Papilliferous Keratoameloblastoma: A Rare Case Report

Abstract

Ameloblastoma is true odontogenic tumor of epithelial origin, which is described as locally aggressive with varying chances of recurrence. It is believed to derive from enamel organ, remnants of dental lamina, lining of odontogenic cysts, or basal cells of oral epithelium. Radiologically, it may present as unilocular or multilocular radiolucency commonly. Although conventional ameloblastoma presents typical histological features as described by Vickers and Gorlin, few unusual variants have been reported with different histological patterns. However, the clinical and biological behavior of these lesser known variants has not been established yet due to the scarcity of cases reported. Here, we report an extremely rare case of papilliferous ameloblastoma in a young male patient with 2-year follow-up and presenting with unusual histological presentation than conventional ameloblastoma.

Keywords: *Ameloblastoma, keratoameloblastoma, papilliferous*

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Introduction

Ameloblastoma is the second most common odontogenic tumor arising from odontogenic epithelium and bearing the most clinical significance, following odontomas.[1] It was first described by Cusack in 1827; however, the term ameloblastoma was coined by Ivey and Churchill in 1930.[2] It was aptly defined by Robinson in 1937 as a tumor that is usually "unicentric, nonfunctional, intermittent in growth, anatomically benign, and clinically persistent."[2,3] Although most ameloblastomas are histologically benign and lack cytological atypia, they are locally aggressive and destructive, with evidence of inconsistent rates of recurrence.[4] The WHO describes four variants of ameloblastoma, i.e., solid-multicystic follicular, plexiform, desmoplastic, and unicystic.[5] However, many unusual histological variants have been reported in the past that mimic the conventional ameloblastomas but present with a widely variable histological presentation. Only a few cases of such histological variants though have been reported in the literature which has made it difficult to predict the clinical course and biological behavior of these variants. We report a case of papilliferous keratoameloblastoma (KA) which is a very uncommon histopathological variant of ameloblastoma.

Case Report

An 18-year-old male patient reported with a chief complaint of swelling in the right side of the mandible since 3 months. Extraorally, a diffuse swelling was present near base of the mandible till the inferior margin, hard in consistency with regular margins, and was not associated with pain, paresthesia, or discharge. The locoregional lymph nodes were not palpable. On intraoral examination, swelling was present in the right buccal vestibular region in the premolar-molar region in the right vestibular area without any signs of ulceration or paresthesia. The teeth in the region showed a positive response to stimulus on vitality testing. The orthopantomograph showed a well-defined radiolucent unilocular osteolytic lesion extending from the distal periradicular area of 43 till right posterior body angle region corresponding to mesial crown outline of 48 anteroposteriorly and from the alveolar crest till the inferior mandibular cortex superoinferiorly [Figure 1]. The three-dimensional cone-beam computed tomography reconstruction showed a lesion measuring 5.9 cm × 3.2 cm × 2.7 cm in size with fine curved and linear bony septae in the center of the osteolytic area. The lesion was expansile with perforation of the buccal and lingual cortical plates and thinning of the inferior border of the mandible. The histopathological examination of

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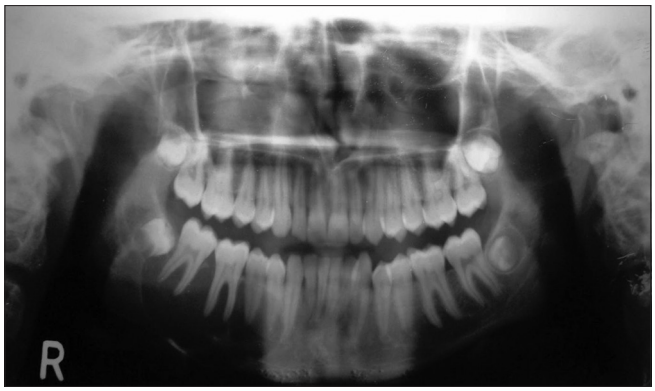

Figure 1: Orthopantomograph of the patient showed a radiolucent multilocular radiolucency on the right side of mandible

