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Case Report

Keratoameloblastoma of the lower jaw: A rare challenging entity

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ABSTRACT

Ameloblastoma is the second most common benign odontogenic tumor with various histopathologic features. Except for the unicystic type of ameloblastoma, the different microscopic patterns of this tumor show no significant correlation with long-term clinical behavior. During recent decades, additional challenging subtypes of ameloblastoma, including "Keratoameloblastoma" (KA), have been introduced in the literature. Here, we present a case of KA and discuss the important diagnostic microscopic features.

Key Words: Ameloblastoma, jaw neoplasms, odontogenic tumors

INTRODUCTION

Ameloblastoma is the most common odontogenic epithelial tumor excluding odontoma. This entity is a benign, slow-growing, and locally invasive neoplasm that probably arises from intraosseous remnants of odontogenic tissues.[1] They can occur in either jaw, but the posterior region of the mandible is the most common site. Histopathologically, ameloblastomas typically consist of islands, nests, cords, or strands of odontogenic epithelium rimmed by columnar cells, demonstrating nuclei located in the apical half of the cell body away from the basement membrane (reverse polarity appearance). A wide variety of histologic patterns are recognized, including follicular, cystic, acanthomatous, plexiform, desmoplastic, granular cell, and basal cells.[2,3]

Acanthomatous change with attempted keratinization may be observed in ameloblastomas, usually in the form of squamous metaplasia of the central stellate reticulum-like cells of the tumor islands.[4] The term "keratoameloblastoma" (KA) has been applied to rare examples of ameloblastoma that exhibit a markedly greater amount of keratin production throughout the tumor. This term was first introduced by Pindborg in 1970, and Altini *et al*. reported the first verified case.[5,6]

This article's goal is to describe the clinical, radiological, and histological characteristics of a new instance of KA.

CASE REPORT

In January 2023, a 54-year-old female patient with no significant past medical or dental history was referred to a private dental office with an expansile swelling of the right side of her face. Radiographic

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examination revealed a well-defined, unilocular lesion with buccal and lingual expansion in the posterior mandible [Figure 1].

The clinicoradiographic differential diagnosis included a "unicystic ameloblastoma" and "developmental odontogenic cysts" - especially odontogenic keratocyst. Then, an excisional biopsy was performed, and the specimen was sent to the pathology laboratory for histological diagnosis.

The gross examination of the specimen showed multiple fragmented pieces of soft gray-brown cyst-like tissue measuring 4 cm × 3 cm × 2 cm in the aggregate, as well as two pieces of bony tissue measuring 1 cm × 1 cm × 0.7 cm.

The tissues were completely embedded, and an odontogenic tumor with cords and nests of odontogenic epithelium inside a dense connective tissue stroma was found during the histological inspection of the H- and E-stained sections. Numerous proliferating follicular islands showed palisaded ameloblast-like cells with prominent reverse nuclear polarity at the periphery. The central epithelium consisted of loosely arranged cells resembling the stellate reticulum of the enamel organ. The low-power magnification showed multiple cystic spaces, which were filled with keratinized material [Figure 2]. High-power examination of the islands showed bland-appearing cells with no evidence of mitotic activity. Therefore, a final diagnosis of "KA" was made. Additional surgical treatment including peripheral ostectomy was performed to minimize the risk of recurrence of the tumor, with a plan for close long-term follow-up.

Furthermore, no evidence of recurrence is observed after 6-month follow-up.

DISCUSSION

Ameloblastoma is a benign, but locally invasive epithelial neoplasm that resembles the enamel organ. In some instances, an aggressive clinicoradiographic presentation of ameloblastoma might be challenging to differentiate from other primary or metastatic neoplasms. Then, microscopic features are often helpful to make a definite diagnosis.[7]

A rare form of ameloblastoma known as "KA" exhibits a keratinization pattern described as showing a lamellar appearance.[8,9] All reported cases in English literature are summarized in Table 1. The World Health Organization (WHO) described histopathologic findings of KA as a separate entity that was described as "ameloblastoma with considerable keratinization" in 1992, although this entity is not included in the most recent WHO classification for odontogenic tumors. Due to keratinization, WHO recognized this lesion as belonging to the histopathological spectrum of "Acanthomatous ameloblastoma."[15,17] We believe that beyond the various patterns of ameloblastoma, it is crucial to differentiate KA from other keratin-forming jaw lesions [Table 2].

