Case Report

A Rare and Unusual Variant of Ameloblastoma

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Abstract

Keratoameloblastoma (KA) is a rare histological variant of ameloblastoma usually characterized by extensive production of keratin within the odontogenic epithelium. Pindborg and Weinmann, in 1958, just introduced the term KA. Later in 1992, the WHO categorized it as acanthomatous ameloblastoma with the presence of areas of keratinization. Only a handful number of cases are documented, and it has to be differentiated from other keratin-producing tumors for rendering appropriate treatment. This review aims to discuss a case report of KA which presented as a challenge to the clinicians.

Keywords: *Complex type, keratoameloblastoma, pathogenesis*

Introduction

The term keratoameloblastoma (KA) was first proposed in 1958 by Pindborg and Weinmann. In 1970, Pindborg described an unusual type of ameloblastoma which comprised partly of keratinizing cysts and partly of tumor papilliferous appearance (papilliferous KA). Later, the WHO in 1992 described it as acanthomatous ameloblastoma (AA) with considerable areas of keratinization. KA can be described as a rare histologic subtype characterized by extensive keratin formation present within the odontogenic epithelium. This review aims to discuss a case report of KA which presented as a challenge to the clinicians.

Case Report

A 74-year-old female reported the chief complaint of swelling in her right lower half of the face for the past 3 months. The patient also revealed that the swelling gradually increased and attained the present size. On extraoral examination, a firm swelling on the right side of the body of the mandible extending from the right lip commissure till the angle of the mandible was noticed. No tenderness or sinus opening was evident on palpation. Intraorally, a smooth-surfaced swelling was present in the edentulous region extending from 45 to 47. There was no evidence of pus discharge or sinus opening. No clinical images were obtained due to the lack of patient consent.

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Based on the history and clinical examination, a provisional diagnosis of ossifying fibroma was given. However, it is necessary to include the differential diagnosis such as multilocular cyst (like odontogenic keratocyst [OKC]), ameloblastoma central giant cell granuloma, giant cell lesion of hyperthyroidism, odontogenic myxoma, central hemangioma, metastatic tumors, and rare lesions such as mucoepidermoid carcinoma and calcifying epithelial odontogenic tumor.

Following this, fine-needle aspiration was done as chairside investigation, and the result was negative. Then, the patient was subjected to radiological examination. Orthopantomogram revealed well-defined multilocular radiolucency presented in the right side body of the mandible extending from 44 to 48 region. Anteroposteriorly lesion extended periapically from the distal aspect of 43–48 region. Superoinferiorly lesion occupied the entire body of the mandible, and the inferior border showed mild bony erosion. Apart from this, 43–44 was severely attired without any evidence of dental caries. Further, cone-beam computed tomography (CT) axial overview showed bicortical expansion in the right body of the mandible, and bony erosion was evidently seen in the right inferior border of the mandible [Figures 1 and 2].

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An incisional biopsy was taken, and after routine processing and staining, under scanner view of 4x magnification, the soft-tissue specimen revealed numerous odontogenic epithelial islands with keratin formation arranged in Pacinian-like stacks manner which is interspersed in dense fibrous hypercellular connective tissue stroma was seen. Under higher magnification of ×40, the presence of epithelial islands which were compressed and producing thin extension in straggling manner into the connective tissue was evident. Along with that, few odontogenic epithelial islands exhibited dysplastic features such as hyperchromatism and pleomorphism. Focal areas showed spicules of metastatic new bone formation with active osteoblastic rimming and numerous osteocytes within it which may be due to the inductive effect of tumor lesion. It is suggested that tumor cells cause stimulation of osteoblasts for the formation of new bone, and this process is called osteoplasia. Based on these findings, a final diagnosis of KA with malignant features was given. Subsequently, segmental resection of the right body of the mandible was done, and the specimen was sent for histopathological diagnosis. After routine processing and H and E staining, the histopathological features were in confirmatory with the incisional biopsy diagnosis of KA with malignant features [Figures 3-5].

Discussion

Ameloblastoma is defined as a true neoplasm of enamel organ type tissue which does not undergo differentiation to the point of enamel formation. In the year 1937, Robinson described it as an usually unicentric, nonfunctional, intermittent growth anatomically benign and clinically persistent. The tumor originates from the residual epithelium of the tooth germ, epithelium of odontogenic cysts, and epithelium of the enamel organ.[1,2] The new version of the WHO classification categorizes ameloblastoma into three groups: Conventional, unicystic, and peripheral. The phrase solid/multicystic was dropped because it could be mistaken for unicystic. Desmoplastic ameloblastoma was also categorized as a histological subtype rather than a clinical-pathological entity because it acts like any other ameloblastoma, despite its clinical and radiographic characteristics [Figure 6].[3]

