Journal of Advanced Medical and Dental Sciences Research

@Society of Scientific Research and Studies *NLM ID: 101716117*

Journal home page: www.jamdsr.com doi: 10.21276/jamdsr Indian Citation Index (ICI) Index Copernicus value = 100

(e) ISSN Online: 2321-9599; (p) ISSN Print: 2348-6805

Case Report

Calcifying odontogenic cyst of the maxilla – Case report and literature review

Yusra Fatima[1], Daniya Naved[2], Banpreet Kaur[3], Nagaraju Kamarthi[4]

[1,2,3]Intern, Department of Oral Medicine and Radiology, Swami Vivekanand Subharti University, Meerut, India; [4]Professor and Head, Department of Oral Medicine and Radiology, Subharti dental College, Meerut, India

ABSTRACT:

Developmental cysts of dental origin are pathological fluid-filled cavities that may or may not present with an epithelial encasing. Unless secondarily infected, they present no evidence of pus formation or collection. Calcifying epithelial odontogenic cyst (CEOC), now formally revised by the World Health Organisation (WHO) as calcifying odontogenic cyst (COC), has complex histopathological features with diverse morphology that behaves aggressively like a tumor. Being racial and gender-neutral, it is rarely encountered, with peak incidence being in the second or third decade of life. CEOC can be seen as a radiolucent lesion with well-defined or poorly-defined edges and may be associated with unerupted teeth. Calcification is crucial to CEOC radiography interpretation. CEOC is characterised by a fibrous odontogenic epithelial wall and lining, with basal cells (cuboidal or columnar)reminiscent of ameloblasts. Conservative surgical enucleation is the preferred treatment for CEOC; however, recurrence is not uncommon. This article presents a case of a 34-year-old North Indian male patient who reported the lesion in the maxillary anterior region in relation to the right incisors and premolar teeth. The condition was resolved by surgical extirpation, and the patient was put on a standard follow-up procedure for five years.

Key words: odontogenic cysts, odontogenic tumor, ameloblastoma, epithelial tumors, radioopaque lesions.

Received: 13 June, 2025 Accepted: 28 June, 2025 Published: 02 July, 2025

Corresponding author: Yusra Fatima, Intern, Department of Oral Medicine and Radiology, Swami Vivekanand Subharti University, Meerut, India

This article may be cited as: Fatima Y, Naved D, Kaur B, Kamarthi N. Calcifying odontogenic cyst of the maxilla – Case report and literature review J Adv Med Dent Scie Res 2025; 13(7):30-35.

INTRODUCTION

The calcifying epithelial odontogenic cyst (CEOC) [synonym: dentinogenic ghost cell tumour, calcifying odontogenic cyst/tumour (ghost cell)], first reported in 1962 (Gorlin cyst),[1] is a contentious lesion arising from odontogenic epithelium with both cystic and solid neoplastic features. The diverse range of clinical behaviors and histological characteristics of benign cysts of odontogenic origin has resulted in many nomenclatures and classification schemes. It wasn't until 1932 that CEOC made its literary debut in Germany.[2] Originally identified as a nonneoplastic cystic lesion in 1971, it was reclassified as an odontogenic tumour in 2005 by the World Health Organization (WHO).[2, 3] In year 2017, a new term calcifying odontogenic cyst (COC) was implemented by World Health Organization (WHO) replacing previous designation and exclusively referring the lesion as a benign cystic type of odontogenic tumor, without neoplastic characteristics or behaviour.[4,5] The nature of the ghost cell lesions remains a topic of debate. Despite the lesion representing approximately only two percent of all odontogenic jaw pathologies and its rare occurrence,[6] CEOC is recognized as individual pathology whose differentiation from similar pathology is must. It is a slow-growing cystic lesion that affects the mandible and maxilla equally, and it is painless from a clinical standpoint. It typically develops intra-osseously and affects the front of the jaws. With a mean age of 30.3 years and no preference for gender, the peak occurrence happens in the second and third decades of life.[7]The radiographic appearance of CEOC is generally seen as a radiolucent lesion with well-defined or poorly defined edges and may be associated with unerupted teeth.[8] Calcification is a crucial radiographic characteristic for CEOC interpretation, observed in almost half of reported cases.[9]Fibrous odontogenic

