



## Ameloblastoma : Notorious Tumor of The Jaw - Report of A Case

### KEYWORDS

Ameloblastoma, Jaw neoplasms, odontogenic tumor, mandible

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**ABSTRACT** Ameloblastoma is an uncommon odontogenic neoplasm that accounts for approximately 10% of all tumors originating from gnathic bones. Its behavior has often been described as benign but locally aggressive, along with a high recurrence rates and also the capability of attaining large sizes leading to tremendous facial disfigurement. Clinical and imaging findings aid in the differential diagnosis of ameloblastomas; however, histopathological evaluation is essential for its definitive diagnosis.

The objective of this case report is to describe a case of ameloblastoma, treated successfully by segmental resection, and provide a brief review of the current state of knowledge on this notorious benign tumor of the jaw.

### INTRODUCTION

Ameloblastoma is a enigmatic group of oral neoplasm of odontogenic epithelium, especially of enamel organ-type tissue that has not undergone differentiation to the point of hard tissue formation.<sup>1,2</sup> These tumors are often asymptomatic and often persists as a slow growing, painless swelling, causing expansion of the cortical bones, perforation of the lingual and/or buccal plates and infiltration of soft tissues, more frequently occurring in the posterior mandible<sup>3</sup>. Ameloblastoma has been categorized broadly into three clinicopathological groups : cystic(unicystic),solid( multicystic) and peripheral (extraosseous). Histological there are six types of ameloblastoma: follicular, plexiform, acanthomatous, granular cell, desmoplastic, basal cell, with the first two being most common. The behavior of the solid or multicystic ameloblastoma is markedly distinct from its counterpart. It has been found to have a more aggressive biological behavior with divesting morbidity and higher recurrence rate than the classic unicystic ameloblastoma, and most of all require a rapid and precise approach in implementation of treatment<sup>4,5</sup>. Importance of ameloblastoma lies in its potential to grow into enormous size with resulting bone deformity. We are here by reporting a case of a 24 year old young male diagnosed as ameloblastoma of the mandible in which wide margin surgical excision of the tumor by segmental resection of the right hemimandible was performed with spanning of the bony defect with titanium reconstruction plates to achieve a favorable aesthetic and functional outcome for the patient .

### CASE REPORT

A 24 year old male patient reported to a private dental clinic with complaint of swelling on the right side of the face for the past 1 year. A history of progressive increase in size of the swelling not associated with pain ,after the patient underwent extraction of 47 was elicited. There was no history of similar swelling in the body. Extraoral examination revealed a diffuse swelling in the right mandibular body region approximately 5×4 cm in dimension over lower jaw extending from the zygomatic region to the inferior border of mandible supero inferiorly, and from the corner of mouth to the angle of mandible anteroposteriorly causing facial asymmetry. The skin over the swelling was normal, without visible pulsation or secondary changes. On palpation the swelling was found to be non tender, non-compressible, non reducible and firm in consistency. In-

traorally, there was buccal and lingual expansion extending from 46 to 48 with 47 missing, there was obliteration of the alveolobuccal and alveololingual sulcus.

On roentgenographic examination, of panoramic view of the jaw showed a well defined, round, unilocular radiolucency extending from 46 to 48 region along with Knife edge resorption of mesial and distal root of 47 .The base of the mandible and the anterior border of the ramus was damaged and thinned . (Figure.1)

The 3D reconstruction following CT scan showed an expansile lesion along with destruction of buccal corticle plate from distal aspect of 46 up to the remolar area of the right mandibular region (Figure 2). A provisional diagnosis of ameloblastoma , odontogenic keratocyst or residual cyst was considered . No cervical lymphadenopathy was present and neurosensory testing revealed normal mandibular nerve function and no other focal neurological deficit. In general examination the patient was well developed and well nourished and was appearing distressed about his possible diagnosis.

Surgical excision of the tumor by segmental resection of the right hemimandible, under general anesthesia was planned after which The tumor mass was removed along with wide margin , spanning of the boney defect with titanium reconstruction plates to achieve a favorable aesthetic and functional outcome for the patient .

