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Chordoma of Nasopharynx - A Rare Case Report

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Abstract: ***Chordomas are rare, locally aggressive malignant tumors originating from embryonic notochord remnants, with an overall incidence of less than 0.1 cases per 100,000 individuals. While they typically occur along the central craniospinal axis, presentation as an extra-osseous primary mass in the nasopharynx is exceptionally rare. Here is a case of 54 year old male presenting with voice change and nasal blockage since 6 months. Imaging revealed a large (46 x 57 x 47 mm) expansile, cystic, and solid lesion in the lower clivus, protruding into the nasopharynx and causing bony erosion. Histologically diagnostic hallmark was the presence of pathognomonic physaliphorous cells-large, epithelioid cells featuring a prominent bubbly, vacuolated cytoplasm. Immunohistochemistry shows positivity for EMA, S100, CK19, and CK8/18. This definitive histopathological and IHC profile excluded major morphologic mimics such as nasopharyngeal carcinoma and chondrosarcoma, securing the diagnosis of conventional chordoma.***

Keywords: Nasopharyrnx, Chordoma.

1. Introduction

Chordomas are rare, locally aggressive malignant tumors that originate from the remnants of the primitive embryonic notochord. While they account for approximately 1% to 4% of all primary malignant bone tumors, their overall incidence is extremely low, estimated at less than 0.1 cases per 100,000 individuals annually. The disease predominantly affects adults between 40 and 60 years of age and is approximately twice as common in males as it is in females.

These tumors typically develop along the central craniospinal axis. The most frequent anatomical sites are the sacrococcygeal region, which accounts for 50% to 60% of cases, and the spheno-occipital or craniocervical skull base (the clivus), making up 25% to 35% of cases. However, chordomas can occasionally present in highly unusual, extra-osseous locations such as the maxilla, mandible, nasal cavity, and the nasopharynx. Primary nasopharyngeal chordomas are exceptionally rare and arise in the soft tissues of the nasopharynx, sometimes without any bony involvement or destruction of the adjacent clivus.

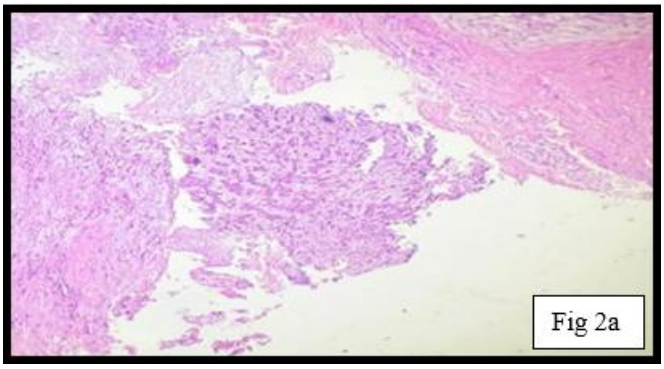
2. Case History

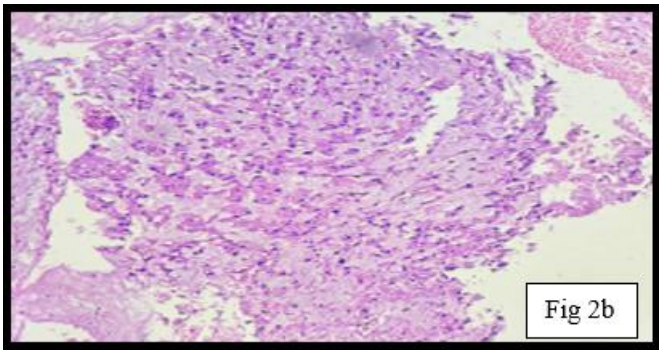
A 54 year male patient presented with chief complaint of voice change and nasal blockage since 6 months. On examination a smooth cystic in appearance mass over nasopharynx extending below till oropharynx was seen. His hemoglobin was 8.5g/dl, and CBC, coagulation profile and biochemical parameters were within normal limits. On CT scan a wall-enhancing cystic lesion measuring 5 x 4.4 x 3.8 cm (CC x T x AP) was seen in prevertebral space in midline extending from clivus upto C2 vertebral level. Internal septations with calcification are seen. Posteriorly it is extending between clivus and anterior arch of C1 vertebra with resultant bony scalloping and abutting dura. MRI showed Focal fairly large well defined expansile abnormal marrow signal intensity lesion (measuring approximately 46 (W) x 57 (AP) x 47 (CC) mm in size), hyperintense on T2 with heterogeneity within, intermediate to hyperintense septations and nodularity within, moderate restricted diffusion on DWI in solid component, with heterogeneous enhancement, was seen in lower part of clivus, protruding into nasopharynx. It was causing erosion of anterior and posterior cortices and showed focal fairly large nodular component projecting anteriorly into nasopharynx causing significant narrowing of the lumen.

