Journal Homepage: -www.journalijar.com

INTERNATIONAL JOURNAL OF ADVANCED RESEARCH (IJAR)

Article DOI:10.21474/IJAR01/14363
DOI URL: http://dx.doi.org/10.21474/IJAR01/14363

CASE REPORT

CALCIFYING EPITHELIAL ODONTOGENIC TUMOR OF POSTERIOR MAXILLA: A RARE CASE REPORT

Mohammad Imran Khan, Abhisheik Khare, Sameera Shamim Khan and Iqbal Ali

Manuscript Info

Manuscript History
Received: 05 January 2022
Final Accepted: 09 February 2022
Published: March 2022

Key words:-
Calcifying Epithelial Odontogenic Tumor, Amyloid, Liesegang Rings

Abstract

Calcifying epithelial odontogenic tumor (CEOT), also known as Pindborg tumor, is a rare benign but locally aggressive odontogenic neoplasm, accounts for<1% of all odontogenic tumors.The tumor has a recurrence rate of 10%–20% and so periodic follow-up is necessary. Here is reported the case of a 21-year-old female patient with a CEOT in the right maxillary region of the jaw.

Introduction:-

Odontogenic tumors represent a spectrum of lesions ranging from malignant and benign neoplasms to dental hamartoma. The odontogenic tumors are unique to jaw because it is arising from odontogenic residues either odontogenic epithelium, ectomesenchyme or from both components[1,2]. WHO classified Odontogenic Tumors based on which component involved in the development of a tumor. They classify into three categories epithelial origin, mesenchymal origin, and mixed origin[3]. The calcifying epithelial odontogenic tumor is an epithelial origin locally aggressive benign odontogenic tumor[1].

The CEOT can be located either intra-osseously or extra-osseously. When occurring intra-osseously (by far the most common), it may occasionally show local invasiveness. The intraosseous CEOT often presents as a painless mass with slow growth. When located in the maxilla, patients may sometimes complain of nasal congestion, epistaxis, and headache. The characteristic radiographic appearance is of an irregular unilocular or multilocular radiolucent area containing radiopaque masses of varying size and opacity. In many cases, especially in tumors of relatively short duration, the calcified concrements are minute and may be undetectable on radiographs. When an unerupted tooth is associated with the tumor, the radiopacities tend to be located close to the tooth crown[4].

Calcifying epithelial odontogenic tumor (CEOT) is a rare odontogenic tumor that accounts for less than 1% of all odontogenic tumors. Thoma and Goldman first described CEOT as “adenoid adamantoblastoma” in 1946, but it was only acknowledged as a distinct entity by Pindborg in 1958 and in 1971 the term “Calcifying epithelial odontogenic tumor” was generally accepted and adopted by the WHO. 1,2 It is a benign, slow-growing, locally invasive odontogenic tumor. It generally occurs in patients between 20-60 years of age, with a mean age of diagnosis of 40. It affects men and women equally. The most common location of the tumor is the mandibular premolar and molar region (68%) and, less frequently, the maxilla. Half of the cases are associated with an impacted tooth[5-7]. Till now 400 cases of CEOT are reported in the literature[1].

Case Report

A 21 years old female patient reported to the department of oral and maxillofacial surgery with chief complaint of decayed tooth, pain and swelling in upper right back region of jaw for one year. As the swelling was asymptomatic

Corresponding Author:- Mohammad Imran Khan

and not cause much disfigurement of the face, the patient did not consult to the clinician. With due time swelling size increased and associated with intermittent pain. Her past medical and family history was non-contributory. On examination her face was asymmetrical, with slight extraoral swelling was seen.

Intraorally a well-defined expansile swelling seen in the maxillary posterior region extended from 15-to 16 region, lesion entirely positioned buccal to first molar which was firm, tender on palpation and bony hard in consistency. Figure 1: (a) lesion on right posterior region of maxilla.

Overlying skin appeared normal and on palpation the swelling was bony hard and there was mild tenderness present. There was no involvement of the lymph nodes. Intraoral examination revealed root stumps of 16,14,13,12,11 and decayed 27.

