A rare combined unicystic ameloblastoma and odontogenic keratocyst found in the same lesion

Brian M Berezowski BDS, MDent (MFOS), FDSRCS, PhD, FFD (SA), FFDRCS (IRE), FCMFOS (SA), FDSRCPS (Glasg), FDSRCS (Edin), PGD (FO) Odont[1]
Vincent M Phillips BDS, MChD, FC Path. (SA) Oral Path, DiP MAX-FAC Radiol, PhD, DSC[2]
Horst Luckey, Dr med dent[3]

Abstract

English: A young male presented with a bad taste in his mouth and mobile right mandibular molar teeth. Intraoral examination revealed that the areas 18 and 28 were both tender to palpation. The tooth 38 was just protruding through the mucosa and pericoronitis was present. The tooth 47 was mobile and the 48 was not clinically visible. There was possible lingual as well as buccal expansion in the posterior part of the right mandible.
The orthopantomograph taken by the dentist before the consultation showed a well demarcated radiolucent area resorbing the distal root of the 47 and involved the unerupted horizontally impacted 48 and impinging on the inferior alveolar canal in that area.
Microscopic examination of the tissue of the molar region 47–48 showed a cystic structure lined with a stratified epithelium which varied in thickness from a unicystic layer to a multiple epithelial layer. The epithelium showed signs of keratin production in one area with palisading of the basal cells.
Adjacent areas of the epithelium showed signs of ameloblastic change within the superficial cells.
Epithelial lined daughter cysts were present in the surrounding fibrous tissues. The histological features showed the presence of an odontogenic keratocyst with daughter cysts. The lining epithelium of the cyst also showed unicystic ameloblastoma morphology.

German: Ein junger Mann stellte sich mit einem schlechten Geschmack in seinem Mund und mit beweglichen rechten Unterkieferbackenzähnen vor. Die intraorale Untersuchung ergab, dass die Gebiete 18 und 28 empfindlich auf Palpation waren. Der 38er ragte gerade durch die Schleimhaut und eine Perikoronitis war vorhanden. Der 47er war beweglich und der 48er klinisch nicht sichtbar. Es gab einen Verdacht auf eine mögliche linguale sowie bukkale Ausdehnung im hinteren Teil des rechten Unterkiefers.
Das vom Zahnarzt vor der Konsultation aufgenommene Orthopantomogramm zeigte einen gut abgegrenzten, radioluzenten Bereich, mit Hinweis auf eine Resorption der distalen Wurzel des 47ers, welche den retinierten, horizontal verlagerten 48er mit einschloss und sich bis zum unteren Alveolarkanal ausdehnte.
Die mikroskopische Untersuchung der Molarenregion des Gewebes 47–48 ergab eine zystische Struktur, die mit einem geschichteten Epithel ausgekleidet war, dessen Dicke von einer unizystischen Schicht bis zu einer multiplen Epithelschicht variierte. Das Epithel zeigte in einem Bereich Anzeichen einer Keratinproduktion mit Palisadierung der Basalzellen.
Benachbarte Bereiche des Epithels zeigten Anzeichen einer ameloblastischen Veränderung innerhalb der oberflächlichen Zellen.
Epithelgeschichtete Tochterzysten waren in den umgebenden fibrösen Geweben vorhanden. Die histologischen Merkmale zeigten das Vorhandensein einer odontogenen Keratozyste mit Tochterzysten. Das Auskleidungsepithel der Zyste zeigte auch eine unizystische Ameloblastom-Morphologie.

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Case report

A 28-year-old male had not been seen by his dentist in six years. He was suffering a bad taste in his mouth and tooth 47 had become slightly mobile. He suffered occasional headaches. His past medical history was good. His general health was normal, he was not allergic to any medicine and had no allergies that he was aware of. Extra-oral examination showed that the head and neck lymphatics were normal, the TM joints functioned well, and the cranial nerves examined were all normal. There was no swelling or asymmetry of his face.

