Case Report

Ameloblastic carcinoma of maxilla – case report and review

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Abstract

Background: Ameloblastic carcinoma is a rare malignant odontogenic tumor that occurs more commonly in the mandible than in maxilla. **Case Report**: Herein, we report a 70 year old male presenting with right maxillary swelling and intraoral ulcer. CT scan findings were suggestive of Maxillary Carcinoma. Histopathological examination of incisional biopsy was suggestive of Ameloblastic carcinoma, hence advised excision. Histopathological examination of the hemimaxillectomy specimen revealed features of Ameloblastic Carcinoma. **Conclusion:** Ameloblastic carcinoma is associated with high rates of recurrence and metastasis. So, in addition to surgical treatment, long term follow up to monitor potential pulmonary involvement and lymph node metastasis is required. Hence, **i**t is of utmost importance to diagnose Ameloblastic carcinoma.

Keywords: Ameloblastic Carcinoma, Maxilla.

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INTRODUCTION

Primary carcinoma of Jaw bones is rare including odontogenic origin and also entrapped salivary gland epithelium. Ameloblastic carcinoma may arise de novo or in preexisting ameloblastoma or odontogenic cyst. The current concept accepted widely states that ameloblastoma in which there is histological evidence of malignancy in the primary tumor or recurrent tumor regardless of whether it has metastasized can be termed as ameloblastic carcinoma.[1] Ameloblastic carcinoma is a rare malignant odontogenic tumor, particularly very rare in maxilla. In an evidence based literature review of ameloblastic carcinomas of maxilla over the last 60 years only 26 cases were found with mean age being 54.4 years [2] male to female ratio being 2.7:1 and predominant complaint was swelling particularly in left maxilla. The maxillary mandibular ratio of ameloblastoma is 5:1, most commonly occurring in the mandibular molar region [3,4]. More than 50% recurrence occurs within first 5 years after primary surgery.[3] With respect to malignancy, we should differentiate between malignant ameloblastoma and ameloblastic carcinoma. Ameloblastic carcinoma reveals malignant histopathological features independent of presence of metastasis [5] while malignant ameloblastomas metastasize as well differentiated benign cells, [6] particular to the lungs [7,8]

CASE REPORT

In ENT outpatient department, 70 years old male presented with swelling of the right cheek involving middle one third of right side of face in the right maxillary region of 3 months duration. Swelling had gradually increased in size and was associated with pain, difficulty in swallowing. On general examination patient was moderately nourished. Extra oral examination revealed an irregular swelling of size 6x5cm over the right zygomatic region. Swelling was hard and fixed; skin over the swelling was normal. Intraoral examination revealed a swelling of size 4x4cm involving right sided of hard palate, alveolar margin of right maxilla and fullness of right gingivolabial sulcus. No palpable lymphnodes were identified on neck examination. Hence a provisional

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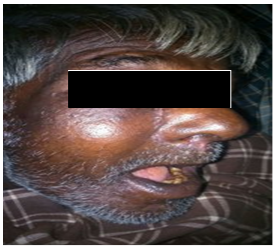
diagnosis of squamous cell carcinoma was given. Computerised tomography CT scan revealed enlargement of right maxillary sinus and complete filling of the soft tissue with dense heterogenous opacities of the mass lesion causing erosion of medial, posterior and anterior walls. Mass extended medially into nasal cavity upto septum, posteriorly into infra termporal fossa and anteriorly extended upto skin - features suggestive of carcinoma maxilla. Left Maxilla was normal. Incisional Biopsy was taken from the lesion and was subjected for histopathological examination. Microscopically the incised tissue exhibited marked cellular pleomorphism, hyperchromatism, loss of the nuclear/nucleolus and nucleus/cytoplasm and areas of necrosis confirming the malignant nature of the epithelial origin**.** Some areas exhibited follicular configuration and also revealed basal cells resembling ameloblasts, occasionally arranged in palisades. The most central cells were arranged more loosely resembling the stellate reticulum of the enamel organ which is unusual in a typical squamous cell carcinoma. Hence diagnosis of ameloblastic carcinoma was made. Advised excision. Under GA tumor was found occupying right maxillary cavity. Right maxillectomy was done and sent for histopathological examination. Gross examination of specimen showed a skin lined soft tissue with teeth measuring 6x5x3cm with ulceroproliferative growth. Cut section showed greyish white friable growth involving whole region. Multiple sections were taken and microscopic examination of the growth showed tumor composed of bony spicules with sheets of pleomorphic cells with hyperchromatic nuclei and increased mitosis admixed with comedo necrosis at few foci and extensive areas of haemorrhage with perineural invasion. Islands individually resembling enamel organ type of tissue with palisading of peripheral tall columnar cells and change in polarity of nucleus was present. The histological picture was consistent with follicular ameloblastoma, with marked malignant changes. So, histopathological diagnosis of ameloblastic carcinoma was made.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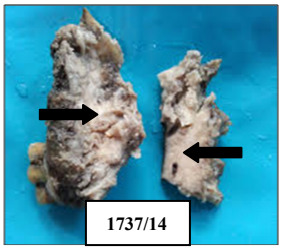