Grossly we received creamish brown multiple soft tissue bits measuring total about 3.5x3x0.5 cm and all tissue was submitted. On microscopic examination sections show respiratory type mucosal epithelium with underlying fibromuscular tissue infiltrated by multinodular neoplasm. Neoplasm composed of sheets, cords and singly scattered epithelioid vacuolated cells with bubbly cytoplasm. Neoplastic cells are surrounded by abundant myxoid stroma. Immunohistochemistry was done to confirm the case, with EMA, S100, CK19, CK8/18 which were positive.

Figure 1: Focal fairly large well defined expansile abnormal marrow signal intensity lesion (measures approximately 46

(W) x 57 (AP) x 47 (CC) mm in size), hyperintense on T2 with heterogeneity within, intermediate to hyperintense septations and nodularity within, moderate restricted diffusion on DWI in solid component, with heterogeneous enhancement, is seen in lower part of clivus, protruding into nasopharynx.





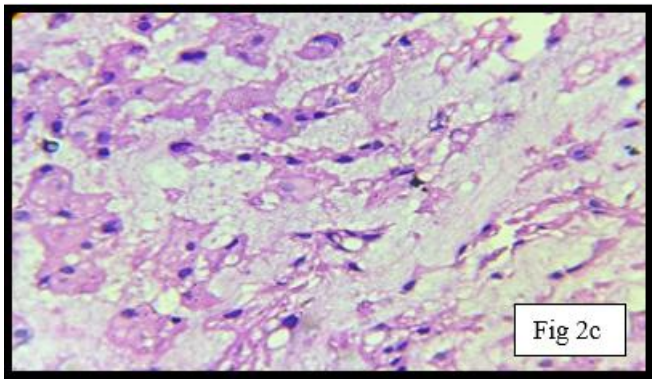
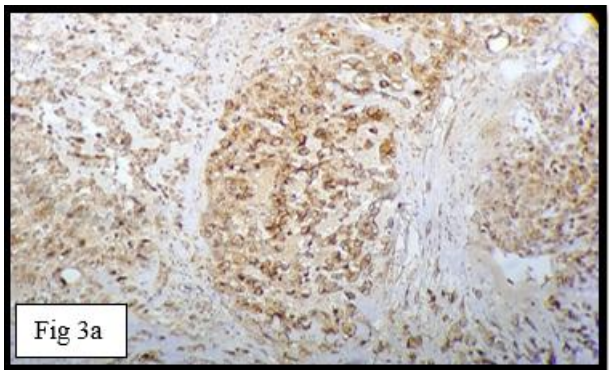
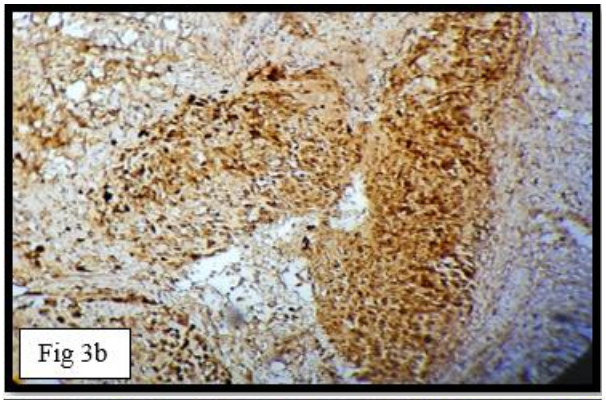
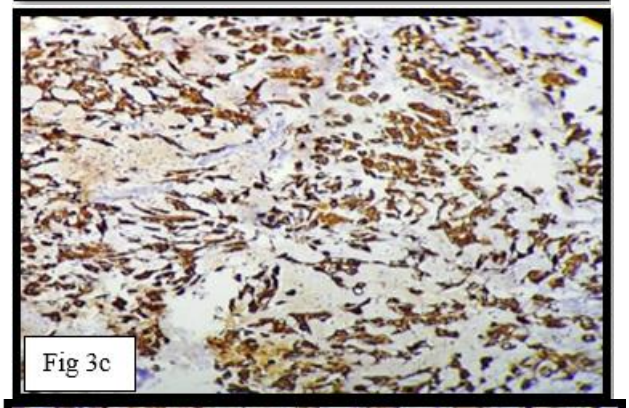


Figure 2 (a): Scanner view shows myxoid stroma and tumor cells arranged in sheets and nests. Fig 2 (b): 10x view. Fig 2 (c): Physaliphorous cells showing abundant eosinophilic vacuolated cytoplasm and moderate pleomorphic nuclei.