Intra-oral examination revealed that the areas 18 and 28 were both tender to palpation. The tooth 38 was just protruding through the mucosa and pericoronitis was present. The tooth 47 was mobile and the 48 was not clinically visible. There was possible lingual as well as buccal expansion in the posterior part of the right mandible. Tongue function as well as glossopharyngeal and vagal nerve function was normal and the mucosae largely intact.

The orthopantomograph taken by the dentist before the consultation showed a well demarcated radiolucent area resorbing the distal root of the 47 and involving the unerupted horizontally impacted 48 (Fig. 1). The lesion was impinging on the inferior alveolar canal in that area. It also showed a radiolucency around tooth 38 and that teeth 18 and 28 were high and in the maxillary antrum. As the lesion at the right angle of the mandible was extensive and close to the inferior alveolar canal, the patient was warned about possible inferior alveolar and lingual nerve paraesthesia and anaesthesia as well as fracture of the mandible.

From the orthopantomograph taken by the same dentist six years previously, no lesion was evident in the right angle of the mandible and the roots of the 47 looked normal and no radiolucency was visible.

The CT scan of the mandible and maxilla was ordered in all three planes of orientation, namely coronal, axial, and sagittal to confirm the extent of the lesion. Coronal views showed the well-defined radiolucency associated with the impacted 48 stretching almost to the inferior alveolar nerve at the lower border of the mandible (Fig. 2). Another coronal view showed that the 47 had been displaced lingually with an adjacent radiolucency. Axial orientation showed a well-defined radiolucency of the mandible in the area 48 eroding the distal root of the 47 (Fig. 3). Another view in the axial orientation showed a slight bulging of the buccal and lingual cortices. The sagittal view showed a horizontally impacted 48 and root resorption of 47 (Fig. 4). There was a large radiolucency below both teeth.

The report from the radiologist confirmed the findings of a radiolucency around the impacted 48 and reported that it was most likely a dentigerous cyst. The differential diagnosis included ameloblastoma, keratocyst or dentigerous cyst associated with the impacted 48.

Following the radiological examination, it was decided to remove the 48 and 47 as well as the 38 but leave the upper wisdom teeth at the patient's request.

At operation under general anaesthesia with naso-endotracheal intubation, local anaesthetic with adrenaline was given into both the left and right angles of the mandible to minimise haemorrhage. The 38 was removed first and a large area with the tooth displaced into the vertical ramus in which bone resorption was found. This proved to be a paradental cyst on histological examination. On the right-hand side there was severe pericoronitis of the soft tissues associated with 47 and 48 teeth. Both the 47 and 48 were removed. Tooth 47 showed shortened resorbed roots and both the 47 and 48 were mobile. There was bone resorption in the area, but the inferior alveolar nerve was intact. The complete lesion was enucleated from distal to the 48 to the distal aspect of the 46 and the area was thoroughly decorticated. No fluid was aspirated as one would have expected from a dentigerous cyst. The 47 and 48 and the associated soft tissues as well as the lesion were sent for histological examination. The clinical diagnosis was either of a keratocyst or unicystic ameloblastoma. The pathology report confirmed the presence of a paradental cyst in the area 38 and an odontogenic keratocyst

