Intracranial Extension Of Giant Multicystic Maxillomandibular Ameloblastoma: A Case Report
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Abstract
Maxillomandibular Ameloblastoma (A) is a slow growing but locally aggressive odontogenic tumour with a high recurrence rate, which can cause severe facial disfigurement and functional impairment. We report a case of histologically confirmed giant multicystic maxillomandibular A with intracranial extension in a 35-year- old woman of the Yoruba ethnic group in Nigeria, which measured 24 x 19 x 15 cm in diameter. Intracranial extension of the multicystic subtype of A is extremely rare. This to the best of our knowledge is probably the sixth recorded intracranial extension of a multicystic A. She had a recurrence after a year of mandibulectomy. She delayed to seek medical attention for five years after the recurrence, during which the A grew to an enormous size with the complaints of pain, severe headache and loss of vision in the right eye. The radiologist plays a crucial role in the diagnosis and treatment of A. We advocate early radical surgery from the onset to prevent recurrence in multicystic A, and long term follow-up for early detection of possible relapses.

INTRODUCTION
Ameloblastoma (A) is an odontogenic epithelial tumour affecting the mandible and maxilla, which is slow growing but locally aggressive1. It can cause severe facial disfigurement and functional impairment1. It is the second most common odontogenic neoplasm after odontoma2. It has a high rate of recurrence of up to 80%; and recurrences have occurred as late as 50 years after resection3-4. There are three histological variants of A namely plexiform, follicular, and granular cell variants. A is much more common in the mandible than in the maxilla, especially in the premolar-molar region, exhibit no sex predilection, and the most common histologic type is follicular A5. Based on biological behavior and histopathologic characteristics, maxillomandibular A has been divided into three subtypes, namely solid or multicystic A, unicystic A, and malignant A6. Intracranial extension or metastasis of all the three subtypes of A is extremely rare. To the best of our knowledge, intracranial extension of multicystic A has only been reported in five patients in the medical literature. Oka K et al reported the first case of mandibular A with intracranial extension in 19867. Phillip et al described intracranial extension of A in a patient in 19928. Zarbo et al reported intracranial metastasis in a patient with A in 20039. Hughes et al from the United States of America described a giant A that measured 15.2 x 11.4 x 12.0 cm with no intracranial extension10. Dunn J L et al reported a giant A which measured 17 x 15 x 13 on CT, but no intracranial extension was observed11. We report a case of giant maxillomandibular A with intracranial extension in a Nigerian woman.

CASE REPORT
TM is a 35 year old woman who presented at the Oral and Maxillofacial Surgery Unit of the Lagos State University Teaching Hospital, Ikeja, Lagos, Nigeria, with a ten year history of right cranio-mandibular swelling (Figures 1 and 2). She noticed this was associated with pain and severe headache, a few years ago. There was also progressive loss of vision in the right eye. The swelling was first observed in the right lower jaw at the onset. She had right segmental mandibulectomy four years after the onset of symptoms at a private hospital in Lagos, and the diagnosis of A was histologically confirmed. She had a recurrence of tumour growth a year after surgery. The swelling has since been slowly and progressively increasing in size to the current size. She is married with children.

Physical examination revealed a depressed-looking woman, in no obvious distress, mildly pale, afebrile and not jaundiced. There was total loss of vision in the right eye with no light perception. Extra-oral examination showed a mass located at the right craniomandibular region with proptosis.
The mass was non-tender, measured 24 x 19 x 15 cm in diameter, and was fluctuant to soft in consistency. Intra-oral evaluation showed a mild right mandibular buccal swelling with missing 48-43 teeth. A clinical diagnosis of Recurrent A was made.

Computed tomography (CT) scan showed a large, expansile, predominantly cystic mass measuring 165 x 117 x 112 mm in diameter in the right infratemporal retromaxillary region with internal hypodense, nonenhancing component, and enhancing peripheral and solid components. This is reminiscent of the soap bubble appearance. Fluid-debris levels were noted in some areas of the mass, but there was no gas to suggest infection. Pressure effect on the ipsilateral maxillary antrum and ethmoidal sinuses was noted, with extension into the pterygopalatine fossa posteriorly, and the right lateropharyngeal and masticator spaces medially. There was proptosis with erosion of the bony wall of the orbit, pterygoid plates, and squamous temporal bone on the right. The postnasal space was obliterated with the mass expanding the sphenoidal sinuses, optic canal, and superior orbital fissure on the right. The sella was also eroded along with the ipsilateral clinoid process. Extra-axial, intracranial extension of the mass through the eroded squamous temporal bone, into the right middle cranial fossa was observed, with mass effect on the right lateral ventricle and septum pellucidum. The ipsilateral Sylvian fissure, basal cisterns, and the cerebral sulci were all effaced. Right hemi-mandibulectomy was noted. The CT diagnosis was Recurrent A (Figures 3 – 13).

She had a successful trucut biopsy under CT guidance, and the tissue was sent for histological examination. Histological sections showed irregular tissue mass of scattered follicles and sheets of neoplastic epithelial cells presenting ameloblastomatous feature of peripheral palisading tall columnar cells, and central stellate reticulum-like cells. Some of these cells showed evidence of squamous metaplasia. The histological diagnosis was Follicular A with Acanthomatous changes (Figure 14).

