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CASE REPORT

RECURRENT BASAL CELL AMELOBLASTOMA OF THE MAXILLA: A rare histological variant and systematic review of literature

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ABSTRACT:

Ameloblastoma is a benign but locally invasive tumour of odontogenic epithelial origin with several histological variants. Basal cell ameloblastoma is the rarest histological subtype with only a few cases reported till date. It shows remarkable similarity to basaloid squamous cell carcinoma (BSCC), cutaneous basal cell carcinoma (BCC) and solid-type adenoid cystic carcinoma (ACC). This report describes an interesting case of basal cell ameloblastoma of the left maxilla in a 37 year old man. He had undergone enucleation of a maxillary lesion 2 years back without a pre-operative tissue diagnosis, which was later established to be an ameloblastoma. The tumor recurred after 6 months and was managed by a standard partial (infrastructure) maxillectomy. We review the literature on this rare entity with emphasis on the appropriate treatment modality.

KEYWORDS- Basal cell ameloblastoma, maxilla, weber fergusson incision, resection

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INTRODUCTION:

Odontogenic tumors comprise a diverse group of lesions with varied clinical behaviour and histological patterns. Ameloblastoma, though the most common odontogenic tumor, is a relatively rare tumor occurring in the jaws representing approximately one percent of oral tumours. The incidence of maxillary ameloblastomas has been variably reported in literature

although the generally accepted figure is 20%.[1]It is a benign but locally invasive tumor with the maxillary ameloblastoma being more aggressive than its mandibular counterpart.

Among the several histopathological subtypes of the tumor, basal cell variant is the least common type with only 13 cases reported in the English literature so far. Only 4 of them have been reported to occur in the maxilla.The scarce literature on this variant does not allow for an accurate prediction of its clinicopathological behaviour. Although, surgery is considered the primary treatment, there is a lack of consensus on the appropriate method of surgical removal. Most authors, however, agree that wide resection with appropriate margins should be the treatment of choice for this tumor.

In this paper, we describe a case of recurrent basal cell ameloblastoma of the maxilla and discuss the relevant literature on this rare entity.

CASE DESCRIPTION:

A 37 year old male patient presented to our OPD with complaints of palatal swelling and left sided epistaxis for around 2 years. He had been evaluated at an outside centre 2 years back where after a physical examination, a CT scan of the paranasal sinuses was done. The CT films, however, were not available for review. Neither fine needle aspiration cytology nor an incisional biopsy was done to obtain a histological diagnosis before the patient was taken up for surgery. Operative notes revealed that the patient underwent a combined endoscopic and Caldwell-Luc approach. A cystic tumor was found in the maxillary sinus which was eroding the anterior and inferior walls. Enucleation of the mass was done and it was delivered from the anterior antral window. Post-operatively histopathological examination of the specimen had revealed it to be an ameloblastoma. No further treatment was done and the patient was asymptomatic for a period of 6 months after which he again developed similar symptoms of epistaxis and palatal swelling.

The patient was then referred to our hospital, a tertiary care referral centre. On examination, the palatal bulge was found to extend from 1st premolar to the retromolar trigone. Medially, it extended upto midline. The bulge was covered by normal palatal mucosa; it was hard and non-tender on palpation (Figure 1). Anterior rhinoscopy revealed a reddish irregularity over the left inferior turbinate. Synechiae were present between the nasal septum and lateral nasal wall in the region of inferior and middle turbinate.

A fresh CT scan of the nose and paranasal sinuses revealed a 4X5X5 cm sized soft tissue density lesion involving the left upper alveolar arch in the premolar and molar region, and extending into the maxillary antrum. The tumor extended medially into the left nasal cavity and caused erosion of the inferior turbinate. Posteriorly, it caused erosion of the posterior wall of maxilla with extension into the masticator space and the pterygoid fossa (Figure 2-4). Slides from the previous surgery were reviewed at our centre and the diagnosis of ameloblastoma was confirmed.

