

An unusual presentation of an odontogenic tumor: A diagnostic quandary

Priya Thomas¹, Aneesh Sebastian², Rekha Krishnapillai¹, Sapna Chandran³

¹Department of Oral Pathology and Microbiology, Annoor Dental College and Hospital, Ernakulam, ²Department of Oral and Maxillofacial Surgery, PMS College of Dental Science and Research, Thiruvananthapuram, ³Department of Oral Pathology and Microbiology, Mar Baselios Dental College, Kothamangalam, Kerala, India

Abstract

Squamous odontogenic tumor (SOT) is a rare benign neoplasm and may be located to multiple sites in the oral cavity mouth. As per the literature, there have been <50 reported cases. The tumor is often asymptomatic, although it can present with symptoms of pain and tooth mobility. Peripheral odontogenic tumor is a rare entity derived from either epithelial or mesenchymal portions of the tooth-forming apparatus. Lesions are common to gingiva and alveolar mucosa. Peripheral granular cell ameloblastoma (GCA) is considered to be even rarer. The purpose of the study is to report a case of SOT with a synchronous association with peripheral GCA of cystic nature in the mandible. The occurrence of SOT with ameloblastoma has not been reported as per the literature search.

Keywords: Granular cell ameloblastoma, lysosomes, peripheral ameloblastoma, squamous odontogenic tumor

Address for correspondence: Dr. Priya Thomas, Department of Oral and Maxillofacial Pathology, Annoor Dental College and Hospital, Muvattupuzha, Ernakulam - 686 673, Kerala, India.
E-mail: priyathomask@gmail.com

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INTRODUCTION

Squamous odontogenic tumor (SOT) is a rare intraosseous benign neoplasm which was first described by Pullon *et al.* (1975).^[1] From then, around 50 cases have been reported till date. It is an odontogenic epithelial tumor with locally invasive nature originating from the periodontium. SOT has been identified to occur in three forms: intraosseous, mural (mural SOT-like proliferation in a cyst) and extraosseous form; aggressive and multifocal variants have also been reported.^[2,3] It has also been reported to be associated with calcifying epithelial odontogenic tumor.^[4] The controversy exists in its histopathological diagnosis with acanthomatous and desmoplastic ameloblastoma.

Peripheral ameloblastoma is a rare odontogenic soft tissue tumor, derived from the tooth-forming apparatus. The lesions are localized to alveolar mucosa and gingiva. They affect mainly the middle age groups. Granular cell ameloblastoma (GCA) is a well-recognized entity, but its occurrence within a cystic ameloblastoma is infrequent.^[5] GCA is a rare variant with <5% of the lesions,^[6] and only very few cases of GCA have been reported in the anterior region of the mandible.^[7]

Here, we report a case of SOT arising from the mandible with an associated peripheral cystic odontogenic tumor on the lingual aspect.

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CASE REPORT

A 54-year-old male patient reported with a chief complaint of pain, swelling and mobility of the lower right back tooth for 4 months. On inspection, the lesion was a well-defined oval intraoral swelling of 2 cm × 1 cm on the buccal aspect of 43, 44 and 45 [Figure 1a]. On palpation, the swelling was firm to hard in consistency. 44 exhibited Grade I mobility and found to be nonvital. The patient had an unremarkable medical history.

Coexisting with the intraosseous lesion, an oval shape, soft tissue swelling of size 3 cm × 1 cm was evident on the lingual aspect of the attached gingiva of 41, 42, 43 and 44. No clinical attachment was exhibited by the two lesions [Figure 1b].

Orthopantomogram (OPG) and intraoral periapical radiograph (IOPA) revealed a triangular multilocular radiolucency in relation to 43,44 regions. Associated bone loss and divergence of roots of 43 and 44 were also observed. Root resorption was present for 44 [Figure 1c and d]. Buccal cortical plate expansion was also noticed. A provisional diagnosis of lateral periodontal cyst, odontogenic myxoma and peripheral ameloblastoma was considered.

