih EK, Snak Y, Rinonce HT, Wasita B, Antoro EL, Amr SS, et al. Primary Chordoma of the Nasopharynx: A Rare Case Report and Review of the Literature. *Case Rep Pathol*. 2019;p. 3826521. doi:10.1155/2019/3826521.
3. Cha YJ, Suh YL. Chordomas: histopathological study in view of anatomical location. *J Korean Med Sci*. 2019;34(13):e107. doi:10.3346/jkms.2019.34.e107.
4. Santegoeds RG, Temel Y, Beckervordersandforth JC, Van Overbeeke J, Hoeberigs CM. State-of-the-art imaging in human chordoma of the skull base. *Curr Radiol Rep*. 2018;6(5):16. doi:10.1007/s40134-018-0275-7.
5. Mitra B, Sengupta S, Rai A, Mehta J, Quader AR, Roy S. Chordoma in Nasopharynx in a 70-Year-Old Female: A Rare Occurrence. *Int J Otolaryngol Head Neck Surg*. 2014;3(6):342. doi:10.4236/ijohns.2014.36061.
6. Lynn-Macrae A, Haines I, Gk, Altman KW. Primary chordoma of the lateral nasal wall: case report and review. *Ear Nose Throat J*. 2005;84(9):593–5.
7. Gupta R, Khurana N, Singh S, Meher R. Primary chordoma of the nasal cavity: a rare site of presentation. *Pathology*. 2007;39(2):273–5.
8. Yan ZY, Yang BT, Wang ZC, Xian JF, Li M. Primary chordoma in the nasal cavity and nasopharynx: CT and MR imaging findings. *Am J Neuroradiol*. 2010;31(2):246–50.
9. Mou Z, Liu Z. Primary chordoma of the nose. *Chinese Med J*. 2003;116(1):154–6.
10. Kataria SP, Batra A, Singh G, Kumar S, Sen R. Chordoma of skull base presenting as a nasopharyngeal mass. *J Neurosci Rural Pract*.

2013;4(1):95–7.
11. Nguyen RP, Salzman KL, Stambuk HE, Ahuja AT, Harnsberger HR. Extraosseous chordoma of the nasopharynx. *Am J Neuroradiol*. 2009;30(4):803–7.
12. Gupta D, Rathore PK, Chauhan A, Khurana N. Unusual Presentation of Chordoma in Nose. *Indian J Otolaryngol Head Neck Surg*. 2016;68(3):380–3.

Author biography

Pallavi Padamsing Ghunawat, Student https://orcid.org/0009-0007-0115-8220

Madhuvantee Shashikant Sonawane, Student

Shashank Ramesh Godbole, Student

Anitha Padmanabhan, Assistant Professor

Nitin Gadgil, Professor

Cite this article: Ghunawat PP, Sonawane MS, Godbole SR, Padmanabhan A, Gadgil N. Chondroid chordoma of the nasopharynx - A rare case report. *IP J Diagn Pathol Oncol* 2023;8(3):161-164.