East African Medical Journal Vol. 94 No. 2 February 2017
ADENOID CYSTIC CARCINOMA OF THE MANDIBLE: CASE REPORT
H.S. Lawal ,FMCDS, Consultant Maxillofacial Surgeon, Department of Oral and Maxillofacial Surgery, Federal Medical Centre Birini Kudu, Jigawa State, U.K. Omeje, FMCDS, Consultant Maxillofacial Surgeon, Department of Oral and Maxillofacial Unit, Aminu Kano Teaching Hospital, Kano, Kano State, E.D. EKOH, BDS, Dental Officer and E.P. Adeghe, BDS, Dental officer, Department of Oral and Maxillofacial Surgery, Federal Medical Centre Birini Kudu, Jigawa state, Nigeria.

ADENOID CYSTIC CARCINOMA OF THE MANDIBLE : CASE REPORT

H.S. LAWAL, U.K. OMEJE, E.D. EKOH and E.P. ADEGHE

SUMMARY

Primary Intraosseous adenoid cystic carcinoma is a very rare lesion. It most frequently affects the parotid glands, sub-mandibular glands and palate. It presents insidiously and is generally advanced when diagnosed. The importance of early diagnosis, prompt radical surgical intervention, plus immediate post-operative radiotherapy and a long term follow up is emphasised.

INTRODUCTION

Adenoid Cystic Carcinoma (ACC) is an uncommon salivary gland malignancy which can also develop in the mucus glands of the larynx, trachea, bronchus, lungs and mammary glands besides the head and neck region (1). The most frequently affected sites are the parotid gland, sub-mandibular gland and palate, whereas the lower lip, retromolar tonsillar pillar regions and sublingual gland are less frequently affected (2).

Very rarely, ACC may arise centrally within the jawbones comprising less than 0.4% of all salivary gland carcinomas. It usually occurs in the posterior mandible of adults where it causes pain due to perineural invasion (neurotropism) (1,2,3) to the best of our knowledge, only 17 cases of centrally located/primary intraosseous ACC have been reported in literature (2).

We hereby report a case of primary intraosseous adenoid cystic carcinoma involving the mandible of a 43 year old man.

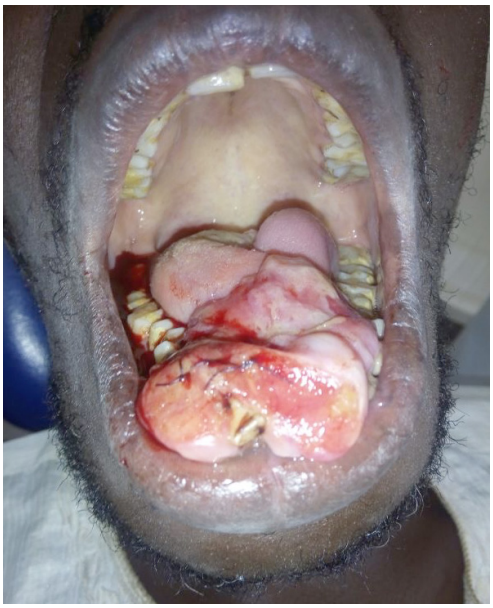
CASE REPORT

A 43 year old male patient presented to the Oral and Maxillofacial Surgery clinic of our hospital with complain of lower anterior jaw swelling of three years duration.(Figure 1) Swelling was said to be painless initially but later became painful with associated lip numbness in the later two months.

Examination of the patient revealed a well circumscribed, non ulcerated bony hard swelling of the mandible extending from the posterior aspect of the mandibular wisdom tooth on the right to around the mandibular first molar on the left side. The associated teeth were mobile (varying degrees of mobility). There was a very minimal bucco-lingual expansion of the mandible but the tongue, floor of the mouth and buccal mucosa all appeared clinically normal at this stage.

Radiographic examination revealed an extensive osteolytic lesion with ill-defined borders extending from the angle of the mandible on the right to the contralateral angle. The submental, sub-mandibular and cervical lymph nodes were not palpably enlarged on examination and chest radiograph was also not remarkable. A provisional diagnosis of Ameloblastoma was made, to rule out primary intra-alveolar carcinoma..

Figure 1
Pre-operative picture of patient with primary Adenoicd cystic carcinoma of the mandible



Radiographic examination revealed an extensive osteolytic lesion with ill-defined borders extending from the angle of the mandible on the right to the contralateral angle. The submental, sub-mandibular and cervical lymph nodes were not palpably enlarged on examination and chest radiograph was also not remarkable. A provisional diagnosis of Ameloblastoma was made, to rule out primary intra-alveolar carcinoma..