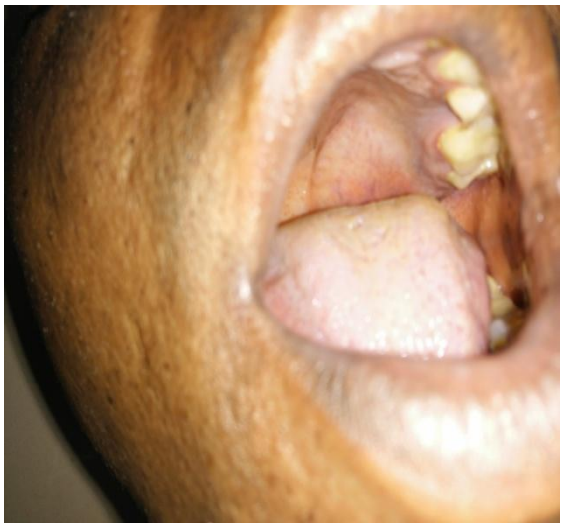

Figure 1 showing the palatal bulge on the left side extending from premolar to nasal cavity and upperalveolar arch

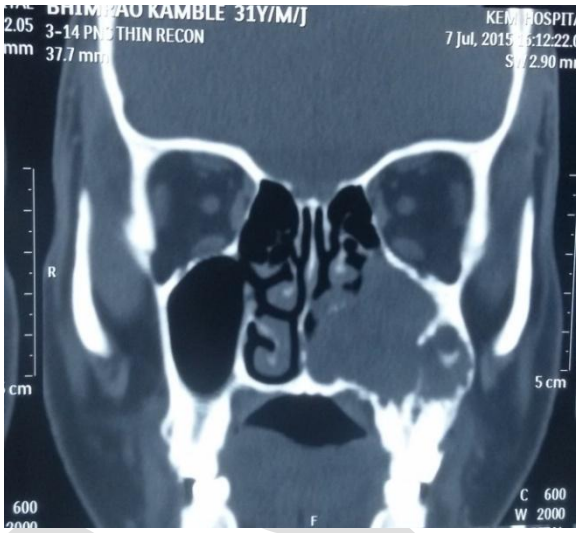

Figure 2 shows the soft tissue lesion in maxillary antrum extending into retromolar trigone

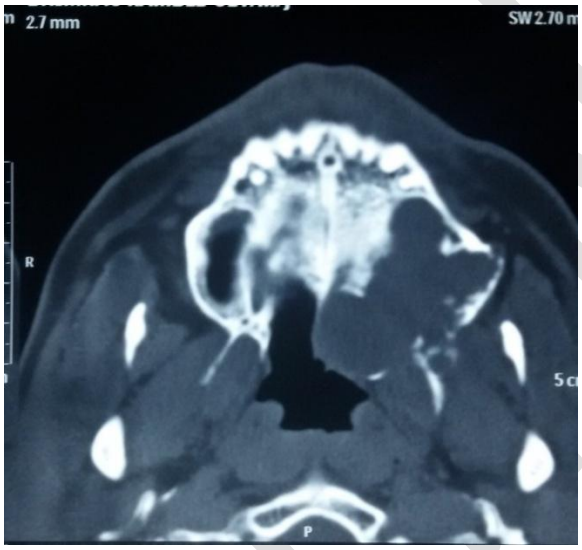

Figure 3 showing the involvement of hard palate and upper alveolar arch Erosion of pterygoidplates and posterior wall of maxilla

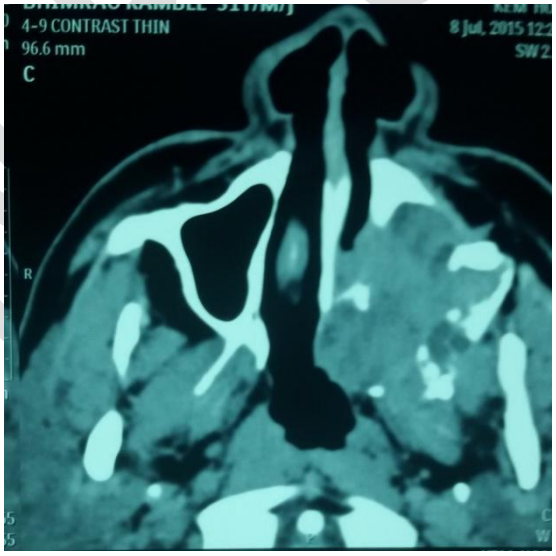

Figure 4. Anterior wall of maxilla is absent (previous surgery).

