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A Rare Case of Ameloblastic Carcinoma involving the Ethmoid and Sphenoidal Sinus

Shayesta Jahan[1], Tejaswini BN[2], Veena R[3], Sowmya BU[4]

Department of Pathology, Strand Life Sciences,Health Care Global Cancer Institute and Research Centre, Bangalore, Karnataka, India

Abstract: ***Ameloblastic Carcinoma is an extremely rare malignant epithelial odontogenic tumour about which there is very little in literature. It is an aggressive neoplasm that is locally invasive and can spread to regional lymph nodes and also to distant sites such as lungs and bones. Histopathology plays a vital role in diagnosing Ameloblastic Carcinoma and differentiating it from Ameloblastoma. Here we present a unique case of Ameloblastic Carcinoma involving the ethmoidal and sphenoidal sinus in a 30 year old female patient which presented a diagnostic challenge. The patient was treated successfully with wide local excision followed by chemo radiation. Given the rarity of the disease and location this report constitutes a valuable contribution to current literature.***

Keywords: Ameloblastic Carcinoma AC, Ameloblastoma AB, Malignant odontogenic tumour,Primary Intra Osseous Carcinoma PIOC

1. Introduction

Ameloblastic carcinoma accounts for 1.5 – 2% of all Odontogenic tumours.[1]It is an extremely rare, aggressive malignant epithelial odontogenic tumour with poor prognosis. The term ameloblastic carcinoma was introduced by Elzay in 1982 to depict a malignant odontogenic tumour that retains the features of ameloblastic differentiation and exhibits cytological features of malignancy in primary or recurrent tumour.[2]

Less than 120 cases of Ameloblastic Carcinoma have been reported in literature .Of these cases, less than five were identified that originated outside the mandible or maxilla[3].

Differentiating Ameloblastic Carcinoma from Ameloblastoma is a challenge due to overlapping clinical features histopathology and different management approaches.[4] In this article we report a rare case of ameloblastic carcinoma of the ethmoidal and sphenoidal sinus involving the left orbit and the left nasal cavity that represented a diagnostic challenge.

2. Case Report

A 30 year old female patient presented with history of self-inflicted trauma over the forehead, after which she noticed excessive watering of the eyes and bulging of the left eye since one month.She was initially treated at a local hospital and then she was referred to our hospital.

Local examination revealed mild swelling in the infraorbital region in the left side with proptosis of left eye. Deviation of the left eye and widening of the interpalpebral region of the left side was noticed.

On direct nasal examination of the left nasal cavity there was a sub mucosal growth present in the left lateral wall of size 3x2 cm and there was congestion over the swelling.

Clinical diagnosis of Osteosarcomaof orbit was made.Ophthalmology reference was taken and work up including PET CT and MRI scan was done.

Brain MRI was done which showed enhanced mass lesion in the left infra orbital wall extending into left extra canal space with no obvious intra cranial extension which was suggestive of neoplastic bone lesion.

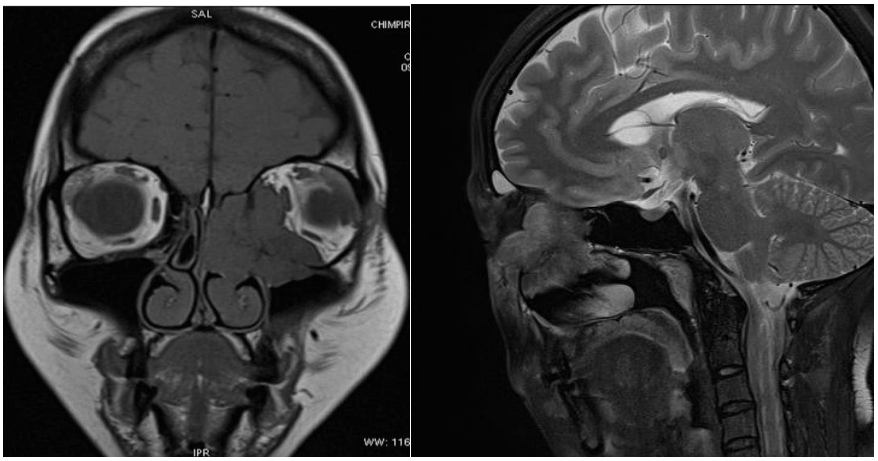


Figure 1: T2W MRI image (a-coronal – sagittal)showing hyperintense lesion lobulated T2/FLAIR intermediate signal intensity mass involving the medial, inferior extra conal compartments of left orbit, left maxillary and anterior ethmoid sinuses, left nasolacrimal ducts and left nasal cavity with proptosis of left eyeball. No intracranial extension was seen.