Intraosseous well-differentiated squamous cell carcinoma should be considered an important differential diagnosis. Significant cellular atypia with pearl-like keratinization and lack of microscopic features of ameloblastoma (such as palisading and

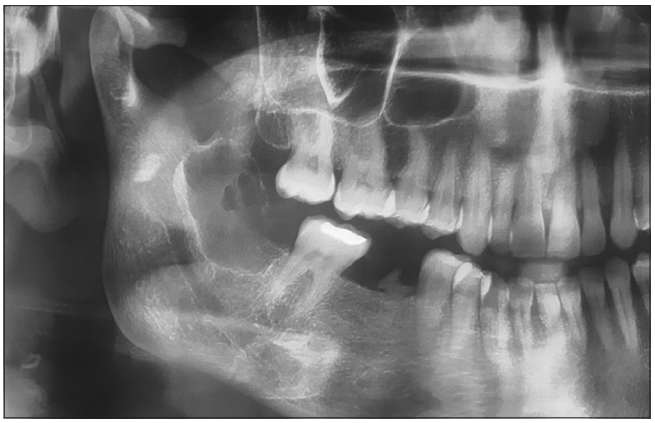

Figure 1: Panoramic view shows a well-defined lesion on the posterior area of the right mandibular bone.

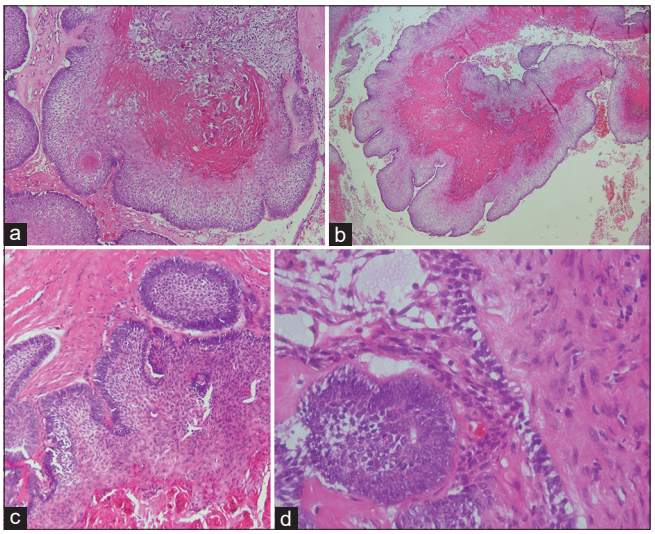

Figure 2: (a and b) Prominent keratinization at the superficial layers of the epithelium (×100). (c and d) Palisading and reverse polarity at the peripheral columnar cells and stellate-like cells at the central area of the nests (c: ×100 and d: x400).

Table 1: Summary of keratoameloblastoma cases reported in English literature

Case	Age	Site	Clinical findings	Radiographic findings	Follow up
1. Pindborg, 1970[5]	57	Posterior mandible	-	Multilocular radiolucent	-
2. Altini *et al*., 1976[6]	28	Anterior maxilla	Swelling	Multilocular radiolucent	-
3. Altini *et al*., 1991[10]	76	Posterior mandible	Swelling	Multilocular radiolucent	No evidence of recurrence at 1 year
4. Siar and Ng 1993[11]	30	Anterior mandible	Swelling	Multilocular radiolucent	-
5. Siar and Ng 1993[11]	35	Posterior mandible	Swelling	-	-
6. Siar and Ng 1993[11]	35	Posterior maxilla	Swelling	Mixed	-
7. Siar and Ng 1993[11]	39	Anterior mandible	-	Unilocular radiolucent	-
8. Norval *et al*., 1994[12]	26	Posterior mandible	Painful swelling	Multilocular radiolucent	-
9. Said-al-Naief *et al*., 1997[13]	26	Posterior maxilla	Mass from socket	Unilocular radiolucent	Recurrence at 7 months
10. Kaku *et al*., 2000[14]	35	Posterior mandible	Pain	Multilocular radiolucent	
11. Takeda *et al*., 2001[8]	76	Posterior mandible	Swelling	Multilocular radiolucent	-
12. Collini *et al*., 2002[7]	62	Posterior mandible	Swelling	Irregular with soft tissue extension, internal calcifications	Two recurrences at 39 and 58 months
13. Whitt *et al*., 2007[15]	45	Anterior maxilla	Painful swelling, tooth mobility	Unilocular, internal calcifications	No evidence of recurrence at 10 months
14. Adeyemi *et al*., 2010[16]	38	Posterior mandible	Swelling, tooth mobility	Multilocular radiolucent	No evidence of recurrence at 24 months
15. Sisto and Olsen 2012[17]	35	Posterior mandible	Painful swelling	Multilocular mixed	-
16. Ketabi *et al*., 2013[18]	21	Anterior mandible	Painful swelling, tooth mobility	Unilocular radiolucent	No evidence of recurrence at 12 months
17. Mohanty *et al*., 2013[19]	46	Posterior mandible	Swelling	Multilocular radiolucent	-
18. Raj *et al*., 2014[20]	22	Posterior maxilla	Painful swelling, paraesthesia	Unilocular radiolucent	No evidence of recurrence at 24 months
19. Lee *et al*., 2015[9]	56	Posterior maxilla	Swelling	Unilocular, internal calcification	Multiple recurrences at 3, 6, 17, 21, 33, 46 months
20. Bedi *et al*., 2015[21]	27	Posterior mandible	Painful swelling	Unilocular radiolucent	Recurrence at 5 years
21. Palaskar *et al*., 2015[22]	65	Anterior mandible	Painful swelling	Unilocular radiolucent	Recurrence at 4 months
22. Konda *et al*., 2016[23]	44	Posterior mandible	Painful swelling, tooth mobility	Unilocular radiolucent	No evidence of recurrence at 1 year
23. Prabhakar *et al*., 2020[24]	74	Posterior mandible	Swelling	Multilocular radiolucent	No recurrence at 6 months
24. Prabhakar *et al*., 2020[24]	42	Anterior maxilla	Swelling	Multilocular, internal calcifications	-
25. Kuberappa *et al*., 2020[25]	65	Anterior and posterior mandible	Painful swelling	Multilocular radiolucent	No evidence of recurrence at 2 months
26. Robinson *et al*., 2022[26]	41	Posterior maxilla	Swelling	Unilocular radiolucent	Recurrence at 6 years
27. Robinson *et al*., 2022[26]	40	Posterior maxilla	Swelling	Unilocular, internal calcification	No evidence of recurrence
28. Robinson *et al*., 2022[26]	31	Posterior mandible	Swelling	Unilocular radiolucent	No evidence of recurrence
29. Robinson *et al*., 2022[26]	45	Anterior and posterior mandible	Swelling	Multilocular, internal calcifications	No evidence of recurrence
30. Robinson *et al*., 2022[26]	29	Anterior and posterior maxilla	Swelling	Multilocular mixed	No evidence of recurrence
31. Robinson *et al*., 2022[26]	45	Anterior and posterior mandible	Swelling	Multilocular, internal calcifications	-
32. Robinson *et al*., 2022[26]	48	Posterior mandible	Swelling	Multilocular, internal calcifications	No evidence of recurrence

