CASE REPORT

Maxillary Keratoameloblastoma: An Infrequent Odontogenic Tumor

Bisna Lulu V.C.[1], Jilssa Shaji[2], Leela Krishnan C [3], S.K. Padmakumar[4], Nivia M.[5], Sreepriya C.[6]

Abstract

Introduction: Ameloblastoma is a benign odontogenic epithelial tumour that exhibits locally aggressive behaviour and a high recurrence rate. Keratoameloblastoma (KA) is a rare variant characterised by marked central keratin formation. Its clinical, radiographic, and histological features remain poorly defined due to limited reported cases and overlap with similar lesions. Ongoing debate exists regarding whether KA is a distinct entity or a subtype of acanthomatous ameloblastoma. It typically presents as jaw swelling, most often in the posterior mandible, with rare maxillary involvement. Histology may show features resembling an odontogenic keratocyst. Approximately 40 cases have been reported, including a few Type 3 Keratoameloblastoma cases.

Clinical Presentation: 32-year-old male patient, with chief complaint of pain and swelling in the gum region associated with the upper left front teeth

Management and Prognosis: Recurrence occurred in 27% of resected cases and 50% of conservatively treated cases. The biological behaviour of keratoameloblastoma remains uncertain, though conventional ameloblastomas show lower recurrence after resection than conservative treatment

Key words: Ameloblastoma, Keratoameloblastoma, Odontogenic keratocyst

Introduction

Ameloblastoma is widely recognised as the common odontogenic tumour in Asian populations, next to odontoma. First described in the early 1900s, it originates from enamel organ–like tissue but does not develop enough to produce enamel. By the 1930s, it came to be understood as a benign yet locally aggressive tumour, and C. W. Ivy and F. Churchill further refined its classification. The tumour most often occurs in the mandible, accounting for about 80% of cases, while the remaining 20% are found in the maxilla[1].

Keratoameloblastoma (KA) was first described by Jens Jørgen Pindborg in 1970, with the earliest well-documented case reported by Altini and colleagues in 1976. In 1992, the World Health Organisation (WHO) briefly defined it as an Ameloblastoma showing prominent keratin formation. Its histological features were better clarified in 1993 through a case series by Siar and Ng[2]

The 2017 WHO Classification of Head and Neck Tumours recognised KA as a histological variant of Ameloblastoma, alongside conventional, unicystic, and peripheral types, while noting considerable microscopic variability. Despite similar biological behaviour among most variants, rare forms like KA remain diagnostically challenging and are still considered uncommon in the 2022 WHO classification[1].

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KA is a rare and debated variant of ameloblastoma, with uncertainty regarding whether it is a variant, a cystic form, or a hybrid[3], marked by prominent keratin formation. Its true frequency is likely underestimated because its features overlap with other keratinising odontogenic lesions, making diagnosis challenging. There is still no clear agreement on whether KA is a separate entity or a subtype of Acanthomatous ameloblastoma, which has led to uncertainty in its classification[2].

Case Report

A 32-year-old male patient reported to the Department with pain and swelling in the gum region associated with the upper left front teeth. The patient stated that it had been present for approximately two weeks and had gradually increased in size, causing discomfort during chewing. He had initially consulted a private clinic nearby. There are no other significant systemic conditions reported.

Extraoral examination did not reveal any facial asymmetry, swelling, or visible deformity. Intraoral examination showed a localised, firm swelling in the left anterior maxillary region extending in relation to teeth 23, 24, and 25. It was mildly tender on palpation and appeared to involve the underlying bone. Percussion testing elicited a positive response in teeth 21 through 25. Clinical mobility testing revealed grade I mobility in the same teeth. Despite these findings, the surrounding periodontal tissues were healthy, and overall oral hygiene was satisfactory.

Radiological examination:

OPG & Multiplanar CBCT images of Maxilla with reformatted axial, sagittal, coronal, panoramic & 3Dimages were obtained and assessed.

OPG: Unilocular radiolucency extending from the mesial aspect of 11 to the distal aspect of 25

CBCT reveals a well-defined, mildly expansile radiolucent lesion with scalloped margins in the left anterior region, involving teeth 11–25 and measuring about 14.8 × 22.7 × 14.8 mm. It extends from the mesial aspect of 11 to the periapical region of 25, with loss of cortication of the nasopalatine canal.