Currently, she is been planned for possible multi-disciplinary team surgery between the maxillofacial surgeon, neurosurgeon, ophthalmologist and the otorhinolaryngologist.
**Figure 3**
Figure 3: Giant Ameloblastoma. Noncontrast axial CT scan, showing erosion of the wall of the right maxillary antrum (up arrow) and encroachment of the right lateropharyngeal space (right arrow) by tumour.

**Figure 4**
Figure 4: Giant Ameloblastoma. Axial bone window CT scan showing extent of tumour (down arrows).

**Figure 5**
Figure 5: Giant Ameloblastoma. Contrast-enhanced axial CT scan, showing multiple cysts (down arrows) in tumour and enhancing solid components (up arrows).

**Figure 6**
Figure 6: Giant Ameloblastoma. Noncontrast-enhanced axial CT scan, showing extra-axial intracranial extension of tumour into the right middle cranial fossa (up arrow).
Figure 7
Figure 7: Giant Ameloblastoma. Contrast-enhanced axial CT scan, showing extra-axial intracranial extension of tumour into the right middle cranial fossa (up arrow).

Figure 9
Figure 9: Giant Ameloblastoma. Axial bone window CT scan showing destruction of the right squamous temporal bone by tumour (left arrow).

Figure 8
Figure 8: Giant Ameloblastoma. Coronal Nonenhanced CT scan, showing intracranial extension of tumour through the eroded squamous portion of the right temporal bone (down arrow) into the right middle cranial fossa.

Figure 10
Figure 10: Giant Ameloblastoma. Three-dimensional volume rendering CT scan of the face in soft tissue window (anterior projection), showing extent of tumour and marked distortion of the face.
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Figure 11
Figure 11: Giant Ameloblastoma. Three-dimensional volume rendering CT scan of the back of the head and neck in soft tissue window (right posterior oblique projection), showing extent of tumour.

Figure 13
Figure 13: Giant Ameloblastoma. Three-dimensional volume rendering CT scan of the face showing extensive destruction of the right hemi-maxilla, and lateral wall of the right orbit (anterior projection) in bone window.

Figure 12
Figure 12: Giant Ameloblastoma. Three-dimensional volume rendering CT scan of the face in soft tissue window (right anterior oblique projection), showing extent of tumour and marked distortion of the face.

Figure 14
Figure 14: Ameloblastoma. Photomicrograph of a hematoxylin-eosin stain of the tissue (original magnification, x400), showing islands of scattered follicles of proliferating odontogenic epithelium (right arrows), and immature fibrous stroma (up arrows)

DISCUSSION
The differential diagnosis of lytic bone lesions of the jaw has been well described\(^1\). The radiologist plays a pivotal role in the diagnosis and treatment of A\(^1\). The recurrence rate is related to surgical technique, age of the patient, and the cystic characteristics of the tumour\(^1\). Effective en-bloc resection of A depends on adequate delineation of the tumour through the combined use of radiography, CT with
three dimensional reformations, magnetic resonance imaging (MRI), and angiography. Higher recurrence rates are observed in older patients, those with multicystic tumours, and those who exhibit soap bubble appearance. Our patient, though young at the time of initial presentation, has a multicystic A with soap bubble configuration. There was no evidence of CT, MRI, or angiographic evaluation to delineate the tumour prior to her surgery. Our patient did not experience local sepsis, bleeding or ulceration from the site of tumour, which is the pattern in giant A; and therefore had no laboratory evidence of leucocytosis, iron deficiency anaemia or hypoproteinaemia.

Although surgery is the mainstay of treatment of A, the extent of surgery is controversial. Radical resections which include marginal and segmental mandibulectomy result in local control rates exceeding 90%, while conservative procedures like enucleation and/or curettage result in local control rates of approximately 80% and 50% for unicystic and multicystic A respectively. Most cases of A are of the multicystic subtype, and these are more difficult to manage than the unicystic subtype. Though radiotherapy may achieve local control in the occasional patient with incompletely resectable tumour, Mendenhall and colleagues advocate wide en bloc resection as the optimal treatment of A. Chemotherapy has no place in the management of A. Sammartino G et al developed a new decisional algorithm in managing A in 2007. They recommended an initial curettage, subsequent post-operative follow-up, and finally early radical surgery for recurrence. This algorithm may suffice for unilocular A which has a relatively lower recurrence rate. For multicystic A, the treatment should be radical surgery ab initio to prevent recurrence. Our patient failed to attend follow-up review for six years. Recurrences have been associated with absconding from reviews especially in our environment in Nigeria. Failure of patients to attend follow-up review promptly and regularly is a mitigating factor against nipping early recurrences in the bud, before they attain enormous size. There are challenges in the surgical management of giant A in developing countries with mortality rates as high as 9%, apart from post-surgical-related psychosomatic problems and reduced quality of life.

CONCLUSION

Only five cases of A with intracranial extension have been previously described in the medical literature, to the best of our knowledge. Intracranial extension or metastasis of all three subtypes of A is extremely rare; and this is probably the sixth recorded intracranial extension of a multicystic subtype A in the literature. We advocate early radical surgery from the onset to prevent recurrence in multicystic A, and long term follow-up for early detection of possible relapses. The surgical management of giant A can be challenging in developing countries like Nigeria.

References

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