**Case Report**

# A case report of a bony swelling in the mandible of a young female patient

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**Abstract:** Ameloblastoma is a benign odontogenic tumour having the tendency to occur in the posterior mandible. It is speculated to be of epithelial in origin. Although it is slow growing in nature but it can slowly infiltrate into the surrounding tissues thereby causing the extensive damage to the jaw bones. Ameloblastoma is considered to be the lesion primarily occurring in elderly male patients. This article presents a case of multicystic ameloblastoma occurring in a young girl highlighting the typical radiographic features of this extensive lesion. Surgical en bloc resection was done for the patient over the right side of the mandible and reconstruction was done using the iliac graft.

Keywords: Odontogenic, Benign, Ameloblastoma, Multilocular, Radiolucency.

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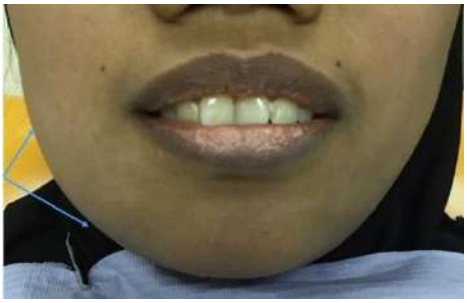
**Introduction:** Ameloblastoma is be the most common benign odontogenic tumor occurring in the jaws. It arises from the remnants of the enamel organ without the formation of enamel. This tumor is characterized by its locally invasive nature and causing the disfigurement of the face. It was defined as "unicentric, nonfunctional, intermittent in growth, anatomically benign and clinically persistent" by Robinson. This tumor has the predilection in $3^{rd}$ -$4^{th}$ decade of life, commonly seen in the males. It is associated with the painless, unilateral swelling of the jaws with mobility and displacement of the teeth. It is also occasionally seen in association with the impacted teeth. [1]

**Case Report:** A 19 year old Indonesian female reported to the SEGi Oral Health center with the complaint of persistent swelling over her right side of face since many years. Her history of present illness revealed that the swelling was gradual in onset, not associated with any pain since 3-4 years. There was no history of trauma given. Patient revealed that the tooth on the right lower jaw became mobile and she got it removed from the govt. clinic 2 years ago. There was no significant medical history. All her vitals were with in normal range. There was an obvious facial asymmetry. A solitary swelling was seen over the right side of the face involving the lower border of the mandible, with diffused borders (Fig. 1). On palpation, swelling was non tender and hard in consistency. Swelling was non mobile and non-fluctuant. There was no local rise in the temperature.

Intraoral examination revealed clinically missing 16, 36, and 46. There was grade 1 mobility irt 44, 45. Significant vestibular obliteration was seen in 46 region. Overlying mucosa showed secondary ulcerations irt 46 region, due to traumatic occlusion from the upper molar. On palpation, swelling was bony hard and non-mobile.

There was buccal and lingual cortical plate expansion irt 46 region. (Fig. 2)



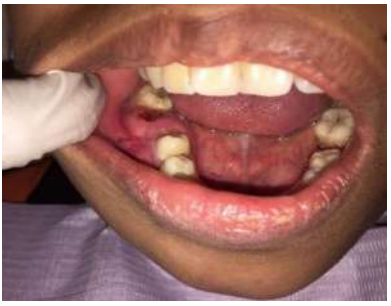


Figure 1: Extra oral Photograph  Figure 2: Intraoral Photograph

Provisional diagnosis of Odontogenic Keratocyst was made based on the patient's age and location of the lesion. An OPG was taken for the patient. The radiograph revealed well defined multilocular radiolucency in left lower side of the mandible. Radiolucency was seen extending from the apical region of 44 to the distal third of the root of 48. Internally, multiple septae were seen with in the radiolucency giving it a soap bubble appearance. Lower border of the mandible appeared to be scalloped. There was mesial migration of 45 and distal migration of 47, with missing 46. Root resorption was seen irt 45 in the apical region. (Fig. 3)