The lesion involves the nasal floor and left maxillary sinus floor, showing thinning and a small perforation, along with sinus mucosal thickening suggestive of inflammation. Inferiorly, it extends between 21 and 25 and approaches the alveolar crest near 23–24. A small radiopaque fleck near 23 indicates focal calcification.

There is expansion, thinning, and focal perforation of both cortical plates, with root resorption in 23 and 24 and blunting of 21. Overall, the findings suggest ameloblastoma, with OKC as a possible differential.

INCISIONAL BIOPSY

MACROSCOPY

Three soft tissue bits, dark brown in colour, soft in consistency, irregular surface of size 1.5 x 1 x 0.9 cm.

MICROSCOPY :

The epithelium exhibits reversal of polarity with ameloblast-like cells and areas of proliferation into the connective tissue, along with keratin formation and microcyst formation.

DIAGNOSIS: KERATOAMELOBLASTOMA

EXCISION BIOPSY (ENUCLEATION)

MACROSCOPY

Multiple soft tissue bits, along with hard tissue bits, were received. Hard tissue bit resembling a canine tooth and bony spicules, soft tissue bit is white in colour, soft to firm in consistency, irregular surface of aggregate size 5.5x 3.5x 2.3cm.

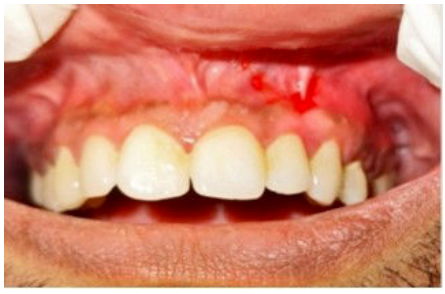

Fig. 1: Swelling seen in relation to the left anterior maxillary region

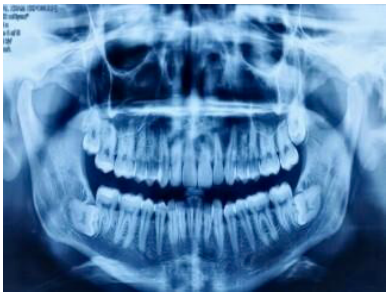

Fig. 2: Orthopantamogram

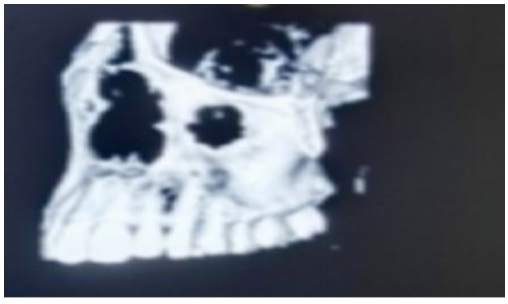

Fig. 3: CBCT

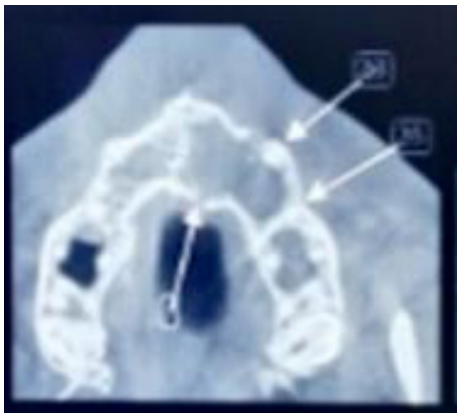

Fig. 4: Axial View

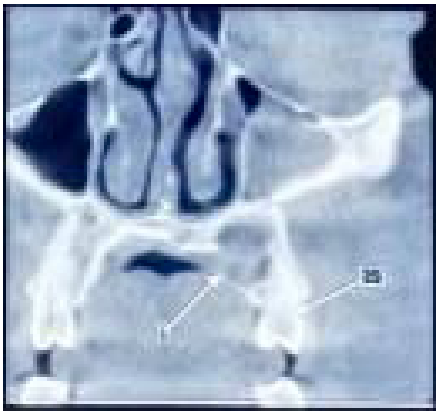

Fig. 5: Coronal View

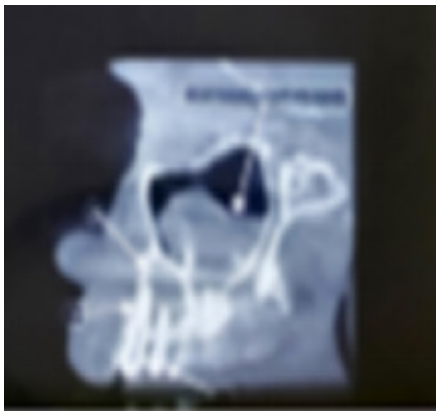

Fig. 6: Sagittal View

Sections show a moderately collagenous fibrous capsule lined by odontogenic epithelium of uneven thickness. The lining epithelium demonstrates surface corrugation, basal cell palisading, parakeratin formation, and focal detachment from the underlying stroma, resembling odontogenic keratocyst. The connective tissue stroma shows multiple odontogenic epithelial islands exhibiting central keratin pearl formation surrounded by ameloblast-like cells, stellate reticulum–like areas, squamous metaplasia with keratin production, and focal microcyst formation. The presence of these ameloblastomatous epithelial islands favours keratoameloblastoma over odontogenic keratocyst.

