



A CASE OF SINONASAL CHONDROSARCOMA–CASE REPORT

SankaraNarayanan Gopalakrishnan K.M Elango Swarna Saravanan

Government Royepettah Hospital Chennai

ABSTRACT

Introduction: Chondrosarcoma of sinonasal region is very rare. Chondrosarcoma make up 10-20% of primary bone tumor. Less than 10% constitute the head and neck region. Chondrosarcomas take origin from the embryonic rest of the cartilaginous matrix of the cranium or primitive mesenchymal cells. First choice of treatment is surgical excision. Radiotherapy also has role in treatment.

Case report: In our case 43 year old female presented with c/o proptosis of right eye, loss of vision since 1 month. All necessary blood investigation was done. CT scan PNS and MRI showed mass occupying the right nasal cavity along with dural involvement and encasement of right ICA by 180 deg. Endoscopically the tumor was excised. The vision improved postoperatively. Histopathology revealed low grade chondrosarcoma. Patient was treated with post op radiotherapy. Patient is under follow up.

Conclusion: Endoscopically nasal mass was removed. Patient showed improvement in vision postoperatively. This case report shows the feasibility of using endoscopic technique to aid the resection of selected sinonasal malignant tumors with orbit and skull base involvement with reasonable tumour clearance. The patient is being followed up regularly and 1 year post treatment there is no evidence of any residual or recurrent lesion.

Key words : chondrosarcoma , nasal cavity, malignant tumors of nose.

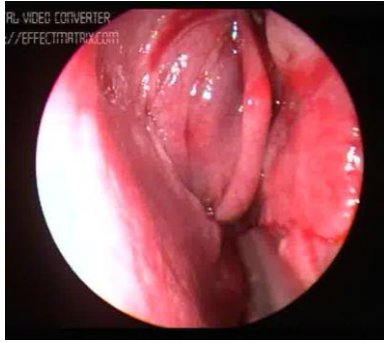
CASE REPORT

43 year old female presented with protrusion of Right eye ball - 1 month and loss of vision in Rt eye for 1 month, no history of loss of smell, no epistaxis, no headache, no nasal discharge, no ear and throat complaints. General examination afebrile, conscious, oriented. Vital signs—normal, CVS-S1S2, RS-NVBS, CNS—NFND. Local examination: Right eye- eccentric proptosis (forward, outward, laterally), vision – complete loss of vision with no perception of light, eyeball movement—restricted medially. Left eye- normal vision 6/6 eyeball movement –normal. Nose – deviated septum to right, no obvious mass lesion seen but there was a bulge in the middle meatus lateral wall.



PRE OPERATIVE PICTURE

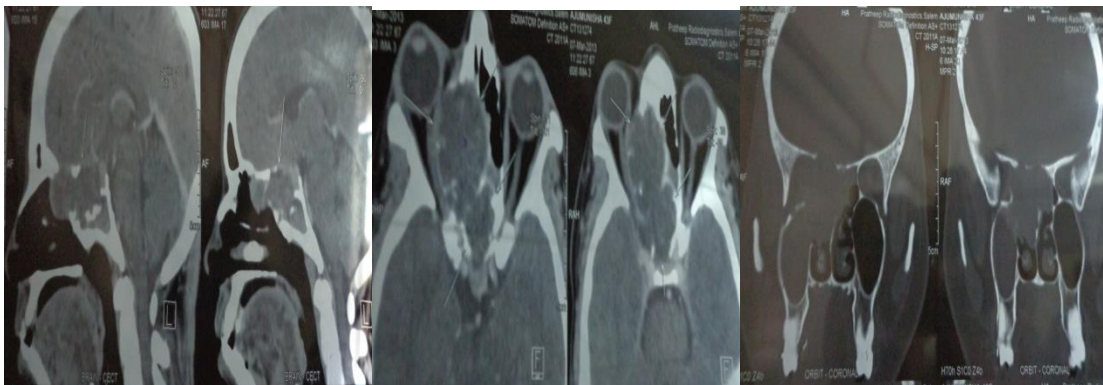
Diagnostic nasal endoscopy: Right side -smooth mass seen occupying the entire middle meatus. Deviated septum to right, Left side – OMC free, inferior turbinate hypertrophy



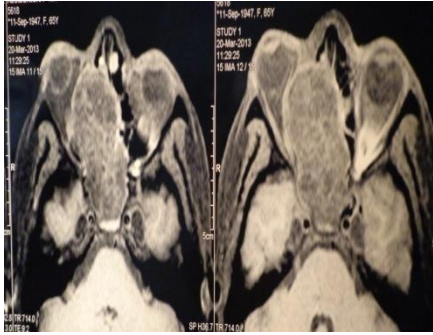
DIAGNOSTIC NASAL ENDOSCOPY PICTURE

Throat – Artificial denture upper jaw palate – no mass lesion. Postnasal exam-choana-free, no mass lesion. Ear- tympanic membrane intact both ears. No palpable neck nodes. All routine blood investigation –normal.