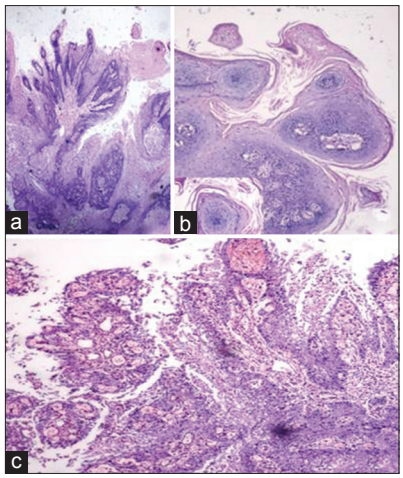

Figure 2: (a) Photomicrograph showing papilliferous histology of the epithelium with thin connective tissue cores (H and E, ×4); (b) Photomicrograph showing keratin on the surface in the form of parallel lamella and keratin flakes (H and E, ×10, Inset ×40), (c) photomicrograph showing acantholytic cells with areas of focal necrosis on the surface (H and E, ×4)

the submitted specimen showed lining epithelium with tall columnar cells with superficial loosely arranged stellate reticulum-like cells with mature fibrocellular stroma and proliferating odontogenic islands. The lining epithelium was thrown into multiple sharp or blunt and rounded verrucopapillary projections into the cystic lumen plugged by keratin [Figure 2a]. These projections were supported by thin connective tissue cores. Keratin was seen deposited on the surface in the form of parallel lamella, and keratin flakes were also present [Figure 2b]. An unusual finding seen in our case was the presence of acantholytic cells with areas of focal necrosis on the surface [Figure 2c]. Multiple odontogenic islands with peripheral columnar odontogenic cells with hyperchromatic nuclei were present in the connective tissue stroma. The supporting connective tissue stroma showed epithelial islands with acanthomatous changes and microcystic degeneration. The connective tissue was densely fibrocellular with numerous dilated and engorged blood vessels. Immunohistochemical staining with cytokeratin 19 (CK 19) showed positivity of the basal and suprabasal cells indicating odontogenic nature of the epithelium, whereas immunohistochemistry with anti-Ki-67 antibody showed intense positivity in the basal and suprabasal cells with infrequent positivity in the superficial cells indicative of high proliferative potential of the lesion. Immunohistochemical staining with anti-p53 antibody showed basal and suprabasal positivity of the lining epithelium suggestive of mutation in the tumor suppressor gene. Based on the histopathologic evaluation, a diagnosis of papilliferous KA was given. The lesion was treated surgically with wide local excision. Postoperative radiograph of the patient shows uneventful healing. The patient has not reported back with any recurrence 2 years after the surgery.

Discussion

Ameloblastomas are a common odontogenic tumor that shows diverse, yet pathognomic histopathological features. However, wide variation has been reported in the past regarding the histopathological presentation of ameloblastoma. Squamous metaplasia has been reported commonly in the acanthomatous variant of ameloblastoma where the central stellate reticulum-like areas are replaced by squamous cells. This keratinization is known to occur in lesions such as odontogenic keratocyst (OKC), acanthomatous ameloblastoma, calcifying odontogenic cyst, squamous odontogenic tumor, and squamous odontogenic carcinoma.[6] Keratin formation has also been reported in a variant of ameloblastoma termed KA which also showed with verrucopapillary projections, which was first described by Pindborg in 1970.[7] Till date, six cases have been reported in English literature with papilliferous histologic components in ameloblastoma [Table 1].

The distinction between acanthomatous ameloblastoma with keratin production and KA is not clear. The WHO in 2005 described keratin with pearl formation in ameloblastoma under the histologic variant of acanthomatous ameloblastoma.[5] In 1992 classification of odontogenic tumors by the WHO, it was defined as ameloblastoma with extensive keratinization.[11] O'Dell and Morgan[12] have described ameloblastoma with keratinization replacing the central stellate reticulum-like areas in ameloblastic follicles as KA. KA has been proposed to be a variant of acanthomatous ameloblastoma by Norval *et al*.[9] with a variable amount of keratin production though the extent and degree of keratinization have not been quantified to distinguish between the two lesions. Whitt *et al*.[13] proposed one criterion to distinguish acanthomatous ameloblastoma from KA that is the presence of keratin in the connective tissue stroma as compared to acanthomatous ameloblastoma which has keratin only in the areas showing squamous metaplasia in the center of odontogenic islands and follicle.