The patient was taken up for left partial (infrastructure) maxillectomy via lateral rhinotomy approach to ensure a wide excision of the mass (Figure 5-7). Because of the involvement of pterygoid fossa by the tumor, the root of pterygoid process was drilled by a cutting burr. Oro-nasal separation was achieved with an obturator (Figure 8). Histopathological examination of the resected specimen revealed the tumor to be a basal cell variant of ameloblastoma. All the

resected margins were negative for involvement. Post-operatively patient achieved a normal oral feeding and a good voice. No recurrence has been observed at 6 months of follow-up.

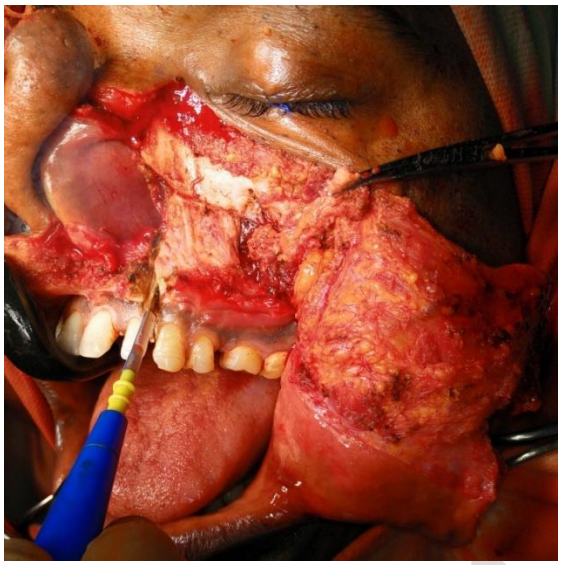

Figure 5. Upper cheek flap elevated after Weber Fergusion incision. Anterior bony cuts made.

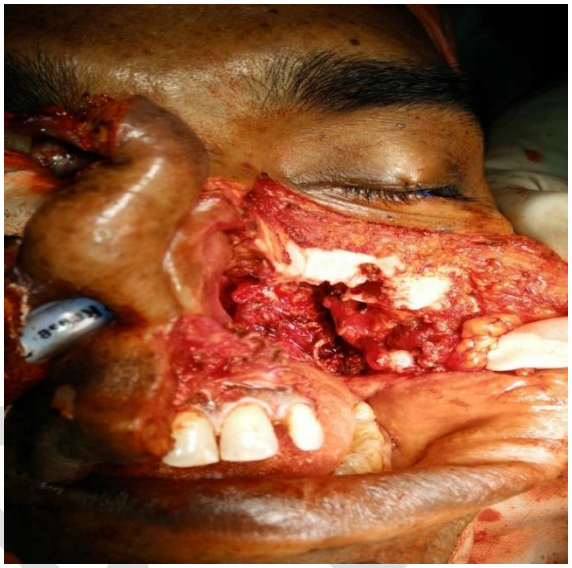

Figure 6. Defect after removal of the maxillectomy specimen.

Figure 7 shows the specimen of left partial maxillectomy with en-bloc excision of tumor.

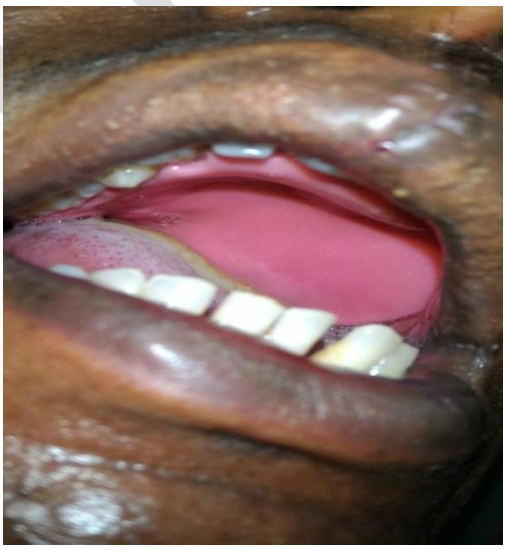

Figure 8 shows the obturator in place to to achieve oro-nasal separation.