A panoramic radiograph showed a well-defined unilocular radiolucency with a corticated margin extending from the root of tooth 15 to the root of 13 area, which measured about 2cm x 1.5cm in diameter. Figure 1: (b) lesion on radiograph involving right first molar with root stump of the tooth.

Gross examination of the surgical specimen included small bit of soft tissue measuring approx. 0.8x0.7x0.5cm in total. Specimen was brownish in color and soft to firm in consistency.

Microscopic features revealed the presence of non-keratinized stratified squamous epithelium showing arcading pattern with moderately collagenous connective tissue stroma. Epithelial islands show dystrophic calcification within the amyloid like material forming concentric rings (Liesegang rings). Numerous blood vessels lined by endothelial cells and few areas of focus show multinucleated giant cells and clear cells.

Special stain:
The amyloid like material was positive for Congo Red special stain.

Discussion:-

The calcifying epithelial odontogenic tumor is classified as an uncommon, benign, odontogenic neoplasm that exclusively epithelial in origin, the etiology of which remain enigmatic. The source of epithelial cells initially suggested by Pindborg was reduced enamel epithelium but today, most investigators believe the cells of origin are stratum intermedium[1].

There have been very few reported cases in the maxilla till date and posterior maxilla being the rarest site. This case adds to a small number of Pindborg tumor cases located in the maxilla. Three distinctive features of the present case are: a rare localization of the lesion, presentation as an ulcerated mass, and a comparatively young age of patient.

According to Pindborg, the characteristic histologic criteria for diagnosis of CEOT are sheets of large polygonal epithelial cells that have well-defined borders and show prominent intercellular bridges. Nuclear pleomorphism varied with size, shape, and number. Cellular abnormalities and mitotic figures are rarely seen. Cytoplasm is abundant and eosinophilic. Varying amounts of extracellular amyloid-like material that stains Congo red positive in polarized light[8,9]. This amyoid like material has been extensively investigated by histochemical, immunohistochemical, ad biochemical methods. It has been identified as unique protein that is produced by this tumor, as well as by the normal odontogenic apparatus and other odontogenic neoplasms and this material has been designated as “odontogenic ameloblast associated protein” (ODAM)[10].

Concentric calcified masses of Liesegang’s rings calcification pattern are pathognomonic for this tumor. Ai-Ru et al. proposed a subclassification into four patterns. Two or more patterns can coexist in the same tumor. The patterns are as follows:

Pattern I: Polyhedral epithelial cells arrangements with deeply eosinophilic cytoplasm and prominent nuclei, distinct cell outlines and intercellular bridges, frequent cell abnormalities, no mitotic figures and calcifications are seen in the fibrous stroma.

Pattern II: Cribriform appearance of the cell arrangements, intercellular bridges might be discrete, few cell abnormalities, and calcification of the eosinophilic material filling cribriform spaces with Liesegang calcification pattern.

Pattern III: Scattered or dense arrangements of the epithelial cells with varying size and multinucleated giant cells are frequent; mucoid material is present in the stroma.

Pattern IV: Nests and cords of epithelial cells containing an abundant eosinophilic cytoplasm or a clear vacuolated cytoplasm, variable amounts of stroma with eosinophilic material and calcifications[8].

The presence of calcifications in CEOT may have prognostic implications. The absence of calcification indicates a poorly differentiated tumor with more chance of recurrence. The differential diagnosis of CEOT includes calcifying epithelial odontogenic cyst, adenomatoid odontogenic tumor, ameloblastoma, ameloblastic fibro-odontoma, odontogenic fibroma and ossifying fibroma[11-13].

