Combined odontogenic tumour-odontogenic keratocyst and ameloblastic fibroma: A case report

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Abstract
Occurence of combined odontogenic tumours is rare. These lesions are also named as hybrid lesions. Histologic features of these lesions are often identical to other odontogenic neoplasms such as ameloblastoma, adenomatoid odontogenic tumor, ameloblastic fibroma (AF), and ameloblastic fibro-odontoma. With variable clinical presentation, ranging from cysts to neoplasms, these lesions show varying degrees of aggressive behaviour. Mostly these combined tumors contain features of one of the odontogenic tumors in combination with either a cyst or tumor. We present a case report of combined odontogenic tumor; an odontogenic keratocyst with an ameloblastic fibroma. Prediction of clinical outcome of such lesions becomes challenging owing to their paucity. It becomes important to understand their salient features so that differentiation from the more common conventional neoplasms distinctly helps to expand classification and provide prognostic criteria.

Keywords: Combined odontogenic tumors, Hybrid odontogenic tumors, Odontogenic keratocyst, Ameloblastic fibroma.

Introduction
Report of Combined odontogenic lesions, or hybrid lesions, within the jaws is rare. Among such reported cases, the most common is the adenomatoid odontogenic tumor (AOT) with calcifying epithelial odontogenic tumor (CEOT).[1]

Case Report
An 18 year old female reported to the department with the chief complaint of pain in the left back region of lower jaw for 2 months. Extraoral examination showed no gross facial asymmetry (Fig. 1). Intraorally the buccal and lingual vestibule in 37 region were seen to be non-obliterated and with intact mucosa (Fig. 2). On palpation the area was tender with slight buccal expansion in respect to 36 37. Adjacent teeth showed no mobility. On aspiration scanty creamy white aspirate was obtained. Based on minimal buccolingual expansion and creamy white aspirate a provisional diagnosis of odontogenic keratocyst (OKC) was made.

OPG showed a unilateral well defined multilocular radiolucency involving mandibular angle and ramus area. The radiolucency measured approximately 6 cm × 3 cm in its largest dimensions and was irregular in shape with well corticated scalloped borders. Anteroposteriorly it extended from apices of 35 36 through the ramus up to sigmoid notch. Also seen was an impacted 38 within the radiolucency (Fig 3).

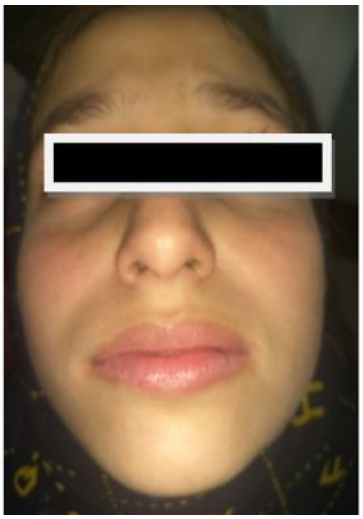

Fig. 1

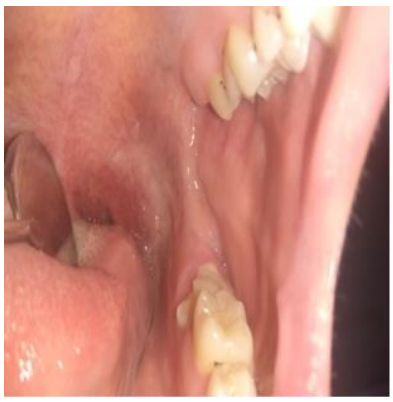

Fig. 2

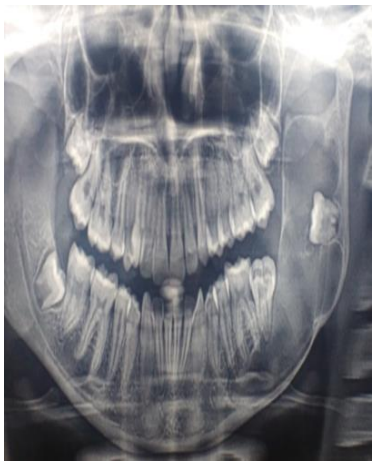

Fig. 3

Ultrasonography showed a hypoechoic lesion measuring 4.8x1.54x2.91cm with heterogenous internal echo, smooth boundary echo, enhanced posterior wall echoes and cystic ultrasound pattern (Fig. 4a &4b). Color Doppler showed the lesion to be avascular (Fig. 4c).