epithelium including ameloblast-like columnar or cuboidal basal cells is the hallmark of CEOC on microscopy. Ghost cells, which are frequently calcified, are seen in the cystic lining, and cells that resemble a stellar reticulum cover the basal cell layer. It is also possible to see melanin deposits in the epithelial lining.[10] In cases of CEOC, conservative surgical enucleation is the treatment of choice. When cystic linings undergo malignant degenerations, recurrences are possible, though rare.[9] We present a clinical case report of an adult male patient with a unique and rare but classical calcification within the cyst.

CLINICAL CASE PRESENTATION

A 34-year-old male patient presented to the outpatient department of Oral Medicine and Diagnosis at one of the Dental College in North India, complaining of swelling and discomfort around his upper right front tooth, which he had been experiencing for the last two months. Associated with the chief complaint was the patient's concern about its steady increase in size from an original lesser bulge to its current expanded stage. The personal history of the patient did not reveal any finding that could be significantly associated with the condition. A three-year past history of trauma in the same region was the only important association. Medical, drug, and dental histories were all within the normal limits of acceptance, with the patient following a regular habit of daily toothbrush and tooth paste use. The extraoral examination demonstrated a slight right-sided asymmetrical face, which was obscured to some extent by the patient's appearance, which included a beard and moustache **(Figure 1 A, B)**. The obliteration of the nasolabial groove on the affected side was visible more towards its approach to the external nostrils (Figure 1 A). Physical and biological extraoral parameters were within typical limits that included the condition of the temporomandibular joint and the border movements of the mandible. The right nostril and alae of the nose were slightly elevated when compared to the left side (Figure 1 A). Intraoral examination disclosed a solitary wide enlargement in the labial vestibule (right side) extending from tooth number 11 to 14 and covering a length of approximately 2 centimeters in length (Figure 1 C). Vertically, the enlargement stretched from vestibular depth to the mucogingival junction (Figure 1 D). The anatomical extension of the swelling started on the labial frenum, which extended to the distal extension of the maxillary right sided first premolar (Figure 1 C). Ipsilateral anterior teeth seemed to have undergone major shifting in the form of mesial and distal rotations with change in axial inclination (Figure 1 C). The swelling when palpated was hard in consistency, non-tender, and did not yield to pressure. Intraorally, the mucosa over the swelling was smooth with a reddish pink hue, easily differentiated from the adjacent paler mucosa. There was no response in the form of sensitivity of lesion-associated teeth on percussion, although the teeth seemed to have drifted. Radiographic investigations included conventional and a cone beam computed tomographic investigation (CBCT), which revealed the location on the right maxilla with a well-defined, non-corticated lesion (Figure 2 A, B, C). Radiographic dimensions disclosed the lesion to be 13.45 mm by 16.37 mm by 25.20 mm at the medial, anterior-posterior, and superior inferior sides, respectively. The overall lesion presented a very well-defined radiolucent mass that had a conglomerate radiopaque mass within its confines (Figure 2 A, C). The perforation of the labial cortical plate was evident on the CBCT (Figure 2 A, C). Overall, the radiographic picture mimicked that of an infected dentigerous cyst with either a mesiodens or odontoma, which was clearly differentiated through history. Because of the expanding direction of the cyst in the anterior direction, there was no evidence of root resorption at the apices or the axial surfaces. Fine needle aspiration yielded approximately 2 milliliters of straw-colored fluid, confirming the cystic lesion. Other differential diagnoses considered were ameloblastoma (unicystic), ameloblastic fibroma, odontome, lateral periodontal cyst, mucocutaneous keratoacanthoma, space infection, bony exostoses, odontogenic tumour (adenomatoid), and CEOC.[11-15]

