Case report:

Untreated Giant Ameloblastoma of Jaw - Diagnostic and Management Dilemma

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Abstract:

Ameloblastoma is a benign locally aggressive odontogenic epithelial neoplasm. It is slow growing tumour and mostly of benign behaviour. The most frequent location is at lower molar region and rarely involve upper jaw or maxillary sinus. Patients are usually asymptomatic until reaching a certain large size. Delayedtreatment is relatively common in our society. Patients mostly seek for treatment when a large tumour size is achieved. Herein we report a case due to its rarity, giant size, imaging challenge and management, which involves radical resection and free flap reconstruction.

Keywords: Giant ameloblastoma, odontogenic epithelial neoplasm, computed tomography.

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Introduction

Ameloblastoma usually presents as painless, slow-growing tumour that causes expansion resulting in perforation of the cortical bone. It has an aggressive behaviour and recurrent course. It is rarely metastatic. In developed countries, ameloblastoma is usually treated early before growing in size. Metastases are known to occur in 2-5% of cases and 80% involvethe lung[1].

Radiographically, it appears as an expansile, radiolucent lesion with thinned cortices, giving characteristic soap bubble-like appearance.

In the recent histologic classification of odontogenic tumours from the World Health Organization (WHO), ameloblastoma is defined as a benign, locally invasive epithelial odontogenic neoplasm of putative enamel organ origin. It represents approximately 10% of all odontogenic tumours[2].

We present a case of giant ameloblastoma of mandible in a 52-year-old Malay gentleman causing facial disfigurement. The challenge in managing this case is a radiological dilemma in differentiating between types of ameloblastoma on plain radiograph and computed tomography (CT) as well as to provide complete tumour removal and reconstruct the bony defect by oral maxillofacial team in order to give a better cosmetic outcome for him.

Case report

A thin built 52-year-old Malay gentleman with no underlying medical illness presented with painless lower jaw swelling in the last19 years which was massively increased in size and appearance. He started experiencing pain over the lower jaw 3 weeks prior he presented to our medical center.

He had history seek of treatment previously during the initial presentation, however he

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refused any surgical intervention and opted for conservative treatment. Subsequently, his lower jaw mass massively growing. Otherwise, he had no problem related to articulation and deglutition. No complaint of breathing difficulty. No fever, skin changesordischarge.

On general examination, he appeared healthy and not cachexic. Clinically, large and elongated chin (Fig.1). Large mass involving entire mandible and hard inconsistency. No lip paraesthesia. Overlying skin was normal. No pus discharge or bleeding seen. Mouth opening limited (2 finger breadth). Both temporomandibular joint movements were good.

Intraoral examination noted raised buccal sulcus from angle to angle and erythematous. No pus discharge seen. Tongue movement intact.

Figure 1: (A) anterior view and (B) lateral view. (Patient presented with lower jaw swelling. Clinically, large and elongated chin.)

A lateral radiograph of mandible shows an ill-defined expansile lytic lesion with soap bubble appearance involving the body and angle of mandible, and erosion of alveolar process. The molar tooth was displaced anteriorly (Fig.2). Maxilla and condylar process of mandible were spared.

Contrasted computed tomography (CT) of neck was done, which revealed large expansile multiloculated lytic lesion involving mental protuberance, body as well as the angle of mandible with cortical disruption and erosion of alveolar process of mandible. It was measured 7.2cm x 11.8cm x10cm with area of central necrosis. Presence of small air locule within the body of left mandible, likely representing infective process. No air fluid level within. Adjacent temporomandibular joints were preserved(Fig.3). Enlarged left submandibular node. No intracranial extension. Bilateral submandibular and parotid glands were normal. No narrowing of trachea. CT thorax showed a solitary lung nodule. Hence, provisional diagnosis was given as ameloblastoma. However, in view of the huge lesion with a solitary lung nodule, ameloblastic carcinoma or malignant ameloblastoma still cannot be totally excluded.