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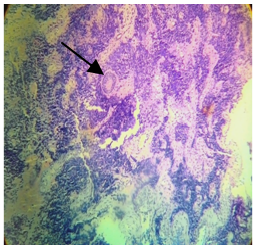
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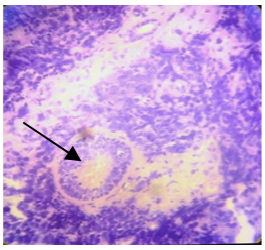
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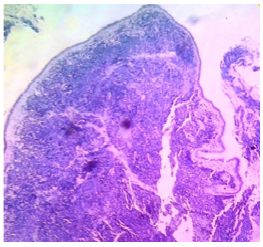
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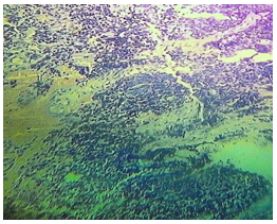
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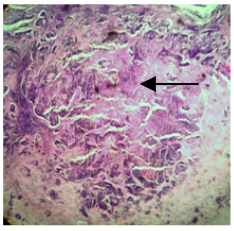
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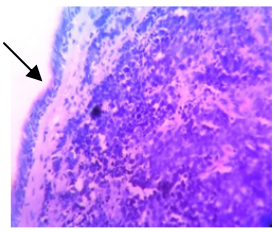
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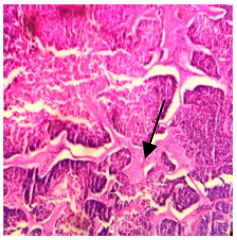
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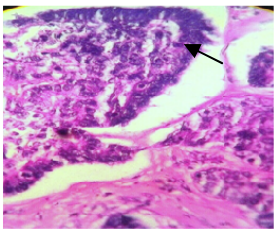
Figure 10

Legend

Figure 1: Patient with right maxillary swelling.
Figure 2: Gross examination of right hemimaxillectomy specimen revealing an irregular grey white growth.
Figure 3: Ameloblastic differentiation (arrow) seen with irregularly arranged pleomorphic epithelial cells in nests and cords. (10X)
Figure 4: Ameloblastic differentiation (arrow) with pleomorphic epithelial cells (40X)
Figure 5: Pleomorphism of irregularly arranged epithelial cells in nests and cords.(10X)
Figure 6: Pleomorphism of irregularly arranged epithelial cells in nests and cords.(40X)
Figure 7: Malignant cells admixed with areas of necrosis(arrow)
Figure 8: Islands resembling enamel organ type of tissue with palisading of peripheral tall columnar cells(arrow)
Figure 9: Bony spicules (arrow) with sheets of malignant cells
Figure 10: Tall columnar cells with marked cellular atypia, pleomorphic nuclei, and mitotic figures with loss of peripheral palisading and nuclear polarity (40X)

DISCUSSION

Maxillary ameloblastic carcinomas are very rare. The origin including its histopathogenesis in still unknown. The predominant and first clinical sign was swelling; bleeding or ulceration occurs in few cases. Progressive types of ameloblastic carcinoma may be associated with degree of aggressiveness, possibly characterized by

*Cortical bone perforation
*Invasion of soft tissue
*Recurrences
*Metastases

Metastases most commonly occur via haematogenous route and rarely through lymphatics with high percentage of pulmonary metastases; lymphnode metastases also occurs rarely. High incidence of pulmonary metastases necessitates detection of pulmonary metastases by computer tomography or PET scan and the need for long term follow up. Increased serum calcium which might be unspecific due to osteolysis is also considered as a predictor of metastases[9]

According to Hall *et al.*, [10] the following criteria were specified for diagnosis of ameloblastic carcinoma:

A.Clinical Criteria

1.Rapid growth
2.Pain
3.Paraesthesia
4.Propensity to perforate cortex.

B.Histopathological parameters suggesting possibility of malignant transformation

1.Presence of sheets or islands or trabeculae of epithelium
2.Absence of stellate reticulum like structures
3. Round to spindled epithelial cells with little or no differentiation towards columnar cell morphology of ameloblastoma.

