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Keratoameloblastoma: a tumor *sui generis* or a chimera?

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The term keratoameloblastoma has been used to describe a histologically heterogeneous group of ameloblastoma variants which have in common the formation of keratin by the ameloblastomatous epithelium. The English language literature contains reports of only 12 cases of keratoameloblastoma, of which 4 cases exhibited a papilliferous component. We report a unique tumor that we believe falls within the broad histopathologic spectrum of keratoameloblastoma. We review the key clinical and histopathologic features of the previously reported cases of keratoameloblastoma and present an additional case that presented as an expansile, radiolucent lesion with internal opacification between the roots of teeth in the left anterior maxillary alveolar ridge of a 45-year-old white male. There is wide variation in the histopathologic appearance of cases reported under the appellation keratoameloblastoma. Our case exhibited a histopathologic feature shared by only 2 of the previously reported cases, notably islands and anastomosing cords of epithelium forming lamellated, pacinian-like stacks of parakeratin that extruded into the collagenous tumor stroma without eliciting a foreign body response. Due to the small number of reported cases, we are unable to accurately assess whether the biologic behavior of keratoameloblastoma differs from other histologic types of ameloblastoma. **(Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2007;104:368-76)**

The appellation keratoameloblastoma has been applied to ameloblastomas that exhibit evidence of keratin production to varying degrees. Keratin formation in ameloblastomas may take various forms. Squamous metaplasia within the stellate reticulum-like areas of ameloblastoma is a well-recognized feature that is the histologic hallmark of the acanthomatous variant of ameloblastoma.[1] It has long been recognized that keratinization in acanthomatous ameloblastomas may progress to keratin pearl formation in the central portion of the epithelial follicles.[2-4]

The English language literature reports 12 lesions classified as keratoameloblastoma. A major distinction in these reports is whether or not the lesion exhibited a papilliferous component. Despite the similarity of names, keratoameloblastoma and papilliferous keratoameloblastoma are distinct morphologically. Pindborg[5] first proposed the term keratoameloblastoma for use as a diagnostic entity, although some[6-8] erroneously attribute this to his 1958 publication,[9] rather than to his 1970 publication.[5] In his report, Pindborg described a histologic variant of ameloblastoma, which he termed papilliferous ameloblastoma.[5] This lesion exhibited a growth pattern consisting of both epithelial follicles exhibiting central microcyst formation with keratinization and follicles exhibiting a papilliferous lining epithelium. Subsequently, 3 additional cases of ameloblastoma with a papilliferous component were reported in 1991,[10] 1994,[11] and 2002,[12] bringing the number of case reports of ameloblastoma with papilliferous features in the English language literature to 4 (Tables I and II). The remaining 8 cases without a papilliferous histologic component were reported under the terms

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Table I. Clinical features of present and previously reported cases of keratoameloblastoma

Author	*Papilliferous component*	*Age, y, gender, race*	*Clinical*	*Radiographic*	*Treatment*	*Follow-up*
Whitt et al.	No	45M Caucasian	Anterior maxillary alveolar process. Eroded buccal and lingual cortex and nasal floor	2.0-cm lucency with internal calcification and an ill-defined margin	Curettage	No evidence of disease at 10 months
Collini et al.[12]	Yes	62M, race unknown	Ramus and condyle of right mandible	5.0 cm × 5.0 cm hard mass. Irregular lucency with soft tissue extension and internal calcification	Hemi-mandibulectomy	Two local recurrences at 39 and 58 months
Takeda et al.[8]	No	76M Japanese	Left mandibular body	Multilocular lucency greater than 4.0 cm	Resection	Unknown
Kaku[14]	No	35M Japanese	Right mandibular body	Lucency between the roots of the first and second molar teeth	Unknown	Unknown
Said-al-Naief et al.[7]	No	26M African American	Right posterior maxilla. Expansion into maxillary sinus	Well-defined lucency between the roots of the first and second molar teeth	Curettage, followed by partial maxillectomy	Recurred within 6 months after curettage. Unknown after maxillectomy.
Norval et al.[11]	Yes, focal	26F, race unknown	Right mandibular body, eroded buccal cortex	Lobulated radiolucency	Segmental resection	Unknown
Siar et al.[6]	No	30M Chinese	Anterior mandible	Multilocular radiolucency	Resection	Unknown
Siar et al.[6]	No	35M Malay	Left mandible	No information	Hemi-mandibulectomy	Unknown
Siar et al.[6]	No	35F Malay	Right maxilla	Ground glass, indistinct borders	Unknown	Unknown
Siar et al.[6]	No	39F Chinese	Left anterior mandible	Cystic radiolucency	Enucleation	Unknown
Altini et al.[10]	Yes	76M Black	Right mandibular body, angle, ramus	Multilocular radiolucency	Hemi-mandibulectomy	No evidence of disease at 12 months
Altini et al.[13]	No	28M Caucasian	Anterior maxilla	Multilocular radiolucency	Wide local excision	Unknown
Pindborg[5]	Yes	57F, race unknown	Right mandibular body and ramus	Multilocular radiolucency	Not reported	Unknown

keratoameloblastoma or keratinizing ameloblastoma in 1976[13] (For reference 13, note that MEDLINE incorrectly cites Altini M as first author of the report by Lurie R, Altini M, and Shear M. We have chosen to cite this report as Altini M, Lurie R, and Shear M, using the incorrect MEDLINE citation, to be consistent with prior citations in the recent literature.), 1993,[6] 1997,[7] 2000,[14] and 2001[8] and generally consisted of odontogenic epithelial follicles exhibiting varying degrees of keratinization (Tables I and II).

The purpose of this article is to present the histopathologic features of a lesion that we believe falls within the histopathologic spectrum of keratoameloblastoma, to illustrate the broad spectrum of histomorphology presently implied by the diagnostic term keratoameloblastoma, and to suggest proper terminology for subsets of this lesion.

CASE REPORT

A 45-year-old white male presented with a lesion of the left anterior maxillary alveolar ridge, between the roots of vital teeth, which had been enlarging for at least 6 months. Clinically, there was both facial and palatal expansion of the alveolar process. The patient's chief complaint was swelling, but in the interim period