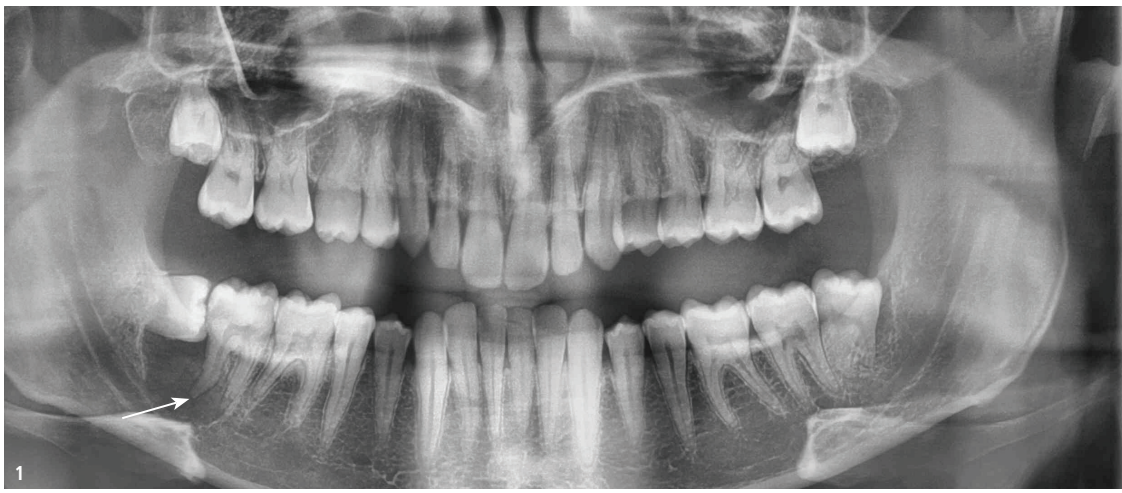
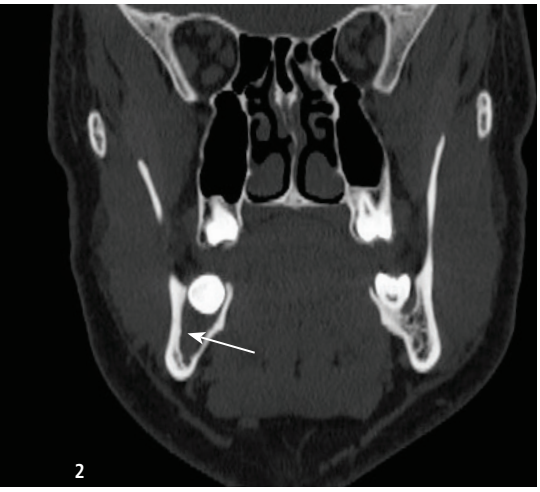
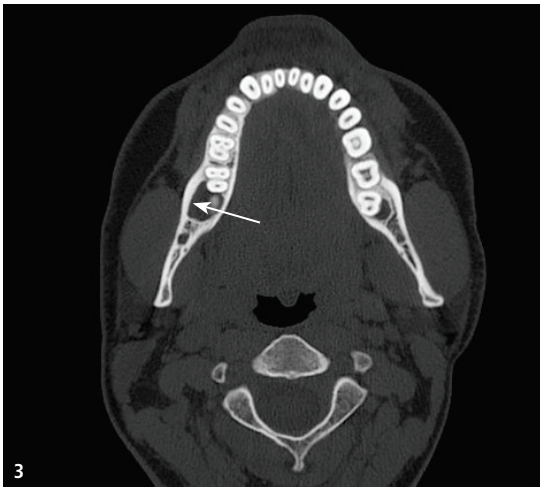
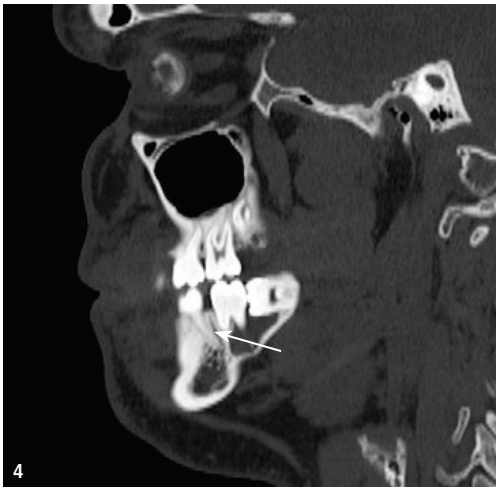