Histopathologically granular cell change and an extensive clear cell component are considered as predictors of metastases and / or aggressive behaviour[11,12,13]

CLASSIFICATION OF MAXILLARY AMELOBLASTIC CARCINOMA

By Elzay 1982[14]

1. Arising from an odontogenic cyst.
2. Arising from an ameloblastoma.
2a.Well Differentiated Malignant ameloblastoma
2b. Poorly Differentiated Ameloblastic Carcinoma
3. Arising de novo
3a.No keratinizing
3b.Keratinising

By Slootweg and Muller 1984 [15]

1. Primary intra osseous carcinoma ex odontogenic cyst
2a. Malignant ameloblastoma
2b. Ameloblastic carcinoma arising de novo, ex ameloblastoma or ex odontogenic cyst.
3. Primary intraosseous carcinoma de novo
3a. No keratinizing
3b. Keratinizing

WHO classification[16]

Metastasizing malignant ameloblastoma
Ameloblastic Carcinoma – Primary type
Ameloblastic Carcinoma – Secondary Type dedifferentiated, intraosseous
Ameloblastic Carcinoma – Secondary Type dedifferentiated, peripheral
Primary intraosseous squamous cell carcinoma – solid type
Primary intraosseous squamous cell carcinoma derived from keratocystic odontogenic tumor
Primary intraosseous squamous cell carcinoma derived from odontogenic cysts
Clear Cell odontogenic carcinoma
Ghost Cell odontogenic carcinoma.

Kruse *et al.* 2009[2]

1. Malignant ameloblastoma
1a.Metastase with features of an ameloblastoma Well Differentiated
1b. Metastase with malignant features Poorly Differentiated
2. Ameloblastic carcinoma arising from an ameloblastoma
2a.without metastase
2b.Metastase with features of an ameloblastoma Well Differentiated
2c. Metastase with malignant features Poorly Differentiated
3a. Ameloblastic carcinoma with unknown origin histology
3b.Metastase with features of an ameloblastoma Well Differentiated
3c.Metastase with malignant features Poorly Differentiated

Differential Diagnosis of Ameloblastic Carcinoma

1.Primary intraosseous squamous cell carcinoma, metastatic carcinoma of Jaw, central high grade mucoepidermoid carcinoma and bony invasion of carcinoma originating from adjacent soft tissues should be excluded in Ameloblastic carcinomas arising de novo where the basic criteria i.e " ameloblastic differentiation" should be present for the diagnosis of Ameloblastic Carcinoma [17]

2. Exclusion of metastasis as invasion of bone by tumor from adjacent soft tissue or paranasal sinus and metastasis in Jaws from visceral neoplasam [18]

3.The squamous odontogenic tumor composed of islands of squamous epithelium that lacks stellate reticulum like zones and peripheral palisading may be mistaken for ameloblastic carcinoma. [19]

4.Squamous cell carcinoma arising in the lining of an odontogenic cyst.

5.Clear cell variant of odontogenic and non-odontogenic neoplasms to be excluded when there is presence of clear cell component in ameloblastic carcinoma.
6.Acanthomatous ameloblastoma and keratoameloblastoma confuse and divert us from ameloblastomatous features [11,20]
7.Basaloid squamous cell carcinoma BSC creates a diagnostic dilemma due to common features of solid nests and strands of tumor cells with peripheral palisading present in both basaloid squamous cell carcinoma and ameloblastic carcinoma. However, the periodic acid – Schiff PAS postivity in microcystic spaces present in BSC is useful to differentiate it from ameloblastic carcinoma.

Treatment

The sponge maxillary bone architecture facilitates the tumor to spread rapidly leading to infiltration of adjacent vital structures. In contrast to mandible, where the growth is decelerated due to thick and compact bone structure.[21] Hence maxillary ameloblastomas/ameloblastic carcinomas should be treated as radically as possible. Curettage of maxillary ameloblastoma is associated with recurrence in almost 100% of cases.[8] A surgical resection with 1015 mm margin free of tumor is recommended. [22] and the resection may be limited related to adjacent pivotal anantomical structures particularly in maxilla. Most of ameloblastic carcinomas are intraosseous, so effectiveness of radiotherapy is considered critically. [23] suggested adjuvant radiotherapy in following cases:
* Patients with positive resection margins
*Multiple positive Lymphnodes.
*Extracapsular spread
*Perineural invasion
*Patients in whom salvage surgery would be inefficient.
Reports about chemotherapy regimen is rare.
Neck dissection should be done only in presence of clinically positive lymph nodes.

CONCLUSION

Maxillary ameloblastic carcinomas are very rare. Patients with ameloblastic carcinoma should undergo radical resection followed by radiotherapy if necessary and must undergo a lifelong follow up including CT/ MRI scans for early detection of recurrence or pulmonary metastasis.

CONSENT

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

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