IMMUNOHISTOCHEMISTRY: CALRETININ- NEGATIVE

DIAGNOSIS: KERATOAMELOBLASTOMA - Simple with OKC-like features

Discussion

Ameloblastoma is a commonly encountered odontogenic tumour of the jaws that, although benign, can behave aggressively and recur if not fully removed. As highlighted in the WHO Classification 2022, correctly identifying KA is important, especially to distinguish it from lesions like keratocystic odontogenic tumour and squamous odontogenic tumour, which do not show the typical features seen in ameloblastoma[1].

There is still some uncertainty about whether KA should be considered a separate subtype. Norval EJ suggested it might just be a form of Acanthomatous ameloblastoma, while Siar and Ng believed its prominent keratinisation makes it distinct enough to stand on its own. The 2017 WHO Classification did not consider Keratoameloblastoma a distinct histological subtype; instead, it was categorised under conventional ameloblastoma owing to its rarity and insufficient evidence to support a separate classification.[2]

Most affected individuals were of Indian origin, followed by patients from Caucasian, Japanese, Malay, and Chinese populations. The reported age span was 21 to 76 years, with a mean age in the 40s. It commonly occurs in the posterior mandible and presents as a symptomatic swelling. Earlier reports showed a strong male preponderance, but the latest data suggest that men and women are now affected almost equally[4].

The lesion usually occurs in the posterior mandible and rarely in the maxilla, presenting as a gradually enlarging, painful swelling. On imaging, it appears as a unilocular or multilocular radiolucency, sometimes with small calcified areas. It may displace teeth, occasionally cause root resorption, and expand bone with possible cortical perforation—though root resorption is less common than in conventional ameloblastoma[2].

Ameloblastoma is known for its marked histological diversity, reflecting the adaptable nature of odontogenic epithelium, though the underlying mechanisms are yet to be fully elucidated.

Keratoameloblastoma is generally classified into four types:

