International Journal of Otorhinolaryngology and Head and Neck Surgery
Gavendra S et al. Int J Otorhinolaryngol Head Neck Surg. 2023 Feb;9(2):194-200
http://www.ijorl.com
pISSN 2454-5929 | eISSN 2454-5937

DOI: https://dx.doi.org/10.18203/issn.2454-5929.ijohns20230108

Case Report

Chondroblastoma- a rare sinonasal tumor: case report and review of literature

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Received: 22 August 2022
Revised: 10 December 2022
Accepted: 04 January 2023

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ABSTRACT

Chondroblastoma is a benign primary bone tumor that typically develops in the epiphyses of long bones. Chondroblastoma of the craniofacial skeleton is extremely rare, with most cases occurring in the squamosal portion of the temporal bone. In this report, we describe a case of chondroblastoma of the nasal cavity presenting with a left sided nasal mass which was excised with open approach. We review the literature on craniofacial chondroblastomas with particular emphasis on sinonasal lesions.

Keywords: Chondroblastoma, Aneurysmal bone cyst, Maxillofacial tumor

INTRODUCTION

Chondroblastoma (CB) is a rare tumour, accounting for less than 1% of all primary bone tumours. It is most commonly localised to the epiphysis or apophysis of the long bones, occurs in the first two decades of life, and is predominantly male.[1-3] It is rare in the head and neck region, with the temporal bone being the most common site of origin, and it occurs in the fourth and fifth decades of life, which is significantly older than the long bones CB.[3] Few cases have been reported from the craniofacial region. Craniofacial chondroblastoma is rare and accounts for 2-7% of all chondroblastomas.[13-15]

Eighty-three percent of patients with cranial chondroblastoma are older than 30 years, while 92% of patients with long bone chondroblastoma are younger than 30 years.[12] It is more common in the sphenoid and ethmoid bones and less common in the maxilla. The tumour is a locally aggressive, well-demarcated, expansive lesion. The tumour matrix has chondroblasts and areas of calcification.

CASE REPORT

A 22-year female presented with complaints of left sided painless disfiguring facial swelling associated with watering of eyes. On examination, there was a diffuse swelling over the left side of external nose and cheek fullness. The facial swelling was firm in consistency with no tenderness or local rise of temperature. There was presence of soft tissue mass in left nasal cavity reaching till vestibule pushing the nasal septum to right. Ophthalmic examination was normal. On oral examination, there was evidence of left sided palatal bulge. Ear examination was normal.

Computed tomography (CT) scan if paranasal sinuses show a well-defined heterogenous progressively enhancing soft tissue mass lesion with its epicentre in left nasal cavity causing its complete obliteration and expansion, showing irregular scattered hemorrhagic cysts.

Magnetic resonance imaging (MRI) scan of paranasal sinuses showed progressively enhancing soft tissue mass

lesion with its epicenter in left nasal cavity causing complete obliteration and expansion showing variegated signal intensity, multiple haemorrhagic foci and irregular scattered haemorrhagic cysts. The tumor showed severe rarefaction and erosion of medial wall of maxillary sinus medially, erosion of posterolateral wall of maxillary sinus laterally, obliteration of nasolacrimal duct anteriorly, abutting the lamina payracea and floor of the orbit with bulge into extraconal compartment (no intracranial extension) superiorly, complete destruction of hard palate inferiorly and no extension into pterygopalatine or infratemporal fossa posteriorly.

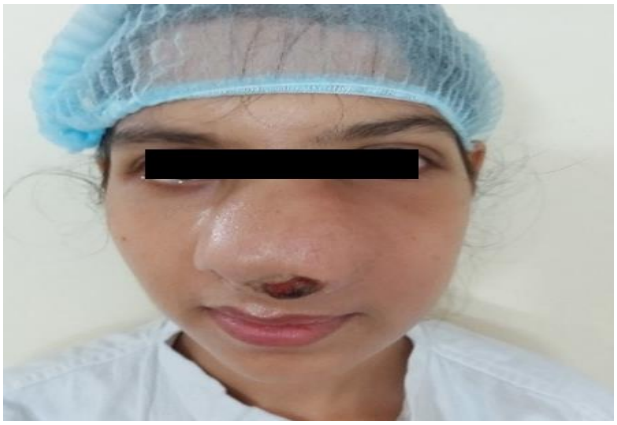

Figure 1: Left sided nasal mass.

