

CASE REPORT

CHONDROID CHORDOMA OF NASAL SEPTUM: A CASE REPORT

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ABSTRACT: Chordomas are rare neoplasms of presumed notochordal origin that arise along the vertebral axis and show a proclivity for the sphenoid-occipital and sacral regions.^(1,2) Chondroid chordoma is a variant of chordoma, with cartilaginous component. We report a histologically proven case of chondroid chordoma, with an exceptional localization to the nasal septum. Chondroid Chordomas are likely to recur and hence diagnostically important to institute effective treatment.

KEYWORDS: Chordoma, Chondroid chordoma, notochord, Immunohistochemistry.

INTRODUCTION: Chordomas account for 2 to 4% of primary bone tumors, being the fourth most common pathology among primary bone cancers. The annual incidence is estimated to 1 per 1000,000, with a peak incidence in the 4th to 7th decade.⁽³⁾ Chondroid chordoma are thought to derive from the notochord. Chondroid chordoma is a subtype of chordoma possessing elements of both chordoma and cartilaginous tissue with better prognosis than classic (Non-chondroid) chordomas. Virchow first described the classical distinguishing feature of chordomas- 'physaliferous cell' a cell with large vacuoles containing mucin and glycogen and with a "bubbly" appearance of the cytoplasm. In 1973, Heffelfinger et al. were the first to describe chondroid chordoma.⁽²⁾ In chondroid chordoma, within features of classical chordoma, areas of chondroid differentiation are observed mimicking a low-grade chondrosarcoma. Differential diagnosis can only be assessed by immunochemistry.

CASE REPORT: A 55yrs male patient came to ENT OPD with complaints of right sided nasal obstruction and mass in the right side of nasal cavity since 1yr.

ON EXAMINATION: Diffuse swelling on the right side of nose with obliteration of naso maxillary groove, on anterior rhinoscopy swelling in the right side of nasal cavity (FIG 1&2), pinkish in colour with smooth surface, probe test showed that the mass was sensitive to touch, non-tender, does not bleed on touch, firm in consistency, immobile, probe could not be passed on the medial aspect of the mass probably arising from septum & the septum was deviated to left.



Fig. 1



Fig. 2

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Radiology- CT scan of PNS: Plain CT scan of PNS was done which showed well defined soft tissue density mass lesion in the anterior aspect of right nasal cavity suggestive of benign lesion. (Fig. 3 & 4).



Fig. 3

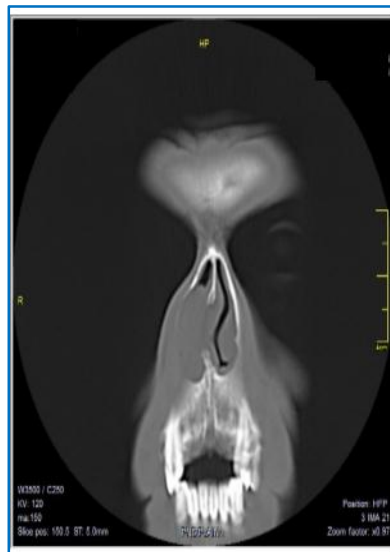


Fig. 4

Other routine investigations were normal, and the patient was posted for complete excision of the mass.

TREATMENT: Killian's incision was taken on the right side of the septum and the mass was excised in to from the septum. The mass was sent for HPE.

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Fig. 5: Gross Specimen

GROSS SPECIMEN: The specimen was a grey white soft tissue mass and measured 3.5 x 3x1.5 cms. in dimension. The external surface appeared mucoid with lobulations. Cut section revealed grey white areas with mucoid areas.

HISTOPATHOLOGICAL EXAMINATION: Sections studied shows a tumour tissue with a lobulated appearance. Pseudostratified ciliated columnar epithelium identified at one focus. The tumour exhibits a lobular pattern separated by fibrous septae. Within the lobule, small round cells without significant cytologic pleomorphism are arranged in a cord like fashion in a myxoid stroma. Many of the tumour cells exhibit vacuolization of the cytoplasm resembling physaliphorous cells. Significant areas of cartilaginous differentiation are also identified. A close differential diagnosis of chondroid chordoma and chondrosarcoma was given and IHC recommended.