Incisional biopsy was performed under local anaesthesia, the result revealed infiltrating nests of malignant epithelial cells forming cribriform pattern. A diagnosis of primary intraosseous adenoid cystic carcinoma was made. Patient was therefore scheduled for subtotal mandibulectomy and restoration of the residual configurational mandibular defect with reconstruction plate followed by post-operative radiotherapy.

Owing to lack of fund, surgery was however not done until six months later at which stage radiographic examination revealed radiological evidence of advanced disease with bilateral involvement of the entire mandibular rami necessitating a more radiacal surgery, thus the patient was consented as such. (Figure 2)

Intraoperatively, there was tethering of the lateral border of the tongue to the mandible in the molar region on the right and an associated pathological fracture at the same point. A total disarticulating mandibulectomy with no form of reconstruction was done.

Figure 2
pre-operative plain radiograph of patient in figure 1

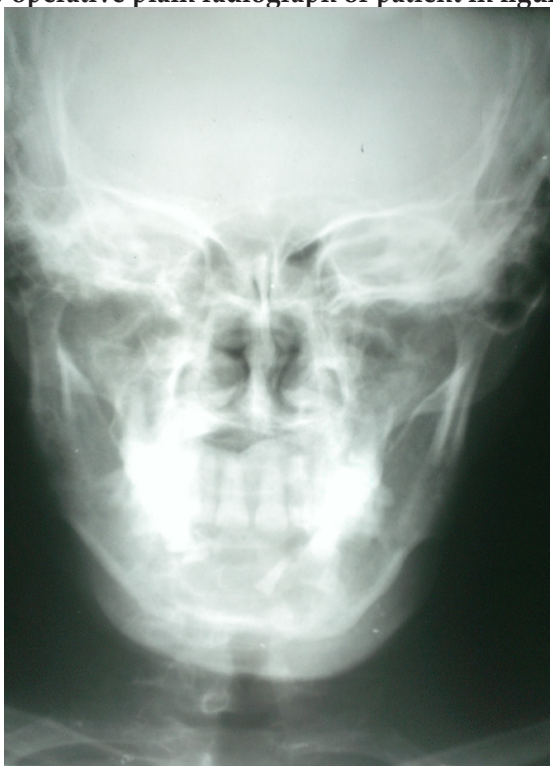
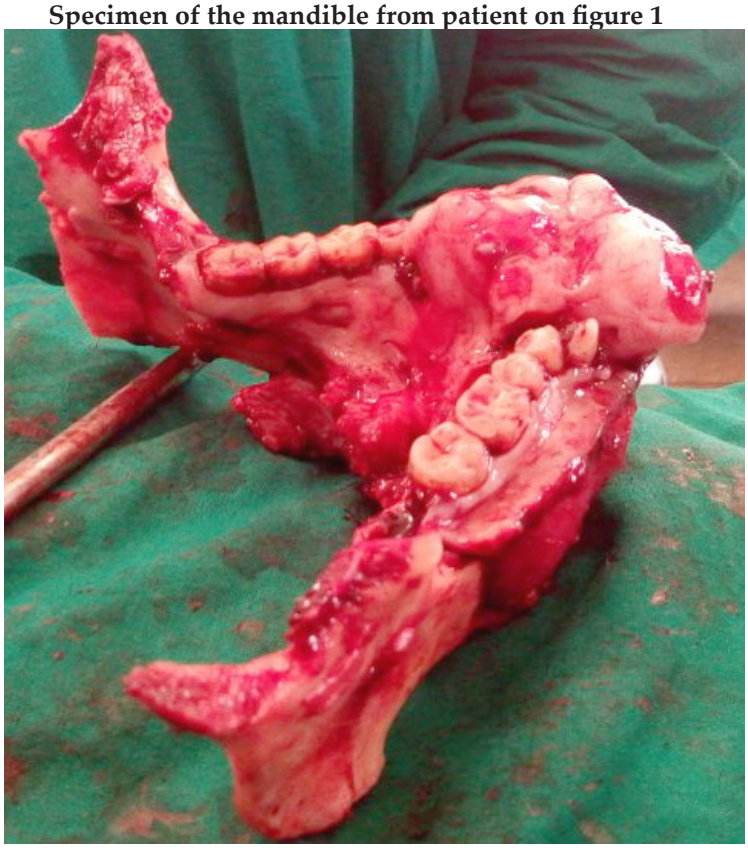