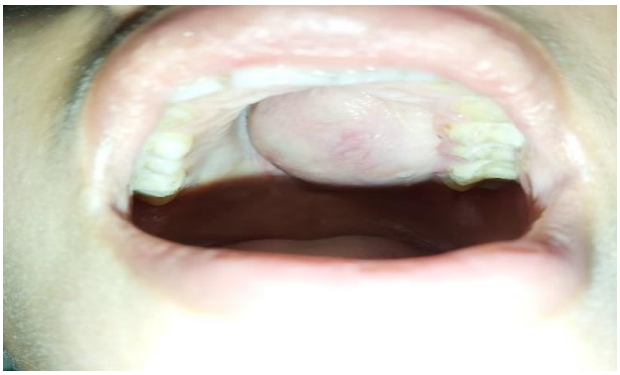

Figure 2: Mass causing a bulge in the palate.

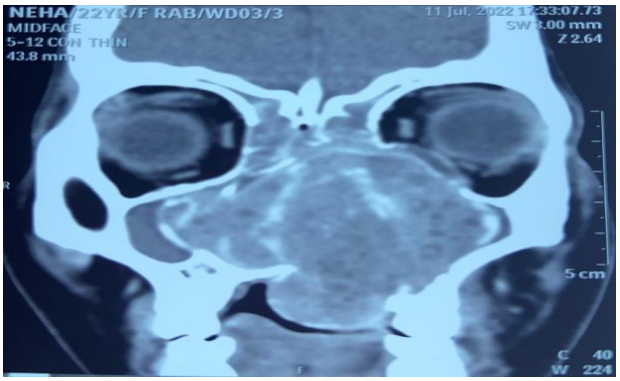

Figure 3: MRI scan of paranasal sinus (coronal view) showing soft tissue mass lesion.

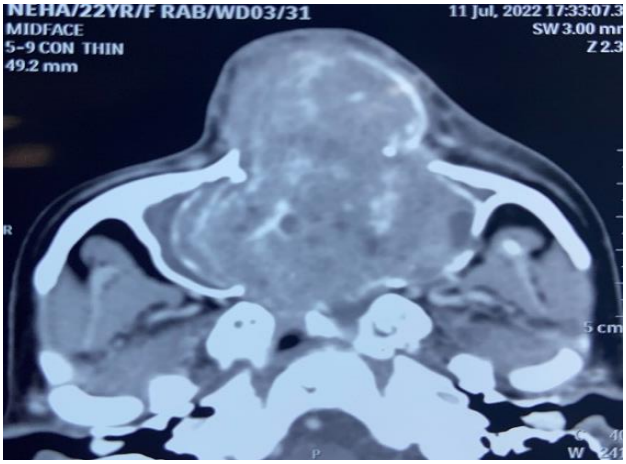

Figure 4: MRI (axial) showing maxillary sinus.

The patient had undergone a biopsy of the lesion at a private hospital, which showed diffuse proliferation of plump to elongated spindle cells intermixed with giant osteoclastic cells and no deposition of malignant osteoid giving it an impression of giant cell tumor.

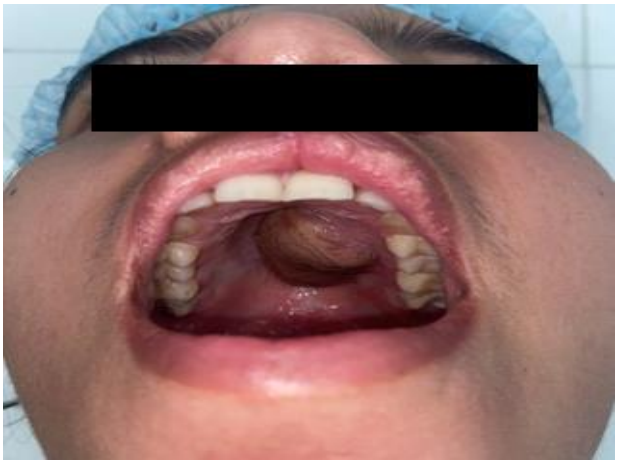

Figure 5: Free ALT flap reconstruction of the post-excision defect.