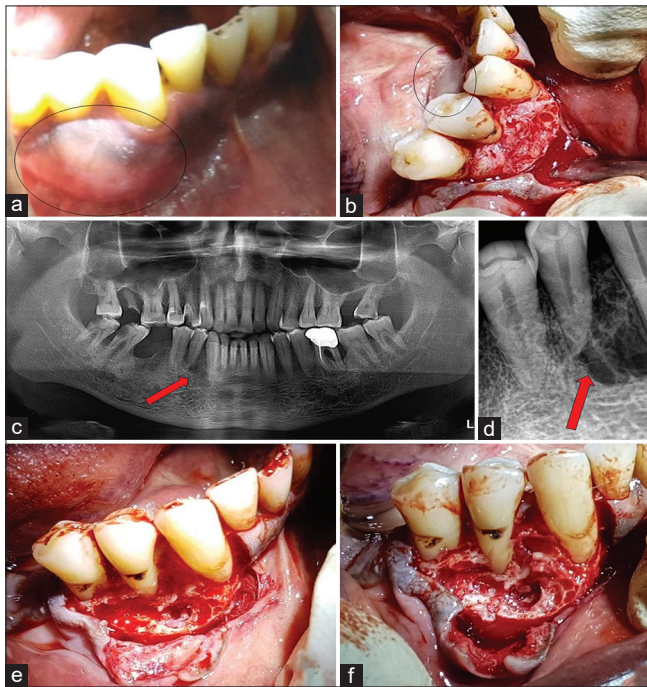


Figure 1: (a) Well-defined oval intraoral swelling on the buccal aspect of 43, 44 and 45; (b) oval shaped, soft tissue swelling on the attached gingiva on the lingual aspect of 41, 42, 43 and 44; (c and d) OPG and IOPA depicting a triangular multilocular radiolucency in relation to 43,44 regions. with divergence of roots 43 and 44; (e and f) Surgical elevation of mucoperiosteal flap with buccal cortical plate perforation and multilocular appearance with irregular bony margins

Intraoperatively, a firm multilocular swelling measuring about 2.5 cm × 3 cm was found in buccal aspect extending from the alveolar process to half way down the lower border of the mandible superiorinferiorly and from the distal side of the right canine region to the mesial side of the right mandibular first premolar tooth anteroposteriorly. Surgical excision was planned under local anesthesia. On elevation of the mucoperiosteal flap, a multilocular appearance with irregular bony margins was noted [Figure 1e and f]. Curettage of the buccal aspect and complete excision of the lingual lesion was done and sent for histopathological examination. Lingual cortical bone was found to be intact.

Histopathological examination revealed numerous variable sized tumor islands of odontogenic epithelium dispersed in a matured connective tissue stroma [Figure 2a]. The islands consisted of peripheral layer of flattened or low cuboidal cells and central squamous cells [Figure 2b and c], along with attempting keratin formation in some foci [Figure 2d]. Cystic degeneration was also observed in some islands.

The lingual lesion showed an ameloblastomatous cystic lining with granular changes in superficial cells associated with a moderately fibrous cystic capsule [Figure 3a-d]. Ameloblastomatous follicles with peripheral tall columnar cells exhibiting reversal of polarity and subnuclear vacuolization with central round to oval-shaped granular cells' changes were observed. The granular cells exhibited coarse eosinophilic granular cytoplasm with peripherally

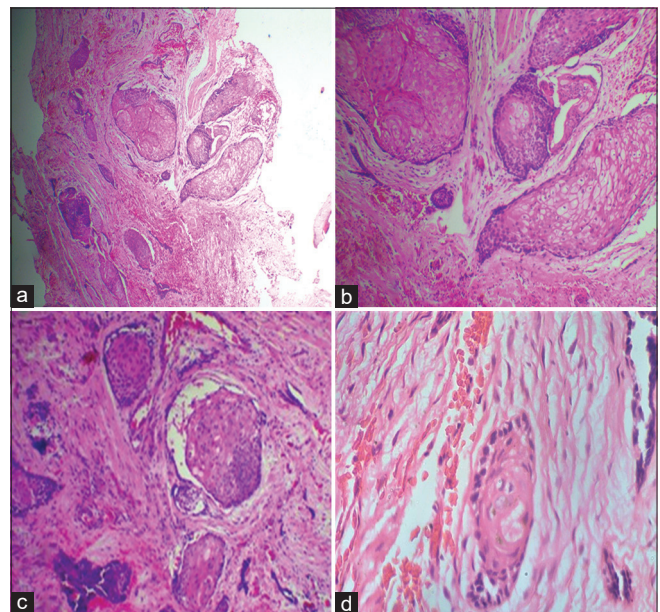


Figure 2: Histopathology of buccal lesion: (a) Tumor islands of odontogenic epithelium dispersed in a matured connective tissue stroma (×4), (b and c) Islands with peripheral flattened or low cuboidal cells and central squamous cells (×10), (d) Tumor island with attempting keratin formation (×10)

arranged pyknotic, hyperchromatic nuclei [Figure 4a and b]. The granular cells were also seen dispersed within the connective stroma [Figure 4c]. The granular cells stained positive for Periodic acid–Schiff staining [Figure 4d].

