Original Research Paper

Oral Pathology



A REVIEW ON AMELOBLASTOMA

Dr. Malavica Sethi M*	*Corresponding Author
Dr. Kamali Prabha R	
Dr. Jessica P	
Dr. P. Pooja Sri	
Dr. Mary Tresa Jeyapriya	
Dr. Sathish Kumar	

ABSTRACT Ameloblastomas are a mysterious class of oral tumours that are significant due to their propensity to get extremely large and cause skeletal deformities. The solid or multicystic (the traditional intraosseous ameloblastoma), unicystic, and peripheral variants are the three clinical subtypes. Although surgery is the basis of care, there is debate regarding how much tissue should be removed. Local control rates after radical resections, such as segmental and marginal mandibulectomy, are higher than 90%. Contrarily, local control rates for unicystic and multicystic ameloblastomas, respectively, are typically 80% and 50% for conservative operations such enucleation and/or curettage. In the rare patient with incompletely resectable illness, limited evidence with radiation suggests that treatment may lower the risk of progression and produce long-term local control.

KEYWORDS : Ameloblastoma, Peripheral, Plexiform, Unicystic , odontogenic tumour

INTRODUCTION

Ameloblastoma is a slow-growing, locally invasive odontogenic epithelial tumour that mostly develops from enamel tissue that has not undergone differentiation.⁽¹⁾ Cusack first identified it in 1827⁽²⁾, and the French physician Louis-Charles Malassez classified it as a "adamantinoma" in 1885.⁽³⁾ Ivey and Churchill renamed it "ameloblastoma" in 1930.⁽⁴⁾ A benign epithelial odontogenic tumour, ameloblastoma was listed by the World Health Organization (WHO) in 2017.⁽⁵⁾ Many ameloblastomas have MAPK pathway gene mutations, with the BRAFV600E mutation being the most prevalent.⁽⁶⁾

Epidemiology Of Ameloblastoma

Ameloblastoma most frequently manifests in patients between the ages of 30 and 60, with a small male preponderance, and the mandible being the most common site of presentation ⁽⁷⁾. Ameloblastoma is either the most prevalent or the second-most prevalent benign odontogenic tumour, according to the majority of epidemiological studies.⁽⁸⁾

Clinical Presentation

The most typical presenting feature of ameloblastoma is a slowly developing, painless enlargement of the maxilla or mandible. Every now and then, ameloblastoma is unintentionally found on radiographs collected for other reasons ⁽⁹⁾. 80 percent of ameloblastoma instances (mostly in the posterior mandibular area) involve the mandible. Also, the posterior molar area is where most cases of maxillary ameloblastoma are seen ⁽¹⁰⁾. Ameloblastoma may also be related to unerupted third molar teeth. The ameloblastoma expands significantly as it grows in the buccolingual direction. Upon presentation, ameloblastomas typically measure around 4 cm in size⁽¹¹⁾. Ameloblastoma pain is a rare symptom, however it can be brought on by bleeding within or near the tumour. Other symptoms of ameloblastoma include malocclusion, facial deformities, soft tissue invasion, or loosening of the teeth ⁽¹²⁾. The paediatric age range is where the unicystic type of ameloblastoma most frequently manifests. Due to its frequent dentigerous association with an

immature tooth, it may develop from a pre-existing dentigerous cyst or a dental follicle. As a result, unicystic ameloblastoma most frequently develops in the third molar region ⁽¹³⁾. The average age at which ameloblastoma metastasizes occurs is 43 years and 16 months, with a little male predisposition. The lungs, followed by lymph nodes, are the most often afflicted secondary sites, and the mandibular cases demonstrated greater metastasis than the maxillary instances.⁽¹⁴⁾

Classification

According to the International Agency for Research on Cancer and the World Health Organization, 2003, ameloblastoma is categorised as a benign tumour with odontogenic epithelium, mature fibrous stroma, and without odontogenic ectomesenchyme. Other categories for ameloblastoma include:

- Solid/multicystic
- Extraosseous/peripheral
- Desmoplastic ameloblastoma
- > Unicystic.

Solid/Multicystic Ameloblastoma

The benign epithelial odontogenic tumour known as the solid or multicystic ameloblastoma affects the jaws.⁽¹⁵⁾ It makes up around 10% of all odontogenic tumours in the jaw and is slowgrowing, locally aggressive.⁽¹⁶⁾ Solid multicystic ameloblastomas (SMA) are growths that only appear on the dental lamina and develop from odontogenic epithelial remnants. SMAs can also develop in nonneoplastic odontogenic cysts, particularly dentigerous and odontogenic keratocysts, due to malignant alterations in the lining or wall.⁽¹⁷⁾

SMAs appear as an expansile, radiolucent, multiloculated cystic lesion on radiographs, which has a recognisable "soap bubble-like" look. ⁽¹⁸⁾ Solid ameloblastoma has six histopathologic subgroups, including (Figure 1)