The patient was then scheduled for surgery, and the mass was removed in Toto via Weber-Fergusson's approach. The specimen was sent for histopathologic examination, which revealed osteogenic sarcoma. Keeping in view the rarity of the incidence, slides were reviewed which then showed it to be chondroblastoma with aneurysmal bone cyst changes. Patient underwent plastic reconstruction using free anterolateral thigh flap for the left partial maxillectomy defect.

Keeping in view the rarity of chondroblastoma in maxillofacial area we have reviewed the data available so far in literature on chondroblastoma involving maxillofacial areas and have compiled it in the table below (Table 1).

Table 1: Review of the data available so far in literature on chondroblastoma involving maxillofacial areas.

Author (year)	Age (years)/ sex	Complaints	CT/MRI s/o growth	Sinuses involved	HPER	Surgery	Outcome and prognosis	Follow up
Martinez-Madrigal et al (1991)	14/F	Left nasal obstruction, epistaxis, exophthalmous, diplopia	Partially calcified tumor	Maxilla and ethmoid sinus	Chondroblastoma. Less cellular areas exhibited an interstitial matrix with chondroid differentiation. Scattered calcifications and focal areas of ossification	Excision	No record	No record
Cho et al (2017)	13/M	Right-sided visual disturbance	Localized multilobulated multiseptated cystic lesion	Sphenoid, ethmoid and frontal sinuses	Chondroblastoma with secondary aneurysmal bone cyst components	Endoscopic excision	Good	No local recurrence
Tang (2020)	40/F	Repeated dizziness and intermittent headache and	Oval lesion with a thick sclerotic margin	Sphenoid sinus	Chondroblastoma	Endoscopic excision	No record	No record
Sivaraju (2016)	25/f	Impairment of vision in both eyes	MRI- skull base tumor	Ethmoid, sphenoid sinus	Chondroblastoma	Open craniotomy and transbasal approach excision (thin rim of the tumor close to cavernous ICAs left behind)	Referred to oncologist for radiotherapy	No record
Wang et al (2016)	5/ MCH	Nasal obstruction, proptosis and decreased vision	CT-soft tissue density lesion with multiple cystic cavities with intraorbital extension	Sphenoid and ethmois sinus	Chondroblastoma with secondary aneurysmal bone cyst	Endoscopic resection	Postoperative MRI s/o no residual	No recurrence

Continued.

Author (year)	Age (years)/ sex	Complaints	CT/MRI s/o growth	Sinuses involved	HPER	Surgery	Outcome and prognosis	Follow up
Madhu et al (2005)	18/M	Palatal swelling with bulging of eye	CECT s/o irregular heterogeneous mass lesion (orbital extension)	Maxilla	Chondroblastoma	No record	No record	No record
Elkhatib et al (2018)	19/M	Vision disturbance, headache	CT- large expansile mass (intracranial extension)	Sphenoid	Chondroblastoma Osteoclastic giant cells scattered throughout the tissue	Endoscopic resection with left optic nerve decompression	Vision regained within two weeks	Residual tumor follow-up with imaging
Arzoo et al (2018)	58/F	Obstructive sleep apnoea symptoms	CT-large nasal mass with dehiscence and erosions in the cribriform plate	Ethmoid	Chondroblastoma with atypical features	Endoscopic resection	Postoperative MRI s/o no residual mass	Six months post op, complete remission
Al-Dewachi et al (1980)	13/f	Painless mass for 4 months	CT s/o mass	Maxilla	Chondroblastoma	Surgical resection	Good	8 mo. post op no evidence of disease
Badia et al (1985)	17/f	NR	CT s/o mass	Maxilla	Chondroblastoma	Surgical resection	Good	6 mo. post op no evidence of disease
Burgin et al (2010)	30/f	Headache	CT s/o mass	Sphenoid sinus	Chondroblastoma	Endoscopic resection	No record	Tumor left on ICA on 6 monthly follow up
Miyake et al (1984)	1.7/f	Seizure	CT s/o mass	Frontal region	Chondroblastoma	Surgical resection	No record	No record

