CASE REPORT

Giant ameloblastoma mortality; a consequence of ignorance, poverty and fear

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SUMMARY

Ameloblastoma is a benign tumour of odontogenic origin. Although locally invasive, delay in treatment can lead to severe disfiguring of the facial region. In this report, we present two cases, a man and a woman, of a large ameloblastoma that presented with the typical radiographic features of variably sized radiolucent loculations. Patients presented initially with relatively small tumours but due to fear of surgery, ignorance and possibly poverty, tumour growth became enormous. The female patient died while awaiting consent for surgery and the male patient died at home due to complications closely associated with fear and growth of tumour. Literatures on mortality of ameloblastoma are reviewed. The need for more public enlightenment on the nature and treatment of facial tumours is highlighted.

BACKGROUND

Ameloblastoma, being largely a benign lesion, has good prognosis when treated early. However, ignorance and fear in our patients and relations led to delay in accepting surgical intervention thereby leading to preventable death of the patients.

CASE PRESENTATION

Ameloblastoma of the jaws is the most commonly encountered odontogenic tumour in Africa^{1–3} and Asia.^{4–5} It may arise from the enamel organ, remnants of dental lamina, the lining of an odontogenic (dentigerous) cyst or possibly from the basal epithelial cells of the oral mucosa.⁶ Ameloblastoma is rare worldwide.⁷ Its peak incidence is in the third to fourth decades of life and has an equal sex predilection. Approximately 80% of ameloblastomas occur in the mandible, usually in the posterior region.⁸ A notable exception is African Blacks, in whom any region of the mandible may be involved.⁹ It often presents as a slow growing tumour expanding the cortical bones, sometimes perforating them and involving the adjacent soft tissues. Patients may present with malocclusion, loose teeth or more rarely with parasthesia and pain. The associated unerupted tooth/teeth may be displaced and resorption of the roots of adjacent teeth is common.¹⁰ Owing to its initial painless nature and slow growth, many affected patients present to the hospital when the tumour has become enormous, hence the giant size. This is the case with the two patients presented here. From these two cases, we found that lack of resources, fear of surgery and even ignorance contributed to the delay in presentation. Consequently, there is high morbidity and mortality associated with this

benign lesion in our environment. Although a benign lesion the architectural pattern of the ameloblastoma is such that the border of the tumour within the cancellous bone lies beyond the apparent macroscopic surface and the radiographic boundaries of the lesion,¹¹ hence many authors suggest radical surgery as treatment.

Various factors such as, the dimensions and the location of the lesion, the growth rate and the relationship with the nearby structures, the histological type, the clinical characteristics, the recurrences, the general conditions of health and the age of the patient have influenced the decision to use conservative or radical approach to treatment.¹² However, the histological aspect has a decisive role as the solid multicystic ameloblastoma has a higher recurrence rate than the unicystic-intramural ameloblastoma. Some authors¹³ found the location of the lesion to be an important risk factor for the recurrence of ameloblastoma; the posterior of the jaw having recurrence that is more frequent. Some other authors¹⁴ consider ameloblastoma as a locally destructive and invasive tumour that can recur despite adequate surgical removal.

Few cases of ameloblastomas with metastases have been reported. Although it is locally aggressive with a high recurrence rate (50–72%), the fact that ameloblastoma rarely metastasises explains why it is considered benign.¹⁵ The lung is the most frequent site of metastases, occurring in up to 88% of disseminated cases. The next most frequent metastatic sites are regional lymph nodes, pleura, vertebra, skull, diaphragm, liver and parotid glands.¹⁶

Delayed treatment can result in significant morbidity from facial deformity and infiltration of adjacent tissues. Mortality can occur from invasion of vital structures in the head and neck, super infection, recurrent and even distant metastases.⁷ The case series presented here depicts the adverse effect of delay in presenting to hospital for treatment due to fear of surgery, ignorance and sociocultural belief, with consequent loss of lives.

Case 1

A 29-year-old woman, from a polygamous background came to the hospital with a 5-year history of swelling in the mandible. Swelling was slow growing, painless with no associated weight loss. Extraoral examination showed a healthy looking young woman with a firm swelling on the anterior part of mandible with some extension bilaterally measuring 9 cm × 6 cm (figure 1). The overlying skin appeared normal. Intraoral examination showed a full complement of upper teeth. There

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Figure 1 Case 1: lesion at presentation.

was a buccolingual swelling extending from lower right second molar region to the lower left first molar region. All the lower anterior teeth were mobile and displaced labially and lingually. Overlying mucosa appeared normal but highly haemorrhagic towards the posterior region. A provisional diagnosis of ameloblastoma was made with differential diagnosis of an odontogenic keratocyst. Aspiration of the lesion yielded a blood-stained straw-coloured aspirate. Investigations carried out included radiographs (PA skull and oblique lateral), and incisional biopsy. Radiographs showed multilocular radiolucency extending from the distal aspect of lower right second molar to the mesial aspect of lower left second molar. Other vital signs included blood pressure 110/60 mm Hg, pulse 78/min and respiration 17/min. Histology report revealed the follicular type of ameloblastoma ([figure 2](#)). The microscopic examination showed a benign lesion composed of epithelial islands consisting of outer palisaded ameloblast-like cells and inner stellate cells surrounded by fibrocollagenous connective tissue stroma. The mode of treatment was explained to the patient and her relations. The patient was booked for surgical excision and autogenous bone graft under general anaesthesia but she did not show up for admission.