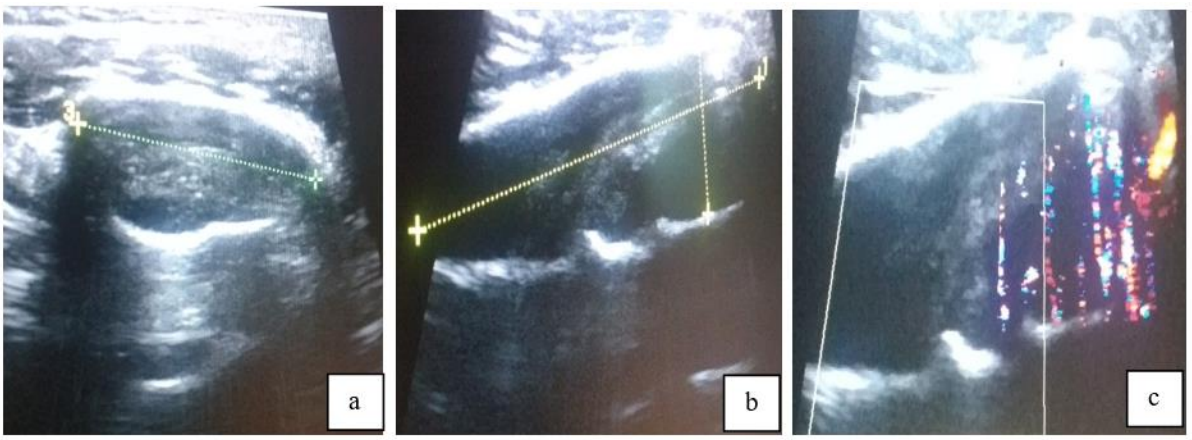

Fig. 4 a,b,c

An incisional biopsy was done and the specimen sent for histopathological examination. Contrary to the provisional diagnosis HPE showed the features of both odontogenic keratocyst (Fig 5a) and ameloblastic fibroma (Fig. 5b) – a compound odontogenic tumor.

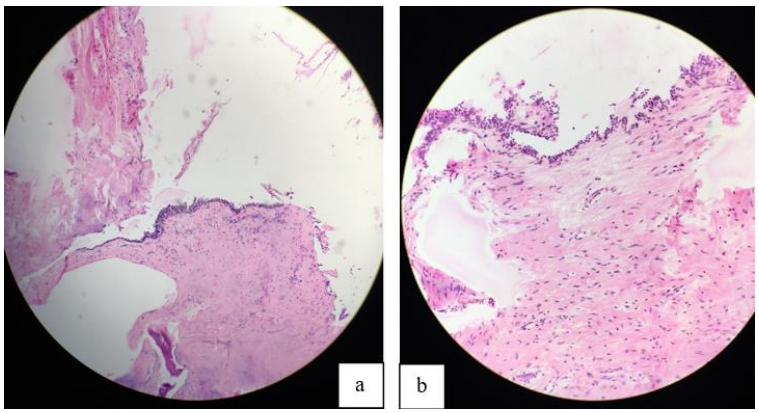

Fig. 5 a,b

Discussion

First described by Philipsen in 1956,[2] the odontogenic keratocyst (OKC) is now designated by the World Health Organization (WHO) as a keratocystic odontogenic tumour (KCOT) and is defined as "a benign uni-or multicystic, intraosseous tumour of odontogenic origin, with a characteristic lining of parakeratinized stratified squamous epithelium and potential for aggressive, infiltrative behaviour." WHO "recommends the term keratocystic odontogenic tumour as it better reflects its neoplastic nature."[3] The origin of KCOT is believed to be from cell rests of the dental lamina.[4-6] With a reported male predominance KCOT has a wide age range, peak incidence being in the second and third decades. The mandible is affected more frequently than the maxilla with 65-83% of KCOTs in the mandible. Both in maxilla and mandible, it has a predilection for the posterior part of the jaw.[4,7-9] A significant number of cases are diagnosed incidentally during routine dental examination. Most commonly presenting signs and symptoms include swelling, pain, and paresthesia. Infected cases present with discharge, abscess, trismus, and cellulitis.[4,9,10]