Oncoview PET CT scan was done which showed a 3.6 x 3.5 x 3.3 cm ill-definedmetabolically active mass with sun burst periosteal reaction epicentered in inferomedial wall of left orbit to left anterior ethmoid air cells, left nasal cavity, left maxillary sinus, left nasolacrimal duct with soft tissue extension to extraconal compartment of left orbit along medial and inferior aspects causing proptosis of left eye ball which favoured a malignant etiologypossibly Osteosarcoma.There was no evidence of pulmonarymetastases.

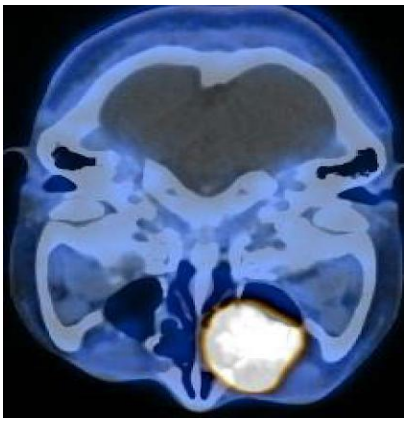


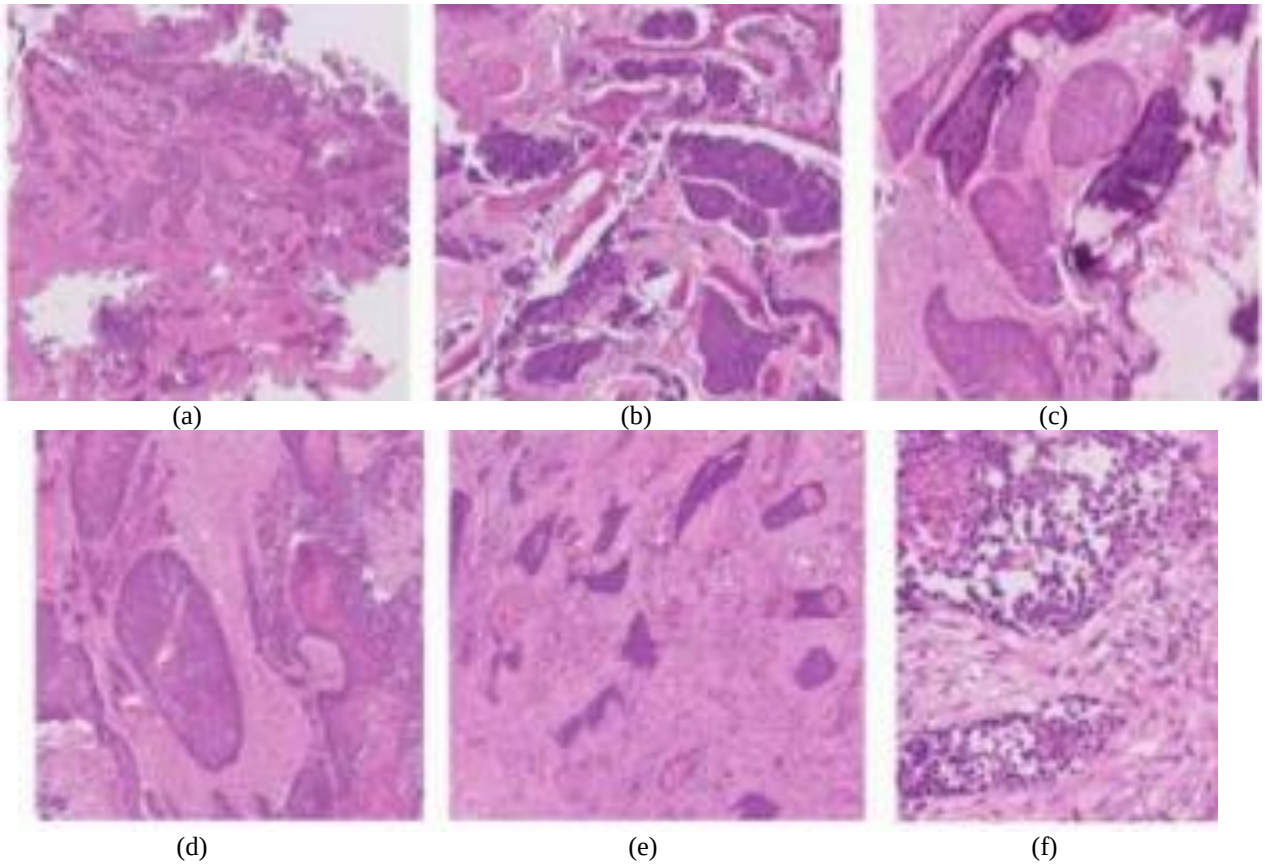
Figure 2: PET-CT showing lobulated metabolically active mass

The patient underwent wide local excision of the lesion by combined trans nasal and trans orbital approach.