The patient was informed and educated about the condition and various other possibilities associated with it. The patient was informed that surgical enucleation of the cyst could be considered as a treatment option pending histological confirmation. After obtaining written informed consent, the patient was taken to the oral surgery department to have surgical enucleation done. Following the post-pandemic infection control guidelines and strict aseptic precautions,[16,17] the surgery was performed by giving a crevicular incision under local anesthesia in the maxillary right anterior region. The lesion was exposed and enucleated by careful dissection. Haematoxylin and eosin staining revealed fibrous connective tissue reinforcing the cystic epithelial lining, which was revealed by histological investigation of the cyst lining (Figure 2D). Isolated patches of reverse-polarity cells that looked like ameloblast-like cells and tall columnar cells arranged in a palisading pattern were present in the basal cell layer of the odontogenic epithelium. In the layer above the epithelium, there were cells that looked like stellate reticulum and ghost cells, as well as various quantities of eosinophilic structures. A fibrous connective tissue stroma, hyalinized beneath the epithelium and containing several bundles of collagen fibers, lay atop a thin layer of odontogenic epithelial islands (Figure 2D). Areas of an eosinophilic and basophilic material that were also seen suggested dysplastic dentin (dentinoid). The connective tissue capsules also showed a mild lymphocytic infiltrate, along with a small number of bony trabeculae. Features were suggestive of a calcifying epithelial

odontogenic cyst with an odontome. Patient was instructed regarding wound care and advised to use a post-surgical mouthwash to help keeping the oral cavity bacteria free.[18]The patient experienced uneventful postoperative healing and received advice for periodic follow-up. The long-term follow-up was scheduled for a minimum period of 5 years, during which the patient had to report every six months, at which time a radiographic verification would be conducted for recurrence of the lesion.