Figure 2 (a) histopathological appearance 10x, (b) 40x showing calcification ring formation

To explain the pathogenesis Peacock et al conducted a study on seven cases of CEOT. They conclude that the sonic hedgehog pathway (SHH) to be involved in the development of CEOT and they also noticed mutation in PTCH1. PTCH1 is a tumor suppressor gene within the Sonic hedgehog pathway. The sonic hedgehog pathway regulates the development of multiple organ systems, including odontogenesis by controlling cell to cell interaction and cell proliferation in tissue. PTCH1 mutation first detected in nevoid basal cell carcinoma syndrome and has also been found in both CEOT and keratocystic odontogenic tumors. However, the clinical significance of these mutations is unknown. CEOT affected a wide range of ages, most commonly occurs between 20 and 60 years of age, with a peak incidence in the 5th decade and equal distribution between both gender. However, some cases reported before 20 years of age, present case also reported CEOT affected a 13-year-old female. It commonly presented as asymptomatic, slow-growing, and locally aggressive lesion. Initial clinical signs/symptoms of the lesion are a local expansion of bone with the migration of the teeth of the affected region. Later the overlying mucosa becomes so inflamed that even a slight trauma can lead to bleeding from that side. The premolar and molar region of the mandible is the most prevalent site for its occurrence. Although the case also reported in maxilla but less frequent as compared to mandible. The further central intraosseous form is the most common, larger, and more invasive and about 50% of cases show signs of cortical plate perforation. A study conducted by Bruno R. et al according to results the distribution of 247 cases of CEOT among different ethnic groups/races is as follows white-90, Asian-55, Indian -54, blacks -29, Hispanics- 8, Persian/Iranian -6, and 5 in Turkish[1,13,14].

Treatment

The treatment plan is dependent on multiple factors such as the size of the tumor, location of the tumor, general condition of patient and operator skill. Small, well-defined, intra-bony mandibular lesions can be treated by simple enucleation or curettage followed by judicious removal of a thin layer of bone next to the tumor. Larger tumors may need segmental resection, hemi-mandibulectomy and hemi-maxillectomy, followed by bone grafting or distraction osteogenesis[16].

In our case, surgical excision along with the associated teeth was done.

The recurrence rate varies from 10% to 20% with the clear cell variant having a higher recurrence rate of around 15%. Malignant transformation is rare.

Thus, our report describes a case of CEOT with atypical clinical and radiographic features, which include occurrence in the posterior maxilla, root resorption of the teeth associated with the lesion and absence of calcifications radiographically.

