

# B4. Ameloblastoma

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## Review of Ameloblastoma Case

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### ABSTRACT

Ameloblastoma is a benign epithelial neoplasm and ranges from 10% of the entire odontogenic tumor. Ameloblastoma is characterized by a slow growth pattern and can grow to a very large size and cause severe facial deformities. These tumors are most common at the age of the third and fourth decades, and most often occur in the posterior mandibular, especially in the third molar tooth, as well as associated with the impacted follicular or dental cyst. Classification of Ameloblastoma According to WHO distinguishes into benign ameloblastoma which include: (1) solid/multicystic ameloblastoma, (2) unicystic Ameloblastoma, (3) the peripheral (or extraosseous) ameloblastoma, (4) the desmoplastic ameloblastoma and malignant ameloblastoma based on the frequency sequence which includes: (a) metastasizing ameloblastoma, (b) Primary ameloblastic carcinoma, (c) Secondary intraosseous ameloblastic carcinoma, (d) secondary peripheral ameloblastic carcinoma. A radiological examination that can be conducted to diagnose ameloblastoma is plain photo, CT Scan and

MRI. The image of Meloblastoma radiography may vary. Some of the depictions of luent lesions are firmly, unilocular, well-orticated, which often relate to Corona tooth impacted or no eruption, so that it cannot be distinguished by odontogenic keratosis and dentigerous cysts on radiography. Some of the other, multilocular, internal septa and honey comb or soap appearance bubbles are often similar to the large odontogenic keratosis. However, only histopatological findings can help to determine the malignant tumors and the alteration of carcinomatose.

**Keywords:** Ameloblastoma, Neoplasma, Odontogenic tumor

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### INTRODUCTION

Ameloblastoma is a benign epithelial neoplasm and ranges from 10% of the entire odontogenic tumor.<sup>1</sup> Ameloblastoma is characterized by a slow growth pattern and can grow to a very large size and cause severe facial deformities.<sup>2</sup> These tumors are most common at the age of the third and fourth decades, and most often occur in the posterior mandibular, especially in the third molar tooth, as well as associated with the impacted follicular or dental cyst.<sup>1,3</sup>

Radiological tests that can be conducted to diagnose ameloblastoma are plain film, CT Scan and MRI. The description of radiographic ameloblastoma may vary.<sup>1,4,5</sup> Some reveal the image of radiolucency lesions firmly, unilocular, well-orticated, which often relate to Corona tooth impacted or no eruption, so that it can not be distinguished by odontogenic keratosis and dentigerous cysts on radiography.<sup>14</sup> Some of the other, multilocular, internal septa and honey comb or soap appearance bubbles are often similar to the large odontogenic keratosis. However, only histopatological findings can help determining the malignant tumors and the alteration of carcinomatose.<sup>1,4,5</sup>

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### CASE REPORT

A 25-year-old male was presented in the dental clinic with a complaint of asymmetric swelling on the right lower jaw. Patient stated that swelling was gradual on onset and

progressed in size in a 2-year course. There is surgery history of have been diagnosed as suspected ameloblastoma since 3 years in the other hospital and since then there is an unhealed extraction socket for which patient had consulted several dental practitioners by whom he was treated unsuccessfully without investigations and proper diagnosis. He did not have any history of other disease. Physical diagnosis: Patient was physically healthy and mentally alert. Asymmetrical face with bulging was seen on the right cheek. Extra orally there was ill defined solitary spherical swelling involving the right angle of mandible measuring  $\pm 7 \times 9$  cm which was noncompressible, nonreducible, nonmobile, nonfluctuant, and fixed to the underlying structure with the right mandible. No pain with open mouth 2 fingers. Intra orally there was buccal plate bulging and expansion from region of molar to posterior. The color some with the color of the surrounding mucosa tissue, unclear boundaries. Teeth 7 and 8 lower right jaw missing / post extraction, an ulcer appeared on the mucosa of the region. Clinical provisional diagnosis established was current ameloblastoma of right mandible. The patient was sent to the radiology department for further evaluation with contrast multi-detector computed tomography (MDCT) scan. CT scan results reveal, there is a large lytic lesions involving the mandible of which is mostly around body of the right mandible and ramus. It measures  $\pm 6.5 \times 8.5 \times 5.5$  cm. The expanded mandibular cortex is partially thickened and almost visible. No visible periosteal reaction. These lesions have cystic multistocular components and large amounts of internal

calcification are recorded in the ventral area (Figure 1 and 2) also internal solid enhancers post contrast (Figure 3 and 4). Visualisation of the lesion with Volume-rendered MDCT (Figure 5 and 6). None of the teeth involved in the lesion. The muscles and soft tissues surround this lesion were morphologically normal. This lesion morphology was similar to ameloblastoma.

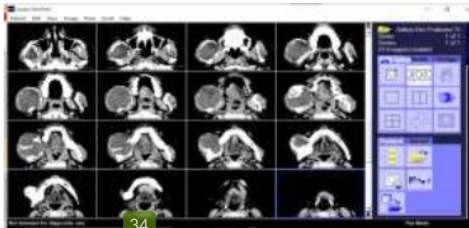


Figure 1: Non-contrast MDCT-scan axial view

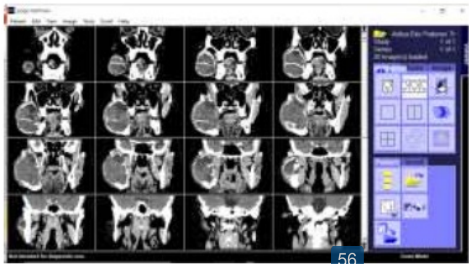


Figure 2: Non-contrast MDCT-scan coronal view

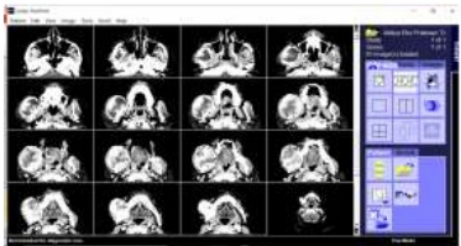


Figure 3: Contrast MDCT-scan axial view.