However, the patient presented 3 years later, with her relations who aided her to the hospital, as she was too weak to walk alone. Enquiry revealed that she had been under native and herbal treatment. Further enquiry revealed that most of the immediate family relations (who had consulted oracle) were made to believe and have the fear that patient would die during the surgical treatment, hence the move for alternative herbal treatment. She had not been feeding well due to lack of

Figure 2 Case 1: photomicrograph of the lesion.

Figure 3 Case 1: size of the lesion 3 years later.

resources and tumour size. Examination showed a chronically ill-looking woman. The patient was not in obvious respiratory distress but uncomfortable due to the large size of the tumour. The patient was pale, anicteric and afebrile. There was facial asymmetry due to a massive swelling extending from the right ramus to the left body of the mandible. It was oval in shape and also extended from the right preauricular region of the face protruding downward to overshadow the neck, the clavicle and the upper part of the chest. It measured 50 cm × 28 cm, and it was about two-third the size of the patient's head ([figure 3](#)). The overlying skin appeared shiny. Numerous scarification marks dominated the skin on the body of the mandible. There were no palpable lymph nodes. There was a sinus discharging pus at the right molar region. The lower and upper lips were incompetent. Intraorally, the oral hygiene was poor with frank halitosis. The lesion obliterated the floor of the oral cavity and the buccal sulcus on both sides. Overlying mucosa appeared very haemorrhagic. All the molars and premolars on the right side were absent. The lower left molars, premolars and canine were very mobile. There were indentations of upper teeth on the mucosa overlying the lesion. Investigations revealed the following: haemoglobin 9 g/dL and white cell count 12 000/dL. Radiographic examination showed multilocular radiolucency extending from the right condyle to the left ramus of the mandible. There was resorption of the roots of premolar and canine ([figure 4](#)). The patient could not afford a CT scan. She was admitted to the surgical ward. To combat the infection, she was given a full course of antibiotics and analgesics. As part of preparation for surgery the patient was transfused with two units of blood. Anaesthetist reviewed the patient and necessary pre-operative preparations were made in readiness for surgery. However, the relations of the patient were reluctant to give consent for surgery due to fear of death during surgical operation. About 3 weeks later, there was sudden profuse bleeding from the sinus track on the lesion and attempts at salvage failed and the patient died.

Case 2

A 56-year-old man presented with a massive swelling on the mandible of 11 years duration. He gave a history that 9 years earlier, he visited one of the teaching hospitals in the Western part of the country where the lesion was diagnosed as ameloblastoma after an incisional biopsy. Then, the swelling was like the size of a tennis ball. He was booked for a surgical excision of the lesion under general anaesthesia. On the morning of the surgery day, while waiting in the surgical ward, someone, a visitor confronted him with the question, 'Have you tried other alternatives before coming for this operation?' This question brought fear into him, made him postpone the surgery in order to look for the 'other

Figure 4 Case 1: radiograph of the lesion.

alternatives'. Nine years later, he visited our centre as he could not find other alternatives. His facial expression portrayed a hidden anxiety and fear associated with his condition. Extraoral examination (figure 5) showed an obvious facial asymmetry with swelling on the mandible which extended from the right body to the left ramus. It was oval in shape and measured about 42 cm × 30 cm. Anteriorly, it protruded forwards casting a shadow over the upper part of the neck and the chest region (figure 6). Overlying skin appeared normal. Lesion was hard to touch at the right and left body of the mandible. It was firm at the angle of the mouth and egg shell crackling effect at the anterior region. No labial paraesthesia was elicited. Intraoral region showed a full complement of upper teeth. On the lower jaw only the lower left molars were present. There was a buccolingual expansion of the mandible extending from the right second molar to the left ascending ramus. Overlying mucosa appeared normal. Associated teeth were mobile. There was no area of ulceration over the lesion. A provisional diagnosis of giant ameloblastoma was made. Aspiration biopsy yielded a straw-coloured fluid. Radiological investigation showed a multilocular radiolucency extending from the distal aspect of lower right third molar to angle of the left mandible and truncation of the roots of buried lower left third molar (figure 7).