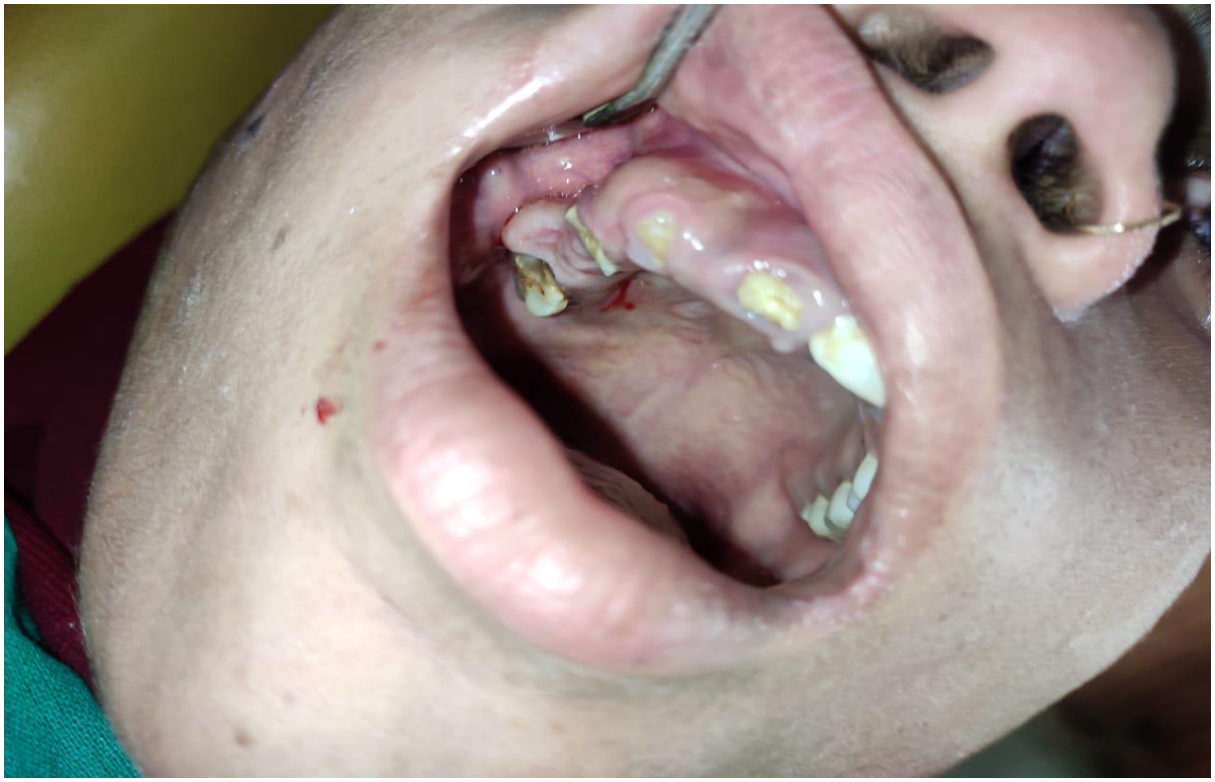

Figure 1(a):-

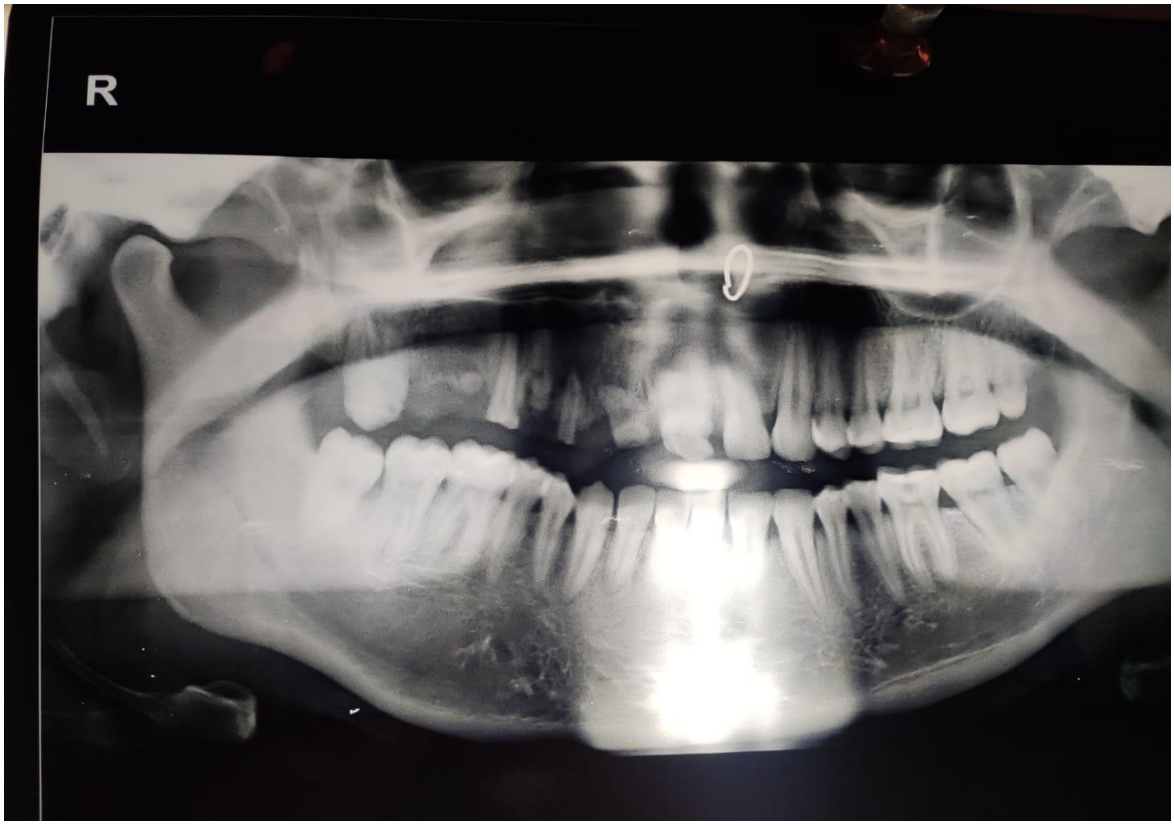

Figure 1 (b):-

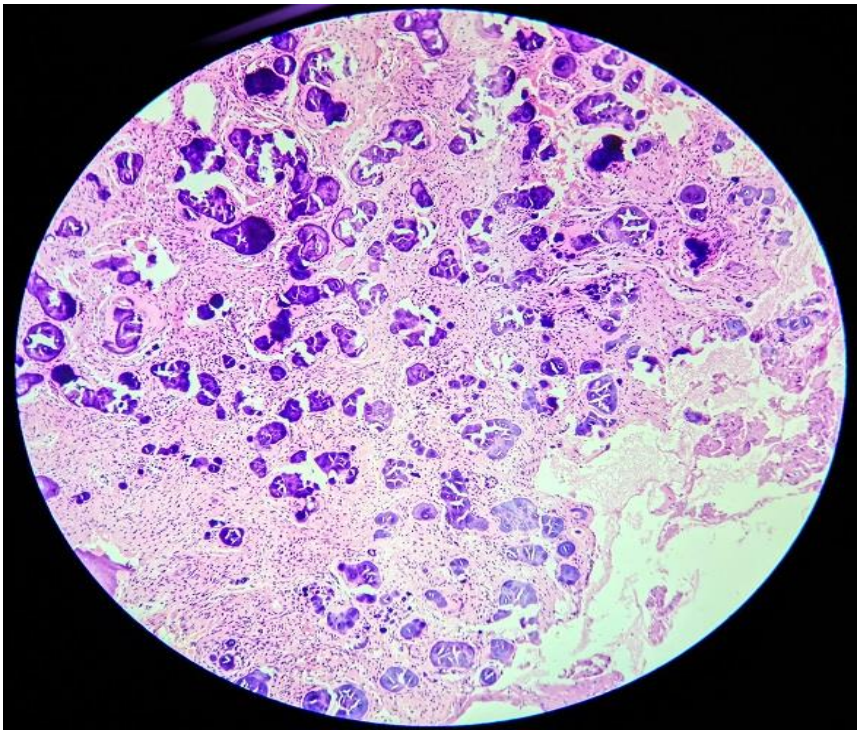

Figure 2(a):-

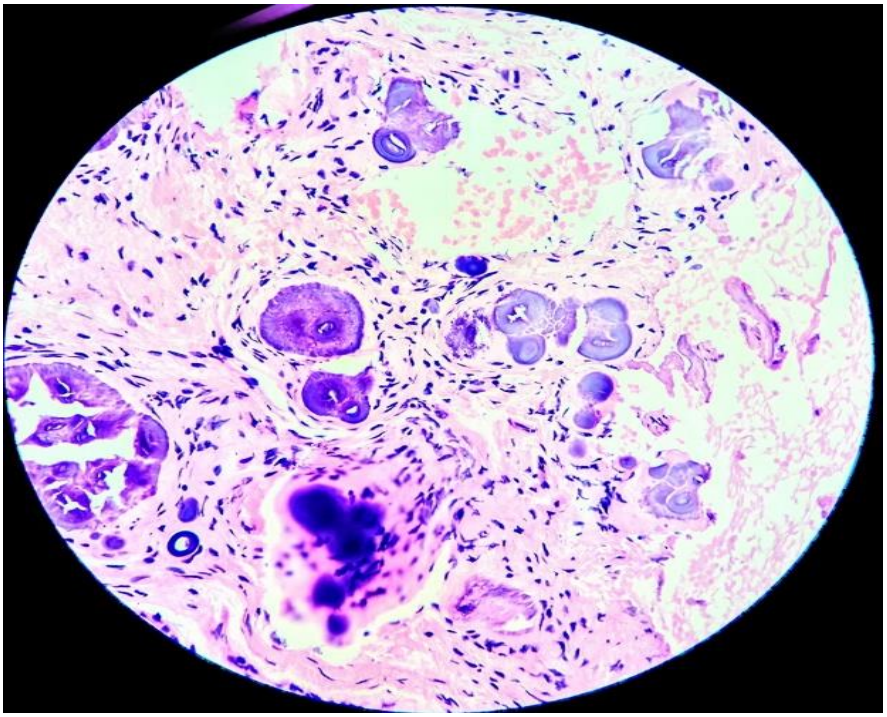

Figure 2 (b):-

Conclusion:-

CEOT also known as Pindborg tumor is typically a benign yet locally aggressive tumor more commonly seen in middle-aged adults, but can be seen in children, albeit rarely. Amyloid like matrix is a unique component that may be associated with tumor maturation and differentiation, and possibly lower risk of malignant transformation. Thorough clinical and radiological examination and careful histopathological interpretation are important for

