A Rare Case of Glandular Odontogenic Cyst With a Brief Review Of Literature

Glandular odontogenic cyst (GOC) is an extremely rare cyst of jaw bone with only 182 cases documented so far in the English literature. GOC is considered to be a locally aggressive lesion with high rate of recurrence. Anterior mandible is the most common site of involvement for GOC, involvement of the maxillary bone which is a rare occurrence. The clinical and radiographic finding of GOC are varied & often pathognomonic. As clinical & radiographic findings are overlapping with those of other odontogenic cyst, a careful histopathological examination is needed to arrive at a definite diagnosis of GOC. In the present paper we reported a rare case of GOC in a 23yrs old male involving maxillary bone along with a review of literature focussing on clinical & radiological presentation and treatment outcome.

Keywords: Odontogenic Cyst, Glandular Odontogenic Cyst, Jaws.
INTRODUCTION
Glandular odontogenic cyst (GOC) is an extremely rare jaw bone cyst of odontogenic origin which was first described in 1988 by Gardner et al.1 Glandular odontogenic cyst (GOC) was first documented as ‘sialo-odontogenic cyst’ by Padayachee and Van Wyk2 in 1987. After careful analysis, they concluded both GOC and sialo-odontogenic cyst as separate entities.3

In the 1992 World Health Organization (WHO) typing of odontogenic tumors, has categorised to represent one of three possibilities. (I) A true cyst of glandular origin from either entrapped salivary gland primodial or undifferentiated primitive epithelial rests that differentiates into glandular epithelium. (II) An odontogenic primodial origin cyst in which the epithelial lining undergoes prosoplasia (metaplasia from a less specific differentiation to a more specific differentiation) into glandular epithelium. (III) Low-grade mucoepidermoid carcinoma that forms an initial single cystic space instead of the usual multicystic spaces.4

GOC is relatively rare lesion with a frequency rate of 0.012-1.3% of all the jaw cysts and its prevalence rate is 0.17%.5 It has two clinically important attributes: A “high recurrence rate” and an “aggressive growth potential.” The lesion is usually asymptomatic with mandible involved twice as common as maxilla5

The present paper reviews 183 cases of GOCs reported in the English literature focusing on its clinical & Radiological presentation as well as treatment outcome, along with the report of a rare case of maxillary involvement of GOC in a young adult male.

CASE REPORT
A 29-year-old male patient reported to the outpatient Department of Oral Medicine and Radiology with a complaint of swelling on the right side of face since 8-9 months. Swelling was associated with mild discomfort was gradual in onset & progressive in nature. Patient presented with facial asymmetry due to a diffuse extra oral swelling on the right middle one third of face approximately 5×3 cms (fig 1). The swelling was non-tender, firm to hard in consistency & non-compressible. Intraorally a diffuse swelling was noted in maxillary posterior region (3×4 cms) causing obliteration of buccal vestibule. The swelling extended palatally upto the midline was soft to firm in consistency & non-tender (fig 2). There was no history of trauma, pain, paraesthesia or any discharge associated. Right submandibular lymphadenopathy with signs of inflammation was noted. Teeth in the region of swelling (14, 15, 16, 17) were non responsive to pulp vitality test, while teeth irr 11, 12, 13 showed delayed response. Based on case history and clinical findings, a provisional diagnosis of ‘Odontogenic cyst involving the right maxilla’ was given. The differential diagnosis of OKC, Lateral Periodontal cyst, Cystic Ameloblastoma, AOT, Mucoepidermoid cyst, COC and compound odontoma was considered.

The patient further underwent intraoral & extraoral radiographic evaluation along with CT & FNAC.

The intraoral periapical radiograph revealed a well-defined multilocular radiolucency measuring 5×3 cm with scalloped margins (Fig 3a & 3b). Maxillary occlusal radiograph revealed multilocular radiolucent lesion crossing the midline (fig 4a, 4b) along with buccal cortex expansion on right side. Orthopantomogram (OPG) also showed a multilocular radiolucency with corticated & scalloped inferior borders, superiorly with loss of sinus floor (fig 5)

CT was performed which, revealed well-defined unilateral expansile lesion on the right maxilla with hyper-intense borders superiorly compressing the maxillary sinus (Fig 6a). Axial view showed the lesion crossing the mid-line with buccal cortical plate expansion & erosion along with breach in the continuity of right alveolus (Fig 6b). 3D reconstruction images
Fig. 1: Extra oral photograph showing a diffuse swelling in the right middle 1/3\textsuperscript{rd} of face.

Fig 2 (a & b) Intraoral photograph showing diffuse swelling on the right buccal vestibule & palatal crossing the midline
Fig 3. (a & b) IOPA'R showing well-defined multilocular radiolucent lesion with saclloped margins.