Fig. 1: Preoperative orthopantomograph showing large radiolucency of the 47/48 area and the shortening of the distal root of tooth 47 extending to the distal root of 46. – **Fig. 2:** Preoperative coronal CT scan showing marked radiolucency of right mandible adjacent to tooth 48 (arrow) and displacement of wisdom tooth 38 lingually. – **Fig. 3:** Preoperative axial CT scan showing radiolucency of the right mandible with slight displacement of lingual cortex, highly suggestive of ameloblastoma. – **Fig. 4:** Preoperative sagittal scan showing large radiolucency and horizontally impacted tooth 48 and shortening of the roots of tooth 47 extending to distal tooth 46.







with unicystic ameloblastoma morphology in the epithelial lining of the area 47–48. A week after operation the patient was healing well. It was stressed again that possible recurrences may occur and regular follow-up and checks will be needed. A three-monthly check-up was organised. He was seen by his dentist three months later and an orthopantomograph was taken. This showed the area was starting to show bone formation (Fig. 5). Four months later another orthopantomograph was taken by his dentist, and this showed that there was considerable bone formation in the cavity with no distinct radiolucency or a sign of recurrence at that stage. The patient has unfortunately not returned for assessment and follow-up as instructed.

The microscopic examination of the tissue of the molar region 47–48 showed a cystic structure lined with a stratified epithelium which varied in thickness from a unicystic layer to a multiple epithelial layer. The epithelium showed signs of keratin production in one area with palisading of the basal cells (Fig. 6). Adjacent areas of the epithelium show signs of ameloblastic change within the superficial cells. The epithelium of the cyst showed keratocystic odontogenic tumour-like thickening. There was proliferation of the basal cells forming rete pegs extending into the surrounding fibrous tissue stimulated by the chronic inflammatory cell infiltrate (Fig. 7). The surrounding fibrovascular connective tissue showed epithelial lined "daughter cysts" associated with the keratocystic odontogenic tumour (Fig. 8). The luminal layer of cells of the cyst showed an eosinophilic change in morphology resembling a unicystic ameloblastic appearance (Fig. 9). The picture of the cyst lesion showed signs of an odontogenic keratocyst in one area, but there appeared to be ameloblastic change in areas of the epithelium suggesting a possible keratocystic odontogenic tumour with a unicystic ameloblastoma. It was suggested that the patient be followed up on a regular basis to ascertain any recurrence of the lesion.

Fig. 5: The three month postoperative Orthopantomogram showing some bone filling the radiolucent area at the right angle of the mandible.

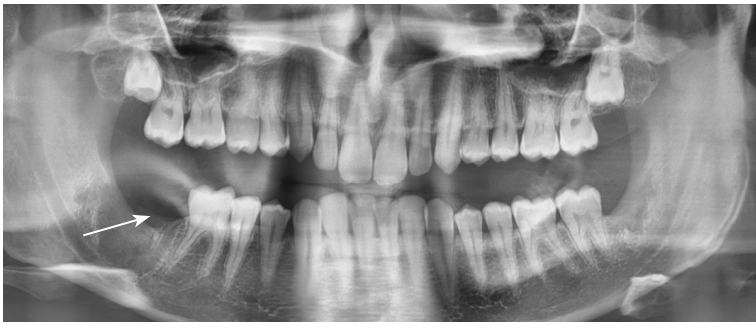
Review of the literature

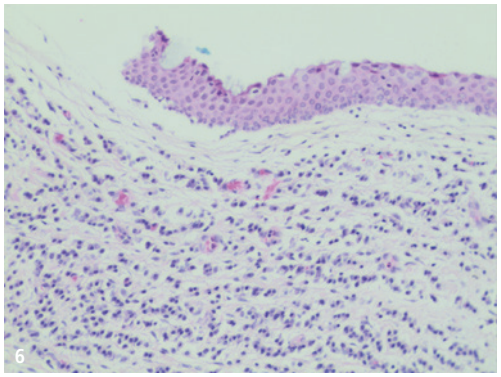
A review of the literature showed that there were very few reports of both an odontogenic keratocyst and unicystic ameloblastoma occurring simultaneously in the same patient.