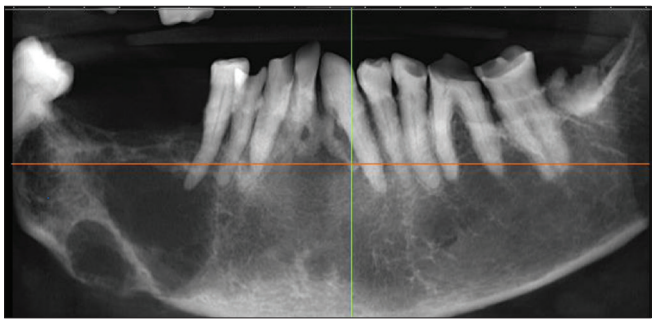

Figure 1: Orthopantomogram revealing multilocular radiolucency

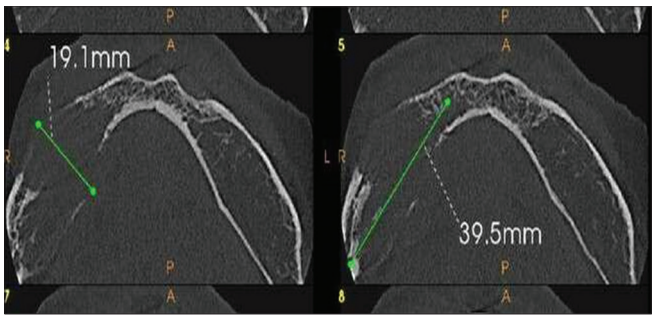

Figure 2: Cone-beam computed tomography showing bicortical expansion in the axial overview

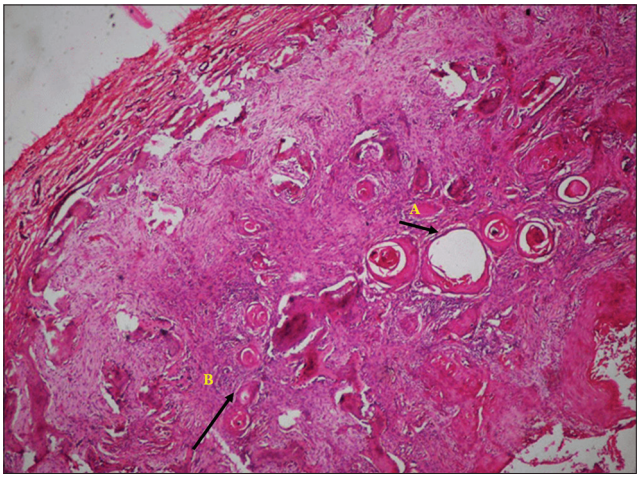

Figure 3: Under scanner view. (A) Keratinising cyst, (B) Numerous epithelial islands with keratin arranged in Pacinian like stacks are seen in fibrocellular connective tissue stroma

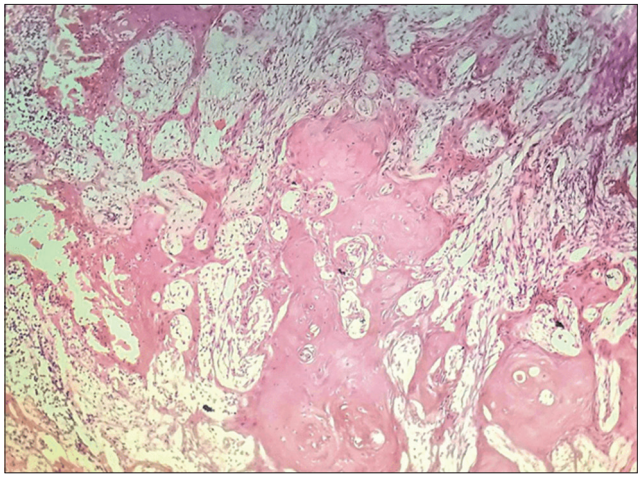

Figure 4: Under higher magnification (×40). Epithelial islands are compressed with straggling extension into the connective tissue, and few epithelial cells in the islands exhibit hyperchromatism, pleomorphism

KA is a rare histological variant of ameloblastoma usually characterized by extensive production of keratin within the odontogenic epithelium. Pindborg and Weinmann, in 1958, just introduced the term KA.[4]

