

Unicystic ameloblastoma in conjunction with peripheral ameloblastoma: A unique case report presenting with diverse histological patterns

Sravani Ghattamaneni, Shilpa Nallamala, Venkateswara Rao Guttikonda

Department of Oral Pathology and Microbiology, Mamata Dental College, Khammam, Telangana, India

Abstract

Ameloblastomas are benign epithelial odontogenic neoplasms which are locally aggressive with an insidious growth pattern. Based on the clinical, radiographic, histopathology and behavioral and prognostic aspects, four variants of ameloblastoma are distinguished. They include solid/multicystic ameloblastoma, unicystic ameloblastoma (UA), peripheral ameloblastoma (PA) and desmoplastic ameloblastoma. UA and PA are two variants that have no clinical or radiological resemblance to each other. A case of simultaneous occurrence of these two lesions displaying an array of different histopathological entities together, with a note on pathological insight, has been reported in a 59-year-old adult male patient.

Keywords: Odontogenic gingival epithelial hamartomas, peripheral ameloblastoma, unicystic ameloblastoma

Address for correspondence: Dr. Sravani Ghattamaneni, Department of Oral Pathology and Microbiology, Mamata Dental College, Giriprasad Nagar, Khammam, Telangana, India.

E-mail: drsravani8@gmail.com

Received: 09.05.2017, **Accepted:** 24.05.2017

INTRODUCTION

Ameloblastomas are a group of relatively rare epithelial odontogenic tumors which are benign in nature, locally aggressive with an insidious growth pattern and exhibit diverse clinicoradiographic and histological patterns. Tumors may originate from the rests of dental lamina, enamel organ, lining or walls of nonneoplastic odontogenic cyst and nevertheless also from the basal layer of oral epithelium.^[1]

Unicystic ameloblastoma (UA) is a term that is derived from its macroscopic and microscopic appearance of being presenting as a large monocystic cavity with a lining that is focally ameloblastomatous. Robinson and Martinez in 1977 first introduced the concept of UA.^[2] Despite the term “unicystic”

radiographically, the lesion not only appears unilocular but also multilocular defect in the jaw bones.^[3] Histological UA is subdivided into Subgroup 1: luminal UA; Subgroup 1.2: luminal and intraluminal; Subgroup 1.2.3: luminal, intraluminal and intramural; Subgroup 1.3: luminal and intramural. Posterior mandible is the most commonly affected site, irrespective of the histological variant.^[3]

Peripheral ameloblastoma (PA), also known as extraosseous ameloblastoma, is a rare variant and exhibits the same histological characteristics as solid/multicystic ameloblastoma. Kuru in 1911 first reported PA, but it was Stanely and Krogh in 1959 who first defined the clinical and histopathological characteristics of this lesion.^[4] The lesions are painless, sessile, firm in consistency, pink to

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How to cite this article: Ghattamaneni S, Nallamala S, Guttikonda VR. Unicystic ameloblastoma in conjunction with peripheral ameloblastoma: A unique case report presenting with diverse histological patterns. *J Oral Maxillofac Pathol* 2017;21:267-72.

Access this article online

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10.4103/jomfp.JOMFP_115_15

dark red showing exophytic growth pattern with different surface characteristics ranging from smooth to granular, pebbly or warty in appearance. Size of the lesion may range from 1 to 2 cm. Gingiva of the mandibular premolar region is the most often affected site.^[5,6] Radiographically, no bony destruction is observed except for larger lesions where there is noticeable saucerization of the underlying periosteum.^[5-7]

The present case is unique as there is occurrence of two tumor entities which are different not only in clinical, radiographical and histological facets but also with respect to behavior and prognosis. Presentation of UA with luminal, intraluminal and mural proliferations with coexisting peripheral ameloblastic variant is reported for the first time in the literature in a 59-year-old male patient.