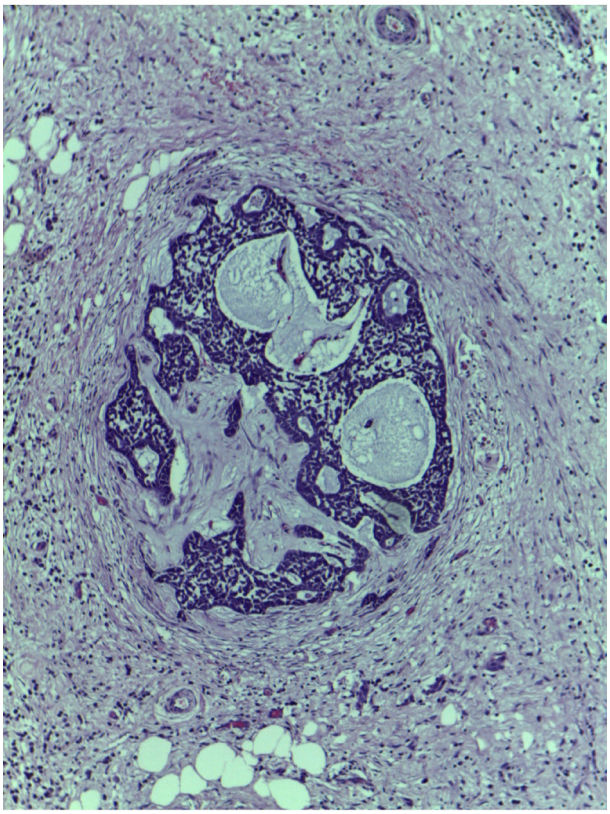
Figure 3
Specimen of the mandible from patient on figure 1



Histology of the surgical sample confirmed the tumour as primary intraosseous ACC with negative tumour margins. Patient had immediate post-operative radiotherapy. He has been followed up for 16 months with no evidence of recurrent disease.

Figure 4
Immediate post-operative photograph of patient with tongue stich in place

Figure 5
photomicrograph of histopathological specimen



DISCUSSION

Primary intraosseous salivary gland carcinomas are very rare and constitute less than 0.4% of all salivary gland carcinomas (4). Mucoepidermoid carcinomas are the most frequently reported type of primary central salivary gland carcinomas of the mandible followed by adenoid cystic carcinoma (ACC) (1-4).

ACC presents insidiously, grows slowly compared to squamous cell carcinoma and therefore tends to be locally advanced at the time of diagnosis (1,4,5).The extensive nature of the present case might have been a result of the insidious nature of ACC.

The pathogenesis of central ACC and other central salivary gland neoplasms is unknown but various theories (4) for their origin have been proposed which include: 1. Development from enclaved retromolar mucous glands during embryogenesis(2). Sub-mandibular and sublingual glands closely apposed in bony defects or cavities in the lingual cortex of the mandible or 3. Fragments of sub-mandibular or sub-lingual glands that have undergone embryologic evagination.

Central ACC is usually seen in the posterior molar/ ramus of adults with peak incidence in the fourth to the sixth decade of life (1, 3). It is also noted not to have sex predilection (1,3) although Batsakis *et al (*4) reported a greater female predilection. Swelling and pain are the most common clinical features as documented in the literature (1-5). These features are consistent with those of the present case. Batsakis (4) in 1979 proposed a diagnostic criteria for central salivary gland neoplasms: I. Radiologic evidence of osteolysis, II. Presence of intact cortical plates, III. Presence of intact mucous membrane overlying the lesion, IV. Absence of any primary tumour within major or minor salivary gland, V. Histological confirmation of the typical architecture and morphological features of a salivary gland tumour. These criteria were observed in the present case, we therefore concluded that the lesion first developed in the right posterior molar/ramus region of the mandible before spreading extensively in the medullary cavity to other parts of the mandible. Adenoid Cystic Carcinoma is well known for its prolonged clinical course and its tendency for delayed onset of distant metastasis, usually to the lungs (1-4) . This might explain the absence of both regional and distant metastasis in the present case despite the extensive nature of the tumour within the mandible.

Histologically, ACC can be categorised into three growth patterns ; cribriform, tubular and the solid pattern. The cribriform and tubular pattern of growth are associated with better prognosis (1-4).

Brookstone and Huvos (6) have however argued that clinical staging is a more reliable prognostic tool than histological staging. According to them, lesions that are located within undisturbed, intact bone and overlying periosteum with no evidence of cortical bone expansion offer the best prognosis and is therefore categorised into stage I. Stage II disease is characterised by lesions surrounded by intact cortical bone that has undergone some degree of expansion but no cortical bone perforation. An instance of cortical perforation, breakdown of overlying periosteum or nodal metastatic spread is categorised as stage III and has the worst prognosis.

