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Ameloblastoma of right angle of mandible in a 30 year old female: A Case report

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Abstract

Ameloblastoma is a true neoplasm of odontogenic epithelial origin. It is the second most common odontogenic neoplasm, and only odontoma outnumbers it in reported frequency of occurrence. Its incidence, approximately 1% of all oral tumours and 18% of all odontogenic tumours, combined with its clinical behaviour, make ameloblastoma the most significant odontogenic neoplasm. A painless expansion of the jaws is the most common clinical presentation; neurosensory changes are uncommon, even with large tumours. Slow growth is the rule, with untreated tumours leading to tremendous facial disfigurement especially for solid ameloblastoma. It is characterised by high recurrence rate and potential to undergo malignant transformation and to metastasize in rare cases. We report a case of follicular ameloblastoma in a 30 year old female.

Keywords: Ameloblastoma, Multilocular, Mandibular neoplasms, Odontogenic tumors.

Introduction

Ameloblastoma is derived from the early English word amel, meaning enamel and the Greek word blastos, meaning germis a rare, benign tumor of odontogenic epithelium (ameloblasts) much more commonly appearing in mandible than maxilla.¹ It was recognized in 1827 by Cusack.² This type of odontogenic neoplasm was designated as an adamantinoma in 1885 by the French physician Louis-Charles Malassez.³ It was finally renamed to the modern name ameloblastoma in 1930 by Ivey and Churchill.⁴ According to WHO 1992 definition Ameloblastoma is defined usually as:-unicentric, nonfunctional, intermittent in growth & anatomically benign, locally invasive polymorphic neoplasm consisting of proliferating odontogenic epithelium, which usually has a follicular or plexiform pattern, lying in a fibrous stromal. It accounts for 5-15% of all intraosseous ameloblastoma. Ameloblastomas are benign tumors

whose importance lies in its potential to grow into enormous size with resulting bone deformity. They are typically classified as unicystic, multicystic, peripheral, and malignant subtypes. The peripheral subtype composes 2% of all ameloblastomas. Of all ameloblastomas in younger patients, unicystic ameloblastomas represent 6% of the cases. A solid or multicystic ameloblastoma is a benign epithelial tumor of odontogenic origin showing a strong tendency to recurrence and local aggression. Solid/multicystic, peripheral, desmoplastic, or unicystic ameloblastomas are other subtypes of ameloblastoma.⁵

Clinical Features: Ameloblastomas occur with equal frequency in both sexes. The age range is usually between the first and the seventh decade of life with a mean in the fourth decade.⁶ They are often associated with the presence of unerupted teeth. Symptoms

include painless swelling, facial deformity if severe enough, and pain if the swelling impinges on other structures, loose teeth, ulcers, and periodontal diseases.

Lesions will occur in the mandible and maxilla, although 75% occur in the ascending ramus area and will result in extensive and grotesque deformities of the mandible and maxilla. In the maxilla it can extend into the maxillary sinus and floor of the nose. The lesion has a tendency to expand the bony cortices because slow growth rate of the lesion allows time for periosteum to develop thin shell of bone ahead of the expanding lesion. This shell of bone cracks when palpated and this phenomenon is referred to as "Egg Shell Cracking" or crepitus, an important diagnostic feature.⁷

Causes The cause of ameloblastoma is not understood. Causes may include injury to the jaw, infections of the teeth or gingiva, or inflammation of these same areas. Infections by viruses or lack of protein or minerals in the diet are also suspected of causing the growth or development of this tumor.⁸

Molecular biology: There is evidence that suppression of matrix metalloproteinase-2 may inhibit the local invasiveness of ameloblastoma, however, this was only demonstrated in vitro. There is also some research suggesting that $\alpha_5\beta_1$ integrin may participate in the local invasiveness of ameloblastomas. A recent study discovered a high frequency of BRAF V600E mutations (15 of 24 samples, 63%) in solid/multicystic ameloblastoma. These data suggests drugs targeting mutant BRAF as potential novel therapies for ameloblastoma.⁹

Roentographic features: Ameloblastoma is tentatively diagnosed through radiographic examination and must be confirmed by histological examination. Radiographically, it appears as lucency in the bone of varying size and features—sometimes it is a single, well-demarcated lesion whereas it often demonstrates as a multiloculated "soap bubble" appearance.