CASE REPORT

A 59-year-old male patient reported with a complaint of gradually enlarging swelling on the right lower half of the face for the past 6 months. The swelling gradually grew and attained the present size. The patient also gave a history of a solitary slow growing soft tissue ovoid swelling, on the lingual side of right back tooth region for the past 10 years. The patient is known hypertensive with a habit of smoking tobacco for the past 30 years. Extraorally, a diffused swelling was noticed on the right lower half of the face giving rise to facial asymmetry. Right submandibular lymph node was tender and soft in consistency. Intraoral examination revealed a swelling on the right side of the body of the mandible extending from 43 to 47, measuring about 5 cm × 4 cm in size, oval in shape with well-defined margins. On palpation, the inspeitory findings were confirmed; the swelling was hard in consistency anteriorly and firm posteriorly and was nontender. Another soft tissue swelling arising in the lingual aspect from the gingiva in relation to the teeth 43, 44, 45 was noticed. On inspection, the swelling was about 2 cm × 2 cm in size, round to oval in shape, erythematous showing pebbly surface and well-defined margins. On palpation, it was sessile, firm in consistency and nontender. There was no discharge noticed from both the swellings. Obliteration of the buccal vestibule was seen from 43 to 48 region. Based on the patient's chief complaint and clinical examination, a provisional diagnosis of ameloblastoma was made and a differential diagnosis of odontogenic keratocyst was proposed.

An orthopantomogram revealed two oval-shaped radiolucent areas separated by a radiopaque septum. Smaller radiolucent area was extending from roots of 43, 44, 45 and larger one in the region of 46, 47 with evidence of

root resorption of respective teeth. Lower border of the mandible was intact [Figure 1].

Computed tomography in axial plane demonstrated an extensive expansile lytic lesion of about 6 cm anteroposteriorly and 4 cm transversely in the region of the body of the mandible. There was an evidence of break in the buccal and lingual cortex posteriorly.

En bloc resection along with involved lymph node was done under general anesthesia, and the specimen was sent for histopathological examination. Grossing of the specimen revealed numerous intraluminal projections into the cystic cavity. Multiple histological sections were prepared from the resected mandible representing all the areas and were thoroughly analyzed.

Microscopically, the tumor obtained from the posterior most region showed a cystic lining composed of ameloblastic epithelium with intraluminal proliferations resembling a plexiform pattern [Figure 2]. Mural extensions of the tumor were also noticed into the underlying connective tissue showing predominantly plexiform pattern in association with focal areas of papillomatous extensions arising with a stalk-like attachment and growing into the connective tissue cores [Figure 3]. Areas of cystic degeneration in the connective tissue were also noticed.

Biopsy from area of the soft tissue swelling revealed the presence of hyperplastic parakeratinised stratified squamous epithelium exhibiting extension of the basal layer of the epithelium into the underlying connective tissue forming ameloblastic islands arranged in the form of trabecular pattern [Figure 4]. Ameloblastic follicles towards the periphery of lesion were also noticed. These follicles appear to be separated from the overlying epithelium by a pseudocapsule formed by connective tissue [Figure 5].



Figure 1: Two oval-shaped radiolucencies extending from 41 to 46 with a radiopaque septum and causing noticeable root resorption of teeth 46, 47

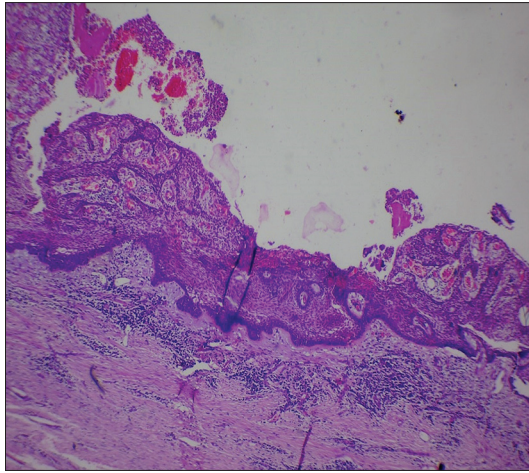


Figure 2: Intraluminal proliferation of cystic lining displaying a plexiform pattern, (x10)