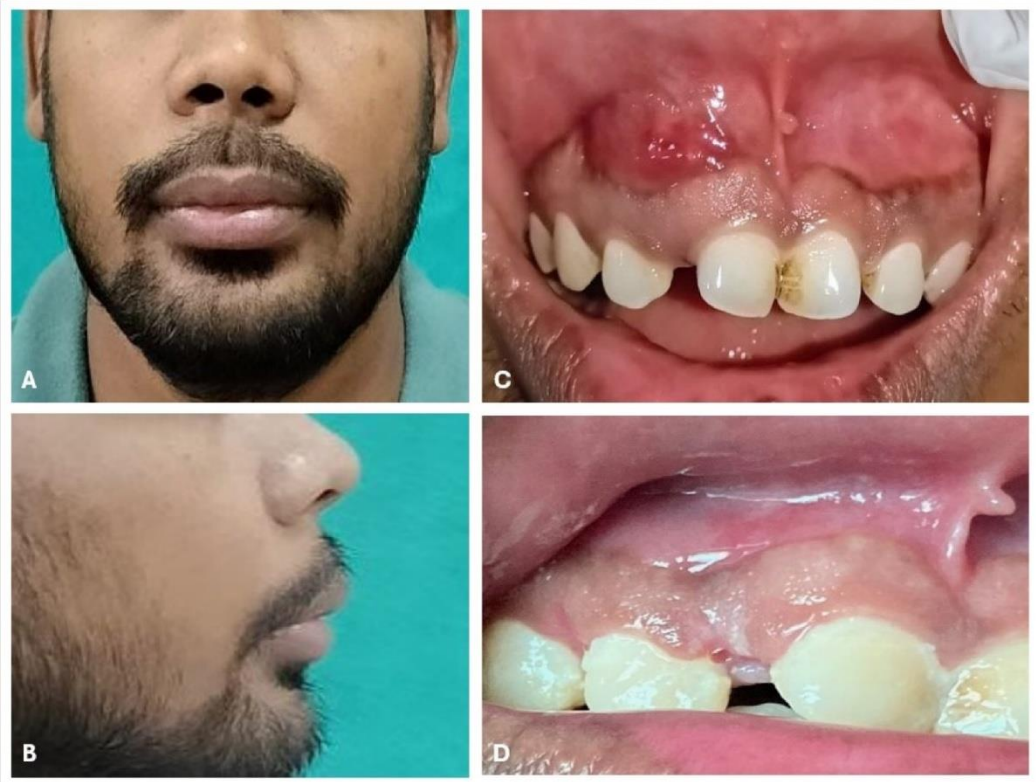

Figure 1: (A) Frontal view of the patient showing right side facial asymmetry with obliteration of nasio labial sulcus and shift in the inter alae axis (B) Lateral view showing normal facial profile with swelling on right side obliterating demarcation landmarks (C) Intra oral view showing bright red lesion in the labial vestibule extending from distal of maxillary right central incisor to the maxillary first premolar region (D) Intra oral view showing the growth upto the junction between attached gingiva and vestibular mucosa.

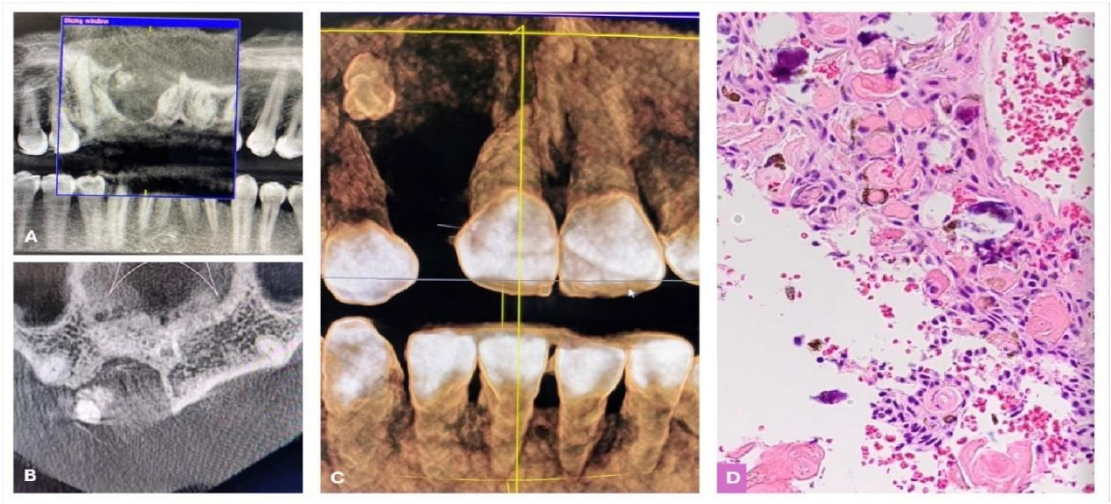

Figure 2: (A) Cone beam computed tomographic view showing the involved region highlighted within the window with a radiopaque conglomerate mass towards the periphery of the lesion (B) Transverse view showing the relation of the mass with the lesion and the adjacent structures (C) CBCT showing the resorption of the bone surrounding the adjacent teeth (D) Histopathological photograph showing relevant features