- > Follicular
- Plexiform
- Acanthomatous

VOLUME - 12, ISSUE - 04, APRIL - 2023 • PRINT ISSN No. 2277 - 8160 • DOI : 10.36106/gjra

- ➢ Basal cell
- Granular cell
- Desmoplastic

It is usual to see mixtures of several histological patterns, and the lesions are frequently categorised according to the predominate pattern present. The rate of recurrence varies depending on the histologic subtypes, with the follicular pattern type having the highest recurrence rate at 29.5% and the accanthomatous type having the lowest recurrence rate at 4.5%. The moderately to densely collagenized connective tissue stroma is home to the proliferating islands, strands, and cords of the neoplasm's epithelial component. The creation of the enamel organ can be modelled as a noticeable budding growth pattern with little, spherical extensions of epithelium extending from bigger islands. Vickers and Gorlin's characteristic histological pattern for ameloblastoma is characterised by a peripheral layer of tall columnar cells with hyperchromasia and reverse nuclear polarity.⁽¹⁹⁾

Peripheral Ameloblastoma

An ameloblastoma that is restricted to the gingival or alveolar mucosa is known as a peripheral ameloblastoma (PA). It does not affect the underlying bone; instead, it invades the nearby tissues, primarily the gingival connective tissue. ⁽²⁰⁾ The vestiges of the dental lamina, the so-called "glands of Serres," the vestibular lamina's odontogenic remnants, the mucosal epithelium's basal cell layer, and pluripotent cells from small salivary glands are the sources of the PA. ⁽²¹⁾

Desmoplastic Ameloblastoma

Eversole et al. first described desmoplastic ameloblastoma in 1984, and it was just recently included to the WHO's list of head and neck malignancies (WHO-2005). ⁽²²⁾ Because to the peculiar histomorphology of this tumour, which includes substantial stromal collagenization or desmoplasia, the label "ameloblastoma with prominent desmoplasia" or DA has been proposed.⁽²³⁾

Unicystic Ameloblastoma

Unicystic ameloblastoma (UA) is a subtype of ameloblastoma that manifests as a cyst with odontogenic cyst-like radiologic and clinical features. A characteristic ameloblastomatous epithelium, with or without luminal and/or mural tumour growth, can be seen lining a portion of the cyst cavity upon histologic analysis. The label "UA" ⁽²⁴⁾was first used by Robinson and Martinez in 1977, however it was also referred to as "cystogenic ameloblastoma" in the second edition of the WHO's worldwide histologic categorization of odontogenic tumours. Unicystic ameloblastomas make around 5–15% of all cases.⁽²⁵⁾

In addition to having the characteristic well-differentiated benign histology of the solid/multicystic type at the primary site, metastasizing ameloblastoma also exhibits similar foci of benign histology in areas far from the primary site, which are referred to as metastases.⁽³³⁾



Figure 1: Four Histological Subtypes Of Ameloblastoma A) Unicystic B)Follicular C)Plexiform D)Desmoplastic

Treatment

Surgery is the preferred form of treatment for ameloblastoma. Some treatments, such as radiotherapy or chemotherapy, have a narrow range of use. Surgery With the least amount of morbidity in the donor area, surgical treatment of ameloblastomas aims to reduce recurrences, restore good function, and improve appearance. Currently, complete en bloc resection (radical surgery) with an acceptable margin of safety is advised for treating typical ameloblastomas (solid/multicystic type), which are categorised as segmental or marginal osteotomies for the mandible and partial or complete maxillectomy for the maxilla. A large excision with a 1 to 1.5 cm bone margin is advised for solid/multicystic ameloblastomas due to the high incidence of recurrence after conservative surgery.⁽³⁰⁾

Vascularized free bone grafts from the fibula, ilium, scapula, or radius are the norm for mandibular repair; the preferred flap is the fibular free flap, which also has the benefit of reconstructing long segment mandibular abnormalities.⁽³¹⁾

Non-operative therapy: Localized ameloblastoma cannot be treated with systemic chemotherapy; on the other hand, chemotherapy is still the only option for treating metastatic ameloblastoma.

Radiotherapy plays a limited part in the treatment of ameloblastoma, much like chemotherapy does. It may be used in individuals who have microscopic or large residual disease after surgery, are not good surgical candidates, or have resectable disease.⁽³²⁾

Prognosis

The patient's age, the location and size of the tumour, the histological type, the extent of the disease, and the stage of the disease all affect the prognosis for ameloblastoma ⁽³³⁾Because of the weak barrier provided by the thin maxillary cortical bone, maxillary ameloblastomas tend to be more aggressive and more likely to recur than mandibular ones. Those with untreated maxillary ameloblastoma that has spread to their central nervous system or those who have seen many recurrences may pass away.

CONCLUSIONS

The most frequent odontogenic tumour of the mandible and maxilla is ameloblastoma. This slow-growing, locally invasive tumour first manifests as a painless enlargement of the jaw or maxilla. Ameloblastoma is best treated with vigorous en bloc excision and concurrent rebuilding. Large tissue abnormalities and the high rate of recurrence have long been problems in the management of ameloblastomas. Current molecular advancements overwhelmingly point to the potential for tailored therapy in ameloblastomas that would produce superior results.

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