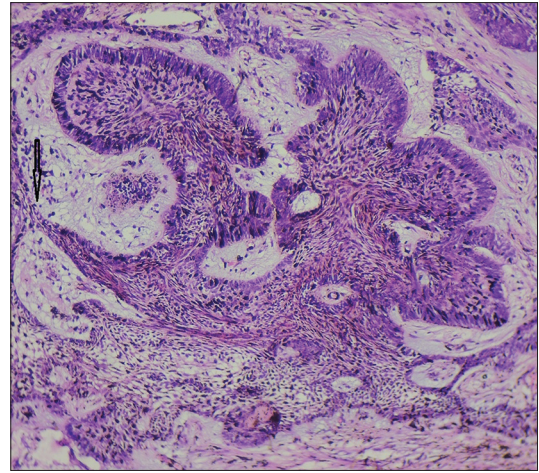


Figure 3: Papillomatous projections growing into connective tissue cores with a stalk-like attachment, (x40)

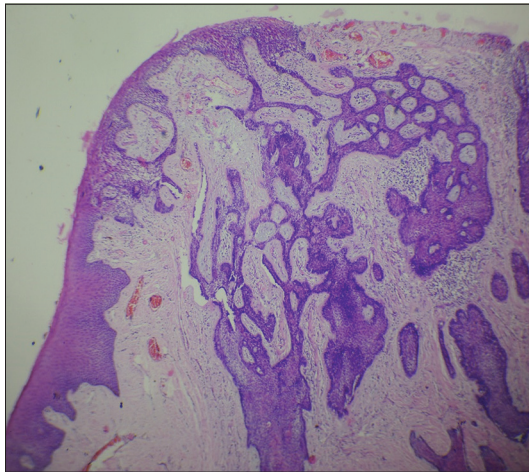


Figure 4: Stratified squamous parakeratinized epithelium showing focal areas of basal cell proliferation forming a trabecular pattern, (x4)

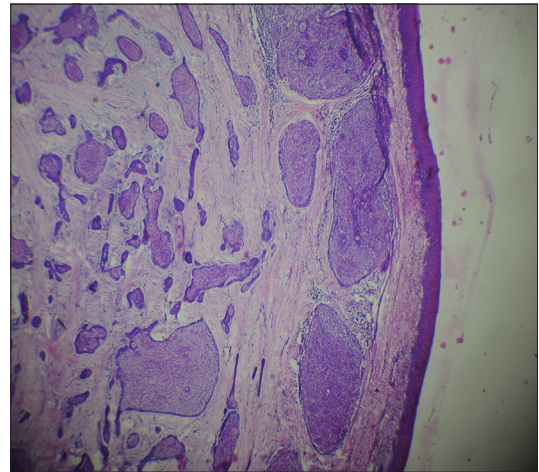


Figure 5: Ameloblastomatous follicles appear to be separated from overlying epithelium by a pseudocapsule, (x4)

Connective tissue stroma showed follicles of ameloblastoma with an admixture of areas of stellate reticulum and polygonal cells of squamous type with prominent intercellular bridges and foci of keratinization indicating acanthomatous change [Figure 6]. Stroma consisted of dense collagenized fibrous tissue toward the periphery, while it was more basophilic and myxoid in deeper sections [Figure 7]. Cystic spaces, engorged blood vessels with red blood cells were also present in the stroma. Biopsy of lymph node showed normal architecture with no abnormality.

Based on the above findings, a diagnosis of plexiform UA of Subtype 1.2.3 showing papilliferous differentiation with concomitant occurrence of an acanthomatous PA was made.

DISCUSSION

UA is a distinctive type of ameloblastoma and accounts for 5%–22% of all ameloblastomas. Based on the profile of

193 cases, Philipsen and Reichart divided the UAs, clinically, into dentigerous variant (associated with unerupted tooth) and nondentigerous variant. Radiographically, it is divided into unilocular or multilocular types. However, a predominance of unilocular configuration is reported. Most of the cases of nondentigerous type occur in 5th–8th decades of life with males being more commonly affected. The present case is to be considered as a multilocular UA of nondentigerous type.