DISCUSSION

The formal classification of CEOC, now better represented as COC, is based on two hypothetical theories: monistic and dualistic. The "monistic" theory considers CEOCs to be neoplastic, even though most lesions seem benign and cystic. The "dualistic" theory suggests that the lesion exists in two different forms—a cystic and neoplastic form. Even though most academics today suggest the dualistic hypothesis, the World Health Organisation (1992) favoured the monistic theory and viewed CEOC as an odontogenic tumor.[4] For avoiding confusion, we address the condition in this paper as CEOC. The rarity of such cases being reported (CEOC represents about 1% of jaw cysts), makes this clinical report unique. It has been described to occur in children also,[19] with greater incidence being reported in the second decade,[20] and a bimodal distribution of the second peak of incidence in later decades of life [6th and 7th]. We report a case in the third decade of life of an Indian male, which is in agreement with its negligible gender or racial predilection. The lesion in our case was located in the anterior maxillary region, which further substantiates its general specific predilection for the anterior jaw region between the incisor and canine region in either the maxilla or mandible.[21,22] The lesion we report is an intraosseous lesion and should be considered as a central variant, although both variants are possible with such a condition. Although CEOC has a tendency to cross the midline,[23] our patient did not have such a clinical feature. Peripheral lesions, either in the maxilla or mandible, eventually involve periodontal structures mostly anterior to the molar region. Central lesions present as asymptomatic bony swelling with expansion in either the labial or lingual direction.[24]There have also been reports of perforation of the overlying compact bone, extending into soft tissues.[21] The cystic lining, when in contact with the apices or axial surfaces of the adjacent teeth, can cause a shift in their axial inclination, as seen in this case. The patient claimed that his teeth had shifted from their previous positions before the swelling, which was also one of his major concerns. One of the anterior teeth that was rotated was not related to the condition, as the patient reported it to be there before noticing the swelling. Researchers have reported root resorption at the apices when the cyst's fluid content exerts substantial pressure.[20,21]

One-third to half of the cases that have been reported show up on x-rays as multilocular or unilocular radiolucency's with calcifications of different densities. This is because they start in the jaws, which is where teeth are located.[8,9,21] Although radiolucent lesions can be uneven and have poorly defined boundaries, they often have a regular outline. About one-third of these cases involve an impacted tooth,[21] which was not seen in this case. Increased mineralization on microscopy has also been reported, which may not be clearly evident on radiographs,[25] which was also observed in this case. However, a central radiopaque mass was clearly visible on the radiographic images. The histopathological features observed in this case follow the classical description of such a lesion as defined by WHO classification (1992).[20] We observed various cell types (ameloblast, ghost, and stellate reticulum) along with fibers of collagen and hyalinization. The central conglomerate radiopaque mass observed on the radiographs is due to the result of the epithelial lining of CEOC being able to induce the formation of dental tissues. Ghost cells also have a liking for calcification and may, in addition, represent abnormal keratinization. This formation of osteoid/dentinoid associated with ghost cells is characteristic of CEOC, although it was earlier termed an inflammatory response,[1] which later was found out to be granulation tissue that gets calcified after laying down of juxta epithelial osteoid.[26] Ghost cells may result from coagulative necrosis, dystrophic calcification, or aberrant keratinization of the odontogenic epithelium. CEOCs are not the only lesions with ghost cells, since they can also be found in other odontogenic tumours (craniopharyngiomas, ameloblastomas,).[27,28] These lesions can also develop several forms of dystrophic calcification.[21,24,29]Ghost cells in CEOCs exhibit unique immunological localization of proteins associated with enamel, small tonofilaments, and dense bundles of fibers. There is no identification of the endoplasmic reticulum, mitochondria, Golgi apparatus, or ribosomes, but the cell membranes remain intact with junctional complexes. [21,30] Immunohistochemical profiles of these cysts have shown cytokeratin profile, B-cell lymphoma 2 (bcl-2) protein family, and Mel-CAM (CD146 or MUC18) expression, suggesting potential involvement in CEOC development.[31]The slow and non-aggressive behavior of the lesion does not warrant unnecessary excision of adjacent normal tissue. Surgical cyst enucleation is more likely the primary mode of treatment. From clinical diagnostic point of view, digital radiography especially with the use of artificial intelligence has made it possible to identify these lesions clearly, especially if any calcifications are present.[32,33] Adjustments of brightness and contrast with automatic options provides clinicians with a much clearer radiographic picture to arrive to a diagnosis, or conduct further investigations.

CONCLUSION

While CEOC's are encountered rarely in clinical practice, they are, however, a major concern for patients due to their apprehensions of being a tumour. They are significant to be considered for a differential diagnosis of an intraosseous swelling in the maxillofacial region. Clinical, radiographic, and histopathological investigations are extremely compulsory for ruling out malignancies. Patient assurance and education are synergistic to surgical correction through enucleation. Future recurrences are

prevented by careful histopathological evaluation and clinical follow-up.