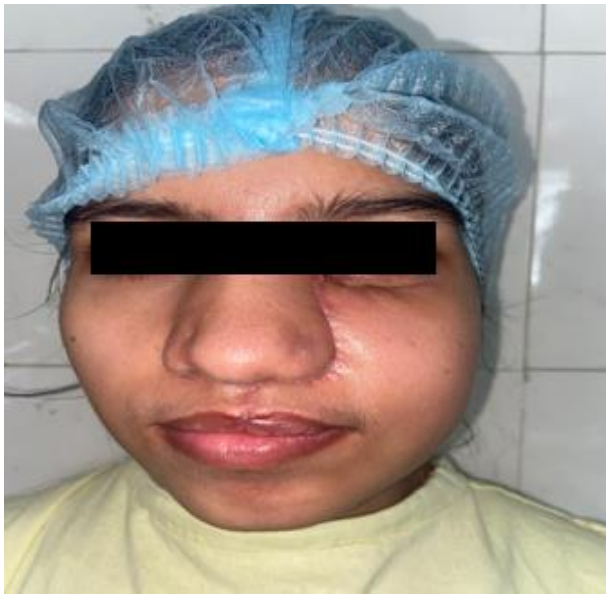

Figure 6: Post operative facial scar.

DISCUSSION

The phenomenon was first described by Codman in 1931 as an "epiphyseal chondromatous large cell tumour of the proximal humerus." The term chondroblastoma was first used by Jaffe and Lichtenstein in 1942.[8] It is a benign, cartilage-producing tumour that most commonly occurs in the epiphyses of growing patients. Typical locations are the knee, rib, and pelvis. Most patients are 10-15 years old, with males predominating. In the temporal bone, chondroblastoma is very rare. Patients with cranial and temporal bone disease are older (40-50 years). It is a locally aggressive tumour.[9] Although locally destructive and prone to recurrence, it is considered a benign tumour which rarely metastasises.[25]

From a histopathological point of view, chondroblastoma is a tumour of immature cartilage with sheetlike proliferation of small to intermediate-sized round polygonal cells. The cytoplasm is eosinophilic, although focally, clear cell change can be seen. The nucleus is centrally placed and relatively large (15–20 lm), and often a central, longitudinal nuclear groove (‘‘coffee bean’’ nucleus) can be seen. Nucleoli are small. Cellular atypia with enlarged, irregular, and sometimes hyperchromatic nuclei may be present, especially in tumors located in the skull and facial bones. Mitoses are occasionally found, although they are not numerous, with an average count of 1 to 3 mitotic figures per 10 high-power fields. Atypical mitotic figures should not be seen, and if present, tend to exclude chondroblastoma from the differential diagnosis. It is a biological intermediate between chondroma and chondrosarcoma. Its biological behaviour is among the low-grade tumours.[10] Hematoxylin-eosin staining is characterised by cartilage with immature cells. The diagnosis of chondroblastoma is inevitably complemented by positivity of CD-68, vimentin, and S-100 protein.[7] Grossly, chondroblastoma is sharply separated from the adjacent bone and contains a mixture of soft, friable, grey-yellow material and hemorrhage. Small calcifications provide a gritty and chalky cut surface. Occasionally, areas of rubbery blue-grey chondroid matrix are seen. Necrosis and hemorrhagic cystic cavities (secondary aneurysmal bone cyst formation).

In addition to the above, variable numbers of multinucleated giant cells are often present as are foci of hemosiderin deposition. The latter occurs more commonly in the tumors located in the skull and facial bones.[5]

Matrix formation must be seen to confirm a diagnosis of chondroblastoma. Bluish or purple granular calcium deposits are seen in approximately one-third of cases, most commonly in long bone tumors. The calcifications may be seen in the cytoplasm or stroma, where they demonstrate a delicate pericellular lacelike or “chicken-wire” appearance. Secondary aneurysmal bone cyst formation is also commonly encountered, especially in the tumors of the hands or feet, where more than half of the cases show this feature. Although it is usually limited to microscopic foci, ABC formation may sometimes be so dramatic that the underlying Chondroblastoma may be overlooked.