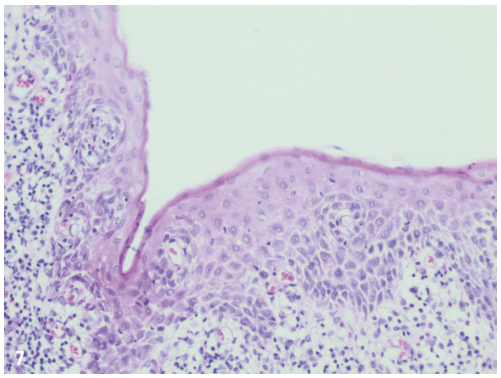
The article by Gupta et al.[1] stated that there were very few reports in the english literature of the simultaneous occurrence of the unicystic ameloblastoma and keratocyst. They advocated, odontogenic keratocyst be treated by marsupialisation, marsupialisation followed by curettage, or excision of the lesion and the placement of Carnoy's solution.

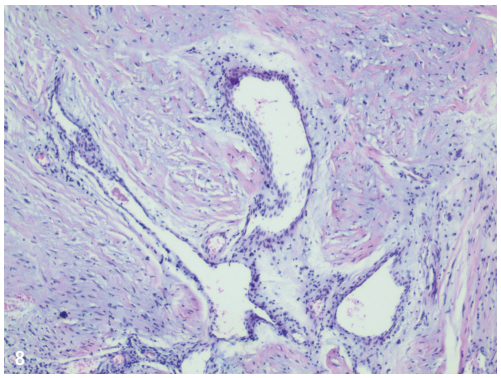
Unicystic ameloblastoma was considered by Ming-Hsuan Hsu et al.[2] They considered it a variant of ameloblastomas and usually occurred in younger patients. It exhibited slow growth with relative local aggressiveness and occurred in the posterior part of the mandible. They showed that computerised tomography demonstrated both buccal and lingual expansion and stressed that long-term follow-up was mandatory.

Vasha et al.[3] investigated the occurrence of some cysts, behaving like tumours and vice versa. They investigated the presence of P63 protein in solid ameloblastomas, unicystic ameloblastomas, and odontogenic keratocyst. They concluded that the higher expression of this gene in odontogenic cysts and tumours correlated with their aggressive behaviour.









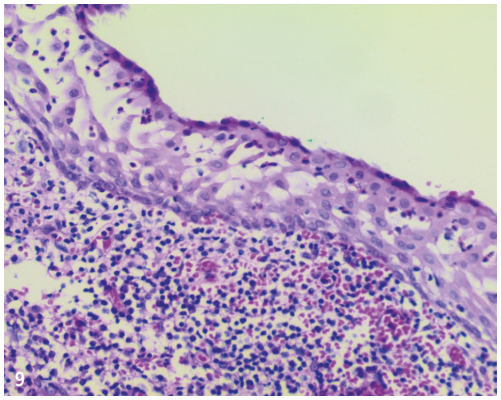


Fig. 6 and 7: The thin stratified epithelium of the typical odontogenic keratocyst with palisading of the basal cells and detachment of the epithelium from the surrounding fibrous tissues (100 x). – **Fig. 8:** Epithelial lined daughter cysts in the surrounding fibrous tissues associated with the lesion in the region 48 (100 x). – **Fig. 9:** The morphological change of the epithelium resembling a unicystic ameloblastoma. The epithelium shows palisading of the basal cells and an eosinophilic layer of luminal cells adjacent to the stratum spinosum layer (arrow; 200 x).