The excised specimen was then subjected to histopathological examination, which was diagnosed as plexiform ameloblastoma . On microscopic examination, the H & E section studied showed epithelium arranged as a tangled network of anastomosing strands which was seen predominantly . The cords or sheets of epithelium were bounded by columnar or cuboidal ameloblast like cells along with basal cells showing reversal of polarity and hyperchromatism. Stellate reticulum like cells were seen in the centre undergoing cystic degeneration surrounding more loosely arranged epithelial cells. The supporting stroma was loosely arranged and vascular. (Figure 3a,3b). Healing was uneventful, and sutures were removed on 7th postoperative day. Patient has been kept under periodic follow-up of 6 months .No recurrence had been reported .till date.

## Discussion

Ameloblastoma is a benign epithelial odontogenic tumor but is often aggressive and destructive, with the capacity to attain great size, erode bone and invade adjacent structures, chiefly interfering with both function and facial esthetics. It is the second most common odontogenic neoplasm and has been very aptly described by Robinson as being a tumor that is usually unicentric, non-functional, intermittent in growth, anatomically benign and clinically persistent<sup>1</sup>. Gusack in 1827 was the first person to Report the tumor of jaws and the first detailed description of ameloblastoma was given by Flakson in 1879. In 1885 Mallassez coined the term "Admantinoma" as this term suggested formation of enamel which is not found in this tumor Churchill in 1934 suggested alternative name of Ameloblastoma which became popular , well accepted and the preferred terminology till date.<sup>1,4,7,8</sup>

According to classification of odontogenic tumors approved by WHO in 2003,ameloblastoma falls under the category of 'Benign Neoplasm and Tumor-like lesions arising from the Odontogenic apparatus showing odontogenic epithelium with mature fibrous stroma, without ectomesenchyme'<sup>9</sup>

As far as etiology is concerned, It has been postulated that the epithelium of origin is derived from one of the following sources like : cell rests of the enamel organ, epithelium of odontogenic cysts, disturbances of the developing enamel organ,basal cells of the surface epithelium or heterotopic epithelium in other parts of the body.<sup>1</sup>

Leon Barnes has categorized ameloblastomas into four types that is based on their behavioral pattern, anatomical location, histological features and radiographic appearance; as solid (multicystic), unicystic, desmoplastic and peripheral varieties. Out of the four types, first three are intraosseous/ central, while the peripheral is extraosseous. Peripheral ameloblastoma manifest as a sessile or pedunculated slow growing mass that is confined to the gingival or alveolar mucosa with no involment of underlying bone where as intra osseous ameloblastoma arise inside the jaw. Unicystic ameloblastomas are those which have been referred as mural ameloblastomas, luminal ameloblastomas, and ameloblastomas arising from the lining of dentigerous cysts.<sup>1,9,10</sup>

Clinically Ameloblastoma has slight predilection for males and often seen in blacks .Most patients are between 20-40 years with highest incidence noted in 33. The average age of occurrence as reported in literature is 38.9 years .The tumor can occur in young children<sup>7,1</sup>. Ameloblastoma develops in all areas of jaw but mandible has five times higher occurrence of the tumor as compared to the maxilla. Within the mandible, the molar angle, ramus area is involved three times more commonly than are premolar and anterior regions combined. In the maxilla, they most usually occur in the molar area, but may be seen occasionally in the anterior region, maxillary sinus and nasal cavity<sup>8</sup>. Maxillary lesions are more dangerous than mandibular lesions due to tendency for the former lesion to spread more extensively in the more porous maxillary bone and possibility of involvement of the cranial bones.<sup>1</sup>