Resorption of roots of involved teeth can be seen in some cases, but is not unique to ameloblastoma. The disease is most often found in the posterior body and angle of the mandible, but can occur anywhere in either the maxilla or mandible. Ameloblastoma is often associated with bony-impacted wisdom teeth—one of the many reasons some dentists recommend having them extracted.⁹

Histopathology will show cells that have the tendency to move the nucleus away from the basement membrane. This process is referred to as "Reverse Polarization". The follicular type will have outer arrangement of columnar or palisaded ameloblast like cells and inner zone of triangular shaped cells resembling stellate reticulum in bell stage. The central cells sometimes degenerate to form central microcysts. The plexiform type has epithelium that proliferates in a "Fish Net Pattern". The plexiform ameloblastoma shows epithelium proliferating in a 'cord like fashion', hence the name 'plexiform'. There are layers of cells in between the proliferating epithelium with well-formed desmosomal junctions, simulating spindle cell layers. The six different histopathological variants of ameloblastoma are desmoplastic, granular cell, basal cell, plexiform, follicular, and acanthomatous. The acanthomatous variant is extremely rare. One-third of ameloblastomas are plexiform, one-third are follicular. Other variants such as acanthomatous occur in older patients.¹⁰

Case report: A 30 year old female patient reported to department of Oral Medicine, Radiology & Diagnosis with the complaint of a progressively enlarging hard mass over the right angle of mandible for the past 3 months (**Fig-1**). The swelling started as a painless swelling which grew slowly to attain the present size. Patient gives history of recurrence of swelling as she got similar swelling enucleated two years back in the same region. While operating for lesion patient mandibular right posterior teeth 46 47 48 were extracted.



Fig 1 Frontal Profile of the patient

There was no history of similar swelling in the body. On extra oral examination, a single ovoid swelling about 5-6 cm in size was seen in the right body and angle of mandible. **(Fig 2)** 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. Intraorally, there was buccal expansion extending from distal surface of 45 to retromolar region. Panoramic radiographic was taken which revealed 6-7cm multilocular, cystic-appearing lesion extending from distal surface of 45 to retromolar region a little less than halfway of ramus of mandible with inferior border of mandible intact. **(Fig 3)** Thinning of lower border of mandible is evident. A provisional diagnosis of ameloblastoma was made. No cervical lymphadenopathy is present and neurosensory testing reveals normal mandibular nerve function and no other focal neurological deficit. Considering the site, age and it being multilocular & recurrent

differential diagnosis of odontogenic keratocyst, aneurysmal bone cyst and central hemangioma was considered. For diagnosis of this multilocular and expansile lesion, aspiration followed by an incisional biopsy was performed with local anaesthesia. Needle aspiration negative for blood or any clear fluids and therefore suggestive of a mass lesion. A typical buccal mucoperiosteal third molar incision was reflected. A large sample of the cystic lining was taken from two different locations. The wound was closed with 3-0 chromic interrupted sutures, and a specimen was sent for histopathological examination. Histopathologically hematoxylin & eosin stained section shows follicles lined by columnar cells with central loosely arranged stellate reticulum like cells. Some follicles are showing cystic degeneration along with squamous metaplasia at places. Areas of necrosis and nonviable bone are also evident. This is consistent with the diagnosis of a multicystic, follicular ameloblastoma. **(Fig 4)**



Fig 2 Lateral Profile of the patient. Extra oral photographs showed mild facial swelling over the body of the left mandible.



Fig 3. Panoramic radiographic of the patient revealing a large multilocular radiolucency at right angle of mandible.