DISCUSSION:

Ameloblastoma, previously known as admantinoma, is a benign but locally aggressive tumor of the jaw that tends to arise from the odontogenic epithelium. The first detailed description of this

tumor was published by Falkson in 1879, but the term 'ameloblastoma' was coined by Churchill in 1933.[2]Although, it is the most common odontogenic tumor, it represents only around one per cent of oral tumors. Around 80 per cent of ameloblastomas are reported to occur in the mandible.[1]The incidence of maxillary ameloblastomas has been variably reported in the literature. Although, it is generally accepted that only 20 per cent of ameloblastomas occur in the maxilla[1], some reports indicate an incidence as low as one per cent, and of those, 47 per cent occur in the molar region, 15 per cent in the antrum and the floor of the nose, 9 per cent in the premolar areas, 9 per cent in the canine regions and 2 per cent in the palate.[1, 3]Iordanidis reported an incidence of 5 percent in his clinical practice.[4]

The classification of the ameloblastoma in the past was poorly defined; the current concept is to classify ameloblastoma into solid or multicystic, unicystic, and peripheral types. This classification has direct bearing on the pathological behaviour of these variants. Solid or multicystic variants are locally aggressive, and recur if inadequately excised.[5]Unicystic ameloblastoma was identified as distinct entity with less aggressive behaviour.[6]

The most common clinical symptom of the maxillary ameloblastoma is a painless swelling of the involved part of the jaw. Other symptoms that may occur are nasal obstruction, epistaxis, sinusitis or symptoms due to extension of disease into orbit. This was also the case in our patient who presented with the chief complaints of palatal mass and epistaxis.

Ameloblastomas, in general, are considered benign but locally invasive neoplasms. However, maxillary ameloblastoma is a relatively more aggressive and persistent lesion, presumably because the thin and fragile bone of the maxilla, unlike the thick continuing cortical plates of the mandible, allows a relatively unimpeded spread of the tumour to the surrounding structures, such as the maxillary sinus, nasal cavity, orbit and occasionally the cranial base.[7, 8] The maxillary bone has a richer blood supply compared to the mandible which could also contribute to the accelerated spread of this tumor in this location.[9] This was also evident in our case where the tumor had caused erosion of the posterior wall of maxilla to extend into the masticator space. Maxillary ameloblastoma is more cellular than the mandibular counterpart and is further characterized by less distinct peripheral palisading, fewer columnar cells, frequent focal acanthomatous metaplasia and more cellular stroma.[9] While planning the treatment of ameloblastoma, it is important to understand the growth characteristics and removing the full extension of tumor, including the surrounding tissue.

Treatment of ameloblastoma is primarily surgical. The surgical options range from curettage, enucleation and cryosurgery to wide local excision, which may require segmental resection in the mandible, and partial maxillectomy in the maxilla. There seems to be a lack of consensus regarding the most appropriate method of surgical removal of ameloblastomas. Proponents of conservative approach argue that ameloblastoma though locally invasive is essentially a benign tumor. Many authors have recommended enucleation with preservation of periosteum which is important for bone regeneration especially in children.[10] However, proponents of radical

approach are of the considered view that conservative surgical options such as curettage and enucleation result in unacceptably high recurrence rates; the recurrence rate up to 55–90% have been reported in the literature.[11] Curettage is followed by local recurrence in 90% of mandibular and all maxillary ameloblastomas because of insufficient removal of tumors.[12]Sehdev et al reported recurrence after the conservative approach (curettage) in more than 90% of 92 ameloblastomas.[13] Ameloblastoma has a persistent and slow growth, spreading into marrow spaces with pseudopods without concomitant resorption of the trabecular bone. As a result, the margins of the tumour are not clearly evident radiographically or grossly during operation, and the lesion frequently recurs after inadequate surgical removal, showing a locally malignant pattern.[14]This fact is also amply evident in our patient who presented with a recurrence after a conservative approach with enucleation.