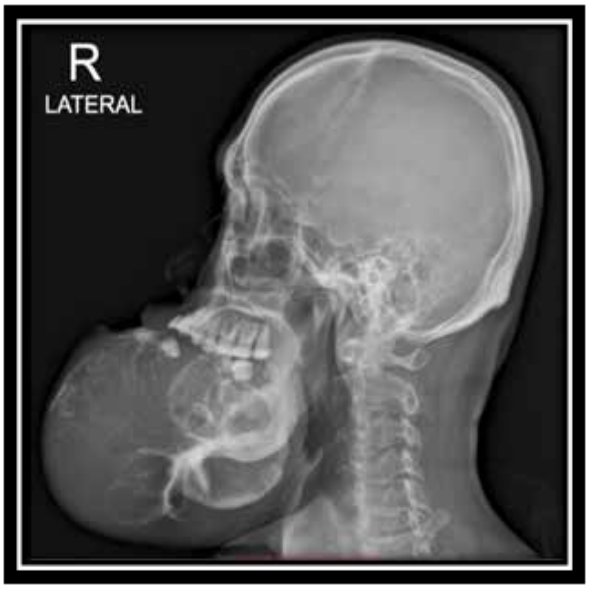

Figure 2: Mandible radiograph (lateral) (An ill-defined expansile lytic intramedullary lesion with soap bubble appearance involving the body and angle of mandible. Erosion of alveolar process. The molar tooth was displaced anteriorly.)

The patient was then subjected to radical neck resection, resection of mandible followed by mandible reconstruction with free subcutaneous fibular flap on the same day. Plating of mandible performed. Fibula bone harvested and moulded to the plate. Total 13 screws applied to his mandible reconstruction. Intraoperatively, temporary tracheostomy performed, and he was admitted to the surgical intensive care unit (SICU) for post operation. Clinically, he was stable throughout his intra and post operation.

Histopathological examination showed follicles and anastomosing cords of odontogenic epithelial islands with peripheral palisading of basal columnar cells within fibrous connective tissue stroma. Low and high magnification of central part of tumour islands lined by palisading columnar cells Squamous differentiation and cystic degeneration noted. Based on these findings, the final microscopic diagnosis was reported as acanthomatous ameloblastoma (Fig.4 and 5).

Postoperative period was also uneventful. He recovered well from extensive procedures without complication.