The present case in a male patient in the right mandibular body region is similar in presentation as compared to

Table 1: Reported cases of papilliferous keratoameloblastoma in literature

Case	Authors	Age/gender	Site	Radiographic features	Histopathological features
1	Pindborg[7]	57/female	Right mandibular body and ramus	Multilocular radiolucency	Ameloblastoma consisting partly of keratinizing cyst and partly of tumor islands with papilliferous histology; suggested the term papilliferous ameloblastoma
2	Altini *et al*.[8]	76/female	Right mandible	Multilocular radiolucency	True papillary regions with cores of connective tissue with large rounded cells with centrally placed nucleus. Sheets of cystic odontogenic epithelial follicles filled with necrotic debris and sometimes parakeratin squamous epithelium
3	Norval *et al*.[9]	26/female	Right mandible	Lobulated radiolucency	Cystic follicles containing orthokeratin, parakeratin, desquamated squames, and necrotic material and dystrophic calcifications, papilliferous areas also present
4	Takeda *et al*.[4]	76/male	Left body of mandible	Multilocular radiolucency	Papilliferous type found with hair-like extensions and hard tissue formation
5	Collini *et al*.[10]	62/male	Right ramus and condyle of mandible	Irregular radiolucency with calcification	Papillary projections; containing necrotic debris. Cribriform, solid, and tubular patterns were also present
6	Mohanty *et al*.[3]	46/male	Right body of the mandible	Multilocular radiolucency	Cystic spaces filled with the necrotic debris and lined by papillary keratin lined infolding of odontogenic epithelium resembling ameloblastoma with connective tissue core

previously reported cases except the age of the patient which is comparatively younger (second decade) as compared to mean age of presentation of KA with papilliferous proliferation which occur at a relatively older age (mean age of occurrence in the sixth decade). The right side of the mandible is commonly involved as compared to the left (2:1) with most cases reported in the posterior body-ramus region.[14] Our case differs from the previous cases in showing multiple papilliferous projections with acantholytic cells with necrotic and hemorrhagic material in the lumen and dilated, congested blood vessels. The lesion also showed multiple solid islands of odontogenic epithelium with hyperchromatic nuclei in the connective tissue stroma.

Whitt *et al*. have described four variants of ameloblastoma showing keratin formation, namely, papilliferous histology (odontogenic epithelium is thrown into papillary projections into the cystic spaces); simple histology (epithelial follicles filled with parakeratin or orthokeratin and lined by ameloblast-like cells with reversal of polarity); simple histology with OKC-like features; and complex histology (epithelial follicles filled with parakeratin or orthokeratin, keratin in connective tissue stroma with possibility of hard tissue formation).[13] Corio *et al*. have described keratinizing ameloblastic carcinoma showing the typical histologic features of malignancy in their series of ameloblastic carcinomas, which included pleomorphism, increased nuclear/cytoplasmic ratio, nuclear hyperchromatism, increased number of mitotic figures, abnormal mitotic figures, and necrosis.[15] Our case fulfills the criteria of papilliferous type of KA under the types described by Whitt *et al*.[13]

The present case exhibited positivity for CK 19, an elevated level of mitotic activity, altered p53 profile, and high proliferation index immunohistochemically with Ki-67. It can be inferred that the lesion is comparatively more aggressive locally as compared to conventional ameloblastoma and should be excised extensively to avoid local recurrence. However, it is still unclear how the production of keratin in histopathologic variants of ameloblastoma affects the biological behavior and prognosis of such lesions. The probable reason could be attributed to less number of reported cases with follow-up of such variant to comment definitively on its behavior.

Conclusion

Papilliferous KAs refer to a variant of ameloblastoma that is rarely reported and its biological behavior is poorly understood. The present case shows higher proliferative activity in the basal and suprabasal cells of the tumor providing an indication of its local aggressiveness as compared to the conventional ameloblastomas. At present, such lesions are treated in a manner similar to conventional ameloblastomas though it lacks any authoritative evidence. Report of more such cases with longer follow-up duration and molecular profiling is required in the future to completely understand the spectrum of clinical and histological features, biological behavior, and prognosis of such cases.

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Conflicts of interest

There are no conflicts of interest.

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