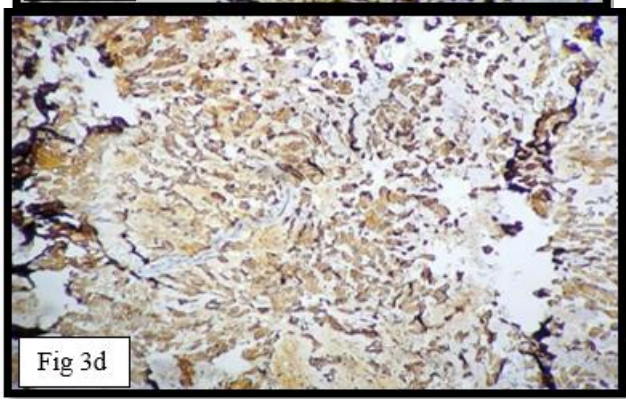


Figure 3 (a): EMA shows strong cytoplasmic staining, Fig 3(b): S100 shows strong nuclear and cytoplasmic positivity, Fig 3 (c): CK19 shows strong cytoplasmic staining, Fig 3 (d)**:** CK8/18 weak cytoplasmic staining

3. Discussions

Chordomas are rare, slow-growing but locally aggressive malignant bone tumors that arise from the embryonic remnants of the notochord [1]. While they typically occur along the axial skeleton, the skull base and clivus (spheno-occipital region) account for 25% to 35% of all cases. However, the presentation of a chordoma as an expansile mass in the nasopharynx of a 54-year-old male is an exceptionally rare, atypical extra-osseous manifestation [2] These extra-osseous nasopharyngeal chordomas are theorized to originate from notochordal remnants along the medial basal canal, which represents the cephalad exit tract of the notochord into the midline nasopharyngeal soft tissues during embryogenesis [3]

The histomorphology of a conventional chordoma is distinct. The tumor is characterized by a lobular architecture, with cords, solid sheets, and lobules of neoplastic cells separated by prominent fibrous septa. The cells are embedded in a rich,

extracellular myxoid or mucinous stroma. The hallmark diagnostic feature is the presence of pathognomonic **physaliphorous (Greek for "bubble-bearing") cells**. These are large, polygonal cells with abundant, clear to eosinophilic cytoplasm containing prominent vacuoles that are laden with mucin or glycogen.[4] To complete the histopathologic evaluation, one must exclude the chondroid variant (which shows hyaline cartilage differentiation) and the dedifferentiated variant (which shows high-grade sarcomatous spindle cells), confirming that the absence of these features secures the diagnosis of a conventional chordoma [5]. Immunohistochemistry (IHC) is paramount in confirming the diagnosis of chordoma and excluding morphologic mimics Notochordal neoplastic cells demonstrate a unique dual epithelial-mesenchymal immunophenotype, showing robust positivity for epithelial markers (such as cytokeratins and epithelial membrane antigen) as well as mesenchymal markers (such as S100 protein). Also Brachyury has found to be very useful marker, but due to its unavailability we have not performed it. [6]

From a pathology perspective, a nasopharyngeal chordoma must be differentiated from several other lesions that present as midline nasopharyngeal masses, as the clinical and radiological findings are often unspecific. **Nasopharyngeal Carcinoma (NPC):** Clinically the most suspected lesion. Histologically, NPC consists of infiltrating islands of atypical epithelial cells (round, oval, or spindle). While NPC is positive for cytokeratins, it lacks the physaliphorous cells, the abundant myxoid matrix, and will be completely negative for S-100. **Chondrosarcoma:** A major morphological mimic, especially if we are dealing with a chondroid chordoma variant (which accounts for up to 33% of cranial chordomas and features both chordomatous and chondromatous elements). Chondrosarcomas exhibit pleomorphic, hyperchromatic cells but lack true large cytoplasmic vacuoles. Crucially, chondrosarcomas are negative for epithelial markers (EMA, Cytokeratins) making the IHC panel definitive for distinguishing the two. **Chordoid Meningioma:** Can present with cords of polygonal cells and vacuolated cytoplasm resembling physaliphorous cells in a myxoid stroma. However, chordoid meningiomas are negative for cytokeratins. **Benign Cystic or Inflammatory Lesions:** Radiologically, extra-osseous chordomas may mimic a Tornwaldt cyst or abscess. Microscopically, this is easily resolved, as cysts are lined by simple cuboidal or flattened epithelium without neoplastic physaliphorous cells, and abscesses are defined by necrotic tissue and heavy leukocyte infiltration.[3]

Providing an accurate diagnosis of nasopharyngeal chordoma heavily alters patient management. Unlike NPC, which is highly responsive to chemoradiotherapy, chordomas are relatively radioresistant and chemoresistant. The primary and most effective treatment is gross total surgical resection with negative margins, often followed by adjuvant high-dose proton beam radiation. Our patient has undergone total surgical resection, further follow up was not possible as patient was referred to higher center.

4. Conclusion

This case underscores the critical importance of integrating classic histomorphology with a targeted immunohistochemical panel when evaluating unspecific midline nasopharyngeal masses. Ultimately, delivering an accurate and timely histopathological diagnosis is vital as it fundamentally alters patient management. It redirects the clinical team away from chemoradiotherapy—to which chordomas are relatively resistant—and toward the primary, most effective treatment: **gross total surgical resection.**

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