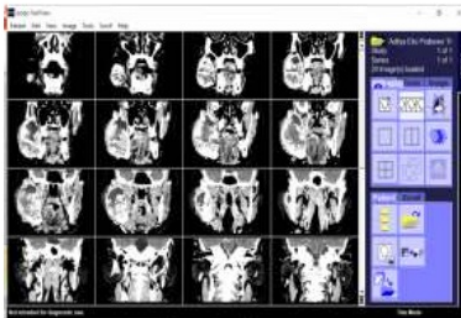


Figure 4: Non-contrast MSCT-scan coronal view

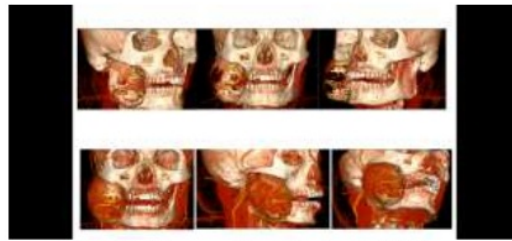


Figure 6: Volume-rendered Contrast MDCT image

## LITERATURE REVIEW

Ameloblastoma is derived from the English word "amel" meaning enamel and Greek word "blastos" which means seed, is a benign odontogenic tumor that is most common, derived from epithelium, is aggressive locally and has a high local recursive incident even after excision on the margins.<sup>56</sup> It was first introduced by Gorlin, who identified Cusack as the first person with this abnormality in 1827.<sup>17</sup> This neoplasm is derived from the enamel-forming cells of the odontogenic epithelium that failed to undergo regression during embryonal development, but the causal factor of specific causes of neoplasms in general is clearly unknown.<sup>1</sup> However, some scholars have assumed that some causative factors are considered to be the cause of a histodifferentiation disorder in Ameloblastoma including: (1) non-specific irritative factors such as extraction, caries, trauma, infection, inflammation, or a tooth eruption, (2) nutritional deficit disorder and (3) viral pathogenesis.<sup>13</sup>

According to Shafer 1974, it is likely that the source of ameloblastoma is (a) the remnants of enamel cells, remaining dental lamina or the rest of the coating of Hertwig's, the reminder of the epithelium malases, (b) odontogenic epithelium, especially dentigerous and odontoma cysts, (c) The developmental disorders of the enamel organs, (d) the basal cells of the jaw surface epithelium, and (e) the heterotopic epithelium. The assertion that the source of the ameloblastoma comes from the epithelium of odontogenic cysts especially the dentigerous cysts supported by Stanley and Diehl who reported retrospectively 33% and 17% of the entire ameloblastoma arise in or incorporated with dentigerous cysts.<sup>13</sup>

Ameloblastoma is characterized by a slow growth pattern and can grow to a very large size and cause severe facial deformities. This disorder is usually asymptomatic and does not cause a change in sensory the nerve functions.<sup>1,2</sup>

Ameloblastoma is the most common odontogenic tumor in the developing world and has an incidence rate of approximately 14% of all tumors and jaw cysts. Global incidence of ameloblastoma is 0.5 cases per million people per year, and it is an odontogenic tumor often encountered in Africa and China.<sup>8,9</sup> In the Western Hemisphere, Ameloblastoma is the most common odontogenic tumor number two occurring after Odontoma, but the African-American population is five times more likely to develop ameloblastoma compared to Caucasian populations.<sup>9,10</sup> Most patients with an ameloblastoma aged between 30 and 60 years, but the average age at the time of the diagnosis varies from continent to continent which is estimated to be each about 42.3 and 30.4 years in Europe and Africa. Only 10 - 15% of cases of ameloblastoma occur in the child population, but this can reach 25% in Africa and Asia.<sup>9,11</sup>

Ameloblastoma maxilla and ameloblastoma extraosseus occur in age groups that are slightly older than the Unicystic Ameloblastoma group, while granular cell ameloblastoma occurs in younger age groups. This disorder is about the same gender prevalence and there are no specific dominant races. Schafer's research related to the synonymates shows the average age of sufferers, i.e. 6 and above and almost all patients are men. The explanation of this is likely that the synonym Ameloblastoma requires a longer period of time before reaching the size of the tumor that can cause symptoms. These tumors may have existed in previous ages but were not clinically visible and not specific symptoms.<sup>1</sup>

Many variants of the prevalence ameloblastoma's in Indonesia. However, the same as previously explained the differences that occur in the incidence of men and women are not so significant, namely by 48% and 52% with an average age of 39.7 years. From these data 25% were diagnosed with unique ameloblastoma, 57% multicystic follicular ameloblastoma, and 18% unspecified multicystic ameloblastoma. Tumor sites occur in 4 regions namely the maxillary posterior (10.4%), anterior maxillary (2.1%), posterior mandible (81.56) and anterior mandible (6.3%).<sup>12</sup> Ameloblastoma is most common in the posterior mandible, especially in the region of third molars, and is associated with follicular cysts or impacted teeth. Most ameloblastomas occur<sup>23</sup> in the ramus and mandibular posterior corpus in 80% of cases. In the mandible, the ramus area of the molar angle is more often three times more affected than the pre molar and anterior areas. About 15-20% of cases are reported to originate from maxilla with only about 2% originating anteriorly from the premolar. In the maxilla, the most commonly affected area is the molar area, but sometimes it can also be found in the anterior region, the maxillary sinus, the rice cavity, the orbit and sometimes to the base of the cranium.<sup>11,13,14</sup>

These cases were treated conservatively (62.5%), and radically (37.5%). The conservative treatments performed were enucleation (37.25%) and curettage (62.8%), whereas radical treatments were marginal resection and segmental resection.<sup>11,12</sup>