reverse polarity at the periphery and stellate-like reticulum at the central area of tumoral nests) can be helpful to rule out variants in ameloblastoma family. In addition, clinicoradiographic findings typically demonstrate an invasive malignant behavior in squamous cell carcinoma.

There are several variations and histologic forms of conventional ameloblastoma, including plexiform, follicular, acanthomatous, basal cell, and granular cell.[16] Acanthomatous ameloblastoma, KA, and the solid variant of odontogenic keratocyst (SOKC) can have similar histologic features, and keratinous material is produced by all three lesions. In acanthomatous ameloblastomas, squamous metaplasia occurs in the central cells of the tumor islands, which sometimes may result in focal

Table 2: Summary of jaw lesions with keratinization in differential diagnosis with keratoameloblastoma

Lesion	Microscopic features	Clinicopathologic findings	Therapeutic approach
Squamous cell carcinoma	Cellular atypia Pearl-like keratinization No evidence of ameloblast-like features	Destructive behavior of malignancy	According to surgical guidelines for malignancies
OKC	No evidence of solid component No evidence of reverse polarity at basal layer	Cystic behavior with prominent intraosseous extension	Enucleation and curettage with/without primary marsupialization
Solid OKC*	No evidence of reverse polarity at basal layer	Similar to KA	From curettage and osteotomy to resection
Acanthomatous ameloblastoma	Prominent squamous metaplasia at neoplastic nests Pearl-like keratinization	Similar to KA	Resection

*Regarding the rarity of case reports, there is no agreement on the therapeutic approach. KA: Keratoameloblastoma; OKC: Odontogenic keratocyst

overt keratin production. Although this subtype demonstrates common microscopic features of ameloblastoma, including basal cells' palisading and reverse polarity, the degree of keratinization of the neoplastic cells is not as prominent as that observed in KA and SOKC.

To differentiate KA from SOKC, no significant, well-defined criteria have been documented, mostly because of the rarity of SOKC cases. However, the reverse polarity of columnar cells at the periphery and loosely arranged cells resembling the stellate reticulum toward the center of the tumoral nests are not usually observed in SOKC.[10,22] In addition, OKC with prominent daughter cysts formation may show overlapping features with KA and SOKC.

In conclusion, KA should be recognized as a separate subtype of ameloblastoma (not a separate entity) with distinguishable microscopic features from other cystic/neoplastic lesions of the jaw. In addition, it seems that there is no clinical significance for KA in comparison with other subtypes of solid ameloblastoma.

Consent

The patient noticed that her name and photo will not be published in the report.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that his name and initials will not be published and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

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Conflicts of interest

The authors of this manuscript declare that they have no conflicts of interest, real or perceived, financial or non-financial in this article.

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