Fig 4. (a & b) Occlusal radiograph showing multilocular radiolucent lesion crossing the midline with buccal cortical expansion.
Fig 5. Panoramic radiograph showing multilocular radiolucent lesion extending up to maxillary sinus

Fig 6. (a) Coronal section showing well-defined unilateral expansile lesion compressing the right maxillary sinus. (b) Axial section showing well-defined unilateral expansile lesion crossing the midline. (c) 3D reconstruction showing erosions of right maxilla & medial wall of maxillary sinus
Fig 7. Showing yellow turbid fluid

Fig 8. Intra-operative photograph showing enucleated lesion
revealed erosion of bone in right maxilla & medial wall of maxillary sinus. (fig 6c)

FNAC yielded in a yellow turbid fluid and reports suggestive of an “Cyst contents with secondary infection” (fig 7)

Complete enucleation and curettage were performed under general anaesthesia. Cystic lining was removed and closure was done (fig 8a). Specimen was sent for histopathological examinations where the epithelial lining showed non-keratinised glandular epithetlum with cuboidal cells, bulbous nucleus, few mucous cells, connective tissue comprised of thick bundles of collagen fibres with focal areas of inflammatory cell-infiltrate, all these features are suggestive of “Glandular Odontogenic Cyst”.

Post-operative the patient was followed up & recurrence was noted after 7 months.

DISCUSSION

The GOC is an extremely rare jaw bone cyst ranging from 0.12% to 0.13% of all jaw cyst and its prevalence is 0.17%. Literature shows that GOC may mimic a wide clinicopathological spectrum which ranges from lateral periodontal cyst to destructive malignant Lesions such as mucoepidermoid carcinoma. A literature review was performed by using the search terms “Glandular Odontogenic Cysts”, “Sialo-Odontogenic Cyst” on the Pub Med journals. Further literature was searched via Pub med, Springer online, Wiley online and by searching the World Wide Web. The search included abstracts and online publications from year 1987 to 2017 along with also included the present case. Till date 183 cases of GOC have been reported in the literature among which 72% cases were involving mandible with maxillary bone was involved only in 28% of cases. Anterior jaw bone (119) was twice more commonly involved than posterior (57). The mean age at diagnosis for all cases (N = 183) was 45.9 years. The age range was between 11 and 86 years. Majority of cases were diagnosed in 4th 5th & 6th decades. In the present case the patient presented to us in the 2nd decade. Mohammad Faisal et al., reported a case of highly aggressive GOC in a paediatric patient of 11 years, extensively involving the anterior and posterior mandible. Keeping in mind the clinical history the reported cases of GOC’s, this seems to be the only reported case of a GOC that probably developed in the first decade of life. The lesion was discretely more prevalent in men than women (2:1). The main clinical finding in this cyst is a painless local swelling, however in the present case pain was present, which may be due to compression of Neurovascular bundle or secondary infection.

Of the 183 cases reported 55.6% cases showed unilocular radiolucencies while 44.4% cases presented as multilocular. Expansion of the cortical bone (76.8%) was seen as a predominant feature followed by breach or perforation (56.6%) which was also noted in our case. Most of the GOC were reported to be treated by conservative modalities such as enucleation, curettage, peripheral ostectomy, chemical cauterization with crayons solution, cryotherapy, etc for 158 cases, with as many as 20 cases treated by marginal resection or partial jaw resection. Mean period for follow-up was estimated to be 0.25 – 8.75 years, with a recurrence reported in 27.8% cases.

Due to overlapping clinical and radiographic features of GOC with other odontogenic cysts. A careful histopathological examination is needed to arrive at a definite diagnosis of GOC. Kaplan et al have proposed major and minor histopathological criteria for GOC. The following major and minor criteria must be present for diagnosis:

The major criteria include:

1. Squamous epithelial lining, with a flat interface with the connective tissue wall, lacking basal palisading.

2. Epithelium exhibiting variations in thickness along the cystic lining with or without epithelial “spheres” or “whorls” or focal luminal proliferation.
3. Cuboidal eosinophilic cells or “hob-nail” cells.
4. Mucous cells with intraepithelial mucous pools, with or without crypts lined by mucous-producing cells.
5. Intraepithelial glandular, microcystic or duct-like structures.

The minor criteria include:
1. Papillary proliferation of the lining epithelium.
2. Ciliated cells.
3. Multicystic or multiluminal architecture.
4. Clear or vacuolated cells in the basal or spinous layers.

Literature review revealed that majority of GOC cases showed more than 4 to 5 histological characteristics. Several authors have reported GOC to have aggressive growth potential & a high rate of recurrence (35.7%) which could be attributed to the cell kinetics in the epithelial lining, multicellular nature of the lesion, thin epithelial lining that may be difficult to separate from underlying connective tissue capsule making it more difficult to remove during surgery. Another factor could be a more conservative approach carried out in the management of GOC. Recurrence was noted in the present case after 7 months.

Treatment of choice of GOC is still controversial & ranges from curettage, enucleation, marsupialisation, en block dissection, excision to partial osteotomy. The present case underwent enucleation due to its clinical & pathological behaviour, with follow up that was uneventful, however recurrence was noted after 7 months & the patient was advised for partial maxillectomy. Thereafter the patient was lost further followup.

With the present data, it is suggestive that a more aggressive approach in treating GOC and a careful long-term follow-up would be required.

CONCLUSION
This case report of GOC will add to the existing knowledge of these rare odontogenic cysts. Our case correlates with the existing literature that GOC affects men more commonly than women, but differ in the site involvement where in the present case maxilla was involved, which is a rarest entity. Definite diagnosis of this cyst is possible only by histopathologic evaluation as the clinical and radiological findings are overlapping with other odontogenic cysts. Aggressive behaviour and the tendency for recurrence have been reported in the literature & so were also observed in the current case. Hence long term follow up is advocated.

REFERENCES