It, thus, appears that the best surgical method for the treatment of a maxillary ameloblastoma is a limited or wide excision of the tumour[15] with a 10-15 mm margin of normal bone, if available.[14] Thus, considering the above and the fact that our patient had already presented to us with a recurrence, we chose to perform a standard partial (infrastructure) maxillectomy in this case along with drilling of the pterygoid base to ensure a wide en-bloc excision of the tumor. Ameloblastoma is generally considered to be a radioresistant tumor although there is evidence to suggest a palliative role in advanced cases to diminish the volume of tumor. It may reduce the risk of progression and result in long-term local control in such incompletely resectable tumors.[16] A case of an advanced maxillary ameloblastoma is reported to have been successfully treated with a combination of chemotherapy and radiotherapy.[17] At present, however, there seems to be no evidence to suggest that post-operative radiotherapy may offer any advantage as an adjuvant modality in terms of loco-regional control or overall survival in a patient with complete excision of the tumor with negative margins.

Histologic variants of the tumour include follicular, plexiform, acanthomatous, granular cell, desmoplastic and basal cell types. Histopathological examination of the tumor in our patient revealed it to be a basal cell variant of ameloblastoma. Basal cell ameloblastoma (BCA) is the least common variant among all and accounts for only 2.02% of the histologic types.[18]We searched for cases of BCA reported in literature through PubMed and Google and found nine published articles comprising 13 cases of BCA (Table 1).[4, 19-26]Nine of them occurred in mandible and only four in the maxilla. It seems to be equally distributed between the two genders. This variant shows a predilection for mandible as is the case for ameloblastomas in general. Our case appears to be the first reported recurrence of basal cell ameloblastoma of the maxilla.

Table 1.Reported cases of basal cell variant of ameloblastoma till date.

S.no.	Author	Number of cases reported	Age/Sex	Site of tumor
1.	Kameyama Y et al[19]	2	3rd decade 7th decade	Maxilla Mandible
2.	Iordanidis et al[4]	1	63/F	Maxilla
3.	HirotaM et al[20]	1	17/F	Maxilla
4.	Kehinde E Adebiyi et al[21]	2	4th decade/M 4th decade/M	Mandible Mandible
5.	FatemaSaify et al.[22]	1	12/M	Mandible
6.	Giraddi et al[23]	3	55/M 17/F 38/F	Mandible Mandible Maxilla
7.	Shakya et al.[24]	1	50/F	Mandible
8.	Pendyala et al[25]	1	72/M	Mandible
9.	Sridhar et al[26]	1	27/M	Mandible

The microscopic features of basal cell ameloblastoma are similar to those of several malignant tumors, including basaloid squamous cell carcinoma (BSCC), cutaneous basal cell carcinoma (BCC) and solid-type adenoid cystic carcinoma (ACC).[26] The number of cases of basal cell variant of ameloblastoma reported in literature is too few to objectively predict its clinicopathological behaviour in terms of local control and prognosis.In all the reported cases, a partial maxillectomy was done and in none of them was recurrence reported. Our patient had presented with a recurrence of ameloblastoma after a surgery done outside in the form of an enucleation procedure. This tends to suggest that the basal cell ameloblastoma is an aggressive variant which requires an en-bloc resection with wide margins to avoid a local recurrence. This is evidenced by the fact that after performing a standard partial maxillectomy, no further recurrence has been noted in our patient. Long term follow-up would be required for this patient as we do not yet have data on long term local control or late recurrences. With this case report, we hope to add to the scarce literature on this rare entity.

CONCLUSION:

Basal cell ameloblastoma is a rare histological variant with a predilection to occur in the mandible. Maxillary tumors are very rare. Pre-operative diagnosis should be established by a FNAC or incisional biopsy. We do not recommend conservative approaches like enucleation or curettage for these tumors. En-bloc excision of the tumor with wide margins is the appropriate surgical modality that can be achieved by a partial maxillectomy in most cases. Long term follow-up is recommended in the absence of sufficient data.

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