A histopathological final diagnosis of squamous odontogenic tumor (SOT) was established along with a synchronous association with peripheral cystic GCA. No recurrence on re-examination was observed following a 2 years of follow-up.

DISCUSSION

Intraosseous SOT originates from the cell rests of Malassez, whereas its peripheral counterpart arises from the dental lamina remnants (rests of Serres) or the gingival surface epithelium.^[8]

SOTs present in a wide range of age groups varying from 8 to 74 years (average age 38).^[9] There is no site predilection for location to either the mandible or maxilla. In maxilla, lesions are more prone to anterior regions, whereas mandibular lesions toward the posterior (bicuspid–molar) areas.^[9] Our case occurred in the anterior–posterior between (canine–premolar) mandibular regions. Maxillary lesions are reported to be more aggressive than mandibular ones.^[10]

There is no gender predilection and both remain equally affected. Clinically, the tumor has resemblance to any other pathology and does not stand out with a characteristic feature for diagnosis. The tumor presents as an asymptomatic bony swelling along with tooth mobility, tooth displacement, ulceration of the soft tissue, occasionally with pain and erythema of the lesion.^[11]

Radiographically, the tumor presents as a well-defined triangular/semilunar radiolucency^[11] adjacent to the roots of the teeth and therefore has been contemplated to arise from the rests of Malassez.

Histopathologically, SOT contains multiple, small islands of squamous epithelium with a moderately cuboidal or flattened peripheral basal cell layer in a collagenous fibrous connective tissue stroma. Cystic degeneration of the islands has been reported and some have been shown to contain both prekeratin and laminar calcified masses.^[10,12]

Although SOT has been categorized as a benign neoplasm, it has been known to permeate into adjacent tissues: alveolar bone, overlying gingiva and oral mucosa. Based on this concept, we thought that this lesion could have permeated and formed a cystic lesion on the lingual aspect. However, as per our observation, the two lesions were separate entities both clinically and histopathologically. Therefore, we categorized the occurrence of a synchronous tumor.

Peripheral ameloblastoma, a rare variant of odontogenic tumor, comprises about 1% of all ameloblastomas. Peripheral ameloblastoma presents as a painless, sessile, firm growth of gingiva of size ranging from 0.5 cm to 2.0 cm.

Isomura *et al.*, 2009, reported a case of peripheral ameloblastoma in the buccal mucosa, with histologic findings of a cystic lesion (plexiform type) surrounded by a fibrous capsule with the presence of tumor cells within the capsule. As per their report, their case is the second to be reported with a mural type.^[13] The lingual peripheral

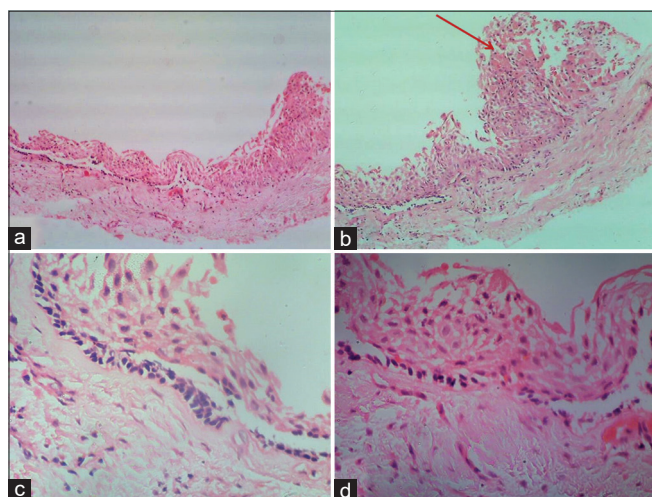


Figure 3: (a) Lingual lesion with cystic epithelial lining and fibrous cystic capsule (×4), (b) Ameloblastomatous cystic lining with granular cell changes in superficially (arrow, ×10), (c and d) Ameloblast-like basal cell layer with loosely arranged superficial polyhedral cells (×40)