The present case can be categorised into stage III disease based on the above criteria and was therefore treated by radical surgery (total mandibulectomy with removal of adjacent soft tissues) with no form of reconstruction as well as immediate post-operative radiotherapy as suggested by various reports (1-7) in the literature. Although resection without reconstruction or the use of only reconstruction plate as a reconstructive modality may not constitute standard of care especially in advanced Centres of Europe, North America and even India where free microvascular transfers are available and accessible. Pedicled musculocutaneous flaps could have been used for plate coverage in resource limited settings like ours, however the need for adjuvant radiotherapy may limit the success of this option. The main limitation in the use of reconstruction plate as the only reconstructive modality is exteriorisation of the plate either into the mouth or extraorally. This occurs in cases where there is poor muscle coverage, which most of the time has been excised following tumour infiltration in ACC.

Extensive soft tissue excision was done in the present case considering the involvement of the lateral aspects of the tongue and the adjoining soft tissues on the right side. The use of frozen section which was not available in our setting would have been very necessary for a more accurate intra-operative determination of tumour free margins, however all macroscopically involved tissues as well as a margin of presumed tumour free borders was excised. These margins were later reported as oncologically tumour free margins.

Reports 1-8 suggest that the best results in the management of central ACC have been the combination of radical surgery with post-operative radiotherapy, therefore post-operative irradiation has been advocated by some surgeons to ensure locoregional control particularly in those with advanced tumours or in those with distant metastasis, as well as those with histologically positive margins.

According to reports (2-7) the long term prognosis for this disease is poor and therefore, patients must be followed for much longer period than five years before assuming that a permanent cure has been obtained. For example, the survival rate for ACC of major salivary glands is between 75-80% at five years, about 40% at ten years and only about 15% or less at 20 years (2,5,7).

In conclusion, the clinical behaviour of central ACC may be seen as a paradox,five tumour growth is insidious and slow but its clinical course is relentless and progressive. Metastatic spread to regional lymph nodes is uncommon but distant spread to the lungs and bones is frequent. Also a five year survival rates are optimistically high but 10 to 20 year survival rates are dismally low, therefore early diagnosis, a prompt radical surgical intervention and adjuvant radiotherapy with a long term follow up is the key to achieving success in the management of this complex disease.

REFERENCES

1. Gumgum, S., Oner, B. and Ustay, C. Rapidly growing adenoid cystic carcinoma in mandible: A case report. *Oral. Oncol. Extra.* 2005:**41**:142-145
2. Thorakkal, S., Vengal, I., Pallikand, M.S. and Sivasankar, S. Primary Intraosseous adenoid cystic carcinoma of the mandible with lung metastasis: A case report. *J. Oral. Sci.* 2008:**1**:95-98
3. Al-Sukhun, J., Lindquist, C., Hietanea, J., Leivo, I. and Pentilla, H. Central adenoid cystic carcinoma of the mandible: A case report and literature review of 16 cases. *Oral. Surg. Oral. Med Oral. Pathol. Oral. Radiol. Endod.* 2006:**101**:304-308.
4. Vinuth, D.P., Agarwal, P., Dhirawani, R.B., Dube, G. Atypical case of primary intraosseous adenoid cystic carcinoma of mandible. *J. Oral. Maxillofac. pathol.* 2013:**3**:436-439.
5. Bradley, P.J. Adenoid cystic carcinoma of the head and neck: a review. *Curr. Opin. Otolaryngol. Head. Neck. Surg*. 2004:**12**:127-132.
6. Brookstone, M.S. and Huvos, A.G. Central salivary gland tumours of the maxilla and mandible:A clinicopathologic study of 11 cases with an analysis of the literature. *J. Oral Maxillofac. surg.* 1992:**50**:229-236.
7. Chen, M.A.,Bucci, K.M.,Weinberg, V.*et al*. Adenoid Cystic Carcinoma of the head and neck treated by surgery with or without post-operative radiation therapy: prognostic features of recurrence. *Int. J. Rad. Oncol. Bio. Phy.* 2006:**66**:152-159.
8. Thiago de Santana Santos, Daniela Guimaraes de Melo, Ana Claudia Amorim Gomes, Emanuel Savio de Souza Andiade, Emanuel Dias de Oliveira Silva. Adenoid Cystic Carcinoma of the mandible. *Braz. J. Otorhinolaryngol.* 2011:**77**:807.