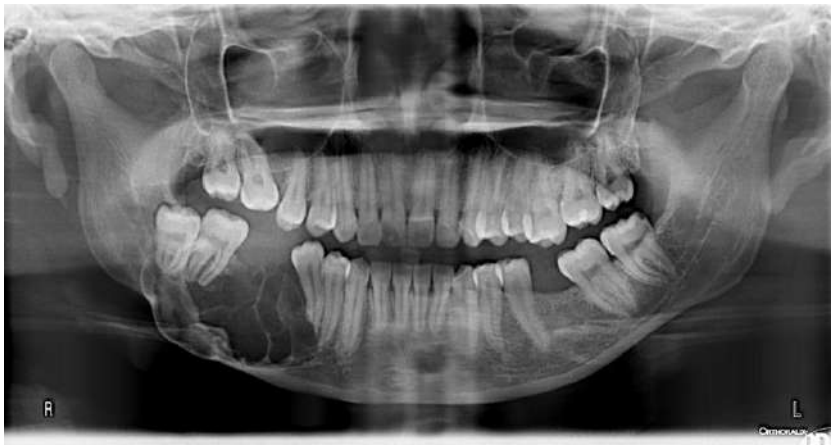


Figure 3: Pre-operative panoramic radiograph of the patient

Radiographic diagnosis of Ameloblastoma was made for the patient. The Odontogenic Keratocyst, Giant cell granuloma, Odontogenic Myxoma, and Ossifying fibroma can be considered in the radiographic differential diagnosis.

Patient was referred to higher center for biopsy and other investigations. Hematological investigations were insignificant. Biopsy confirmed the case as Follicular Ameloblastoma. Patient went back to her home country for the treatment of the lesion. CT scan was done before the surgical removal of the lesion. En Bloc resection of the right side of the mandible was done and reconstruction of mandible was done using iliac crest graft. After 6 months of the surgery, OPG was taken for the patient. (Fig. 4) Patient had the regular appointments with the speech therapist and removable prosthesis on the right side of the mandible (Fig. 5) was planned. Patient is still under the follow up.

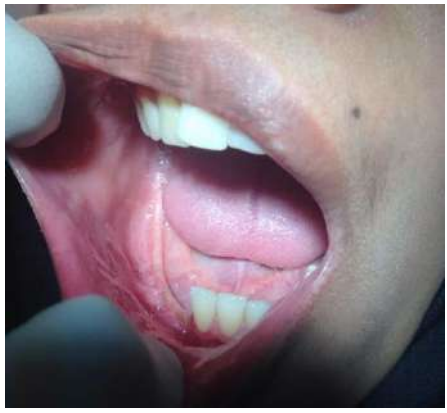


Figure 4: Post-operative intraoral photograph of the patient.

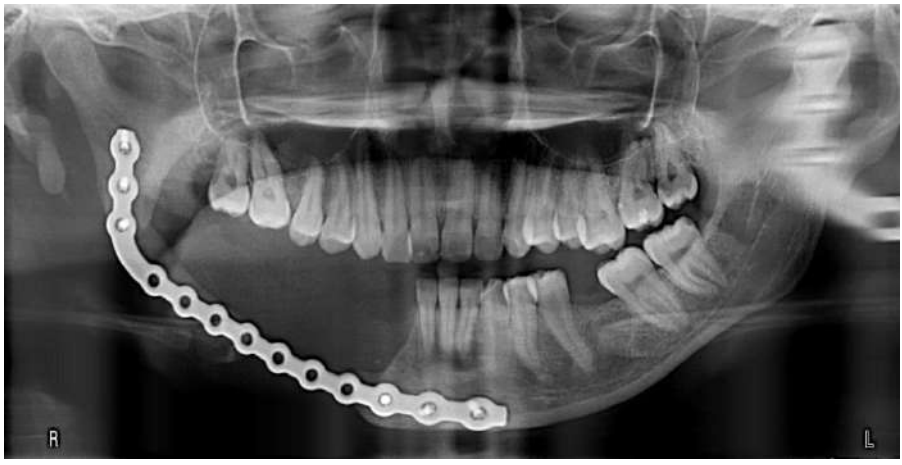



Figure 5: Post-operative panoramic radiograph of the patient

**Discussion:** Odontogenic tumours are the large heterogenous group of lesions involving the jaw bones, arising from the epithelial or ectomesenchymal tissues or both. Odontogenic tumours were first classified by WHO in 1971 following which, many modifications were done in the classification. Ameloblastoma was categorized as the epithelial odontogenic tumor in 1992 by WHO. Latest update on the classification was done in 2017. According to the latest classification