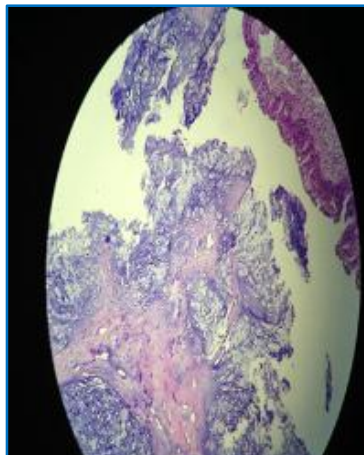


Fig. 6: H&E stain –low power

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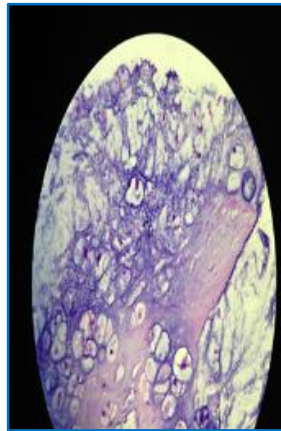
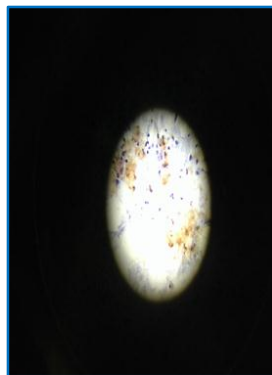
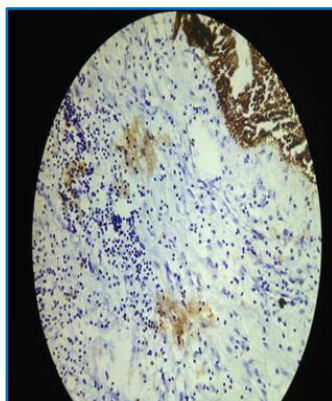


Fig. 7: H&E stain – high power

Immunohistochemistry was done and the specimen was Cyto keratin- positive & S-100 protien – negative.



**Fig. 8: low power
Cytokeratin- positive**



**Fig. 9: high power Cyto
keratin- positive**

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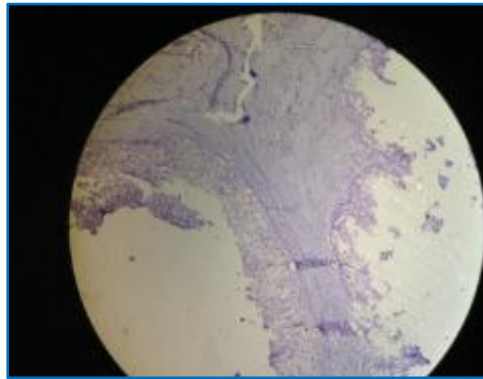


Fig. 10: S-100 protien - negative

Post-operative follow up: The patient was followed up for a period of 6 months and no recurrence was noted.



Fig. 11: Post op follow up

DISCUSSION: In 1973, Heffelfinger et al. were the first to describe chondroid chordoma.⁽²⁾ The pathophysiology of chordoma remains unclear. Chordomas are derived from the primitive notochord. The initial lesions might be benign chordal ectopias called ecchordosis physaliphora which are encountered in asymptomatic adults with an incidence of 1%.⁽³⁾ Chordomas also have a low course evolution. They usually tend to displace surrounding soft tissues more than invading them. Tumors are able to display proteolytic activity and may be widely invasive in bone tissue but generally metastases occur late in the disease course usually years after the initial diagnosis.⁽³⁾ Skull base lesions are less likely to metastasize than sacrococcygeal and vertebral chordomas.

The incidence of metastases in chordomas regardless of their location and their initial size is estimated to be between 3 and 48%.⁽³⁾ Chondroid chordoma are thought to derive from the notochord. The notochord is a transient structure of embryogenesis which arises as a pouch from the mesoderm during gastrulation. It defines the primitive axis of the embryo, conditions its elongation and the formation of the neural tube. In higher vertebrates it regresses completely throughout the column except in the nucleus pulposus. This process explains why chordomas are located along the axial skeleton (From the sacrum to the sphenoidal clivus).⁽⁴⁾ Extra axial localizations remain possible

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but exceptional so that the presentation of this case was very surprising. The classic histopathological characteristic of chordoma is the presence of physaliphorous cells.⁽⁵⁾

These cells contain an intracytoplasmic accumulation of mucopolysaccharides. Their nuclei are vacuolated, eccentric, with prominent nucleoli. Classical chordoma is organized in a lobular pattern, with cords of epithelioid cells within a mucomyxoid matrix. Cells rarely present atypia. In chondroid chordoma, within features of classical chordoma, areas of chondroid differentiation are observed, mimicking a low-grade chondrosarcoma. Differential diagnosis can only be assessed by immunochemistry. Chondroid Chordomas are positive for EMA (Epithelial marker antigen), cytokeratins (CK19) staining. ⁽⁶⁾ Chondroid areas are positive for vimentin staining. ⁽⁶⁾ Chondrosarcomas do not express epithelial markers and are positive only for S-100 protein & vimentine staining. Finally, D2-40 staining has proved to be effective in distinguishing true chondroid tumors from chondrosarcoma, but is not currently used. ⁽⁷⁾

CONCLUSION: Chondroid Chordoma is a rare tumour and its occurrence on nasal septum is exceptionally rare, hence awareness of this tumour is important to avoid misdiagnosis and to institute effective treatment. Chondroid Chordomas are likely to recur & periodic follow up of the patient is required.

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