#### Classification

Ameloblastoma classification according to WHO is divided into benign<sup>54</sup> ameloblastoma which includes: (1) solid/multicystic ameloblastoma, (2) unicystic ameloblastoma (3) peripheral ameloblastoma (or extraosseous), (4) ameloblastoma dense/multicystic, (2) unique<sup>59</sup> ameloblastoma based on sequence (frequency) includes peripheral (or extraosseous) ameloblastoma, (4) ameloblastoma and malignant ameloblastoma based on<sup>3</sup> frequency sequence: a) metastasizing ameloblastoma, (b) primary ameloblastic carcinoma, (c) secondary intraosseous ameloblastic carcinoma, (d) secondary peripheral ameloblastic carcinoma.<sup>15</sup>

But recently, the classification of ameloblastoma has been simplified into 3 types: conventional, unicystic and peripheral. The term solid/ multicystic is discarded, because it can be confused with unicystic types. Of this type, conventional ameloblastoma is the most common,

representing 85% of all ameloblastoma, and occurs mainly in the age of 30 and 40 years, is considered more aggressive because of the high incidence of recurrence. Histologically this can be divided into follicular, plexiform, and granular cell morphological patterns. Other less common histological variants are clear cells and desmoplastic cells. In general, one-third of conventional types have plexiform patterns, one-third of follicular patterns, and the remaining third correspond to other variants.<sup>15,16,17</sup>

Unicystic ameloblastoma (UA) is a neoplastic entity that characterized by a cystic morphological that is covered by ameloblastic epithelium which can present growth tumor to the lumen and connective fibrous tissue. based on histopathological characteristic, it presents 3 histological subtypes, characterized by a pattern of epithelial component proliferation into: luminal, intraluminal and mural. Luminal and intraluminal variants respond satisfactorily to conservative surgical approaches, while the mural variant presents a higher recurrence rate, and is treated in the same manner as conventional ameloblastomas.<sup>18,19,20,21,22</sup>

Peripheral ameloblastoma is the most common<sup>57</sup> variant, accounting for only 1% of cases. It mainly affects patients with an average age of 52 years and most often occurs in the mandibular gingiva. These recurrences rarely occur, even when treated conservatively.<sup>23,24,25</sup>

Clinically, peripheral ameloblastoma appears with solid lesions, the surface can be smooth, granular or papillary, similar to normal mucosa. However, tumors sometimes have a darker color. The recurrence rate of peripheral ameloblastoma is 16-19%, so a follow-up is needed, because there is a possibility for benign peripheral ameloblastoma to reappear to ameloblastic<sup>4</sup> carcinoma.<sup>26,27,28</sup>

Malignant ameloblastoma is characterized by the presence of distant or regional metastases, in contrast to benign histological characteristics which are similar to benign ameloblastic forms. Regional metastases and distant metastases are found in 2% of cases.<sup>29,30,31</sup>

Ameloblastic carcinoma is a primary odontogenic malignant tumor with histological characteristics similar to benign ameloblastic followed by cellular atypia. The primary type can appear de novo, while the secondary type develops through the transformation of intrabony malignancies or peripheral dissemination.<sup>32,33,34,35</sup>

#### Clinical Manifestation

Clinical features in the early stages rarely show complaints, because this tumor is rarely diagnosed early, generally known after 4 to 6 years. Swelling of varying sizes vary so that it can cause facial deformities, the same color as the surrounding tissue, consistency varies there are harsh and sometimes soft parts, firmly limited, appear bone expansion toward buccal and lingual, this tumor extends in all directions urgent and damaging surrounding bones, there is a sign of egg shell cracking or ping pong ball. Phenomena when the tumor mass has pressed the bone cortex and bone thinning, there is no pain and no paresthesias are found, the tumor mucosa does not experience ulceration. Only in some patients with the lump accompanied by pain, reduced sensibility of the nerve distribution area and sometimes ulceration due to tooth compression when the tumor has reached a large size. Teeth in

the tumor area change location and shape. If a secondary infection occurs, ulceration, fistula and even granulation tissue can be found, as well as pain, paresthesias, and signs of inflammation.<sup>1,3,19,30</sup>

In general, ameloblastoma is benign but locally invasive, whereas maxillary ameloblastoma appears as a more aggressive and persistent lesion. This is likely due to the thin, brittle maxilla bone, unlike thick mandibular bone,

which allows uninterrupted spread of tumors to the surrounding structures. A good blood supply to the maxilla when compared with the mandible also contributes to the acceleration of the spread of this local neoplasm. Whereas in patients with primary squamous ameloblastoma in a study revealed mass lesions and nasal obstruction, sinusitis, epistaxis, facial swelling, dizziness, and headache.<sup>1</sup>