ameloblastoma is divided into four categories; conventional, extraosseous / peripheral, unicystic, and metastasizing ameloblastoma. WHO 2017 classification of odontogenic tumours is summarized in Table 1.[1]

Ameloblastoma has been derived from the Greek word 'amel' which means enamel and 'blastos' which means germ. It was first described by Cusack in 1827. In 1937, Robinson described this as a benign tumor that is usually "unicentric, non-functional, intermittent in growth, anatomically benign and clinically persistent." The word ameloblastoma was first coined by Ivey and Churchill in 1939.[2]

Global incidence of occurrence of ameloblastoma is 0.5 cases per 5 million persons per year. Ameloblastoma accounts for 1% of all the tumours and cysts of the jaws and 9%-10% of the odontogenic tumours.[3, 4]

It is a slow growing but locally invasive tumor having the tendency to erode the bone and invading the adjacent structures. Ameloblastoma has the propensity to grow in both the jaws, but most commonly it occurs in the mandible (80%) according to the published literature. It tends to occur in the posterior region of the mandible involving either body or ascending ramus of the mandible, but it can occur anywhere in the either of the jaws.[5]

| 2017 WHO Classification | | | |
|---|---|---|---|
| Benign | | | Malignant |
| Epithelial | Mixed | Mesenchymal | |
| • Ameloblastoma<br>• Ameloblastoma, unicystic type | • Ameloblastic fibroma<br>• Primordial odontog | • Odontogenic fibroma<br>• Odontogenic myxoma/ myxofibroma | **Odontogenic carcinomas**<br>• Ameloblastic |

| | | | |
|---|---|---|---|
| • Ameloblastoma, extraosseous/ peripheral type<br>• Metastasizing (malignant) ameloblastoma<br>• Squamous odontogenic tumor<br>• Calcifying epithelial odontogenic tumor<br>• Adenomatoid odontogenic tumor | enic tumor<br>• Odontoma, Complex type<br>• Odontoma, Compound type<br>• Dentinogenic ghost cell tumor | • Cementoblastoma<br>• Cemento-ossifying fibroma | carcinoma<br>• Primary intraosseous carcinoma, NOS<br>• Sclerosing odontogenic carcinoma<br>• Clear cell odontogenic carcinoma<br>• Ghost cell odontogenic carcinoma<br><br>**Odontogenic carcinosarcoma**<br><br>**Odontogenic sarcomas** |

Ameloblastoma can involve any of the age groups, but peak incidence is reported to occur in 3rd-4th decade of life.[6] Some authors have noticed the peak age of occurrence of ameloblastoma in 2nd and 6th decade of life.[7]

The present case of Ameloblastoma happened to occur in a young female, which makes this case report quite rare. Variable male to female predilection has been reported by some of the authors.[5, 8]

WHO in 2003 classified ameloblastoma as: [9]

- Solid/Multicystic variant
- Unicystic variant
- Peripheral/Extraosseus
- Desmoplastic Ameloblastoma

Recently, it has been proposed that all the conventional lesions present with cystic degeneration both micro/macroscopically. Hence, the term of solid, multicystic ameloblastoma should be discarded and conventional lesions should be appropriately termed as "conventional ameloblastoma".[10]

The most common variant of the ameloblastoma is conventional ameloblastoma, accounting for approximately 90% of all the cases of ameloblastoma. This type of the tumor runs a benign course and slowly expands causing the disfigurement of the face.[11]

