



## Huge Ameloblastic Carcinoma: A Rare Case with PET-MRI Findings

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### Abstract

Ameloblastic Carcinoma (AC) is a very rare malignant odontogenic tumor arising from odontogenic epithelium. The tumor most often involves the mandible. It presents with a rapid and painful sudden swelling in the jaw. It is an aggressive tumor that is locally invasive and can spread to regional lymph nodes or distant areas such as lungs and bones.

We present a case of ameloblastic carcinoma originating from the mandible but filling the maxillary sinus in a 33-year-old female patient who presented with swelling on the right face, together with PET MRI findings.

**Keywords:** PET-MRI; Odontogenic tumor; Ameloblastic carcinoma

### Introduction

Ameloblastic Carcinoma (AC) is a locally aggressive odontogenic malignancy of embryonic origin. It is most commonly identified in the mandible or maxilla [1].

The mean age of AC occurrence is 30.1 years, but a wide range of ages can be affected. There is no proven gender bias, but certain studies have reported a male predominance [2,3].

AC is considered to be a rare malignant odontogenic tumor that has combined histopathological features of ameloblastoma and carcinoma, regardless of the presence or absence of metastasis. Furthermore, AC has been classified into two types, primary and secondary. The former develops de novo and the latter develops by malignant transformation of a pre-existing benign ameloblastoma [4].

AC tends to be aggressive and extends with local destruction. Lymph node involvement and distant metastasis to various regions have also been reported [2,5].

Therefore, diagnostic imaging prior to treatment is extremely important.

### Case Presentation

A 33-year-old female patient, who was referred to our oncology unit, was complaining of intense lower facial pain and progressive swelling for the last two months. Swelling was sudden in onset and gradually increased in size. In the extraoral examination, it was found that the swelling extends to the skin in the right maxillary, orbital and nasal region on the right side of the mandible.

CT and PET-MRI was performed in our own radiology unit. In the right half of the face, an FDG positive (SUVmax: 23) mass lesion, which was thought to originate from the right mandible corpus, causing destruction of the maxilla and mandibular bone with an approximate size of 10 cm × 9 cm was observed in the PET-MRI. There were areas of necrosis in the central part of the mass. The mass was advancing medially towards the oropharyngeal air column and narrowing it. In the superior, it extended to the inferior of the right bulbus oculi and near of the rectus muscle. The mass was filling the right maxillary sinus. Mass extension to the skin was observed anteriorly. It extended into the right nasal cavity. There was no finding suggesting metastasis in other parts of the body in the PET-MRI examination. There was destruction in the maxillary and mandibular bone on CT.

An incisional biopsy was performed on the patient. Histopathological examination revealed a high percentage of cellular stains with cells in a barred arrangement. The cell block revealed a binary basaloid and star-shaped cell population, as well as atypical cells and mitotic figures. Pathology result of incisional biopsy was found to be AE1/AE3 positive, Ki 67 40% positive. CD45, S100, CD3, CD20, Bcl-6 and Cd56 were negative. Therefore, the patient underwent an incisional biopsy,

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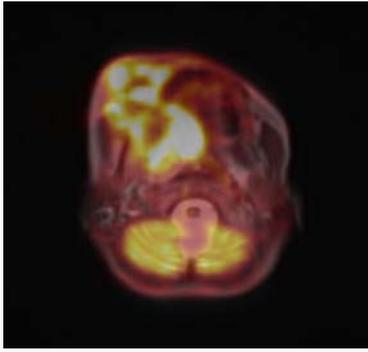
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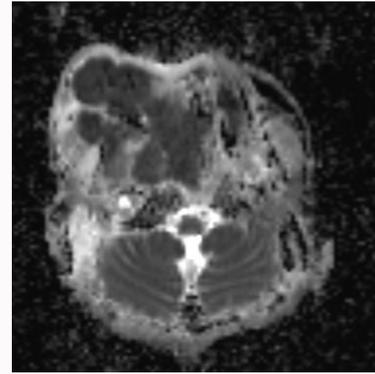
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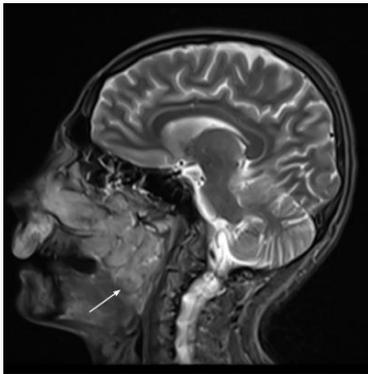
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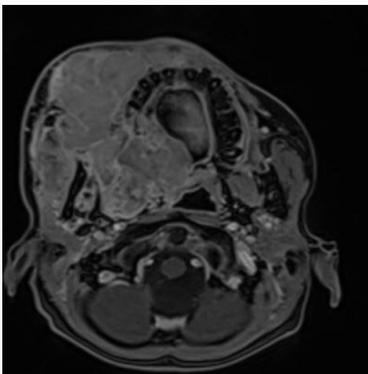
**Figure 1:** Odontogenic ameloblastic carcinoma filling the maxillary sinus showing FDG uptake (SUVmax: 23) on PET-MRI fusion axial images.