Table of Ameloblastoma Types<sup>9, 18</sup>

*Clinicohistologic types of ameloblastoma*

<i>Types of ameloblastoma</i>	<i>Synonyms</i>	<i>Salient features</i>	<i>Conventional radiographic features</i>	<i>Histopathological variants</i>
<b>Benign</b>				
Solid/multicystic	Conventional/Classical ameloblastoma	Mean age: 36 years Male > female Slightly higher in mandible	Unilocular radiolucency Multilocular radiolucency Unerupted tooth Root resorption	Cystic, acanthomatous, granular, basaloid, spindle, clear cell, hemangiomatous
Unicystic	Cystogenic ameloblastoma	Dentigerous type: Mean age: 16.5 years Male > female Non-dentigerous type: Mean age: 35.2 years Female > male Slightly higher in mandible (posteriorly)	Unilocular radiolucency Multilocular radiolucency Unerupted tooth Unilocular radiolucency Multilocular radiolucency	Luminal (plexiform unicystic, intraluminal), mural
Peripheral	Extrasosseous/soft tissue ameloblastoma	Mean age: 51 years Male > female Slightly higher in mandible Exophytic Mean size 1.3 cm	Saucerization	Not applicable
Desmoplastic	Ameloblastoma with pronounced desmoplasia	Mean age: 41.6 years Female = male Maxilla = mandible	Mixed radiolucent/radiopaque Root resorption	Hybrid Desmoplastic ± osteoplasia
<b>Malignant</b>				
Metastasizing	Malignant ameloblastoma	Mean age: 34.4 years Male > female Slightly higher in mandible Distant sites: lungs and other areas	Same as solid/multicystic	Same as solid/multicystic
Primary ameloblastic carcinoma	Not applicable	Mean age: 53 years Male > female Higher in mandible (posteriorly)	Ill-defined multilocular radiolucency Foci of calcification	Not applicable
Secondary ameloblastic carcinoma (intraosseous)	Carcinoma ex intraosseous ameloblastoma	Rapid growth, 7th decade Male > female Slightly higher in mandible	Ill-defined multilocular radiolucency Foci of calcification	Not applicable
Secondary ameloblastic carcinoma (peripheral)	Carcinoma ex peripheral ameloblastoma	Male = female Alveolar bone resorption	Interradicular radiolucency	Not applicable

**Radiograph Manifestation**

Radiological examination that can be done to diagnose ameloblastoma is plain photo, CT scan and MRI. Panoramic radiography is the first step in diagnosing ameloblastoma with radiographic images that vary depending on the type of tumor. CT examination is recommended if the swelling is hard and fixed to the surrounding tissue. CT examination is usually useful for identifying the contour of the lesion, the contents of the lesion, and extension to the soft tissue that helps establish the diagnosis. Plain photos cannot distinguish between tumors and normal soft tissue, they can only distinguish between tumors and normal bone, whereas CT scans and MRIs can show them clearly. MRI is essential in determining the extent of maxillary ameloblastoma so as to determine the prognosis for surgery.<sup>1</sup>

The most frequent radiographic appearance of multicystic ameloblastoma is multilocular lesions, which are often described as soap bubbles when the lesion is large and honeycomb if the lesion is small. There is often oral

and cortical lingual expansion and resorption of tooth roots adjacent to the tumor. Whereas uniquely ameloblastoma appears as unilocular radiolucent lesions with well-defined border around corona of unerupted teeth.<sup>1</sup>

Computed tomography (CT-scan) provides an accurate anatomical picture of tissue pieces in 2 dimensions and 3 dimensions. The advantage of this technique is that there is no overlapping picture and provides a detailed network picture of the area involved. On CT scan ameloblastoma can be found low attenuation cystic area with scattered isoattenuation region, reflecting the presence of soft tissue components. These lesions can also erode the cortex by expanding into the surrounding oral mucosa. Erosion of nearby tooth roots is typical of ameloblastoma and indicates tumor aggressiveness. Nevertheless, only histopathological findings can help determine tumor malignancy and carcinomatous changes.<sup>1</sup>

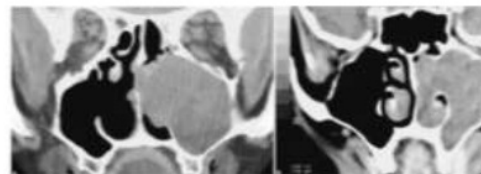
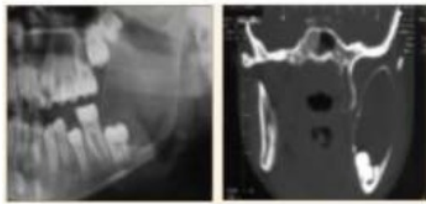


Figure. A Ameloblastoma on the mandibular sinistra at Plainphoto, as a broad lesion and expansionist (B) CT scans of coronal pieces showing widespread lesions of expansion, thinning of the cortex and minimal destruction (C) CT scans show the ameloblastoma on the maxillary sinus and the cavum of rice. Radiography and surgical explorations indicate the underlying sinus base (D) CT scan of the coronal piece of the broad ameloblastoma that satisfies the sinistra maxillary sinus and the cavum rice with lateral sinus wall bone erosion and the base of the Orbita (source Gungum S Hosgoren B, 2005, Schafer DR, 1998)

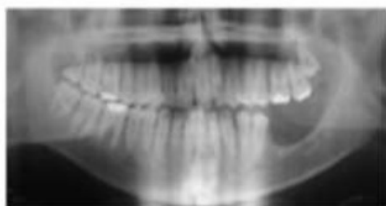


Figure A. Overview of the image found in the mandibular in the OPG before treatment. B. Overview after the appointment of Ameloblastoma after 5 years, not recurrence (source Silva et al. 2018. Vol 66(2). Pp:181-186)

### Histopathology

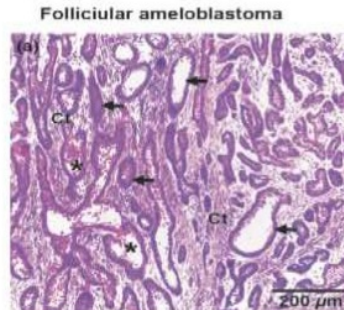
#### 1. Solid/Multicystic/conventional

The most common ameloblastoma is a solid/multicystic/conventional type, found about 91% of all cases of ameloblastoma. Ameloblastoma growth is

slow, categorized benign and occurred in the Decade to 3 or to 4. Histologically, solid/multicystic/conventional is differentiated into two kinds: <sup>9,15</sup>

#### A. Follicular ameloblastoma

Follicular Ameloblastoma often occurs on the posterior maxillary with an overview showing the proliferation of odontogenic epithelium cells resembling the islands. Epithelial nests contain a loose-fitting nucleus resembling stellate reticulum enamel organs. Its essence is surrounded by a single layer of columnar cells such as Ameloblas. <sup>9,15</sup>