Acknowledgement
The authors would like to sincerely thank the teaching and technical staff of the department of oral medicine and diagnosis and department of oral surgery for their valuable guidance and support during the treatment of the case.

Conflict of interest
The authors declare no conflict of interest during treatment of this case or while reporting its results.

REFERENCES

1. Gorlin RJ, Pindborg JJ, Odont , Clausen FP, Vickers RA. The calcifying odontogenic cyst-a possible analogue of the cutaneous calcifying epithelioma of Malherbe. An analysis of fifteen cases. *Oral Surg Oral Med Oral Pathol* 1962;15:1235-43.
2. Toida M. So-called calcifying odontogenic cyst: Review and discussion on the terminology and classification. *J Oral Pathol Med* 1998;27:49-52.
3. Barnes L, Eveson JW, Reichart P, Sidransky D, editors. World Health Organization Classification of Tumors. Pathology and Genetics of Head and Neck Tumors. Lyon: IARC Press; 2005.
4. de Arruda JA, Monteiro JL, Abreu LG, de Oliveira Silva LV, Schuch LF, de Noronha MS, Callou G, Moreno A, Mesquita RA. Calcifying odontogenic cyst, dentinogenic ghost cell tumor, and ghost cell odontogenic carcinoma: a systematic review. *J Oral Pathol Med.* 2018;47(8):721-30.
5. Irani S, Foroughi F. Histologic Variants of Calcifying Odontogenic Cyst: A Study of 52 Cases. The journal of contemporary dental practice. 2017 Aug 1;18(8):688-94.
6. Amboj M, Juneja, M. Ameloblastomatous Gorlin's cyst. *J Oral Sci* 2007;49:319-23.
7. Ledesma-Montes C, Gorlin RJ, Shear M, Prae Torius F, Mosqueda-Taylor A, Altini M, et al. International collaborative study on ghost cell odontogenic tumors. Calcifying cystic odontogenic tumor, dentinogenic ghost cell tumor and ghost cell odontogenic carcinoma. *J Oral Pathol Med* 2008;37:302-8.
8. Sakai VT, Filho CE, Moretti AB, Pereira AA, Hanemann JA, Duque JA. Conservative surgical treatment of an aggressive calcifying cystic odontogenic maxillary tumor in the young permanent dentition. *Pediatr Dent* 2011;33:265-9.
9. Kler S, Palaskar S, Shetty VP, Bhushan A. Intra-osseous calcifying cystic odontogenic tumor. *J Oral Maxillofac Pathol* 2009;13:27-9.
10. Nosrati K, Seyedmajidi M. Ameloblastomatous calcifying odontogenic cyst: A case report of a rare histologic variant. *Arch Iran Med* 2009;12:417-20.
11. Kumar L, Mattoo KA, Singh K, Sahai S, Yadav A. Restoration of oronasal function in sub mucosal cleft by utilising a palatal lift prosthesis: A clinical report. Clinical Dentistry. 2012; 6:26-30.
12. Mattoo KA, Singh M, Singh V. Muco-Cutaneous keratoacanthoma involving maxillary lip. Oral Surgery, Oral Medicine, Oral Radiology. 2014;2(2):21-2.
13. Jain SH, Mattoo KA, Makkar SU. Acute buccal space infection hampering mastication. International Journal of Research in Medical Sciences and Technology. 2015 Mar 19;1(2):3-4.
14. Mattoo KA, Nagaraj K. Herpetiform Ulceration Involving Pharynx–Case Report. Open Access Journal of Dental Sciences. 2017;2(6):000151.
15. Rahman S, Deep A, Mattoo KA. Bony exostoses on hard palate-Prosthetic inferences. Archives of Dentistry and Oral Health V2. I2. 2019:1.