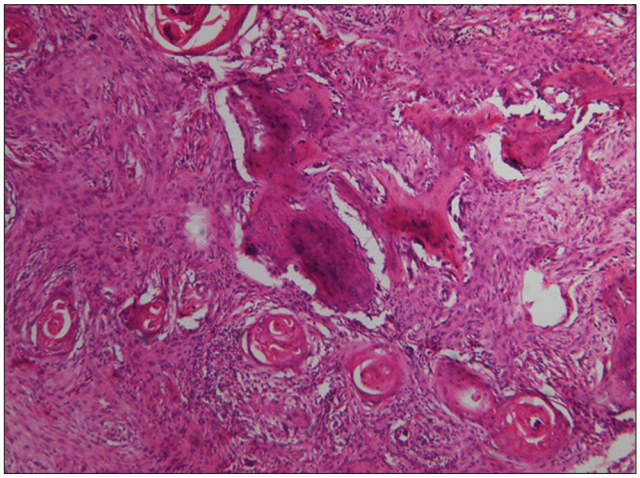

Figure 5: Under higher magnification (×40). Areas of bone formation: (osteoplasia)

Later in 1992, the WHO categorized it as an AA with the presence of areas of keratinization. Clinically, KA shows male predilection in the ratio of 3:1 affecting people of third to seventh decade of life with an average age of 43 years, and it is more common in the posterior mandible. Radiographically, it varies from unilocular to multilocular radiolucency, and sometimes central calcifications appear radiopaque. Few cases have reported mixed radiolucency, and few others exhibiting ground-glass appearance.[5] There is only a handful number of cases reported in the literature totally accounting for 18 cases to date. Based on these cases, it is noted that KA is more prevalent in Asian population and among nine cases four cases have been reported in Indian population [Figure 7].

The pathogenesis revolves like, and normally ameloblastoma can undergo various forms of metaplasia, giving rise to different histological variants such as AA, desmoplastic, granular cell, and basal cell ameloblastoma. The cause of metaplasia here is unknown; however, it is attributed to the multipotentiality of odontogenic

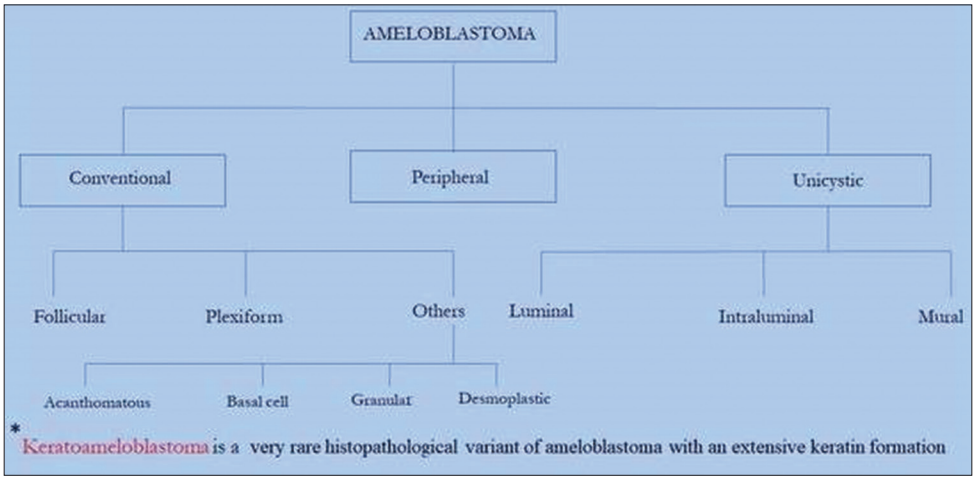

Figure 6: The WHO (2017) Classification of Ameloblastoma

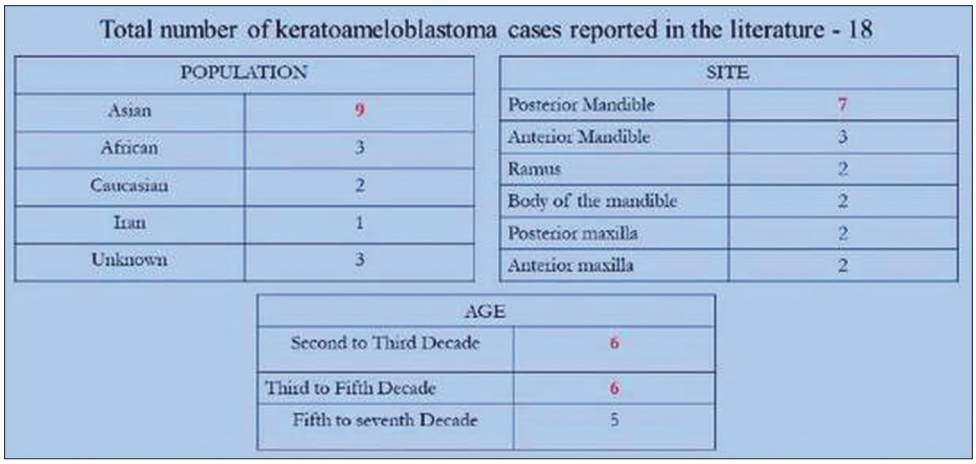

Figure 7: Total number of keratoameloblastoma cases reported to date

epithelium. In KA, the odontogenic epithelial follicles and stellate reticulum cells undergo squamous metaplasia and even progress further to keratin formation. Hence, at a point of time, the epithelial follicles get tightly packed with keratin-like material, and this keratin material gets extruded into the connective tissue stroma leading to the presence of keratin in the connective tissue which is a classic differentiating feature of KA from AA [Figure 8].