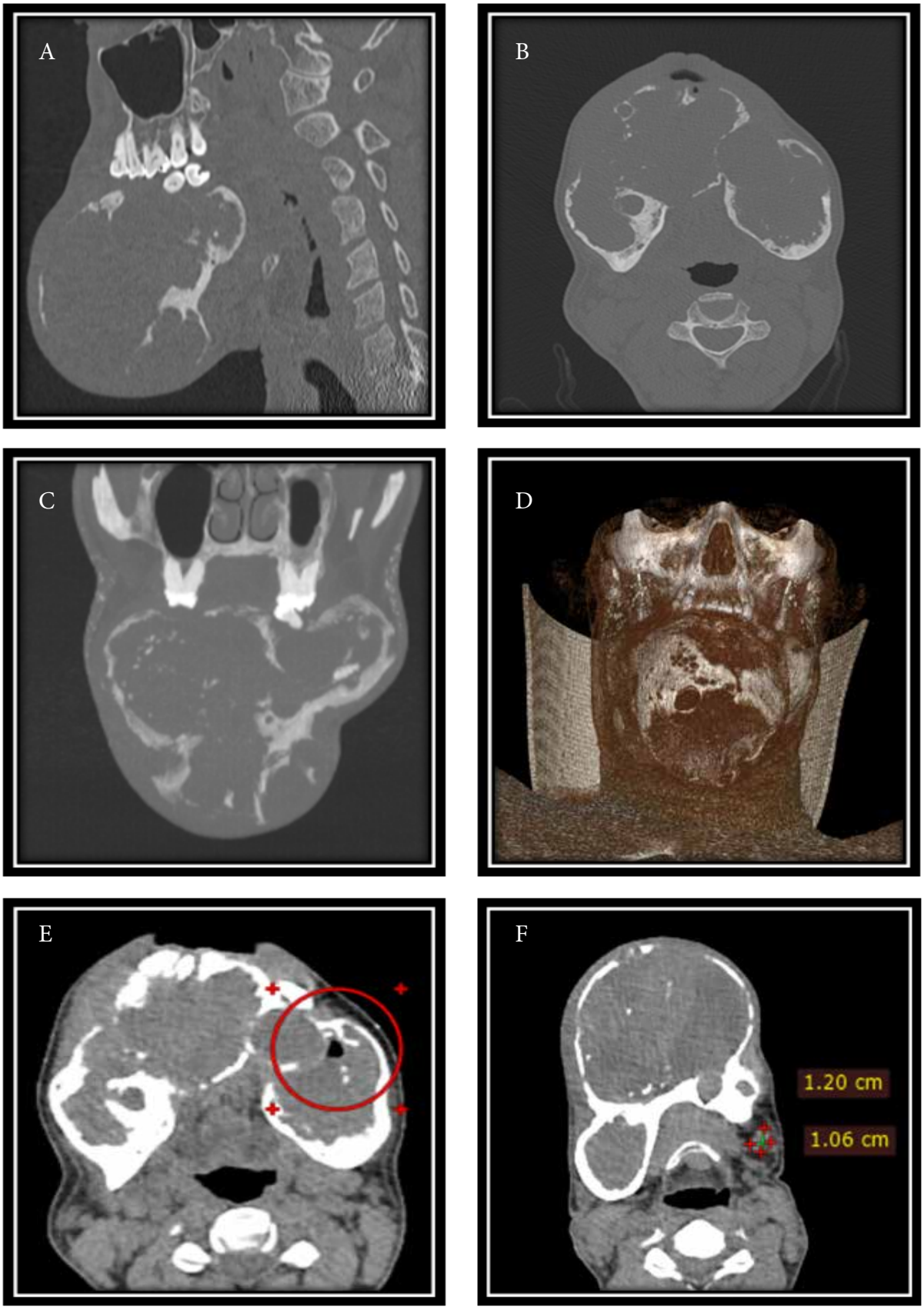

Figure 3: Mandible computed tomography. (CT (A) sagittal, (B) axial and (C) coronal and (D) 3D reconstruction. Large expansile multiloculated lytic lesion inmental protuberance, body and angle of mandible with cortical disruptionand erosion of alveolar process of mandible. No air fluid level within.In soft tissue setting (E) Small air locule within the body of left mandible (F) Enlarged left submandibular node.)

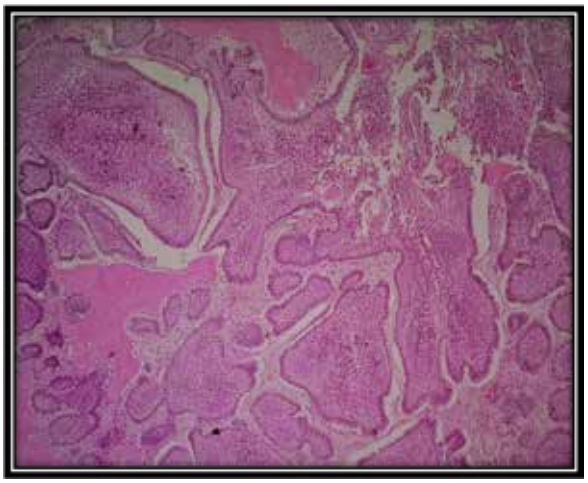

Figure 4: H & E micrograph of the tumour. (Follicles and anastomosing cords of odontogenic epithelial islands with peripheral palisading of basal columnar cells within fibrous connective tissue stroma (objective magnification 10X).)

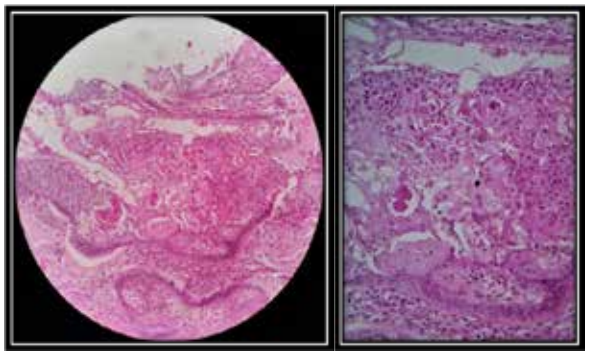

Figure 5: H & E micrograph of the tumour. Low and high magnification of central part of tumour islands linked by palisading columnar cells. Squamous differentiation and cystic degeneration noted (objective magnification 20X and 40X respectively). Based on these findings, the final microscopic diagnosis was reported as acanthomatous ameloblastoma.