16. Al Makramani BM, Al Sanabani F, Mattoo KA. Reviewing infection control in prosthodontic practice with a case of herpes simplex in the times of a corona virus pandemic. Annals of the Romanian Society for Cell Biology. 2021;25(6):10898-904.
17. Mattoo KA, Jain S. Managing prosthodontic (geriatric) patients during the SARS-CoV-2 pandemic. Journal of International Oral Health. 2020 Nov 1;12(Suppl 2):S69-75.
18. Abullais SS, Patel SI, Asiri EA, Jathmi AA, Alkhayri AH, Mousa YM, Ganem AA, Mattoo KA. Comparative evaluation of 3 commercial mouthwash formulations on clinical parameters of chronic gingivitis. Med Sci Monit. 2022;28:e937111-1.
19. Fazeli SR, Giglou KR, Soliman ML, Ezzat WH, Salama A, Zhao Q. Calcifying epithelial odontogenic (Pindborg) tumor in a child: a case report and literature review. *Head Neck Pathol*2019; 13:580-6.
20. Chandran A, Nachiappan S, Selvakumar R, Gunturu S, Lakshmi UV, Bharathi K, et al. Calcifying epithelial odontogenic cyst of maxilla: Report of a case and review and discussion on the terminology and classification. *J Microsc Ultrastruct* 2021;9:98-102.
21. Erasmus JH, Thompson IO, van Rensburg LJ, van der Westhuijzen AJ. Central calcifying odontogenic cyst. A review of the literature and the role of advanced imaging techniques. *Dentomaxillofac Radiol* 1998;27:30-5.
22. Akshatha BK, Manjunath GS, Soundarya N. Calcifying odontogenic cyst associated with compound odontoma–A rare entity. *J Oral Maxillofac Pathol.* 2023;27(Suppl 1):S69-74.
23. Li BB, Gao Yu. Ghost cell odontogenic carcinoma transformed from a dentinogenic ghost cell tumor of maxilla after multiple recurrences. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 2008;107:691-5.
24. Goran K, Klara S, Pavel K, Spomenka M. Calcifying odontogenic Cyst: Gorlin's cyst: Report of two cases. *Coll Antropol* 2004;1:357-62
25. McGowan RH, Browne RM. The calcifying odontogenic cyst: A problem of preoperative diagnosis. *Br J Oral Surg* 1982;20:203-12.
26. Abrams AM, Howell FV. The calcifying odontogenic cyst. Report of four cases. *Oral Surg Oral Med Oral Pathol* 1968;25:594-606.
27. Kumamoto H. Molecular pathology of odontogenic tumors. *J Oral Pathol Med* 2006;35:65-74.
28. Pindborg JJ, Kramer IR, Torloni H. Histological typing of odontogenic tumours, jaw cysts and allied lesions. Geneva, London: World Health Organization, HMSO; 1971. p. 28-35.
29. Hirshberg A, Kaplan I, Buchner A. Calcifying odontogenic cyst associated with odontoma: A possible separate entity (odontocalcifying odontogenic cyst). *J Oral Maxillofac Surg* 1994;52:555-8.
30. Garg A, Malhotra R, Urs AB. Ghost cells unveiled: A comprehensive review. *J Oral Biosci*. 2022;64(2):202-9.

31. Fregnani ER, Pires FR, Quezada RD, Shih IM, Vargas PA, De Almeida OP. Calcifying odontogenic cyst: clinicopathological features and immunohistochemical profile of 10 cases. *J Oral Pathol Med*.2003;32(3):163-70.
32. Sindi AS. The Role and Impact of Artificial Intelligence on the Future of Dental Radiography-A Mini-Review. Advances in Artificial Intelligence and Machine Learning. 2023; 3 (4): 98 [Internet]. Profession; 2020
33. Pandya VS, Morsy MS, Hassan AA, Alshawkani HA, Sindi AS, Mattoo KA, Mehta V, Mathur A, Meto A. Ultraviolet disinfection (UV-D) robots: bridging the gaps in dentistry. Frontiers in Oral Health. 2023 Nov 1;4:1270959.