CT scan finding: solid mass filling the entire Rt ethmoidal sinuses, erosion into body of sphenoid sinus and extradural extension. Mass extending into medial part of Rt orbit compressing the medial rectus and proptosis. Frontal and maxillary sinusitis on the Rt side.



MRI finding: T2 hyperintense mass seen occupying entire Rt anterior and posterior ethmoid sinus with erosion of lamina papyracea and compressing the right optic nerve. Mass extending posteriorly to sphenoid sinus. Encasing Rt ICA by 180deg. Mass destructing nasal septum and involving opposite side posterior ethmoid sinus and sphenoid sinus. Mass showing extra dural involvement.



We were thinking of a sinonasal malignancy and planned for excision and histopathology examination. Radiologist opined as olfactory neuroblastoma/mucoepidermoid carcinoma/ giant cell tumour.

OPERATIVE DETAILS: Endoscopic sinus surgery under general anesthesia. Septal deviation was corrected. Smooth bulge was seen Rt middle meatal area. Right side uncinectomy and middle meatal antrostomy was done. Thick mucus aspirated from Rt maxillary antrum. Friable mass seen occupying entire Rt anterior and posterior ethmoid sinus extending into sphenoid sinus, medial wall of orbit and sphenoidal recess, same was removed in toto. Small defect in the anterior cranial fossa with csf leak was noted, the same was closed in layers with fat, fascia lata graft and mucosal free flap, surgical, gelfoam placed and nose packed with medicated ribbon gauze.



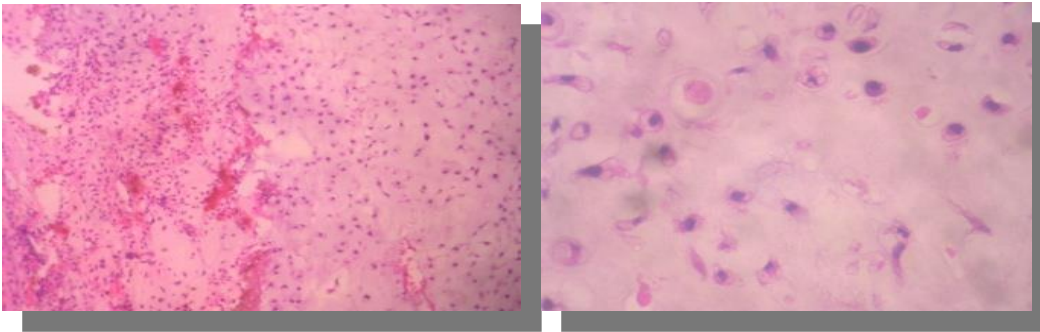
POST OPERATIVE PICTURE

Postoperatively patient vision improved to 1/6. After discussion with ophthalmologist patient was treated with i.v steroids followed by tapering dose of oral steroids, vision improved to 3/6.

HISTOPATHOLOGY



A) Macroscopic picture



1

2

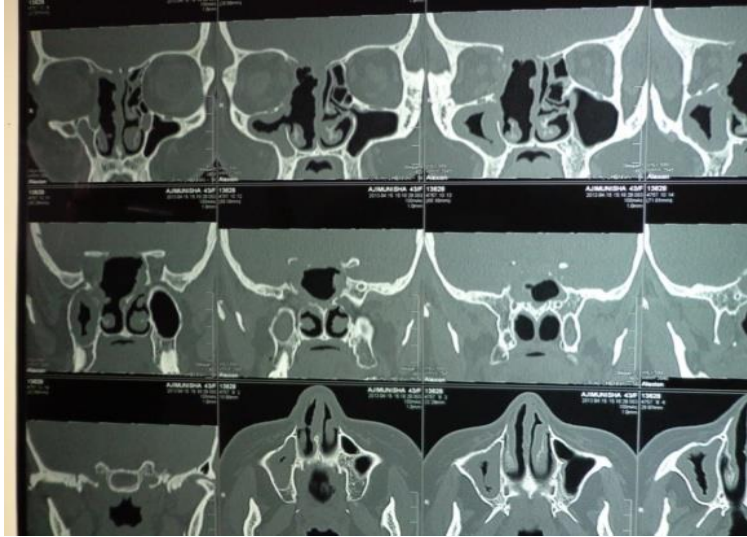
B) Microscopic picture

(1) Respiratory epithelium and lobules of chondrocytes seen.

(2) Chondrocytes showing atypical, binucleation and increased cellularity features were consistent with low grade Chondrosarcoma.

Diagnosis confirmed with immunohistochemistry, positive for s-100 & vimentin .

Postoperatively CT scan revealed mucosal thickening and no residual mass. Post operative radiotherapy was planned after discussion in tumor board.



POST OPERATIVE CT SCAN PICTURE

The patient completed radiotherapy and is under follow up. 1 year post treatment there is no evidence of any residual or recurrent lesion.