Histopathological examination of tissue revealed odontogenic epithelium in plexiform pattern and in islands in a densely fibrous stroma. The basal cells were columnar hyperchromatic arranged in palisaded manner, centrally located stellate reticulum was seen.Focally the cells exhibited nuclear atypia with vesicular nuclei and inconspicuous nucleoli. Squamous differentiation was noted.Focally areas of necrosis was seen.Neoplastic cells were seen infiltrating the bone and adjacent adipose tissue. Lymphovascular invasion and perineural invasion was absent.

Immunohistochemistry showed immunoreactivity for CK7 and P40.The tumour cells were negative for CD56, Calretinin and S100. Ki-67 proliferative index ranged from 20-25%.

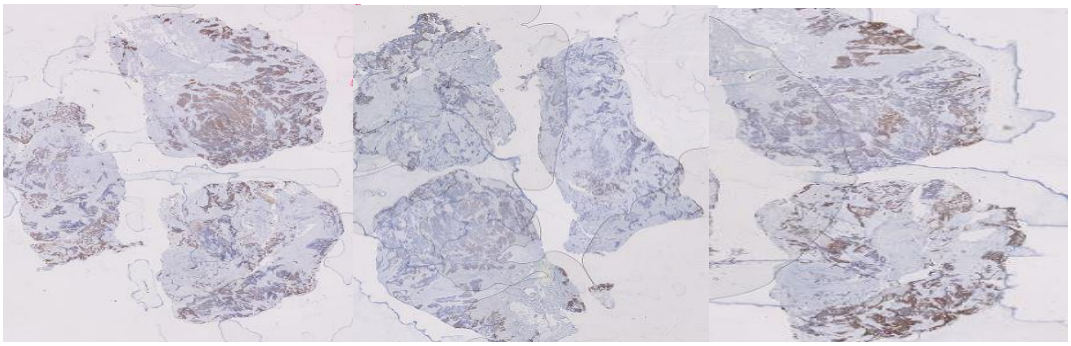
Thus in view of infiltrative pattern and high proliferative index, the diagnosis of Ameloblastic Carcinoma in a background of Ameloblastoma.



(a) (b) (c)

(d) (e) (f)

Figure 3: Photomicrographs showing sheets and nests of ameloblastic epithelium with inflammatory infiltration ((a) H & E, 200X); areas of tumor cells with hyperchromatism, nuclei pleomorphism, and some mitotic figures ((b) H &E, 400X); neoplastic cells seen infiltrating the bone(c,d)H & E, 200X);:Islands of well defined malignant epithelium, with flattened peripheral cells separated by collagenised stroma (e) H & E, 200X); Nests of basaloid cells with periphersl palisading and microcyst formation.((f)H & E ,400X).



Ck7 (g) Ki-67 (h) P40 (i)

Figure 4: Marked positive immunohistochemical expression of CK7 (g), Ki67 (h), and p40 (i)

The patient was planned CT+RT with injection cisplatin100mg /M2 3 weekly for 3 cycles. The patient was given 3 cycles of chemotherapy with Inj Cisplatin and is on regular follow up and is free of recurrence for the past 6months.

3. Discussion

Ameloblastic carcinoma is a rare malignant neoplasm of odontogenic origin.[5] In the WHO 2005 classification, AC was divided into 3 categories; primary type (a), secondary type (dedifferentiated) intraosseous (b) and secondary type (dedifferentiated), peripheral (c). These tumors were classified under "AC" in the WHO 2017 classification based on the morphologic features and similar behaviour between these entities.[6]

The neoplasm is characterized by aggressive behaviour and poor prognosis. It most frequently affects men, with no age group pre-dilection and involves more often the mandible.[7]

The aetiology of Ameloblastic carcinoma is largely unknown and is still controversial. Most cases arise spontaneously without previous history of cancer (de novo) with few cases arising following malignant transformation of ameloblastoma.[8]

The clinical symptoms of AC are more aggressive than ameloblastoma.Distinct features from ameloblastoma are swelling with rapid growth, perforation of cortex, pain, tooth mobility, a non-healing extraction site, ulcer or fistula, facial asymmetry, trismus and paraesthesia.[9]

In the literature, primary orbital ameloblastoma has never been reported; all cases were secondary to invasions from mandible or maxilla.[10]The clinical picture of ameloblastoma depends on the origin of the tumor and also the structures involved. The most common manifestation of mandibular type is painless swelling of jaw, whereas, primary maxillary ameloblastoma presents with swelling of cheek, gingiva, hard palate, nasal obstruction, and epistaxis. The tumor inherently invades local adjacent structures such as paranasal sinuses, orbit and cranial fossa. Invading or compressing the neurovascular structures within orbit and cavernous sinus leads to various ophthalmologic manifestations such as loss or decrease of vision as the most common symptom followed by proptosis and globe displacement, extraocular movement limitation,diplopia, cavernous sinus syndrome, lower lid oedema andptosis. It should be noted that orbital manifestations were almost unilateral: however, there are two reports by Kyrias et al. and Braziset al.[10]with bilateral orbital involvement.

