

Original Research Article

Ameloblastoma –A Case Report with a Systematic Review on Pathogenesis and Recent Trends in Radiographic Assessment

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Abstract: Ameloblastomas a common odontogenic neoplasm is a locally aggressive tumour of the jaw has an unknown etiology but commonly arises from the lining of epithelium of odontogenic cysts, remnants of odontogenic epithelium and basal layer of overlying mucosa. The aim of this report is to describe presentation of an ameloblastomas involving the mandible, and to discuss the differential diagnosis, the radiographic presentation and the management of this lesion.**Keywords:** Ameloblastoma, Posterior mandible, Pathogenesis, Advance imaging

INTRODUCTION

Ameloblastomas common odontogenic neoplasms, is a locally aggressive jaw tumor has an unknown etiology but commonly arises from the lining of epithelium of odontogenic cysts, remnants of odontogenic epithelium and basal layer of overlying mucosa [1]. Ameloblastoma is considered to be aggressive and life threatening if left untreated. Ameloblastoma or adamantinoma or was first described by Cusack in 1827. The lesion has an equal sex predilection with a variable clinical, radiographic and histological appearance [2]. Thus we illustrate a case of ameloblastomam granular cell type in a female patient.

CASE REPORT:

Patient (43/F) reported to the department of oral medicine and radiology with a chief complaint of swelling in the left lower back teeth region for the past 4 months. History revealed that the swelling was increasing in size and pain which was intermittent in nature and present for the past two months. Further history revealed spontaneous exfoliation of the teeth 36, 37 and extraction of 38 done 6 months back by a private dentist, addressing the same complaint. Her medical and surgical history was non-contributory and so was the review on system. On extra oral examination, there was a presence of a single diffuse swelling in relation to the left angle of the mandible measuring approximately 3 X 4 cm in size. The mucosa covering and surrounding the

swelling appears to be normal. No signs of discharge or ulceration were evident. On palpation the swelling is firm in consistency and mildly tender on palpation. It was not compressible and non-reducible (Fig-1). On intraoral examination there was presence of an unhealed extraction socket in relation to 38 regions. The area surrounding the socket appears to be inflamed. On Palpation the alveolus in relation to 38 regions showed bilateral expansion of the cortical plate, extending from the retro molar region to the mesial aspect of 36. The area was tender, with no discharge present (Fig-3). Thus we proceeded with further investigations, her Occlusal radiograph revealed presence of buccal and lingual cortical plate expansion seen irt 36, 37 and 38 region (Fig-4) . OPG revealed no of teeth: 25, presence of multilocular radiolucency with well-defined sclerotic, scalloped margins measuring approximately 4X5 cm in size in relation to 36, 37 and 38 region extending from the alveolus to the inferior border and partially into the ramus of the mandible (Fig-5). Based on the clinical and radiological examination, a provisional diagnosis of ameloblastoma or giant cell lesion of the left of the mandible was made. Further excisional biopsy was carried out. The H & E stain of the biopsied specimen revealed parakeratinized stratified squamous epithelium with underlying fibro cartilagenous connective tissue stroma showing odontogenic islands exhibiting tall columnar cells with reversal of polarity and hyper chromatic nuclei with central stellate reticulum cells,

cystic degeneration and few areas show squamous metaplasia (Fig-6). On the basis of histopathological

and radiological findings, a diagnosis of granular cell ameloblastoma was established.



Fig 1: Profile



Fig 2: ExtraOral



Fig 3: IntraOral



Fig 4: Occlusal Radiograph



Fig 5: OPG

DISCUSSION

Ameloblastomas are odontogenic epithelial neoplasms, also known as ‘cystosarcoma,’ ‘adamantine epithelioma,’ ‘adamantinoma,’ was first described by Cusack in 1827, the term ameloblastoma was later renamed in 1930 by Ivey and Churchill [1, 3]. They are benign slow growing, painless swellings that behave like invasive tumors. They are often locally aggressive, causing death if left untreated. It commonly occurs in the 3rd to 4th decade of life with equal gender and race distribution [2, 3].

Exact etiology of ameloblastoma is unknown. Several etiological factors have been identified, including a) nonspecific traumatizing factors; extraction, caries, trauma, infection, inflammation or tooth eruption; b) Nutritional deficiencies; and c) Viral etiopathogenesis. They can arise from the enamel organ, remnants of the dental lamina, epithelium of dentigerous cysts, or basal cells of oral mucosa epithelium [4, 5].