DISCUSSION

Originally identified by Lichtenstein and Bernstein in 1959¹. Chondrosarcomas take origin from the embryonic rest of the cartilaginous matrix of the cranium or primitive mesenchymal cells. Incidence of Chondrosarcoma comprises approximately 8% of primary bone tumors overall. Less than 10% of chondrosarcomas are found in the head and neck region². Younger population affected commonly. Less than 20% is seen arising in patients over 40 years of age³. Chondrosarcoma can be conventional, other sub types are clear cell, dedifferentiated, mesenchymal⁴.

Commonest site is larynx. Other sites are maxilla, mandible, and skull base². Chondrosarcoma of sinonasal tract is very rare. Chondrosarcoma of nasal cavity can spread to maxillary antrum, orbit, Nasal cavity, sphenoid sinus, anterior cranial fossa. Patient may present with symptoms like Nasal obstruction or discharge, epistaxis, Facial asymmetry, Headache, Restricted ocular movements diplopia, proptosis, Facial pain, dentition changes, Nasal/Aural Fullness⁵.

Chondrosarcomas are graded on a 1 to 3 based on the rate of mitoses, cellularity, and nuclear size. Immunohistochemistry for chondrosarcoma showed S-100 & Vimentin-positive, cytokeratin-negative, Epithelial membrane antigen-negative. This differentiated the tumor from chordoma^{2,6}. Differential diagnosis are chondroma, Meningioma, osteoma, osteosarcomas, fibrous lesions, chordoma. In CT- chondrosarcomas typically appear as a hypodense matrix with scattered small rings forming calcifications and bone erosion. MRI typically demonstrates an iso or low-intensity T1 imaging, high-intensity T2 imaging^{7,8,9,10}. Surgical options available are endoscopic sinus surgery, lateral rhinotomy, medial maxillectomy, maxillectomy, craniofacial resection⁵. Radiotherapy can be given in those cases which are surgically inaccessible, postoperative residue, or recurrent tumors. Radiotherapy using proton beam RT, fractional PBRT, have been used in residual growth and inoperable cases. Chemotherapy is not useful in treating chondrosarcoma^{11,12,13,14}.

Conclusion

Endoscopically mass was removed near toto. Treatment followed with post op RT. Patient showed improvement in vision postoperatively. This case report shows the feasibility of using endoscopic technique to aid the resection of selected malignant tumors of sinonasal region. The patient is being followed up regularly and one year post treatment there is no evidence of any residual or recurrent lesion.

References

- 1) Lichtenstein L, Bernstein D. Unusual benign and malignant chondroid tumors of bone. A survey of some mesenchymal cartilage tumors and malignant chondroblastic tumors, including a few multicentric ones, as well as many atypical benign chondroblastomas and chondromyxoid fibromas. *Cancer* 1959; 12: 1142-57
- 2) Atlas of head & neck pathology 2nd edition – Bruce M Wenig
- 3) Nakashima Y, Unni KK, Shives TC, Swee RG, Dahlin DC. Mesenchymal chondrosarcoma of bone and soft tissue. A review of 111 cases. *Cancer* 1986; 57: 2444-53.

- 4) Fletcher CDM, Unni KK, Mertens F, editors. World Health Organization Classification of Tumours. Pathology and Genetics. Tumours of Soft Tissue and Bone. Lyon, France: IARC Press; 2002. p. 252-254)
- 5) Stell and Maran 4th edition
- 6) Rosenberg AE, Brown GA, Bhan AK, Lee JM. Chondroid chordoma - a variant of chordoma. A morphologic and immunohistochemistry study. *Am J Clin Pathol* 1994;101:36-41.
- 7) Som PM, Curtin HD, eds. Head and neck imaging. 3rd ed. St. Louis, Mo: Mosby, 1996; 224–225.
- 8) Murphey MD, Walker EA, Wilson AJ, Kransdorf MJ, Temple HT, Gannon FH. From the archives of the AFIP: imaging of primary chondrosarcoma—radiologic-pathologic correlation. *RadioGraphics* 2003;23:1245–1278
- 9) De Beuckeleer LH, De Schepper AM, Ramon F. Magnetic resonance imaging of cartilaginous tumors: is it useful or necessary? *Skeletal Radiol* 1996;25:137–144
- 10) Aoki J, Sone S, Fujioka F, et al. MR of enchondroma and chondrosarcoma: rings and arcs of DTPA enhancement. *J Comput Assist Tomogr* 1991;15:1011–1016.
- 11) Proton radiation therapy for chordomas and chondrosarcomas of the skull base. *J Neurosurg* 1999;91:432-439.
- 12) Radiation therapy for chordoma and chondrosarcoma of the skull base and the cervical spine. Prognostic factors and patterns of failure. *Strahlenther Onkol* 2003;179:241-248.
- 13) Results of spot-scanning proton radiation therapy for chordoma and chondrosarcoma of the skull base: The Paul Scherrer Institut experience. *Int J Radiat Oncol Biol Phys* 2005;63:401-409.
- 14) Chondrosarcoma of skull base-report of six cases. Tadashi Morimoto MD, Tomio Sasaki MD, Kintomo Takakura MD Tsuyoshi Ishida MD