A report of two cases of combined odontogenic tumours was made by Neuman et al.[4] The first pertained to ameloblastoma with odontogenic keratocyst and the second, ameloblastic fibro-odontoma with calcifying odontogenic cyst. They said, for their review, they had not found any lesions of a combination of odontogenic keratocyst and ameloblastoma in the english literature.

A case report by Gamoa et al.[5] confirmed, that it was very uncommon for a combined tumour to occur in one patient. They confirmed the aggressive nature of the keratocystic odontogenic tumour with their potential for local destruction, and a high recurrence rate. They went on to describe the ameloblastoma as being slow growing, benign, locally aggressive, and often involving the mandible, with a high recurrence rate after conservative treatment. They were unaware of any lesion of a mixed ameloblastoma and keratocystic odontogenic tumour having been published. They published a case report and histological results where both these entities were identified simultaneously. They stated that the occurrence of the two distinct, but simultaneous lesions in one patient was a very rare finding and because of the high recurrence rate of each, long-term follow-up was advised.

An attempt to differentiate the odontogenic keratocyst and the ameloblastoma by radiographic means was made by Kitisubkanchana et al.[6] They compared a hundred odontogenic keratocysts and a hundred and one ameloblastomas. Their findings stated that most odontogenic keratocysts showed a smooth border of a unilocular shape whilst ameloblastomas showed a scalloped border and are often multilocular. They did not differentiate between unicystic and multicystic ameloblastomas. They concluded that a unilocular lesion with a smooth border, no resorption of the roots or tooth displacement was suggestive of an odontogenic keratocyst rather than an ameloblastoma.

Ahmadian, Friedman and Reich[7] reported that only one case of ameloblastoma combined with odontogenic keratocyst in a non-syndromic patient had been reported in the english literature. They reported four additional cases and said that CD56 highlighted the ameloblastic areas only whilst the odontogenic keratocyst did not show this. They concluded that the lack of staining in the odontogenic keratocyst only was a help identifying these cases of combined lesions and that the odontogenic epithelium might be the cause of such rare occurrences.

The possibility of ameloblastomas arising from odontogenic keratocyst was reported by Razetto et al.[8] They presented a case report of a lesion with both pathologies. In their patient an odontogenic keratocyst was initially diagnosed. A year later, at the postoperative visit, an ameloblastoma in the same area was found.

Discussion

A unicystic ameloblastoma was first described by Robinson and Martinez in 1977.[13] It was considered a variant of ameloblastomas, but it had a relatively benign biologic behaviour. The unicystic ameloblastoma had been given separate consideration for several decades based on its clinical, radiographic, and pathological features. Although its response to treatment in reports from the 1970s and 1980s suggested that this lesion might behave in a less aggressive fashion, recent reports have disputed this concept.[15] Unicystic ameloblastomas accounted for 10 to 46 % of all intraosseous ameloblastomas in various studies.[9, 11, 12] Whether the unicystic ameloblastoma originated de novo as a neoplasm or whether it was the result of neoplastic transformation of non-neoplastic cyst epithelium has long been debated. Both mechanisms probably occur.[15] Unicystic ameloblastomas were seen most often in younger patients, with about 50 per cent of all such tumours diagnosed during the second decade of life.[9, 15] The average age in one large series was 23 years. More than 90 per cent of unicystic ameloblastomas were found in the man-

dible, usually in the posterior regions. The lesion was often asymptomatic, although large lesions might cause painless swelling of the jaws. In many patients, this lesion typically appeared as a circumscribed radiolucency that surrounded the crown of an unerupted mandibular third molar, clinically resembling a dentigerous cyst.[9, 12, 15] Other tumours simply appear as sharply defined radiolucent areas and were usually considered to be keratocysts, radicular or residual cysts, depending on the relationship of the lesion to teeth in the area. In some instances, the radiolucent area may have scalloped margins, but it remained a unicystic ameloblastoma. The surgical findings might suggest that the lesion is a cyst, and the diagnosis of an ameloblastoma was made only after microscopic examination of the specimen.[15]