UNDERSTANDING AMELOBLASTOMA THROUGH TOOTH DEVELOPMENT

The initiation starts with thickening of oral epithelium, which extends into ectomesenchyme as a dental lamina



The cells of dental lamina form a **Bud**-like structure, and this budding cause - condensation of adjacent ectomesenchyme.



. Unequal growth in the different parts of the bud precedes the **Cap Stage**, characterized by a shallow invagination on the deep surface of the epithelial bud (the enamel organ)



At the center of the enamel organ a group of cells form an important signaling center close to inner dental epithelium. This center is known as the enamel knot



The condensed mesenchyme is referred to as dental papilla. Subsequent folding and further growth of the epithelial cap gives rise to the **Bell Stage**



At this stage, enamel organ consists of 4 layers: outer enamel epithelium, stellate reticulum, stratum intermedium and inner enamel epithelium. The inner enamel epithelium produces ameloblasts to form enamel



A higher amount of cells resembling stellate reticulum are usually present in the follicular type compared to the plexiform type of ameloblastoma.



The dental lamina separates from the developing tooth and fragments into discrete clusters of epithelial cells that usually degenerate. However if it persists they are referred to as epithelial pearls or Cell rests of Serres.



The inner and outer enamel epithelium fuse to form the dentin and cementum. It is called the Hertwig's epithelial root (HER) sheath. Remnants of this epithelium is the epithelial rests of Malassez (ERM)



Ameloblastomas arise from epithelium of the enamel organ or dental lamina or epithelial cells of malassez, or heterotopic epithelium. (Epithelium of a previously formed cyst) **Fig-6**



Trauma, chemical insult, impacted teeth or cyst (dentigerous) – leads to abnormal cell growth which infiltrates and destroys surrounding bony tissues leading to ameloblastoma.



The palisading, with polarization of the nuclei in basal cells, is considered as a common characteristic of ameloblastoma and was the main histologic factor that led to identification of ameloblastoma as a neoplasm resembling the dental organ, the fetal tooth-forming structure.



The differentiation level of ameloblastoma cells correspond to the differentiation level of the cells at the cap/bell stage of tooth development and proliferation.



Furthermore proteins that are known to be expressed in the enamel epithelium during the early stage of odontogenesis, including amelogenin, ameloblastin, and tuftelin, are distinctly expressed in ameloblastoma cells



However two other proteins, amelotin and enamelin, which are expressed in the mineralizing stage of enamel formation, are not expressed in tumor cells of ameloblastomas

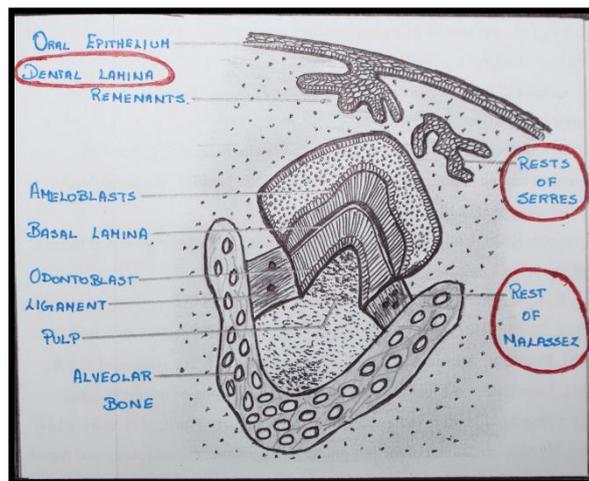


Fig 6: Epithelium leading to formation of ameloblastomas

The benign ameloblastomas can be classified into four histopathological types, i.e. solid/multicystic, extra-osseous/peripheral, desmoplastic, and unicystic type. The solid/multicystic types are the most common and can be subdivided according to their detailed microscopic patterns as acanthomatous, granular cell, basal cell, and desmoplastic type [6]. Although the biological subtypes share many common histological patterns, they are usually classified into two main types, the follicular type and the plexiform type. Both patterns may be present in the same tumor [4, 6].