Figure 5 Case 2: front view of the lesion.

Figure 6 Case 2: side view of the lesion.

The patient could not afford CT scan. He was educated on the general nature of the condition and the need for a second incisional biopsy to confirm the initial diagnosis. He was then booked for incisional biopsy prior to surgical intervention. However, the patient failed to show up for the biopsy as he was afraid of any surgical intervention. About 12 months later, enquiry from a close relation to the patient revealed that the patient died at home from a systemic condition closely associated with fear of surgery.

INVESTIGATIONS

- ▶ Incisional biopsy
- ▶ Aspiration biopsy
- ▶ Radiographs

DIFFERENTIAL DIAGNOSIS

Odontogenic keratocyst

OUTCOME AND FOLLOW-UP

The patients died prior to surgical intervention.

DISCUSSION

Ameloblastoma is a tumour that is slow growing and in most cases, painless. The consequence of this is that those affected

Figure 7 Case 2: radiograph of the lesion.

are usually reluctant to seek medical or dental care. This is one of the reasons why the lesion in the patients reported here became giant in size. One report¹⁷ stated that the most powerful forces known to man are not nuclear weapons, nor nature's awesome wonders, such as the might of an earthquake, the power of the sun or mastery of a hurricane, but the thoughts and ideas of the mind. Moreover, many subconscious have linked a whole load of powerful negative emotions, mainly fear to the very thought of surgery. This is the main issue of the cases presented here. Cases of fear of surgical operations are usually (although not always) caused by an intense negative experience or history from the past. Nevertheless, the mind can also create that fear seemingly without basis or evidence. Being scared of surgery can lead to eating disorders, sleep disturbances, regression to earlier levels of behaviour and even depression.¹⁸ Some patients with intense fear of surgery may experience breathlessness, excessive sweating, nausea, dry mouth, shaking, heart palpitations, inability to speak or think clearly, a fear of becoming mad or losing control, a sensation of detachment from reality or a full-blown anxiety attack. Although our patients did not exhibit all the attributes of fear stated above, the refusal to sign consent (as seen in case 1) and refusal of surgery (case 2) was based on fear.

A report¹⁹ of 109 patients with ameloblastoma showed that certain factors such as prolonged incision and application of native medicine by herbal doctors were responsible for patients' delay in presenting to hospital. In case 1, history revealed that the patient had consulted the native doctor and the numerous scarification marks confirmed prior intervention by native doctors. It is still a common practice in our environment for patients and their relations to consult diviners and spiritual healers before, during and even after visiting the hospital. This has a negative effect on the management of disease conditions as seen in the cases presented here.

Mortality in ameloblastoma is a rare occurrence. Reported cases of mortality have attributed it to metastasis or/and malignant changes of the lesion, especially during a protracted course.²⁰ The female patient in this series had an aggressive increase in the size of the tumour within the space of 3 years. This could be attributed to the multicystic type of ameloblastoma as this tumour is often locally aggressive and has a significant impact on a patient's morbidity and mortality.²¹ The cause of death in the female patient may have been from infection, metastatic spread or fear associated with the tumour size and surgery. The infection was arrested with broad-spectrum antibiotics. A second biopsy could have ruled out or confirm the possibility of malignant transformation or metastasis. Metastatic tumours occur usually in cases of long duration with multiple surgical procedures or radiation therapy²² which was not the case with this patient. However, the concept of haematogenous spread (in which tumour cells were found invading blood vessels) and aspirated neoplastic cells have been suggested²³ as the cause of pulmonary metastases and possibly death. Some authors²¹ suggested that extensive local disease and mandibular focus of the primary tumour also tend to be associated with the development of metastases while the incidence of lymph node metastatic disease argues for lymphatic dissemination as well.

The effect fear has on the physiological operation of the body has been well documented by medical science and psychology. Thoughts, ideas and emotions alter the body's biochemistry and the health of the body. Excess fear or any other negative attribute elicits a negative response in the soul as correlated to negative reactions in the physical body. This is most probably the cause of death of the male patient. He could not come to terms with the surgical

excision of the lesion he had for almost 12 years. Perhaps more enlightenment would have given him the confidence needed to accept the surgical management of the lesion.

Learning points

- ▶ Low socioeconomic status (poverty) of patients hinder access to optimum healthcare in our environment.
- ▶ Fear of surgery is still a major problem among patients and relations. It prolongs the time of presenting to the hospital for treatment.
- ▶ Treatable benign lesion can lead to mortality when treatment is not instituted early.
- ▶ Visit to native doctors/spiritual diviners is a common practice in our environment and this delay patients' decision to seek medical attention.
- ▶ More health enlightenment programmes are required to reduce the fear associated with surgical operations.

Competing interests None.

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