Other features such as tumor necrosis, vascular invasion, cortical breakthrough, and soft issue invasion can also be present in a small percentage of cases. The tumor necrosis is usually composed of bland ghost cells without any inflammation.

Chondroblastoma must be differentiated from other giant cell tumors. The osteoclastic-like giant cells seen in chondroblastomas can have striking histological similarities to giant cell reparative granulomas, aneurysmal bone cysts, giant cell tumors, and chondromyxoid fibromas. Positive staining for S100 is most commonly used to help differentiate from other giant cell tumors, since this protein is expressed on human chondrocytes and related to chondroid tissue formation.[22,41]

Chondroblastoma originated in paranasal sinus is hardly detected in the early stage for the lack of positive clinical presentations. Chondroblastoma on CT imaging is characterized by oval mass with a sclerotic rim and scattered calcification inside the lesion.[2,3] In MRI imaging, chondroblastoma manifests itself as an expansive heterogeneous hypermetabolic mass with 2 distinct components – solid with predominantly low signal in T1 and T2 sequences and multilocular cystic with T1 and T2 elongation and presence of fluid-fluid signals at T2 imaging. Post-contrast saturation is present in the solid component and septum of the cystic component. Possible high signalling in T2 imaging depends on potential haemorrhage in the tumour mass. MRI better defines the infiltration of the dura, cerebral, intracranial, and soft tissue infiltration. There was no observed increase in the signal strength for DWI.[11,12]

The treatment of choice is complete multidisciplinary tumour removal depending on the extent.[9] CBT is intermediate and locally aggressive and rarely metastasizing, radical resection and regular follow-up is recommended to reduce and prevent local recurrence and distant metastasis of CB. Simple curettage is not adequate, as it has been associated with a recurrence rate of 55%.[20,21] Kurokawa et al and Moon et al found no evidence of recurrence 5 and 9 years after following complete en bloc resection of temporal bone chondroblastoma.[17,20] However, some authors have reported recurrence rates approaching 20%, even after complete tumor removal.[22-24] Therefore, surveillance imaging following surgical resection is prudent for early diagnosis of tumor recurrence. In surgical resection, a conservative approach is recommended with a reduction of postoperative morbidity.[8] 80–90% of chondroblastomas are treated with surgery. Local recurrence occurs between 14–18% and is often within two years.

There is insufficient data to predict tumor behavior in craniofacial chondroblastoma. Malignant degeneration to chondrosarcoma is likely rare but has been previously reported and some authors have suggested that lesions with an intratumoral aneurysmal bone cyst may behave more aggressively.[26,27] Radiation therapy is a treatment option for poor surgical candidates, or patients with recurrent or unresectable disease.[18,26] Radiation is not recommended after complete excision, due to the possibility of radiation-induced chondrosarcoma.[18] Metastatic workup is not recommended since metastatic craniofacial chondroblastoma has never been reported (in contrast, pelvic chondroblastoma may spread to the abdomen and lung).[19,28] At present, there is no role for chemotherapy in the management of chondroblastomas.[26]

CONCLUSION

Here, we report this case of chondroblastoma with aneurysmal bone cyst changes of maxilla. It is a rare tumor in maxillofacial region where mandible is the commonest site to get involved and maxilla being a rare site. The case was challenging in terms of diagnosis as well as treatment but showed good results and no recurrence with complete surgical resection and postoperative radiotherapy. The report emphasises on the importance of multidisciplinary approach for this challenging and rare tumor involving surgeons, radiologists, pathologists, plastic surgeons and radiation oncologists.

ACKNOWLEDGEMENTS

Authors would like to acknowledge the support of radiologists, pathologists, plastic surgeons and radiation oncologists for being on board in managing this challenging case.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Gavendra S, Kulkarni P, Shah K, Bradoo R. Chondroblastoma- a rare sinonasal tumor: case report and review of literature. Int J Otorhinolaryngol Head Neck Surg 2023;9:194-200.