The follicular (Ct) tissue contains the islands in the odontogenic epithelium (black arrow) described by peripheral columnar cells indicating reverse polarization. There are islands of cystic degeneration containing loose-like stellate reticulum cell (\*).<sup>9</sup>

#### B. Plexiform ameloblastoma

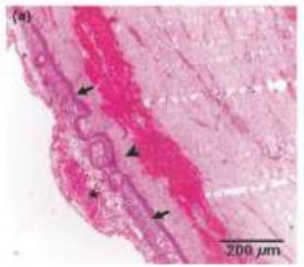
The plexiformis consist of a long epithelial yarn that is beranastomosis or a larger epithelial sheet of odontogens. The threads or epithelium sheets are tied by a columnar and cuboid-like Ameloblas cell that surrounds loosely regulated epithelial cells. <sup>9,15</sup>

Plexiform contains an ameloblastoma connective tissue such as long-string anastomosis of odontogenic epithelium (black Arrow) depicted by columnar peripheral cells displaying reverse and loose polarization resembling stellate reticulum cell. <sup>9</sup>

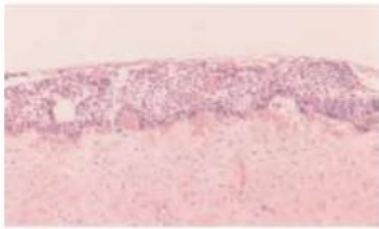
#### 54 The Unicystic Ameloblastoma

Ameloblastoma is the second most common ameloblastoma with a percentage of approximately 10-15% of all cases. 38 most commonly seen at a young age with an average age of 26.1 years, and its main location is the posterior mandible where it often appears as a symptom-less swelling. The majority of Unicystic Ameloblastoma resembles dentigerous cysts because of their relationships with unerupted teeth. The unicystic ameloblastoma in histology is divided into two namely: <sup>9,24</sup>

#### A. Luminal unicystic Ameloblastoma



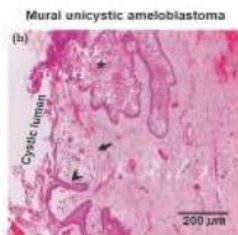
The unicystic luminal Ameloblastoma comes as a cystic lesion coated by the ameloblastomatosse epithelium that protrudes into the lumen in the proliferation of Pleksiformis (referred to as the Intraluminal subtype).<sup>9</sup> The unicystic luminal Ameloblastoma exhibits a fibro-optic wall (black arrow) coated by the ameloblastic epithelium (black arrow) and loose cells such as reticulum stellata (\*).<sup>9</sup>



There is an overview of the basal cell core hypofiliatation of the cystic cavity coating epithelium. Palisading (lined-like fence) and polarization of basal cell nuclei epithelium of cystic cavities and special cytoplasmic vacuolization of basal cells of cystic coatings.<sup>24</sup>

#### B. Unicystic Mural Ameloblastoma

Unicystic murals Ameloblastoma present with the ameloblastomatous epithelium of cells inside the cyst wall. A unicystic mural variant of Ameloblastoma displays follicular or plexyform arrangement of ameloblastomatous epithelium cells in it wall cystic. The extent of unicystic mural of Ameloblastoma is relatively higher when compared to luminal type.<sup>15</sup>

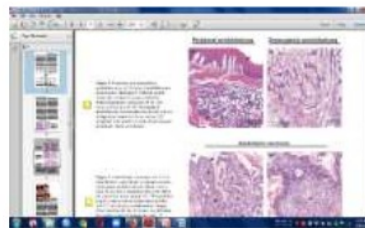
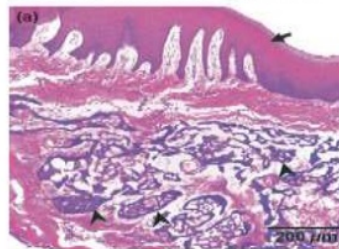


Unicystic murals show a cystic wall fibrous (black arrow), infiltration of the ameloblastic walls into the cystic wall (black arrow), and loose like Reticulumsel (\*).<sup>9</sup>

#### 3. Ameloblastoma Peripheral

The peripheral ameloblastoma is the most rarely found variant, accounting for only 1% of the case of Ameloblastoma. Middle-aged patients with an average age of 52 years are most often exposed. These lesions are more common in the lower jaw of the upper jaw and are found in the posterior of the gingiva or alveolar sulcus. Histologically, the Ameloblastoma device consists of the islands of the Epelium ameloblastic with the same histological pattern as solid/multicystic/Ameloblastoma conventional. 5 solid and Unicystic type is Ameloblastoma intraosseus, while the peripheral type occurs in soft tissues/extraosseus.<sup>1</sup>

#### Peripheral ameloblastoma

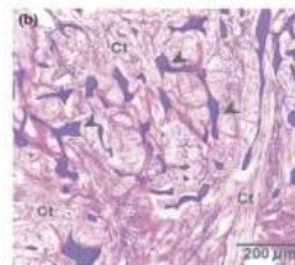


The peripheral ameloblastoma shows the island of odontogenic epithelium in connective tissue overlaid by the stratified oral squamous epithelium (black arrow).<sup>9,17</sup>

#### 4. Ameloblastoma Desmoplastic

Ameloblastoma desmoplastics The growth is slow, there is swelling without pain. Histological depictions of dysplasia broad stroma are patognomonic consists of the islands of odontogenic epithelium with varying forms and sizes proliferating in the connective tissue of collagen. Thick fiber collagen tends to suppress odontogenic island epithelium from the periphery, there by causing strange shapes and sizes.<sup>9,37</sup>

