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Extensive Calcifying Epithelial Odontogenic Tumour of Maxilla

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ABSTRACT
Calcifying epithelial odontogenic tumour (CEOT) presenting at an unusual site is rare. The characteristic cytologic findings in association with radiologic features can help the cytopathologist and the surgeon in rendering a firm preoperative diagnosis of Calcifying epithelial odontogenic tumour even at atypical sites such as maxilla. The purpose of this article is to report an additional case of CEOT and discuss the radiologic appearance, histopathological features, clinical behavior and surgical management. Although it has a predilection for the mandible, a small proportion of cases have been reported to occur in the maxilla and lacks classical clinicoradiologic features. The cytologic features in conjunction with the radiologic picture can be helpful in making a preoperative diagnosis and guiding management. Calcifying epithelial odontogenic tumor (CEOT) is a rare odontogenic tumor of the jaw. Clinically, it is a slowly growing, locally aggressive tumor[1] Histologic criteria for the diagnosis of CEOT have been cited by Franklin and Pindborg[2]. The cytologic features in conjunction with the radiologic picture can be helpful in making a preoperative diagnosis and guiding management.

Keywords : Biopsy, Calcifying epithelial odontogenic tumor, Maxillary odontogenic tumor, Resection.

I. INTRODUCTION

Calcifying epithelial odontogenic tumour (CEOT) is a slowly growing, benign, but nonencapsulated and locally invasive, epithelial, odontogenic neoplasm with a singular histomorphological pattern characterized by irregular sheets and islands of eosinophilic, polyhedral, and often pleomorphic cells, which eventually disintegrate into an eosinophilic, amorphous substance, which stains with amyloid markers and tend to calcify. Like most odontogenic tumors the CEOT occurs as an intraosseous and as a rarer and less aggressive extraosseous variant. The male:female ratio for the intraosseous variant is between 1:1 [1,2] and 1:1.5, and for the extraosseous variant 1:0.8 (based on 11 cases only) [2] .The age range is between 8 and 92 years at the time of diagnosis, with a mean of 36.9 years for both topographic variants. The mean for the intraosseous variant is 38.9 years and for the extraosseous variant 34.4 years, which may be explained by the fact that the extraosseous variant presents as a gingival enlargement, which is likely to be diagnosed at an early stage.

The age peak for men is in the third decade and for women in the fourth decade [2]. The majority of intraosseous cases are diagnosed in patients between 20 and 60 years of age. There is a predilection for peripheral (extraosseous) cases to occur in the anterior segment of the jaws. They present as a gingival swelling covered by mucosa of normal color. The intraosseous tumors on the other hand occur primarily in the mandible and particularly in the premolar and molar regions. The mandible: maxilla ratio is 2:1 [2, 3]. Simultaneous occurrence of CEOT in more than one location has been described, but is exceedingly rare.

The growth rate is slow. The tumor is usually symptomless, apart from a slowly progressive swelling of the jaw. There are a few reports associated with pain, nasal obstruction, epistaxis, and proptosis. An unusual case of maxillary CEOT, which caused displacement of the eye in a 30-year-old woman, was published by Bridle et al. About 60% of the intraosseous tumors are associated with an unerupted permanent tooth, most often a mandibular molar [3].

II. CASE REPORT

A 26-year-old female patient reported to the department of Oral and Maxillofacial surgery, Government dental college & research institute, VIMS campus, Ballari, with a chief complaint of dull aching pain and swelling in left maxillary region since 8 months. On extraoral examination well defined facial swelling around a size of 4.5x6x3cm was present on the left maxillary sinus region and was nontender with normal overlying mucosa. (Fig.1a, 1b and 1c) Correlating with the history and clinical examination panoramic radiograph was advised which revealed a well-defined unilocular radiolucency in the left maxillary sinus with impacted canine anteriorly. (Fig.2) CT scan of the left maxillary sinus showed well defined unilocular radiolucent lesion with

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impacted canine confined within the boundaries of maxillary sinus (Fig.3 and Fig.4).