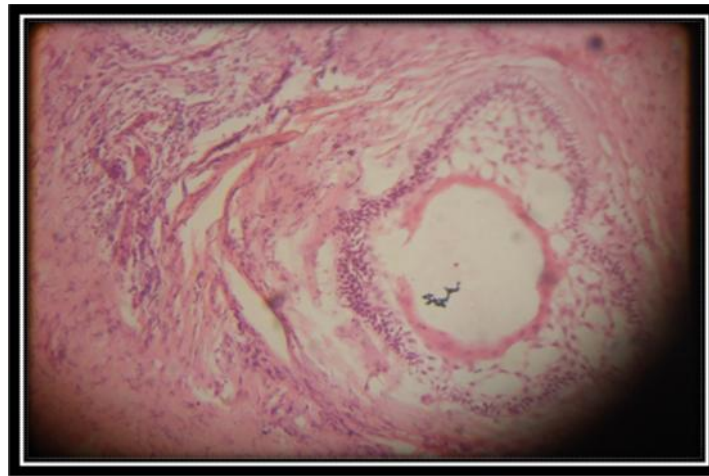


Fig 4 Histopathological slide of the lesion depicting Follicular Ameloblastoma

Discussion: Ameloblastoma is a tumour with a well-known propensity for recurrence. Several factors may influence the rate of recurrence: the clinic radiologic appearance of the tumour, the anatomic site and the adequacy of the initial surgery. Radiologically, the lesions are expansile, with thinning of the cortex in the buccal-lingual plane. The lesions are classically multilocular cystic with a “soap bubble” or “honeycomb” appearance. On occasion, conventional radiographs resembling dentigerous cysts or odontogenic keratocysts.¹¹ The radiographic appearance of ameloblastoma can vary according to the type of tumour. Computed tomography (CT) is usually helpful in determining the contours of the lesion, its contents and its extension into soft tissues. In a patient with a swelling in the jaw, the first step in diagnosis is panoramic radiography. However, if the swelling is hard and fixed to adjacent tissues, CT is preferred. Although the radiation dose is much higher in CT, the necessity of identifying the contours of the lesion, its contents and its extension into the soft tissues, makes it preferable for diagnosis. Plain radiographs do not show interfaces between tumour and normal soft tissue; only interfaces between tumour and normal bone can be seen. The axial view in contrast-enhanced CT images and the coronal and axial views in magnetic resonance imaging (MRI) clearly show both types of interface. Although there are no appreciable differences between MRI and CT for detecting the cystic component of the tumour, for visualizing papillary projections into the cystic cavity, MRI is slightly superior. MRI is essential for establishing the exact extent of an advanced maxillary ameloblastoma and thus determining the prognosis for surgery. Ameloblastomas are treated by curettage, enucleation plus curettage, or by radical surgery.¹² Comparing long term results for 78 ameloblastomas, Nakamura and others reported that the rate of

recurrence is 7.1% after radical surgery and 33.3% after conservative treatment. They recommended wide resection of the jaw as the best treatment for ameloblastoma.¹³

In their series of 26 ameloblastomas, Sampson and Pogrel¹⁴ showed that nearly 31% of tumours recurred after conservative surgery. In present case, the lesion presented as a localised swelling in the mandible and multilocular radiolucency on radiographic examination, which resembled a solid multicystic ameloblastoma or keratocystic odontogenic tumor (KCOT), and was difficult to diagnose clinically and radiographically. The final diagnosis was made on the basis of histopathological examination and also by the correlation of clinical and radiological features. The lesion was surgically removed by department of oral surgery.

Management: While chemotherapy, radiation therapy, curettage and liquid nitrogen have been effective in some cases of ameloblastoma, surgical resection or enucleation remains the most definitive treatment for this condition. The decision to use a radical or conservative approach depends on various factors: 1) the dimensions and the location of the lesion, 2) the growth rate and the relationship with the nearby structures, 3) the histological type, 4) the clinical characteristics, in the recurrences, 5) the general conditions of health and the age of the patient. Because of the invasive nature of the growth, excision of normal tissue near the tumor margin is often required. While not a carcinoma that actually invades adjacent tissues, ameloblastoma is suspected to spread to adjacent areas of the jaw bone via marrow space. Thus, wide surgical margins that are clear of disease are required for a good prognosis. This is very much like surgical treatment of cancer. Often, treatment requires excision of entire portions of the jaw.¹⁵

Recurrence is common, although the recurrence rates for block resection followed by bone graft are lower than those of enucleation and curettage. Follicular variants appear to recur more than plexiform variants. Unicystic tumors recur less frequently than non-unicystic tumors. Persistent follow-up examination is essential for managing ameloblastoma. Follow up should occur at regular intervals for at least 10 years. Follow up is important, because 50% of all recurrences occur within 5 years postoperatively. Recurrence within a bone graft (following resection of the original tumor) does occur, but is less common. Seeding to the bone graft is suspected as a cause of recurrence. The recurrences in these cases seem to stem from the soft tissues, especially the adjacent periosteum. To reduce the likelihood of recurrence within grafted bone, meticulous surgery with attention to the adjacent soft tissues is required.¹⁶

Conclusion

Ameloblastoma is an aggressive tumour of odontogenic origin. There is some difference of opinion about preferable method of treatment, and the only unanimity centres around the fact that complete removal of neoplasm, regardless of how it is accomplished, will result in a cure of the patient. Treatment decisions for ameloblastoma are based on the individual patient situation and the best judgement of the surgeon. Recurrence is the most worrisome long-term complications. This case highlights the importance to correlate the histopathologic findings with clinical and radiographic features to achieve at a correct definitive diagnosis as all such lesions may have prognostically different biologic behaviours and the final diagnosis may alter the therapeutic decision significantly.

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