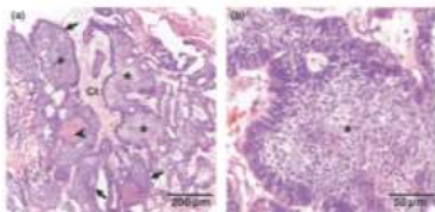
#### Desmoplastic ameloblastoma



Desmoplastic Ameloblastoma shows hyalinized densely stroma connective tissue collagen (Ct) embedded with the islands of the odontogenic string of epithelium (black arrow).<sup>9</sup>

### 5. Carcinoma

A metastatic or malignant ameloblastoma is formerly a benign ameloblastoma that has metastasized to distant and usually lung areas. It is diagnosed based on the clinical picture and metastatic lesions featuring a similar histological feature of ameloblastoma solid/multicystic/conventional. Ameloblastoma was initially docile which lost its differentiation into carcinoma. The ameloblastic carcinoma grows faster and aggressively and can cause pain swelling that melts the cortical bones. Histologically, the ameloblastic carcinoma combines the overall histologic of the pattern of Ameloblastoma with cytological atypia consisting of abnormal mitosis, cellular and hyperchromatism, and focal necrosis.<sup>9</sup>



Ameloblastic carcinoma. (a) Low magnification, containing an odontogenic carcinoma of the island (black Arrow), is coated by cells such as ameloblasts in the connective Tissue stroma (Ct). The Epithelium Islands contain stellate reticulum cell (\*) which displays the changes in the Akantomatose (black arrow). (B) at a higher magnification, cells such as reticulum stellate cell (\*) are surrounded by peripheral-like Ameloblastic cells featuring Hyperchromatism, pleomorphism, and mitotic agents.<sup>9</sup>



Figure of Ameloblastic carcinoma exhibiting a follicular growth pattern with stellate reticulum-like structure or central necrosis. Peripheral columnar cells are palisaded in the follicular islands. (Hematoxylineosin stain; original magnification x100) (Abir et al. 2017. Vol 23. Pp:95-98)

### Diagnose

Ameloblastoma can be diagnosed based on anamnesis, physical, and radiographic examination, and biopsy.<sup>32</sup> To get the final diagnosis of the biopsy, the incision should be

done. Followed by histopathology examination. Ameloblastoma tends to infiltrate trabeculae bone-fringes of lesions before the bone resorptions become apparent radiography.<sup>27</sup>

Fine needle aspiration cytology (FNAC) is a lesions-taking technique with a fine needle to retrieve its contents for cytological diagnosis. FNAC has been widely used for the diagnosis of lymph nodes, salivary glands, and diseases of the thyroid and parathyroid glands. Although FNAC is a simple, fast, and minimally invasive procedure that is different from biopsy.<sup>28,31</sup>

The radiography of the ameloblastoma may vary according to the type of tumor. Computed tomography (CT) usually assists in determining the contour of the contour lesions and extensions to the soft tissues. In patients with swelling in the jaw first step in determining the diagnosis can be performed panoramic radiology. However, if severe swelling of the CT is more likely because it can identify the contour of lesions, content and soft tissue extensions. Compared to CT, Magnetic Resonance Imaging (MRI) is slightly superior to determine the success rate of the diagnosis of Ameloblastoma.<sup>29,35</sup>

### Treatments And Prognosis

The indication of treatment is determined by the extent and magnitude of the tissues involved, the histological structure of the tumor and the benefits to be obtained.<sup>24</sup> According to Ohishi the indication of conservative treatment is in people of young age and Unicystic Ameloblastoma. While the indication of radical treatment is the solid type ameloblastoma with unclear edges, lesions with a description of bubble soap, ineffective lesions with conservative management and large size ameloblastoma.<sup>3,24</sup> Radical management of segmental resection, hemimandibulectomy and marginal resection (Enblok resection). Mandibular reconstruction is reviewed from function and cosmetics, this organ affects the shape of the face, speech function, chewing and swallowing.<sup>8,24</sup>

Some ways can be used, such as by using alloplastic materials, such as bridging titanium plates autogenous and bone grafting for example rib grafts, Krista iliac and tibia and can also be a combination of alloplastic material with autogenous bone grafting. Post-operative treatment of mandibular resection i.e. antibiotic and analgetic medication, it is not necessary to Intermaksila fixation. Avoid physical trauma to the face or jaw because it can cause a mandible fracture. Keep the oral hygiene until the surgical wound heal perfectly. The soft Diet is retained 4-6 weeks. If necessary it can be made of dental prostheses after considering that there has been an internal bone of of mandibular bones, approximately 6 months after surgery.<sup>36,37,38</sup>

The tumor treatment varies from curettage to broad bone resection, with or without reconstruction. Radiotherapy is not indicated due to radioresistance of these lesions. In some literature also found indications for electrocauterization, Krio surgery and use of Schlorosan agent as a treatment option. Reexamination (post-operative follow-up) is important as nearly 50% of the recurrence case occurs in the first five years of the post-operative. Conservative treatments include curettage, enucleation, cryosurgery, and enucleation with curettage or dredging.<sup>39,40,41,42</sup>



### 1. Curettage

Curettage is the removal of the tumor by cutting it from the normal tissues around. The failure of a curettage is due to the dwelling of the periphery of tumors on tissues. This technique can be used for small lesions of unicystic ameloblastoma in the mandibular. Most cases are handled with an intraoral approach, which is usually a buccal, labial, or palatal approach. Stringed curettage with a mukoperiosteal flap with the base is wide enough to ensure the blood supply is not disturbed. The most common flap envelope is applied. Then the incision is made on the gingiva sulcus (for toothed patients) and on the crest alveolar (for the unscalped patient), the flap mukoperiosteal full thickness is opened. Curettage is used to remove lesions from bone cavities. Furthermore, normal bone margins are also discarded by scraping to ensure all tumors are removed, and small bone defects are closed with primary closures; Large bone defect can heal with secondary intention.<sup>1, 3, 25</sup>