In 2007, Joseph C whitt *et al*.[13] described four histological types such as papilliferous, simple, simple with OKC, and complex histology. In papilliferous histology, odontogenic epithelium gets proliferated as papillary projections into the cystic spaces.[6] In simple type, odontogenic islands will be arranged in the form of follicles lined by ameloblast cells showing reversal of polarity, and these follicles are filled with ortho/para keratin. While in simple with OKC type, it shows features of simple histology along with OKC like cystic lining with palisaded basal cell layer giving tombstone or picket fence appearance. In complex type, epithelial follicles are filled with ortho/para keratin, and there may be extrusion of keratin into computed tomography CT stroma arranged in Pacinian-like stacks with or without foreign body reaction. There may be also hard-tissue formation-like cementum or woven bone as an inductive effect of tumor cells. With this, it is clear that our present case belongs to the complex histology of keratoamelobastoma.[7-9]

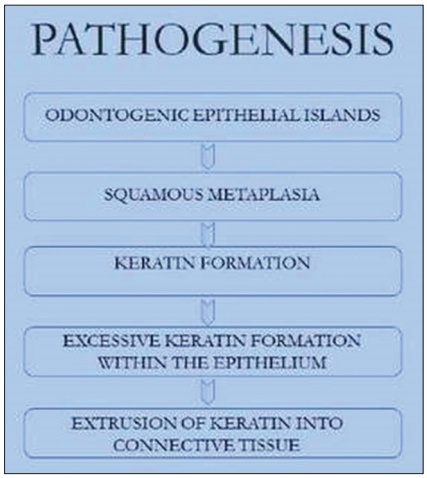

Figure 8: Pathogenesis of keratoameloblastoma

It is also necessary to differentiate KA histopathologically from other keratin-producing neoplasms such as AA, ameloblastic carcinoma, and keratinizing primary intraosseous squamous cell carcinoma (KPISCC) [Figure 9].

To differentiate, in AA characteristically, the odontogenic epithelial follicles exhibit squamous metaplasia and keratin formation present within the follicle. In KPISCC and ameloblastic carcinoma, odontogenic epithelium exhibits dysplastic features such as hyperchromatism, altered nuclear–cytoplasmic ratio, cellular and nuclear pleomorphism, and increased mitotic activity.

Further, Kpiscc reveals a distinct odontogenic pattern with basal-type cells forming alveoli or arranged in a plexiform pattern with palisading of the peripheral cells.[1,10] In ameloblastic carcinoma, the jigsaw puzzle-type nesting of the tumor cells exhibiting malignant features along with that the presence of stellate reticulum, areas of keratinization will be minimally seen. Therefore, by comparing these lesions, KA is more aggressive, and they produce extensive keratinization.

Further, immunohistochemical studies have showed that IHC markers such as cytokeratin 14, 13, 19, anti-ki67,

FEATURES	ACANTHOMATOUS AMELOBLASTOMA	KERATINIZING PRIMARY INTRAOSSEOUS SQUAMOUS CELL CARCINOMA	AMELOBLASTIC CARCINOMA	KERATOAMELOBLASTOMA
EPITHELIUM	Odontogenic epithelium	Dysplastic odontogenic epithelium	Malignant odontogenic epithelium	Odontogenic epithelium
SQUAMOUS METAPLASIA	Present	Rarely present	Present	Rarely present
KERATIN FORMATION	Present within the odontogenic epithelium	Present	Rarely present within the odontogenic epithelium	Lamellar keratin in hair like structures or Pacinian like stacks
STROMAL KERATIN	Absent	Absent	Rarely present	Present
ODONTOGENIC HARD TISSUE FORMATION	Absent	Absent	Absent	Present

Figure 9: Differential diagnosis of keratoameloblastoma

and anti-p53 are positive for KA. Treatment options for KA include segmental resection, hemimandibulectomy. According to the literature, KA has higher recurrence rate of 21% due to high proliferation rate and metastatic nature.[11,12]

Conclusion

KA is a rare aggressive tumor exhibiting features of both ameloblastoma and OKC. Extensive keratinization may mislead diagnosis as primary intraosseous squamous cell carcinoma (particularly when reporting from smaller incisional biopsy specimens). Although only a handful number of cases are documented, it has to be differentiated from other keratin-producing tumors for rendering appropriate treatment.

Declaration of patient consent

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

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