Three histopathological variants of unicystic ameloblastoma have been described. In the first type (luminal unicystic ameloblastoma), the tumour was confined to the luminal surface of the cyst. The lesion consisted of a fibrous cyst wall with a lining composed totally or partially of ameloblastomic epithelium. The lining demonstrated a basal layer of columnar or cuboidal cells with hyperchromatic nuclei that showed reverse polarity and basilar cytoplasmic vacuolisation. The upper epithelial cells were loosely cohesive and resembled stellate reticulum. This finding does not seem to be related to inflammatory edema.[15] The luminal cell layer was often eosinophilic.[10, 14] In the second microscopic variant, one or more ameloblastoma nodules project from the cystic lining into the lumen of the cyst. This type is called an intraluminal unicystic ameloblastoma. These nodules may be relatively small or largely fill the cystic lumen. In some cases, the nodule of tumour that projects into the lumen demonstrates an edematous, plexiform pattern that resembles the plexiform pattern seen in conventional ameloblastomas. These lesions are sometimes referred to as plexiform unicystic ameloblastomas. The intraluminal cellular proliferation does not always meet the strict histopathologic criteria for ameloblastoma, and this may be secondary to inflammation that nearly always accompanies this pattern.[15] The third variant, known as mural unicystic ameloblastoma, the fibrous wall of the cyst is infiltrated by the typical follicular or plexiform ameloblastoma. The extent and depth of the ameloblastoma infiltration may vary considerably.[15]

Conclusion

In this case the cystic lesion had an epithelial lining that varied from a thin layer of five to seven cells in thickness with palisading of the basal cells and an eosinophilic superficial layer resembling an odontogenic keratocyst. Epithelial lined daughter cysts were present in the surrounding fibrous tissue. In other areas the epithelial lining showed the changes seen in unicystic ameloblastoma. The epithelium had basal cell palisading with adjacent stellate reticulum change and an eosinophilic luminal layer (Fig. 9). The morphology of the cystic lesion had characteristics of an odontogenic keratocyst with daughter cysts suggesting a keratocystic odontogenic tumour as well as histological features of a unicystic ameloblastoma. The histological features of the odontogenic keratocyst, the keratocystic odontogenic tumour and the unicystic ameloblastoma appear to be a spectrum of morphological variations that are characteristic of this group of lesions. It may be possible that these histological variations are present in individual cystic lesions of odontogenic origin and may be seen if sufficient sampling of the cysts is undertaken.

All three of these lesions are locally destructive within the bone and if daughter cysts are present a more extensive removal should be undertaken to limit recurrence of the lesion. However, the treatment of these cystic lesions need not be a radical procedure and they require removal, peripheral ostectomy and sometimes if indicated the use of Carnoy's solution. Regular follow-up of the patient is imperative with subsequent radiological images of the site of the lesion.

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Conflict of interest

There is no conflict of interest.

Author affiliations

1. *Brian M Berezowski: Consultant Maxillofacial and Oral Surgeon University of the Western Cape and private practice, Cape Town, South Africa. Visiting Consultant, Department of Maxillofacial and Oral Surgery East Lancashire Teaching Hospitals NHS Trust, Lancashire, United Kingdom.*
2. *Vincent M Phillips: Emeritus Professor of Oral Pathology University of the Western Cape, Honorary Professor University of Cape Town and private practise, Cape Town, South Africa.*
3. *Horst Luckey: Private practice and Past President BDO and EFOSS, Neuwied, Germany.*

ORCID numbers for researchers

BM Berezowski:
0000-0003-1687-0433
VM Phillips:
0000-0003-1432-6274

Brian M. Berezowski
[Infos zum Autor]

Literatur

Kontakt

Dr Brian M. Berezowski PhD
8 Mount Pleasant Road, Newlands
Cape Town, 7700
South Africa
berezbc@mweb.co.za