Radiographically, KCOT presents as a Unilocular or multilocular radiolucency with distinctly corticated, often scalloped, borders. It has minimal buccolingual expansion as the lesion grows anteroposteriorly. The lesion is usually radiolucent with some haziness attributed usually to dense proteinaceous material such as keratin.[11,12-14] In 25-40% of cases, there is an unerupted tooth involved in the lesion.[11,12] Cortical perforation and soft tissue involvement may be assessed by Computed tomography (CT) scans and contrast-enhanced magnetic resonance imaging (MRI).[4] Heterogeneity of growth pattern explains the infiltrative growth in KCOTs in contrast to the expansive growth in other cysts.[15] Histopathology shows thin epithelial lining of six to eight cell layers without rete ridges . This results in epithelial lifting from the fibrous connective tissue and cleft formation, which are considered as artifactual but are characteristic of KCOT.[4] The superficial luminal surface of the epithelium demonstrates wavy parakeratotic epithelial cells; hence, the epithelium is usually described as corrugated parakeratinized epithelium. A prominent palisaded basal layer of hyperchromatic columnar to cuboidal cells are often described as having "picket fence" or "tomb-stone" appearance with keratinaceous material in the cystic cavity.[4,13]

Ameloblastic fibromas (AFs) are a rare variety of benign odontogenic tumors composed of proliferating odontogenic epithelium embedded in a cellular ecto-mesenchymal tissue resembling dental papilla.[16] First described by Kruse in1891and later classified as a separate entity by Thoma and Goldman in 1946[17,18] they are frequently encountered in the posterior mandible with eighty percent cases in the second primary molar or first permanent molar region[19] and 75% associated with an impacted tooth.[20] Primarily considered a tumor of childhood and adolescence, these tumors are frequently diagnosed between the 1st and 2nd decades of life with 75% of cases being diagnosed before the age of 20. Males show a slightly higher prediction than females (M:F= 1.4:1).[21]

AFs usually present with a well-defined unilocular or multilocular radiolucencies.[22] Mostly AFs are encountered as an incidental finding[23,24] reiterating their radiographic significance in the differential diagnosis with entities such as dentigerous cyst, ameloblastoma, odontogenic keratocyst, and ameloblastic fibrosarcoma.[25,26] Microscopically AFs are composed of both the epithelial and connective tissue components; with connective tissue resembling dental papilla. Connective tissue appears myxomatous due to delicate collagen amid spindle and angular cells. The epithelial component is arranged in thin branching cords or small nests with scanty cytoplasm and basophilic nuclei, while stellate reticulum like cells are common in larger nests. Mitoses are not a characteristic feature of ameloblastic fibroma.[27]

The nature of AF still stands enigmatic, as there has been a long debate as to whether ameloblastic fibroma represents a hamartomous growth or is a true benign neoplasm. This controversy further attributes to the difficulties to differentiate between the histology of the neoplastic and the hamartomatous lesions with the histologic features of ameloblastic fibroma.[19]

AFs show high rate of recurrence with more than 45% turning to malignant ameloblastic fibrosarcoma.[28] In addition to detecting the mitotic figures in the histology, immunohistochemical analysis using ki-67, PCNA, and p53 labelling indices would further aid in delineating AFS from AF.[29]

Conclusion

We present two odontogenic lesions with distinct overlapping and combination of histologic features of an odontogenic keratocyst with ameloblastic fibroma. Due to the paucity of cases, the prognosis, clinical behavior and appropriate treatment of such cases is largely unknown. We hope that as more cases are reported in the literature, a better understanding of combined odontogenic lesions will be obtained.

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None.

Conflict of Interest
None.

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