According to Claudin et al the majority of the patients of ameloblastoma are asymptomatic and symptoms appear with tumoral expansion. Bone tends to expand with growth of tumor and palpation may elicit a hard sensation or crepitus. Surrounding bone may become thin so that fluctua-

tion and egg shell crackling may be elicited.<sup>12,9</sup>

Radiographically Ameloblastoma are commonly observed as a radiolucent area, seen in three different patterns. Most common is the multilocular form with various cysts that are in groups or separated by osseous reinforced septa known as soap bubble appearance. The second most common type is a beehive pattern. A third radiographic presentation, which is very important in terms of a differential diagnosis, is the unilocular form<sup>10</sup>. The appearance of septae on the radiograph usually represents differential resorption of the cortical plate by the tumor and not actual separation of tumor portions.<sup>7</sup>

Histologically Ameloblastoma is characterized by the proliferation of epithelial cells arranged on a stroma of connective vascular tissue in locally invading structures that resemble the enamel organ at different stages of differentiation<sup>12</sup>. Diverse histological patterns have been described in the literature and include follicular, plexiform, acanthomatous, papilliferous-keratotic, desmoplastic, granular, vascular and those with dentinoid induction. The tumor found in our patient was an ameloblastoma of the plexiform type<sup>5,6</sup>. The term plexiform refers to the appearance of anastomosing islands of odontogenic epithelium in contrast to a follicular pattern.<sup>10,12</sup>

For a selection of treatment modality for ameloblastomas, the clinical type (solid, multicystic, unicystic, peripheral), localization, size of the tumor, and age of the patient should be assessed. Resection should be wide to include healthy tissue because recurrence is fairly common with this disease<sup>10</sup>.

## Conclusion.

Historically, ameloblastoma has been recognized for over a century and a half. Its frequency, persistent local growth and ability to produce marked deformity before leading to serious debilitation probably account for its early recognition. Recurrence, especially after conservative treatment has also contributed to the awareness of this notorious lesion. The challenges in the management of this tumor are to provide complete excision as recurrence may occur in incomplete removal and also to reconstruct the bony defect in order to give reasonable cosmetic and functional outcome to the patient.

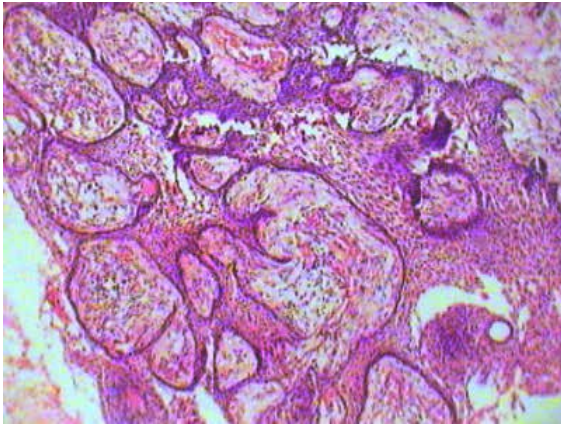
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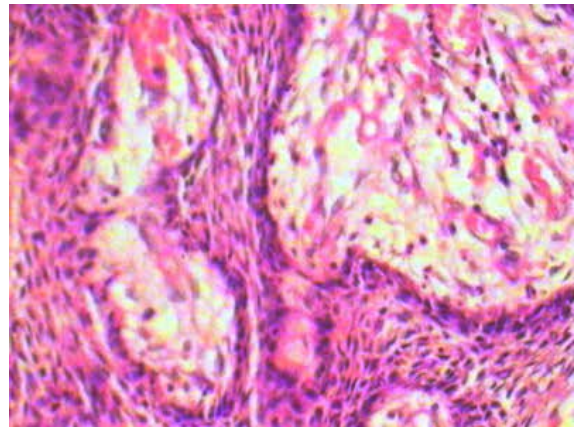
Figure 1: Preoperative 3D reconstruction view following CT scan of the affected mandible



**Figure 2:** Preoperative pantomographic x-ray of mandible showing cystic lesion with unilocular radiolucency .



**Figure 3a**



**Figure 3b**

Figure 3a,b: Photomicrograph showing epithelium arranged as a tangled network of anastomosing strands along with tall columnar cells at the periphery with reverse polarity and Stellate reticulum like cells in the centre. (H & E, Magnification 10 X, 40X)

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