Incisional biopsy of the lesion was performed which revealed histopathological features of CEOT. On microscopic examination well-known sheets and strands of polyhedral, polymorphous epithelial cells with clear intercellular bridges were seen along with huge aggregations of amyloid material and very small foci of calcification were also observed. The closely packed tumour cells exhibited variation in nuclear size and shape where as other areas showed large tumour cells with pyknotic nuclei pressed against the cell membrane and faintly eosinophilic cytoplasm. The differential diagnosis for CEOT include adenomatoid odontogenic tumor (AOT), calcifying odontogenic cyst (COC), ameloblastic fibro odentoma (AFO), odontoma.

The patient was posted for surgical exploration under general anesthesia and the lesion was resected with aggressive curettage using intraoral maxillary vestibular approach and the entire lesion was excised along with impacted canine (Fig.5 and Fig.6). rest of maxilla was preserve. The excised tissue was sent for histopathological examination which reconfirmed with features of CEOT (Fig.7a and 7b) Patient was followed up for one year with no evidence of recurrence (Fig.8).

III. DISCUSSION

The etiology and source of origin of the tumor is unknown. Calcifying epithelial odontogenic tumor (CEOT), is a rare benign odontogenic neoplasm representing about 0.4-3% of all odontogenic tumors. It was believed for a long time that the tumor arises in the reduced enamel epithelium of an embedded tooth. Since not all CEOTs develop in association with an embedded tooth, there must be other sources, and a remnant of the dental lamina is an obvious candidate [3, 4]. Proliferating odontogenic epithelium at the top of the dental follicle of an unerupted tooth at the lower orifice of the gubernaculum dentis is another possibility.

Although the tumor is in some cases relatively easily enucleated, it is nonencapsulated apart from focal areas in some tumors, and it resorbs and infiltrates the surrounding bone [4, 5]. Macroscopically it presents as a firm mass of varying color and at bisecting the specimen usually reveals calcified particles. There may be minute cystic spaces in the tissue, but only one case of unicystic lesion with the tumor developing apparently in the wall of a dentigerous cyst has been published[5,6]

The classic histological picture of an intraosseous CEOT as described by Pindborg shows irregular sheets and islands often with many pointed extensions, which consist of polyhedral epithelial cells with abundant eosinophilic cytoplasm, sharply defined cell borders, and well developed intercellular bridges. The nuclei are round and sometimes slightly lobulated, most of them are strongly basophilic, and they are frequently pleomorphic, but the number of pleomorphic nuclei differs from tumor to tumor. Mitotic figures are rarely encountered, and the nuclear pleomorphism is not a sign of malignancy [6.7]. In a rare case of malignant transformation the number of mitoses is conspicuously increased, tumor cells are found within the vessels, and the Ki-67 index is considerably elevated. Double-nucleated cells may be seen, and the nucleoli may be prominent [7]

Within the sheets of tumor cells are various amounts of rounded, eosinophilic homogeneous masses. The substance can be observed intracellularly in swollen epithelial cells with disintegrated nuclei displaced to the cellular border, which has lost its integrity and presumably represents perished tumor cells, although it has also been interpreted as a secretion product, and even as enamel matrix. Most of the substance is found extracellularly and when it is distributed in foci within a tumor island the pattern is cribriform[7,8].

Extensive amounts of the eosinophilic, homogeneous substance may be seen in the connective tissue at some distance from intact tumor islands. They may still represent areas of perished tumor cells, although they have been interpreted differently. Often small clusters of compressed cells remain in the areas [8].