1. Papilliferous type – shows papillary projections into cystic spaces with abundant keratin.

2. Simple type – consists of typical ameloblastomatous islands with prominent keratinisation.

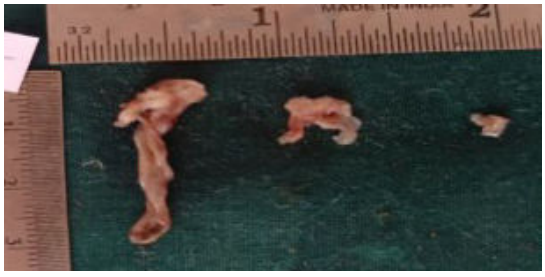

Fig. 7: Grossing of Incision Biopsy

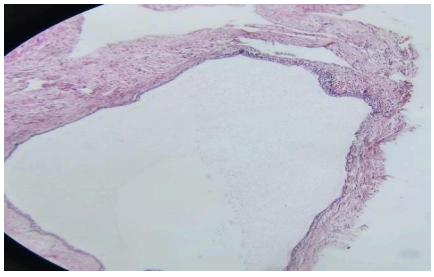

Fig. 8: Parakeratinised stratified squamous epithelium resembling OKC lining

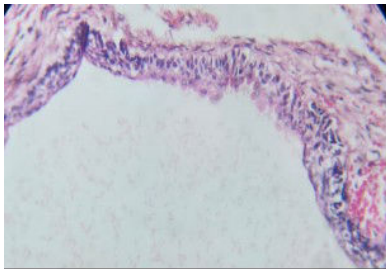

Fig. 9: Ameloblastic Cell Showing Reversal Of Polarity

Fig. 10: Anterior part of swelling

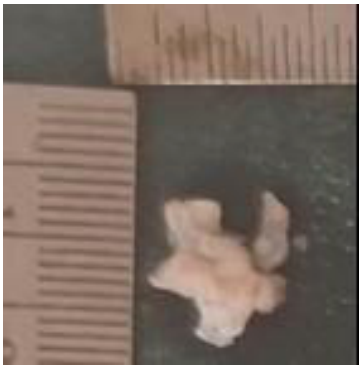

Fig. 11: Palatal part of swelling

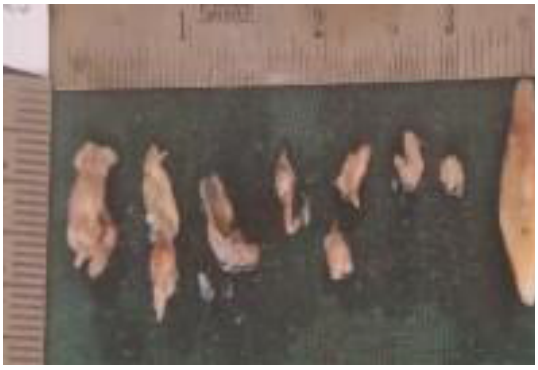

Fig. 12: Superior part of swelling

3. Simple type with OKC-like features – includes areas resembling an OKC alongside classic ameloblastoma features.

4. Complex type – demonstrates a combination of cystic and solid patterns, extensive keratin formation, and occasional hard tissue deposition[5].

Few articles report papilliferous keratoameloblastoma as rare[6], while type 3 Keratoameloblastoma is described in very few literatures, making this case extremely rare

Key features of Keratoameloblastoma show both cystic and solid epithelial follicles within a fibrous stroma. These follicles are made up of loosely arranged cells resembling stellate reticulum, although in this series, the central areas were less prominent, likely due to extensive keratin formation. The peripheral cells are tall, columnar, and display reverse nuclear polarity along with clear subnuclear vacuolization with occasional bone- or cementum-like material[2], are hallmarks that help distinguish KA from OKC.

While the lining may resemble OKC, KA is marked by extensive central parakeratinization, creating a characteristic layered (lamellated) appearance. This keratin can undergo dystrophic calcification, sometimes visible on radiographs. However, overlap with lesions like the solid variant of OKC and those with multiple daughter cysts can make diagnosis difficult, as they may appear similar despite being distinct entities[2].

Studies by Altini and De Villiers have demonstrated that immunohistochemical staining shows calretinin positivity in unicystic and conventional ameloblastomas, predominantly within stellate reticulum-like cells, whereas odontogenic keratocysts typically lack staining. In contrast, the Keratoameloblastoma cases evaluated showed complete absence of calretinin expression. This difference may be attributed to the extensive central keratinization and squamous metaplasia characteristic of Keratoameloblastoma, which replace the stellate reticulum like areas where calretinin is normally expressed[7]. Robinson and colleagues have also reported similar findings. The absence of calretinin may therefore serve as a useful supportive feature in distinguishing Keratoameloblastoma from conventional ameloblastoma[2]. However, because calretinin negativity is also observed in odontogenic keratocysts, its diagnostic specificity is limited and it should be interpreted in conjunction with the overall

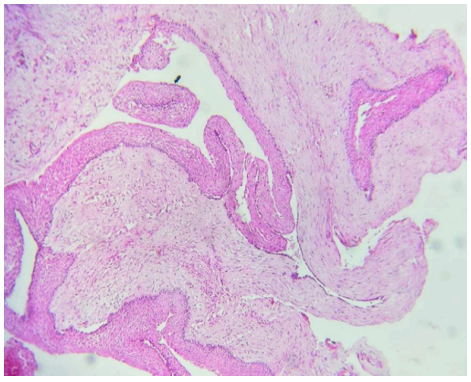

Fig. 13. 4x -Parakeratinised epithelium (OKC lining epithelium), detachment of epithelium from underlying connective tissue.

