Original Article

Malignant Odontogenic Tumors: An Analysis of 15 Cases and Review of the Literature

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Abstract

Introduction: Malignant odontogenic tumors (MOTs) are uncommon primary jaw lesions that represent approximately 6% of all odontogenic tumors. The rarity and complex classification reported for MOTs have also created challenges in their study over the years. **Patients and Methods:** All patients with a histopathological diagnosis of odontogenic tumor from 2003 to 2017 formed the study population. Their departmental and medical records were analyzed. **Results:** A total of 15 MOTs patients were studied. There were 10 males and five females with a M:F ratio of 1.6:1, and the ages ranged from 17 to 80 years with a mean of 48.06 years. The sites were the mandible and maxilla. The mandible to maxilla ratio was 2.25 to 1. **Conclusion:** Ameloblastic carcinoma was the most common MOT.

Keywords: Malignant, odontogenic, tumor

Introduction

Malignant odontogenic tumors (MOTs) are uncommon primary jaw lesions that represent approximately 6% of all odontogenic tumors.[1,2] The rarity and complex classification reported for MOTs have also created challenges in their study over the years.[1] According to Panda *et al*.,[3] for most MOTs, standardized diagnostic criteria are yet to be established. MOTs like their benign counterparts develop from either the epithelial or mesenchymal components though, the resultant carcinomas exhibit an overwhelming prevalence over the sarcomas.[2] The varied MOT recognized in the World Health Organization (WHO) 2005 classification of OTs include metastasizing (malignant) ameloblastoma, ameloblastic carcinoma (AC), primary intraosseous squamous cell carcinoma (PIOSCC), clear cell odontogenic carcinoma (CCOC), ghost cell odontogenic carcinoma, ameloblastic fibrosarcoma (AFS), fibrodentinosarcoma, and fibro-odontosarcoma.[4] There have been some case reports of MOT from Nigeria,[2,5] however, the detailed clinicopathological characterization of these tumors have not been documented.

This study aimed at carrying out a detailed analysis of MOTs in our setting which should improve the current limited understanding of the biologic behavior, clinical management, and resultant outcome of these tumors as well as the overall patient well-being.

Patients and Methods

All patients with a histopathological diagnosis of an odontogenic tumor from 2003 to 2017 formed the study population. These cases were sourced from the departmental and medical records of all cases with orofacial tumors and tumor-like lesions seen at the maxillofacial unit of a tertiary hospital. Relevant data on age, gender, tumor site, clinical features, radiologic appearances, treatments, and follow-up records were retrieved from case notes, operation notes, radiographs and radiology reports, histopathology results, and follow-up records. Only, the MOTs that fulfill the 2005 WHO classification were analyzed.

Results

AC accounted for eight cases which constituted 53.3% of MOTs with an age range of 32–72 years and a mean age of 49.87 years

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and all the cases occurred in males. The mandible was more involved (n = five cases; 62.5%) than the maxilla (n = 3; 37.5%). The diagnosis of six (75%) of the eight cases of AC was reviewed from ameloblastoma based on the patient's clinical presentation with recurrent features of malignancy and convincing histopathological features [Table 1].

Of the eight cases, three had surgical resection with chemoradiation and another three cases had resection only while two cases had no treatment. Only one patient was followed up for 24 months while others defaulted from 6 months. Three patients presented with recurrence 2 years after surgery, while two had recurrence 5 years after initial histopathological diagnosis of ameloblastoma. Metastasis to the lungs and liver was recorded in three patients. The lung metastasis was confirmed by a CT-guided biopsy with a histopathological report of AC. Although, there were palpable cervical lymph nodes, which were excised and subjected to histopathological analysis, and they showed no tumor involvement.

Primary intraosseous carcinoma accounted for four cases and constituted 26.6% of MOTs. All four cases occurred in the mandible with an equal sex distribution of the patients. The patient's ages ranged between 40 and 80 years with a mean of 56.25 years. Three of the patients had mandibular resection with or without chemoradiation, and only one had no treatment. All the patients were followed up for 6 months. Two patients had a recurrence during follow-up [Table 1].

AFS accounted for two MOTs (13.3%) cases. Both cases occurred in females and involved the maxilla with the respective age of 17 and 28 years. Maxillectomy was the treatment of choice for the older female while the younger patient had no surgical treatment due to advanced tumor stage at presentation. She was referred for palliative chemoradiation but she declined. The older female presented with recurrence 5 years later and had a second maxillectomy and chemoradiation.

CCOC represented 6.6% (one case) and it was a female of 52 years with the lesion on the mandible.

Discussion

MOTs are rare malignancies and several documented literature indicated they accounted for 3.4% to 5.6% of odontogenic tumors.[2,6,7] The diagnosis is often incidental either as a clinically malignant tumor whose biopsy result turns out to be an odontogenic neoplasm or through the identification of malignant cellular features in a tumor that has not yet revealed clearly malignant behavior.[8]

Malignant ameloblastoma was the predominant malignancy, followed by AC in the report by Goldenberg *et al*.[9] However, AC (53.3%) was predominant in this case series, followed by primary intra-alveolar carcinoma (26.6%) with all patients being male and an age range of 32–72 years with a mean of

Table 1: Clinical details, presentations, and follow-up of 15 cases of malignant odontogenic tumors