Discussion

Ameloblastoma in mandible can progress to great size if left untreated and cause significant facial disfigurement, teeth displacement and malocclusion. Typically, itpresents asa painless, slow growing mass and often take a few years when patients start to have symptoms. When the tumour reaches a certain amount of size, they might experience pain, facial asymmetry and difficulty in chewing or other symptoms such as lip paraesthesia.

Giant ameloblastoma in the mandible israre. Multicystic ameloblastoma accounts for 10% of all odontogenic tumours inthe jaw[3]. Peak incidence occurs in the third to fifth decadesof life. It can progress to great size if left untreated and especially inpatients who have delayed treatment as in the case of thispatient because fear of surgery and financial concerns. A delayin treatment iscommon in this area because most local patients seekfor an alternative pathway beforethey present to us. He has delayed treatment for nearly nineteen years.

A timely and correct diagnosis with treatment requires a multidisciplinary approach. Standard CT imaging with three-dimensional reconstruction (3D) technique can benefit surgical planning by accurately assessing the extent of the disease. Magnetic resonance imaging (MRI) can also provide additional details regarding extraosseous components, location, solid component of the lesion and involvement of neurovascular structures. These can guide the managing team for surgical resection and mandible reconstruction planning[4]. Angiogram is also recommended to delineate the vascular supply. The embolization may be needed if presence of vascular supply to avoid bleeding during intraoperative.

The radiological diagnosis of giant ameloblastoma, ameloblastic carcinoma or malignant ameloblastoma is great challenge inview similar radiographics appearance. Radiographically, all these lesions can be radiolucent, uni or multiloculated with soap bubble appearance. The majority of tumours have well defined margins. Tooth root Resorption occurs when the tumour is adjacent to a tooth causing erosion of the tooth root and tooth displacement. If the tumour is infected, it will have small ai rloculeas seenin our patient.

Ameloblastic carcinoma is a rare malignant lesion, which is more aggressive than the typical ameloblastoma. Both can appear similar radiographically. However, imaging features such as erosion through the cortex with extension into the surrounding oral mucosa, extensive solid components, mixed solid and cystic components, and the occasional presence of focal dystrophic calcifications are more closely associated with ameloblastic carcinoma. It is also associated with high rate of local recurrence and metastasis, particularly to cervical lymph nodes[2]. Malignant ameloblastoma refers to metastatic benign ameloblastomas[5].

Although giant ameloblastoma israre, it was wellknown for its high recurrence if excision was incomplete. The mainstay of giant ameloblastoma surgical resection with wide free margin. Reconstruction of large mandible defects

following radial resection of giant ameloblastoma is a huge challenge to managing teams because the surgery involves head and neck structures as well as mandible function and cosmetic outcome of the facial appearance post operation. Mandible reconstruction postresection of tumour is very important as it affects patient's quality of life. Encouraging data were published on better outcomes regarding mandibular resection and reconstruction in extensive ameloblastoma with single stage procedures[6,7]. The free fibula flap as a source of vascularized bone in reconstructive surgery is widely used and has been demonstrated to be an ideal flap for mandibular reconstruction.

Aydin *et al.*(2004) and Bilkay *et al.* (2004) prefer fibular flap as first choice or mandibular reconstruction as it offers thick cortical bone which ensures a secure fixation of osteosynthesis plates and screws [8,9]. According to the authors, this is an ideal flap for insertion and osseo integration of dental implants.

The recurrence rate after resection is 13-15%, as opposedto 90-100% after curettage[10]. Eppley *et al.* (2002) and his analysis of 60 mandibular ameloblastoma cases showed no evidence of recurrence in those cases treated with en bloc resection compared to enucleation and curettage. The recurrence rate was very high reaching 25% to50%[11.]

Conclusion

Giant ameloblastoma in the mandible is benign, but locally aggressive odontogenic tumour. It is difficult to differentiate between benign and malignant changes when the lesion is too big. It is always challenging for a diagnostic and management when the patient presents late. The challenge in the management of giant ameloblastoma of the mandible is not merely complete removal of the tumour to prevent recurrence but also providing mandible reconstruction for patient's better quality of life. It needs multidisciplinary approach.

Conflict of interest

No conflict of interest has been disclosed by the authors.

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Authors Contributions

Conception and design: HZS, NAS. *Collection and assembly of data:* HZS, NAS, ATM, NAR, JAA. *Critical revision of the article for important intellectual content:* HZS, NAS, ATM, NAR, JAA.

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