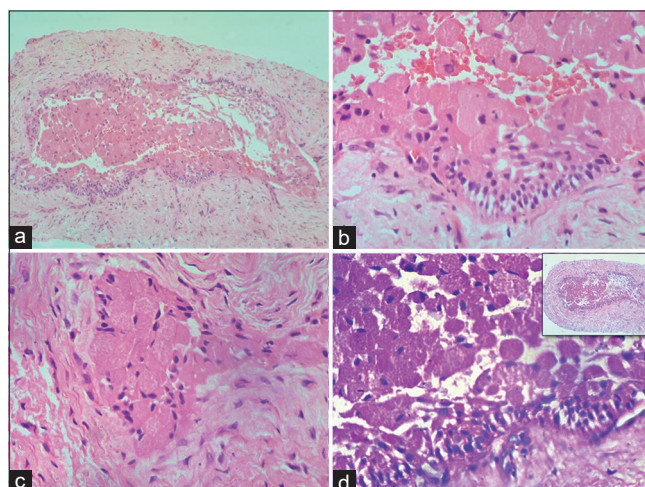


Figure 4: (a) Ameloblastomatous follicles with granular cells changes within the capsule (×10), (b) Peripheral ameloblast-like cells with central granular cells, (c) granular cells dispersed within the connective capsule, (d) periodic acid–Schiff-positive granular cells

gingival lesion in the current case was a cystic variety with tumor islands within the capsule but of a granular cell type.

Factors such as epithelial discohesiveness or the intrinsic production of proteinases (e.g., metalloproteinases and serine proteinases) have been attributed to the reason why certain ameloblastomas become completely cystic.^[14] Our case portrayed a basal layer with ameloblast-like cells following the Vickers–Gorlin criteria such as palisading, hyperchromatic nucleus and reversal of polarity.

The most striking feature of this case was the presence of eosinophilic cells containing granules in the superficial layers along with ameloblastomatous follicles with central granular changes. The granular cell subtype of ameloblastoma is characterized by groups of large cuboidal, columnar or round-shaped granular cells, with pyknotic hyperchromatic nucleus and abundant eosinophilic cytoplasm-filled acidophilic granules that resemble lysosomes, both ultrastructurally and histochemically.^[15]

Granular cell type of unicystic ameloblastoma has been described first as a granular cell odontogenic cyst in 1970 by Gold and Christ.^[16] Abaza in 1989 concluded that such granular cell odontogenic cysts do not exist and they were unicystic GCAs.^[5] Motahary *et al.*^[17] reported a case of unicystic ameloblastoma with granular changes similar to our case.

The granular accumulation within the cytoplasm could be related to lysosomal insufficiency caused by lysosomal overload. This aggregation occurs due to dysfunction of lysosomal enzymes or lysosomal biogenesis.^[18] The reasons for granular change have been debated over time. This change has been attributed to a degenerative process,^[19] metabolic phenomenon,^[16] apoptotic cell death of tumor cells or phagocytosis by neighboring neoplastic cells.^[20]

The recommended treatment for peripheral ameloblastoma differs from the treatment of other forms of ameloblastoma because the tumor is usually small and remains localized to the superficial soft tissue.^[21] GCA is known for its locally aggressive behavior and high recurrence rate.

The occurrence of a peripheral odontogenic tumor in the same region, but progressing anteriorly without a clinical attachment, depicted a remarkable shift from the cases reported. Such a tumor could have occurred due to some kind of inductive or a molecular phenomenon from the major tumor.

CONCLUSION

We designate this case to a synchronous tumor due to the simultaneous occurrence of intrasosseous SOT on the buccal aspect and a peripheral cystic ameloblastoma with granular cell changes on the lingual aspect. Clinically, both the lesions were not interconnected. The clinical, radiographic and histopathological features of the buccal lesion were consistent with SOT. The histopathological features alone rendered a diagnosis of peripheral GCA. As per our knowledge, such a case has not yet been reported in literature.

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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Conflicts of interest

There are no conflicts of interest.

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