confirmed diagnosis. Early surgical intervention is advocated for the treatment of histopathologically confirmed calcifying epithelial odontogenic tumors as the recurrence rate reported are around 10-20 % to prevent the tumour expanding beyond confines of maxilla or mandible.

References:-

1. Sheetal Singar, Ajay Pratap Singh Parihar, Prashanthi Reddy, Ashish Saxena and Arvind Jain; An Aggressive Calcifying Epithelial Odontogenic Tumor: A Rare Case Report; Journal of Advances in Medicine and Medical Research 2021;33:24-30
2. Shafer WG, Hine MK, Levy BM.Shafer's Textbook of Oral Pathology. 8 thedi. New Delhi: Elsevier; 2018.book
3. John M, Merva S. Odontogenic tumors: where are we in 2017?. J Istanbul Univ Fac Dent 2017;51:S10-S30.
4. Saurabh Sabnis, Abhishek Mulay, Pravin Walunjkar; Calcifying epithelial odontogenic tumour: Report of a case and review of literature. MedPulse International Journal of Pathology 2020;13:41-44
5. JayarathiIshwarya K S, V. Ramesh, P. D. Balamurali, Karthikshree V Prashad; Case report and review of calcifying epithelial odontogenic tumor. International Journal of Oral Health Dentistry 2021;7:206-209
6. Lee W, Myung NH, Kim CH. Calcifying epithelial odontogenic tumor: report of three cases with immunohistochemical study. Int J Clin Exp Pathol. 2016;9(5):5733–42
7. 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 and Neck Pathol. 2019;13(4):580– 6.
8. TallaHarshavardhan, Sushma Naag, Dorairaj Parthiban Umamagesh, Esther Priyadarshini; Maxillary Calcifying Epithelial Odontogenic Tumor Presenting at a Postextraction Site: A Case Report and Brief Review of Literature. Journal of Indian Academy of Oral Medicine & Radiology 2017;29:333-6
9. Nascimento GJF, Pereira KMA, Nonaka CFW, Medeiros AMC, Galvao HC. Calcifying epithelial odontogenic tumor of the maxilla. Braz J Otorhinilaryngol2009;75:468
10. Brad W Neville, Douglas D Damm, Carl M Allen, Angela Chi; Oral and Maxillofacial Pathology, Frist south asia edition.
11. Vidya Ajila, R Gopakumar, Shruthi Hegde, Harish Shetty; Calcifying epithelial odontogenic tumor of the posterior maxilla: Journal of health and research 2016;3:74-8
12. Seema Kaushal, Sandeep R Mathur, Maneesh Vijay, Ankur Rustagi; Calcifying epithelial odontogenic tumor (Pindborg tumor) without calcification: A rare entity. 2012;16:110-2
13. K Sri Krishna, Vinod VC, M Venkateswarlu, Sridevi K, GS Prasad Reddy; Pindborg Tumor: Review of Literature and Case Reports. 2011;23:660-663
14. Bruna Barcelos Só, Vinicius Coelho Carrard, Laura de Campos Hildebrand, Marco Antonio Trevizani Martins, Manoela Domingues Martins; Synchronous Calcifying Epithelial Odontogenic Tumor: Case Report and Analysis of the 5 Cases in the Literature. 2020;14:435-441
15. A Zhang, SY Chaw, AA Talacko, WJ Besly, NW Savage, PA Monsour; Central calcifying epithelial odontogenic tumour in the posterior maxilla: a case report. 2016;61: 381-385
16. Neeraj Singh, Sharad Sahai, Sourav Singh, Smita Singh; Calcifying epithelial odontogenic tumor (Pindborg tumor). 2011;2:225-7.