This tumor has the propensity to occur in young individuals, with no sex predilection. The mean age of the diagnosis of this lesion is around 3rd decade of life. This occurs most commonly in the mandibular posterior region and slowly infiltrates in the adjacent structures and erode the bone. There can be egg shell crackling effect also. Displacement and mobility of the teeth is often reported along with this tumor. Becelli et al have reported that 13% of cases of mandibular ameloblastoma tend to have paresthesia of the area innervated by mandibular nerve. Recurrence rate is around 60-80%.[4, 12]

The second most common type of ameloblastoma is the unicystic variant. It accounts for approximately 5-15% of all cases of ameloblatoma. This tumor occurs in relatively younger individuals, diagnosed usually in 2nd decade of life and presents as asymptomatic swelling in the mandibular posterior region. Most common clinical differential diagnosis of unicystic ameloblastoma is dentigerous cyst, as both the lesions occur around the crown of the impacted tooth.[13]

Peripheral variant of ameloblastoma (PA) is usually seen as sessile or pedunculated growth over the gingiva. PA has slight elderly male predilection, commonly involving the mandibular posterior region. 30 % of PA happens to occur in the mandibular premolar area. This variant is usually restricted to the soft tissues of the oral cavity and doesn't erode the underlying bone and is often mistaken for pyogenic granuloma. Other lesions with the similar clinical appearance are fibroma, peripheral giant-cell granuloma or peripheral ossifying fibroma.[14]

Desmoplastic ameloblastoma (DA) accounts for 4-14% of all the ameloblastoma cases. It was first described by Eversole et al.in 1984. DA is usually diagnosed after histopathological confirmation. It commonly occurs in 4th decade of life with equal sex predilection. DA is commonly seen in the anterior region of the jaws and is relatively smaller in size as compared to the other variants of ameloblastoma. It usually presents as the painless swelling of the jaw bones leading to the displacement of the teeth.[16]

Histopathologically, solid/multicystic ameloblastoma have six subtypes: follicular, plexiform, acanthomatous, basal cell, granular and desmoplastic. The follicular type of ameloblastoma have proliferating odontogenic epithelial cells arranged in islands, while plexiform type have epithelial cells arranged in continuous anastomosing strands.[11]

Wright et al, 2014 described two main histopathological variants of unicystic ameloblastoma as the luminal and mural type. Peripheral ameloblastoma have similar histopathological features as multicystic ameloblastoma. DA consists of islands of odontogenic epithelium in varied shapes with highly collagenous CT.[10]

Radiographically, ameloblastoma can present either as unilocular or multilocular radiolucencies with the corticated and scalloped borders. Typical radiographic appearance of ameloblastoma can vary from soap bubble to tennis racket or honey comb appearance because of the numerous internal septae. Ameloblastoma has the tendency to cause the buccal or lingual cortical plate expansion. Displacement of the adjacent teeth with the resorption of the roots is a quite common feature.[16]

DA have diffused borders unlike other variants expressing its infiltrative nature in adjacent bone marrow spaces according to Philipsen et al. This feature of DA makes it appear like a fibro-osseous lesion.[17, 18]

Conventional radiography is the initial diagnostic tool for the smaller lesions but the true extent of the large ameloblastoma can be assessed only by 3-dimensional imaging like CT, CBCT, PET or MRI.[19]

Major treatment modalities for the diagnosed cases of ameloblastoma are: surgical enucleation, marsupialization and wide surgical enbloc resection based on the type and size of the lesion.

Our case was diagnosed as the follicular type of conventional ameloblastoma, so the wide surgical enbloc resection was done for the tumorous lesion.

**Conclusion:** Ameloblastoma is a slow growing yet devastating tumour of the jaws. Early diagnosis of this tumour is crucial considering that it can occur even in the young individuals, as in our case. So, complete examination with proper diagnostic imaging modalities are important in the early diagnosis correlating the clinical and radiological findings with the histopathological report.

**Conflict of Interest:** There is no conflict of interest.

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