### 2. Enucleation

Enucleation is the removal of a cyst from the wrapping layer to its contents. Enucleation indications are odontogenic keratotic lesions that have a high recurrence rate. Enucleation has 2 methods of approach, namely intraoral and extraoral approaches.<sup>1, 3, 25</sup> The intraoral approach is performed by flap incision and elevation, bone removal, and cyst enucleation.<sup>1</sup>

#### 1. Flap incision and elevation

If a cyst involves a tooth, an incision is made around the tooth, both with and without consideration for extraction. The purpose of the incision is to provide good access and facilitate healing, besides the incision is useful in the process of closing the operating area if it requires extraction of 1 tooth or several teeth. If the cyst involves up to the periodontium, then the incision should be made away from the cervical area of the tooth. To facilitate healing in the edentular area, an incision is made along the crest. The ascending and descending arms of the incision extend toward the buccal sulcus and are outside of the swelling. The purpose of this incision is that suturing can be carried out on a healthy bone surface.<sup>1, 3, 4</sup>

#### 2. Bone Removal

The thin bone tissue remains to be preserved. If the lesions are large, after the mukoperiosteal flap, the bones can be penetrated using the elevator periosteal inserted between the cyst sac and the bone. If the bone tissue is untenable, the mukoperiosteum is elevation and the bone tissue underneath is removed using a BUR acrylic to provide good access to the enucleation process.<sup>1, 3, 4</sup>

### 3. The Cyst Enuxation

The cyst should be lifted completely without ripping or pierced it. Perform dissection using blunt instruments. Use a rolled-up gauze layer, then insert between the cyst sac and its bone cavity using hemostat. Another alternative is to marinate the cyst so that the cyst is wrinkle making it easy to secreted. After the cyst has been dilated, further treatment can be performed on the tooth involved, for example: root canal filling, apicectomy, retrograde root filling, or extraction. Recheck the post-enuxation area, perform irrigation, and then can be performed closing with the sewing.<sup>1</sup>

Conservative treatment, such as enucleation, could preserve the bone integrity and allow continued growth of the mandible. Nonetheless, such an approach is found to be associated with a high post-operative recurrence rate in the range of 60–80%, mainly due to the failure to remove the tumour islands that infiltrate the bone walls. In response to the challenge, a modified conservative management (MCM), which consists of enucleation, peripheral ostectomy and chemical cauterisation with the Car-roy's solution, has been used in the Sultanah Bahiyah Hospital, a referral centre for dental ment in northern Malaysia.<sup>43</sup>

The findings suggest that the MCM has a great potential to be used as another alternative to treat ameloblastoma other than radical surgery, and the recurrence rate showed superiority to other conservative techniques used alone or in combination.<sup>43</sup>

The prognosis is usually beneficial even though it can cause deformities. The relatively high relapse number for this type of tumor remains a challenge. The conventional ameloblastoma treated with enucleation or curettage presents a higher rate of recurrence when compared to unicystic asmissional ameloblastoma in the same way. The treatment aimed at ameloblastoma recursive is a radical surgery that provides free survival of the disease for at least 10 years but requires clinical and radiography monitoring for a certain period of time.<sup>3, 15, 24</sup>

Table of Ameloblastoma Types <sup>41</sup>

Author	Type of ameloblastoma	Surgical treatment	Follow-up
Ooi et al. (1)	Unicycstic, Multicycstic	Radical	-
Pogrel (6)	Solid, Multicycstic	Segmental resection + 1 cm margin + soft tissue margin	-
	Unicycstic	Enucleation + support technique Resection + 0.5-1cm margin	-
	Peripheral	Local excision	-
Sham et al. (3)	Solid, Multicycstic	Resection>1 cm margin + gingiva	-
	Aggressive unicycstic	Resection>1 cm margin + gingiva	-
Hertog et al. (2)	-	Biopsy when recurrence	> 10 years
Hammartjord et al. (7)	-	Conservative if follow-up Radical if recurrence risk	10 years
Bianchi et al. (5)	Extended	Radical + reconstruction	-
Simon et al. (8)	Extended	Reconstruction	-
Ramakant et al. (9)	Advanced tumors	Radical	-
Hasegawa et al. (10)	Solid, Multicycstic	Conservative treatment	>10 years
Adebayo et al. (4)	-	-	Long term
Sharma et al. (11)	Extended	Radical + reconstruction	-
Chukwunke et al. (23)	Extended	Radical + reconstruction	-
Hou et al. (14), Cohen A et al. (14)	-	Surgical planification	-

**Differential Diagnosis (DD)**

Diagnosis of appeals to lesions of the maxillary antral includes synonasal lesions, odontogenic tumors, and tumors derived from Salivarius minor Glandula, Antral pseudocyst. Tumors and odontogenic cysts are considered strong DD when lesions are found in the tooth bearing area of the mandible and or when there is a dental structure in the lesions. A definite diagnosis cannot be enforced based on clinical and radiographic but histopatological confirmation is required.<sup>1,34</sup> On the most frequent radiography of the multicystic Ameloblastoma, a multilocular lesion, which is often described as an image of soap bubbles when large lesions and honeycomb images when the lesions are small. Often found oral and cortical lingual expansion and tooth root resorption adjacent to tumors. While the Unicycstic Ameloblastoma appears to be a strict unilocular Lusen lesion is firmly around the corona that does not interrupt the tooth.<sup>4,21,22</sup>

The diagnosis of ameloblastoma is a unilocular dentigerous cyst located around the dental crown not eruption is often indistinguishable. The appearance of internal bone septa is identification for ameloblastoma, but other lesions also have internal septa such as infections Keratocyst, giant cell granuloma, infections myxoma, and ossifying fibroma.<sup>23,33</sup>