Ackermann *et al.* mentioned three histologic groups based on the clinicopathological study of 57 UAs; they include as follows.^[8]

- Group I: Cyst lined by a variable often nondescript epithelium (luminal UA)
- Group II: Cyst showing intraluminal plexiform proliferation of epithelium; (intraluminal/plexiform UA)
- Group III: Cyst with invasion of epithelium into the

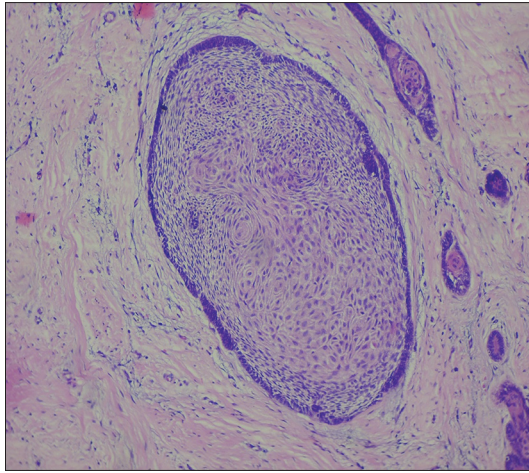


Figure 6: Ameloblastoma follicle displaying acanthomatous changes, (x10)

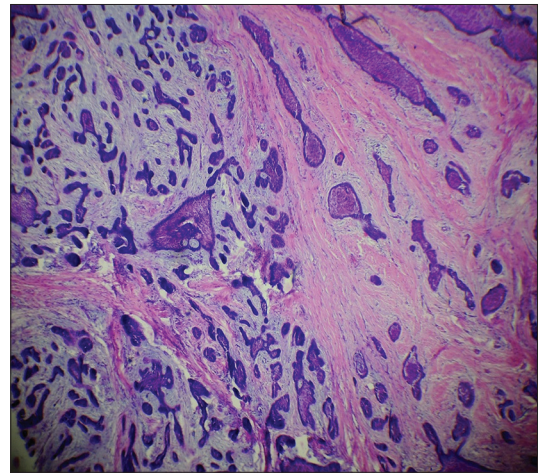


Figure 7: Connective tissue showing myxomatous area in the depth of the lesion, (x10)

cyst wall in either follicular or plexiform patterns. (mural UA).

Ackermann's classification was modified by Philipsen and Reichart^[3] as:

- Subgroup 1: Luminal UA
- Subgroup 1.2: Luminal and intraluminal
- Subgroup 1.2.3: Luminal, intraluminal and intramural
- Subgroup 1.3: Luminal and intramural.

Considering the fact that UA may present with different histological combinations, more aggressive treatment approach is required as the grade progresses.

Gardener in 1981 reported a variant of intraluminal plexiform UA similar to Subgroup 1.2 of Reichart and Philipsen classification. The minimum criteria required for diagnosing UA is focal presence of ameloblastomatous epithelium lining a single macrocystic space which may sometimes mimic a lining of a dentigerous or a radicular cyst.^[9]

The histopathological diagnosis of the present case corresponds to the Subgroup 1.2.3 of Philipsen and Reichart classification. The intraluminal proliferations had a close resemble to the cystic lining of a radicular cyst.

In the present case, the tumor islands in mural tissue partly displayed distinct finger-shaped projections growing into the connective tissue core with a stalk-like attachment. Pindborg in 1970 reported an unusual type of ameloblastoma with islands consisting of keratinizing cysts and papilliferous appearance.^[10] In the present case, there was no evidence of keratinizing cystic epithelium but there was an evidence of papillomatous growth as reported by Pindborg.

Finally, the lesion was considered to be a UA with mural and intraluminal projections showing plexiform pattern predominantly, with focal areas of papillomatous growth.

PA accounts for 2%–10% of all ameloblastomas and represents an exceedingly rare lesion.^[9] Clinically, the lesion resembles epulis (42.6%) and benign tumors (26%) like ossifying fibromas more commonly, less commonly it resembles a pyogenic granuloma or a papilloma. In the present case owing to its clinical appearance, consistency and duration, the lesion was considered to be a fibroma. Biopsy of the lesion confirmed the diagnosis of a PA.