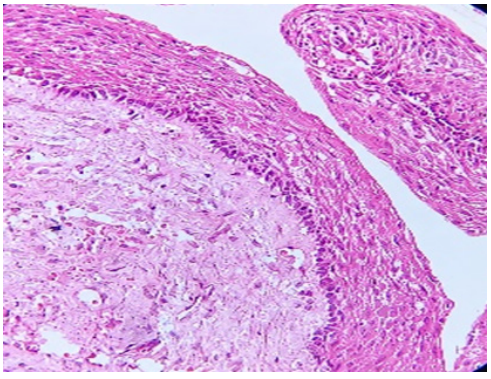

Fig. 14: 40X- Ameloblastic epithelium with peripheral palisading , reversal of polarity and subnuclear vacuolization

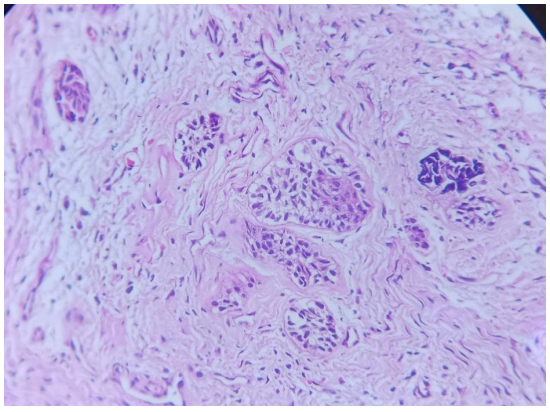

Fig. 15: 40X - Ameloblastic follicles are noted, showing central stellate reticulum like areas with squamous metaplasia exhibit central keratin formation and focal areas of hard tissue formation within the follicles.

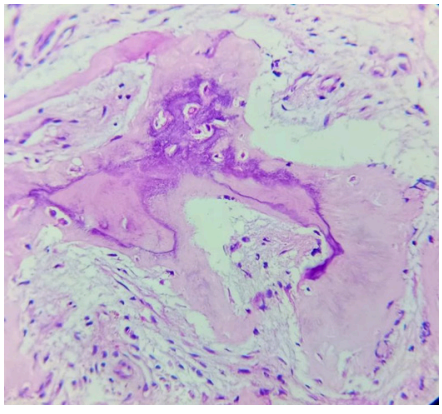

Fig. 16: 40 X- Peripheral reactive bone formation

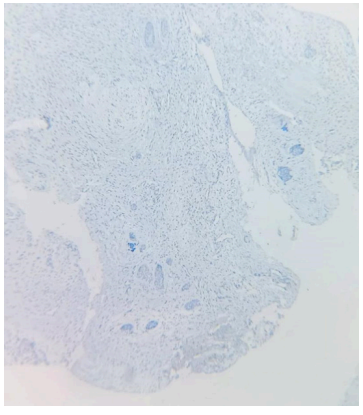

Fig. 17: Immunohistochemistry-Calrectinin Negative

Feature	Keratoameloblastoma	Acanthomatous Ameloblastoma	OKC
Histology	Ameloblastic follicles with keratin formation	Ameloblastic follicles showing squamous metaplasia	Parakeratinized cystic epithelial lining
Keratinization	Extensive and prominent	Focal to moderate	Surface corrugated parakeratin
Stellate reticulum-like cells	Reduced or replaced by keratinized cells	Present with squamous metaplasia	Absent
Calretinin	Negative	Positive	Negative
Diagnostic clue	Extensive keratinization within a typical ameloblastoma	Squamous metaplasia within ameloblastic follicles	Cystic lesion with corrugated parakeratinized lining and no ameloblastic follicles

histopathological findings rather than as an isolated marker.

Extensive keratinization within ameloblastic follicles favors keratoameloblastoma, squamous metaplasia favors acanthomatous ameloblastoma, and a corrugated parakeratinized cyst lining with calretinin negativity favors OKC.

PROGNOSIS AND MANAGEMENT

Recurrence occurred in 27% of cases managed by resection and 50% of those treated conservatively among cases with follow-up. However, due to the small number of cases and inadequate follow-up, firm conclusions cannot be made. The biological behaviour and prognosis of KA remain uncertain. Conventional ameloblastoma is known to have a significantly lower rate of recurrence when managed with surgical resection compared to conservative treatment approaches.

Conclusion

Careful slide evaluation and sound histopathological knowledge are crucial for accurately diagnosing Keratoameloblastoma and distinguishing it from odontogenic keratocyst, ensuring appropriate surgical management.

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