**Figure 4:** Mass showing restriction of diffusion in ADC maps.



**Figure 2:** Extension towards the oropharynx on T2W sagittal MRI image.



**Figure 3:** T1W postcontrast heterogeneous enhancing mass.

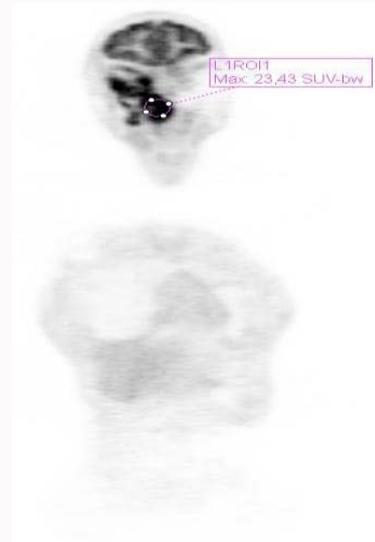
presenting a histopathological report of odontogenic ameloblastic carcinoma (Figures 1-5).

The patient was evaluated as a locally advanced stage and neoadjuvant 2 courses of chemotherapy and radiotherapy were planned.

## Discussion

Ameloblastic carcinoma is an extremely rare odontogenic tumor that originates from the dental enamel or odontogenic epithelium, and shows the histological picture of ameloblastoma with cytological atypia with or without metastases [4].

These tumors commonly occur in the third to fifth decades of life with no preference based on gender and ethnicity, accounting for



**Figure 5:** Coronal PET image (SUVmax: 23).

eighty percent of the cases in the mandible [6].

Most cases of AC arise spontaneously (*de novo*) with few cases arising from a malignant transformation of an existing ameloblastoma or a benign odontogenic cyst [7].

The clinical symptom of AC is more aggressive than ameloblastoma. Distinct features from ameloblastoma are swelling with rapid growth, perforation of the cortex, pain, tooth mobility, a non-healing extraction site, ulcer or fistula, facial asymmetry, trismus and paresthesia [2].

The progression of this aggressive tumour with extensive local destruction and metastatic spread results in a poor long-term prognosis, however treatment modalities for such patients are controversial as there is no clear consensus on the best approach to manage this condition [1].

There is no established consensus on treatment of AC due to the low incidence of the disease, although wide local excision is the mainstay of therapy for the primary tumor. Treatment of local extension and metastases usually involves some combination of chemotherapy and radiation. In rare cases, neoadjuvant radiation or chemotherapy is used to shrink a tumor prior to resection. Radiation therapy alone can be effective, but is reserved for local recurrences or cases where the primary tumor may not be surgically resected.

In cases of systemic metastases, chemotherapy may be indicated, although studies have demonstrated mixed results. Targeted therapy focused at genomic aberrations is experimental [8-10].

ACs can recur locally 0.5 to 11 years after definitive therapy. Distant metastasis is usually fatal and may appear as early as 4 months or as late as 12 years postoperatively. The most common site for a distant metastasis is the lung, followed by bone, liver and brain. Distant metastasis can occur in the absence of a local or regional recurrence [10].

## Conclusion

We reported the evaluation and treatment approach of locally advanced ameloblastic carcinoma, which is a rare tumor, together with PET MRI findings.

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