Odontogenic keratocyst is a lesion that is believed to be derived from dental lamina and other odontogenic epithelium sources. These cysts range from 5-15% of the entire mandibular cyst. Most occur at the age of 2 to 4, although it can occur at all ages. Lumen cysts often contain cheesy material and there are parakeratinized lining lumen. Daughter cyst and kalistic epithelial nests are found outside of primary lesions, so infections Keratocyst has the highest recurrence number of all infections cysts when treated conservatively with curettage. Radiographically this lesion appears to be a unilocular or

multilocular Lusen lesion, with fine and corticated borders, often associated with impacted teeth and can lead to expansion and bone destruction. Although these lesions are most common regarding the corpus and Ramus mandibular, the maxilla is often about the posterior or the canine portion, but can also occur in the anterior mandible or throughout the maxilla. Odontogenic Keratocyst reveals a more aggressive growth than other odontogenic cysts and may have limits to undulated and multilocular sightings, these characteristics that make these lesions difficult to distinguish with ameloblastoma. Odontogenic Keratocyst may lead to thinning the cortex, shifting gears and resorption of root.<sup>1,33,39</sup>

Radiographically these lesions appear to be unilocular or multilocular, with smooth and corticated borders, often associated with impacted teeth and can cause bone expansion and destruction. Although these lesions mostly affect the corpus and ramus of the mandible, the maxilla often affects the posterior or canine region, but can also occur anteriorly in the mandible or in the entire maxilla.<sup>4,21,22</sup>

Odontogenic keratocyst contains septa curvature, but usually keratocyst tends to grow along the bone without any sign of expansion, which is characteristic of ameloblastoma. Giant cell granulomas occur in younger age groups and have more granules or unclear smooth septa. Odontogenic myxoma has septa that looks similar but, usually there is one or two thin sharp, straight septa, which is characteristic of myxoma. Myxomas are not as extensive as ameloblastoma and tend to grow along the bone. The septa of ossifying fibroma is usually wide, granular, and unclear, and there is often a small, irregular

12)ecular pattern.<sup>23,39,44,45,46</sup>  
 Dentigerous cyst is the most common non-inflammatory odontogenic cyst and the most common cause of pericoronal lucency associated with impacted teeth. Dentigerous cysts formed in the layers of the tooth follicles as fluid accumulates between the follicular epithelium and the corona of the developing or unerupted tooth. Most dentigerous cysts occur in

adolescents and young adults and often form in the cervical area of the unerupted mandibular third molars, but can also affect maxillary third molars, caninus maxillar, and bicuspid mandibular. Patients usually do not complain of pain. An important feature of this cyst is its ability for asymptomatic expansion and the potential to shift or adsorb surrounding teeth or bones.<sup>1</sup>

Radiographically, these cysts appear as well-defined lined lesions around the corona of unerupted teeth, usually third molars, round or oval in shape, corticated. The root of the tooth involved is often outside the lesion and in the mandibular bone. Cyst size can have many variation, a diameter of 2 cm or more, can cause expansion into the mandible. The radiographic features of these cysts can be similar to cystic, unilocular odontogenic keratocysts variation. Humongous dentigerous cysts often develop with undulated boundaries because of their expansion, and produce radiographic images similar to those of large odontogenic keratocysts or ameloblastomas. An untreated dentigerous cyst can develop into ameloblastoma in its layers (eg ameloblastoma mu

29).<sup>4,6,21,22,47,48</sup> Odontogenic adenomatoid tumor (AOT) is a benign hamartoma of odontogenic epithelium characterized by slow and progressive growth. These tumors are rare and are typically diagnosed at the age of the second decade, with the majority occurring in adolescent and young adult women and associated with unerupted teeth. About 70% of these tumors occur in the maxilla, and canine region. Radiographically this tumor appears as a unilocular radiolucent lesion with a clear border associated with impacted teeth. Can be found calcification of the amount of which varies and can shift or prevent tooth eruption. If the tumor borders the tooth, the lesion is found more apical to the root of the tooth than a dentigerous cyst.<sup>1</sup> Mucocele is an expansion of sinus space caused by obstruction of the sinus ostium and accumulation of secretions. This disease is the most common cystic lesion involving sinus paranasales and the most significant local complications of sinusitis due to obstruction of the sinus ostium. The most common locations are the frontal sinus and the anterior ethmoid sinus. Usually there is a history of surgery, trauma or recurrent sinusitis. Mucocele can cause maxilla expansion or erosion. Mucocele formation due to inflammatory ostial obstruction in the maxillary sinus is rare, the location of the most frequent inflammatory or allergic process is in the maxillary sinus.<sup>1</sup>

#### Complication

Complications of malignant ameloblastoma are usually due to local invasive or distant metastatic spread. In the case of local complications, it can cause progressive maxillary and maxillary distortion because deformity, pain, and malocclusion. Benign ameloblastoma can metastasize to faraway places usually to the lungs. Ameloblastic carcinoma can develop from initially benign ameloblastoma that loses differentiation into carcinoma. Ameloblastic carcinoma grows faster and more aggressively and can cause painful swelling which pierces the cortical bone. Because of its locally aggressive nature of growth characteristics, ameloblastoma can quickly

become a massive and expansive tumor which causes tooth mobility, tooth movement, and strange facial appearance if the patient suspends treatment.

#### CONCLUSION

Complications of malignant ameloblastoma are usually due to local invasive or distant metastatic spread. In the case of local complications, it can cause progressive maxillary and maxillary distortion that causes deformity, pain, and malocclusion. Benign ameloblastoma can metastasize to faraway places usually to the lungs. Ameloblastic carcinoma can develop from initially benign ameloblastoma that loses differentiation into carcinoma. Ameloblastic carcinoma grows faster and more aggressively and can cause painful swelling which pierces the cortical bone. Because of its locally aggressive nature of growth characteristics, ameloblastoma can quickly become a massive and expansive tumor which causes tooth mobility, tooth movement, and strange facial appearance if the patient suspends treatment.

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## B4. Ameloblastoma

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