Histological diagnosis	Mean age	Sites	Clinical features	Working diagnosis	Treatment/follow-up
Ameloblastic carcinoma	49.8	Mandible Maxilla Temporal region	Mandibular swelling Involvement of the floor of the mouth Maxillary swelling Temporal swelling Ulceration Involvement of the left orbit with loss of vision Middle cranial fossa involvement Presence of reactive lymph nodes History of previous surgery (1-3) Recurrence (1-3)	Recurrent ameloblastoma Ameloblastoma Ameloblastic carcinoma	Mandibulectomy with soft-tissue excision Hemimandibulectomy Extended maxillectomy Total maxillectomy Upper and lower eyelids excision Exenteration Soft-tissue reconstruction Chemotherapy and radiotherapy Follow-up from 3 months to 5 years one death
Ameloblastic fibrosarcoma	22.5	Maxilla	Maxillary swelling Blockage of both nares Bleeding Tooth mobility	Sarcoma	Inoperable Maxillectomy Chemotherapy and radiotherapy
Primary intra alveolar carcinoma	56.25	Mandible	Mandibular swelling Involvement of floor of the mouth Ulceration Mobile teeth Loss of teeth Recurrence History of surgery	Carcinoma Primary intraosseous carcinoma	Resection with soft-tissue excision Resection with disarticulation Chemotherapy Radiotherapy Follow-up 3-6 months
Clear cell odontogenic carcinoma	52	Mandible	Mandibular swelling Extension floor of the mouth	Clear cell odontogenic carcinoma	Died before surgery

49.87 years. The mandible was the most affected in the ratio of 1.67 to 1. The mandibular predominance again is in conformity with the South Indian report,[10] and Goldenberg *et al*.,[9] who also reported that the mandible was more affected as compared to the maxilla.

AC was the most common MOTs in our series (53.3%) and this agrees with a previous report (67%) from Southwest Nigeria though, higher than the report from South India (30.7%) by Pandiar *et al*.[10] AC is more common in the mandible than maxilla from the reports of Pandiar *et al*.[10] and Lawal *et al*.[2] in India and Nigeria, respectively. This observation is comparable with our finding of 62.5% occurrence in the mandible. The mandible as a site of predilection for AC is comparable with the universal predilection of benign ameloblastoma.[5] However, the Iranian report by Taghavi *et al*.[7] had more cases in the maxilla than the mandible. In addition, Fomete *et al*.[5] reported two cases of AC in the maxilla. One of the patients in this series presented with extensive skin ulceration involving the lower jaw. All the patients (100%) with AC in this series were male with a mean age of 49.87 years and a range of 32–72 years. Pandiar *et al*.,[10] in South India, reported an equal sex distribution and a mean age of 51.25 years.

Kruse *et al*. [11] reported metastasis in 34.6% of cases with 29.6% occurring in the lungs while 23.1% had a recurrence of the malignancy. Two patients presented with metastasis to the lungs and one to the liver with a recurrence rate of 33.3%. The lung metastasis confirmation by CT-guided biopsy was the first of its kind in our center and in Northern Nigeria. No nodal metastasis though increased calcium serum level has been considered a predictor of metastasis.

The recommended treatment modalities are surgical resection, radiotherapy, and chemotherapy. About 25% and 12.5% respectively , had mandibular resection with radiochemotherapy and mandibular resection only while 25% had maxillectomy only. There was no treatment given in 25%.

The WHO 2005 defined PIOSCC as a central jaw carcinoma derived from odontogenic epithelial remnants.[4] Lawal *et al*.[12] in their review stated that PIOSCC was first described by Loos as central epidermoid carcinoma of the jaws with the diagnostic criteria of the absence of initial connection with the overlying skin or mucosa and exclusion of metastasis from a distant primary tumor by physical and radiographic examination during at least a 6 months follow-up period. The WHO divides PIOSCC into three groups, namely those arising de novo, those arising from preexisting odontogenic cysts, and those arising from preexisting odontogenic neoplasm.[8] They are rarer than AC, aggressive, and tend to overgrow their precursor lesion, although remnant or even a tooth may be found engulfed by the carcinoma.

Lawal *et al*.,[2] reported a mean age of 63.6 years and a mandibular predominance. In South India, the mandible was also the predominant site.[10] There were four (26.6%) patients in this study with an aged range of 40–80 years and a mean of 56.25 years. The mean in South India was 60.56 years.[10] The mandible was the only site involved in these patients. The treatment modalities were resection with radiotherapy and chemotherapy in 25% while 75% had surgical resection only. There was a 50% recurrence rate.

AFS is the malignant counterpart of ameloblastic fibroma.[8] It is more common in teenage and young adults with a mean age of 27 years, and it is common in the mandible.[1,8] In this study, two patients both females aged 17 and 28 years (mean = 22.5 years) had lesions sited in the maxilla. They both presented with pain and swelling similar to reports by Morgan.[1,8] Lawal *et al*.[2] reported a solitary case of ABFS in a 28-year-old female with the lesion sited on the maxilla as seen in this study. It seems ABFS has a predilection for the maxilla in this part of the world. Of two cases, one had total maxillectomy with recurrence while the one had an inoperable lesion and refused palliative care.

CCOC is an odd malignant tumor of presumed odontogenic origin and considered a separate entity with about 74 cases in the English literature.[1] Morgan,[8] in his review of the literature, stated that it was first recognized as an entity in 1985. It affects a wide age range of patients with the majority in the sixth decade and a predominance female ratio.[1,8] It is seven times more common in the mandible than the maxilla.[1] In this study, we recorded one (6.6%) female patient aged 52 years with the lesion on the mandible. Both age and sex and site are in agreement with the literature.

Conclusion

MOTs occurrence in our environment is well established with ACs being the most common. Their diagnosis was retrospect in most of the cases. We did not record any case of malignant ameloblastoma in this study. AC is most of the time presenting as ameloblastoma.

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Conflicts of interest

There are no confl icts of interest.

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