Clinically it originates within the bone, an exception is the peripheral subtype which arises in the gingival or buccal mucosa and 80 % of the cases occur in the mandible, with a predilection for the posterior mandibular region [7]. Rare sites include sinonasal cavities. The first symptom noticed is facial asymmetry and painless swelling. Pain usually occurs following hemorrhage, commonly after fine needle aspiration and when accompanied with rapid growth it indicates rare malignant ameloblastomas. Tooth displacement and root resorption are reported with desmoplastic ameloblastomas. Paresthesia is uncommon, with rare reported cases of perineural invasion [1, 2, 8].

Radiographically it may appear either as unilocular or multilocular, histologically as unicystic or multicystic. The internal structure has variable appearance ranging from totally radiolucent to mixed, the coarse and curved bony septa originate from normal bone that has been trapped within the tumor it usually creates an internal compartments. That is usually round and of varying size [9]. With growth or expansion of the tumor, there may be coalescence and fusion of the

compartments and as a result, there may be transformation from a multilocular to a monolocular cystic space. On a computed tomography enables visualization of cortical destruction, medullary involvement and soft tissue extension, there by enables identifying extent of the pathology for supporting surgical planning. Areas of low attenuation are seen in the cystic space, enhancement effect primarily in the solid components is seen in contrast-enhanced CT. MRI on the other hand is slightly is slightly superior with its ability to detect the cystic component with a more complete information on the soft tissue involvement and marrow extension . MRI is particularly useful for ameloblastomas arising from the maxilla, as it helps to characterize extension to the orbit, paranasal sinuses, and skull base. Desmoplastic ameloblastomas which has poorly defined soft tissue borders can be often mistaken for fibro-osseous lesion, thus MRI is mainly indicated in such cases [9, 10]. In cases of metastatic ameloblastomas PET-CT is generally, where it mandated as it aids in staging and identifies distant metastasis [10]. Histopathologically ameloblastomas resembles normal odontogenic/enamel epithelium and ectomesenchyme. Microscopic patterns of ameloblastomas include follicular, plexiform, acanthomatous, spindle, basal cell-like, desmoplastic, and granular cell [6, 11]. Treatment with local techniques includes curettage, enucleation or marsupialization. Enucleation can be with or without curettage and involves removing the tumor while avoiding the spilling of neoplasm fluid [12].

Radical treatment includes marginal or en-bloc segmental resection with safety margins and reconstruction of the bony defect. Marginal

mandibulectomy is an 'en bloc' resection of the tumor. In this procedure 1 cm of the adjacent bone and periosteum is removed as it may be invaded by the tumor [11]. Unicystic ameloblastomas have a lower rate of recurrence and enucleation, curettage is usually all that is needed to manage this type. Multicystic ameloblastomas are the most aggressive and have a high recurrence rate following local excision. Since ameloblastomas tend to recur following local treatment, it may be complemented with cryotherapy or diathermy [11, 12].

Cryotherapy is a technique that uses an extremely cold instrument or liquid such as liquid nitrogen to freeze and destroy abnormal skin cells. Diathermy is a procedure that heats and destroys abnormal cells. It uses high-frequency electromagnetic radiation, electric current or ultrasonic waves [13]. Ameloblastomas of the maxilla should be treated as radically as possible [10, 11]. Due to the spongy osteo architecture of the maxilla which facilitates the diffusion of the tumor to the sinus ethmoidalis, pterygoidea fossa, temporal fossa and base of skull. Considering the fact that the conservative treatment of the maxilla bones offers local recurrence in 100% of the cases and a 60% of mortality rate, it is important that the patients with this pathology have a lifelong follow-up. Malignant transformation of ameloblastomas is seen in less than 1% of the cases [14]. Chemotherapy and radiation seem to be contraindicated in ameloblastomas involving the jaw; however the same for distant metastasis is still controversial [15-17].

CONCLUSION

Ameloblastomas are usually benign tumours, with a malignant potential of about 1%. Adequate documentation, thorough history along with clinical examination and interpretations of radiographs are necessary to diagnose these lesions accurately. Radiographic evaluation is necessary to assess the tumor and provide the best treatment possible. For best possible prognosis, the patient should be treated by the specialists in a timely manner. Surgical excision is considered the treatment of choice with minimal recurrence percentage.

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