PA is commonly sessile, exophytic growths mostly seen in elder age groups with male predominance. The most common site of occurrence is from the soft tissues of mandibular premolar region.^[4,9] Etiology of PA is unclear; however, two major sources of origin are considered. First is from the cell rests of Serres or remnants of dental lamina and second from the surface epithelium.

Zhu *et al.* did a comprehensive review of 43 Japanese cases and reported that PA are common in 5th–7th decades. In their study, 70% of the lesions occurred in males and the common site is mandibular premolar region. Their study also revealed that most of the cases showed acanthomatous differentiation followed by plexiform, follicular and mixed type.^[11] The present case is in line with the above demographic variables and histologically displayed a mixed pattern consisting of plexiform, follicular and acanthomatous differentiation.

Microscopically, the lesion showed multifocal extensions of surface epithelium that maintained continuity with the ameloblastomatous tissue giving rise to a trabecular or

plexiform pattern. Abutting tissue displayed areas with a band of collagenous tissue separating the follicles from the surface epithelium. This perplexing appearance suggested that the origin of tumor could be from the surface epithelium as well as from the remnants of dental lamina. Zhu *et al.* suggested that oral epithelium has a potential to differentiate into ameloblasts and to form teeth and odontogenic lesions.^[9,11]

El-Hakim and El-Khashab reported an unusual case of PA in conjunction with UA (mural type) in a 13-year-old male child, the lesion appeared to be developing from the lining of a dentigerous cyst related to an impacted mandibular canine.^[12] The present case is also a similar entity, except for the presence of an impacted tooth.

Odontogenic gingival epithelial hamartomas (OGEHs) are peripherally localized hamartomatous lesions initially described by Baden *et al.*^[13] Clinically, they are asymptomatic, small rounded masses on the lingual aspect of alveolar ridges. Origin of OGEH is considered to be from the basal cell layer of the surface epithelium or from the remnants of the dental lamina. The first theory is based on the fact that provoking stimuli, chronic irritating factors and trauma may trigger the growth of basal layer of the epithelium, mainly in older individuals.^[14]

Based on the reviews of Kim *et al.* and Philipsen and Reichart, evidence points toward the origin of OGEHs from cell rests of Serres.^[14,15] Immunohistochemical analysis of a case of OGEH showed positivity for cytokeratin 19 and ameloblastin protein that suggested the origin from dental lamina.^[14]

OGEHs microscopically show a well-defined circumscription by a fibrous pseudocapsule, pseudoglandular arrangements, hydropic degeneration and squamous metaplasia in the myxoid stroma.^[14] Odontogenic epithelial cells appear scattered in a mature fibrous stroma which becomes loose and slightly myxomatous in the depth of the lesion.^[16] Similar features were noted in the present case.

As suggested by Sciubba and Zola,^[17] the diagnosis of PA should be confined to those OGEH lesions that originate from overlying mucosal epithelium with infiltrative characteristics. On account of this fact, the present case was considered as a PA. Nevertheless, the lesion also exhibited histological features consistent with OGEH.

PA, OGEH and peripheral odontogenic fibromas, all belong to the same histomorphological spectrum of a

hamartomatous lesion and the true nature can only be determined if a large number of cases are available for the study.

Treatment for PA and OGEH is conservative as they are harmless and rarely recur.

CONCLUSION

The present case is unique in nature for the reason that there is simultaneous appearance of the two lesions presenting at two different anatomical locations with different clinical, radiological and diverse histopathological presentations. Further, the peripheral variant exhibited features of both PA and OGEH representing a transitional stage between odontogenic tumor and a developmental anomaly. The present case was a collation of histological entities such as plexiform, follicular, papilliferous and acanthomatous types. As astute oral pathologists, we should be able to postulate such rare variants and do one's bit in contributing cases to the literature. At this juncture, the present case is one that must be taken into account.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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