It is presumed that Ameloblastic Carcinoma constitutes 2% of all Ameloblastoma cases. In a review article by Brazis et al it has been proposed that Ameloblastic Carcinoma ex-ameloblastoma is more prevalent (17%) among cases with orbital invasion.[10]

The radiological features of AC are comparable of Ameloblastoma, unilocular or multilocular radiolucent lesions with lamina dura or tooth apex resorption. However AC may exhibit focal radio opacities, dystrophic calcifications and a poorly defined lesional border.[11]

AC is composed of islands and chords of ameloblastomatous epithelium in an infiltrated pattern. The epithelium may reveal a single outer layer ofameloblastic cells of columnar or cuboidal shape which may or may not exhibit palisading and reverse nuclear polarity. The stellate reticulum within the islands is often condensed and hyper cellular. The characteristic features of AC are nuclear enlargement stippled nucleus, nuclear hyperchromatism, mild pleomorphism, increase mitotic activity with abnormal forms of mitosis. Dyskeratosis, keratin pearl formation, necrosis and dystrophic calcification may be observed in some cases.Seldom, AC may reveal clear cell differentiation.[12]

The differentiation of AC from AB is most important. AC is diagnosed when there is evidence of cytological atypia in a histological pattern of AB. An elevated mitotic rate is especially helpful as AB exhibit less mitosis. IHC may be helpful in distinguishing the two entities. Ki-67 LI is found to be significantly higher in AC than in AB.Mean Ki-67 LI in AC has been reported to be approximately 20%, while in AB it is approximately 4%–5%.[13]In our case Ki 67 was found to be 20-25%.The expression of certain CKs can also help to differentiate between AB and AC, with CK 18 being especially useful. While CK 18 is diffusely positive in all tumor cells in AC, it is only weakly positive in stellate reticulum in AB.[14]

In a study by Martínez et al. comparing histological and immunohistochemical features of ameloblastoma and AC, they suggested that both Ki67 and p53 could be good markers of malignancy .[15]

Our differentials were Basaloid variant of Squamous cell Carcinoma.The distinguishing feature of AC from SCC include the jigsaw puzzle type nesting of tumorcells,the presence of stellate reticulum, and cystic degeneration of cells. The other differential was Primary Intraosseous Carcinoma .The presence of sheets /islands of cells which have undergone metaplastic change with high mitotic activity in AC can be mistaken for PIOC.In PIOC, there is absence of ameloblastic differentiation which is always seen in AC. There is no established consensus on the treatment of AC.Surgery is the most widely used treatment option, with the role of radiotherapy and chemotherapy still not very clear. There is a higher recurrence rate following curettage alone than following partial resection, and therefore a wide surgical resection, with clear margins, is recommended.[15] Radiotherapy has been used in cases with positive surgical margins, perineural infiltration, or soft tissue invasion.[15]Since AC has a high recurrence rate and frequent metastasis in the lung and regional lymph nodes, it is pertinent that the patients be followed long term.

Knowledge regarding AC is limited by the rarity of this disease, and most information is presented in the form of single case reports or case series.[16]This presentation was particularly rare due to the location of the neoplasm and the radiological findings which mimicked an Osteosarcoma.

Since the present case showed areas resembling AB, we postulate that the AC arose in a background of AB. Ameloblastoma demonstrates a wide spectrum of histologic and biological behaviour ranging from benign to malignant. Hence histopathological examination is the key step in planning management and treatment of these cases.

Cases showing carcinomatous transformation require promptsurgery and long term follow up to detect the late recurrence, metastasis or regional lymph nodes. Therefore diagnosis at an early stage, close periodic screening for metastasis and adequate treatment are necessary to improve patient prognosis.

4. Conclusion

Ameloblastic Carcinoma is an uncommon odontogenic tumor that exhibits malignant features in the primary site. Cases of Ameloblastoma show a wide variety of histologic and biologic behaviour ranging from benignity to frank malignancy, thus histopathological examination is a vital tool in determining their nature and treatment plan. Cases showing carcinomatous transformation require early surgery and long term follow up.

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