Vickers et al. [9] demonstrated the substance to react positively to amyloid staining like Congo red and fluorescence with thioflavine T. This finding has been confirmed by numerous later investigators. The eosinophilic, homogeneous, positive amyloid reacting substance eventually becomes calcified. Calcified foci are initially seen as tiny spots in small areas of the substance. With increasing calcification they form spherules showing appositional basophilic concentric rings (Liesegang rings). Eventually the calcified areas coalesce, forming large calcified aggregates. Before calcification the homogeneous substance is faintly PAS-positive, but with progressive calcification the areas become more PAS-positive. Calcified spherules may also be seen scattered in the connective tissue without association to the homogeneous substance. In some tumors formation of rounded islands of hard tissue is seen; it is a

cellular product that contains collagen and has morphology like cementum.

There is a considerable histomorphological variation from tumor to tumor and within the individual tumors. Some are dominated by large irregular epithelial sheets, other show numerous small islands and strands of tumor epithelium in the connective tissue stroma, which always consist of mature collagenous connective tissue. Calcification may be sparse or conspicuous; extensive calcification is primarily seen in large tumors of long duration [9, 10].

The extraosseous variant of CEOT shows principally the same histomorphology, but the tumors cells form rather strands and small islands than large sheets, and the amount of calcified material may be minimal or lacking. The extraosseous variants are undoubtedly diagnosed at an earlier stage than the intraosseous tumors. In a few cases Langerhans cells have been described in the tumor; the cells were S-100 protein-positive and identified ultrastructurally on the finding of rod- and tennisracket-shaped Birckbeck's granules. Their function in the tumor is unknown.

The presence of numerous clear cells in CEOT is uncommon; Krolls and Pindborg reported the first two examples of this rare tumor in 1974; Philipsen and Reichart reviewed 15 cases in 2000, and further cases have been published. In some of the cases the clear cells component has been so dominating that the tumors are probably better classified as CCOC with patterns of CEOT. Such CEOT patterns are also seen in AOTs. There is evidence that CEOTs with numerous clear cells are more aggressive than CEOTs without clear cells [9, 10].

Microscopically the classic CEOT is quite distinctive and not too difficult to diagnose. In absence of calcification and presence of cellular pleomorphism a primary intraosseous squamous cell carcinoma (PIOSCC) or metastatic tumor should be ruled out. In contrast to these the CEOT is characterized by none or few mitotic figures and a low Ki-67 index [9, 10].

In cases with a considerable amount of clear cells the differential diagnoses are CCOC, central mucoepidermoid carcinoma, central acinic cell carcinoma, and metastatic renal carcinoma. Tumors consisting of short strands and small islands of epithelium in abundant fibrous connective tissue stroma and with little or no eosinophilic, homogeneous, amyloid-staining positive substance or calcification may be difficult to distinguish from an epithelium-rich odontogenic fibroma. Both may show foci of calcified material and cementum-like hard tissue. As the connective tissue is the tumor component, it is conspicuously more cellular than the stroma of a CEOT. Amyloid staining (Congo red and fluorescent thioflavine T) in most cases will disclose minute areas of amyloid-positive homogeneous substance in a CEOT; it is not supposed to be present in an odontogenic fibroma. Dunlap reported, however, two cases of what they considered epithelium-rich odontogenic fibromas, which both contained solitary or clustered eosinophilic hyaline droplets, which were weakly positive for amyloid staining [9, 10].

Methods of treatment have varied from simple enucleation or curettage to hemimandibulectomy or hemimaxillectomy. Treatment is dictated to some extent by the location and the size of the tumor. Complete removal is necessary, so resection of the entire mass, with tumor-free surgical margins and long-term follow-up is indicated.

IV. CONCLUSION

CEOT involving the maxilla is a rare odontogenic neoplasm. Along with radiologic correlation, the classical cytologic features help with the preoperative diagnosis of CEOT & aid in guiding management. The characteristic cytologic findings in association with radiologic features can help the cytopathologist in rendering a firm preoperative diagnosis of CEOT even at atypical sites such as the maxilla.

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FIGURE LEGENDS

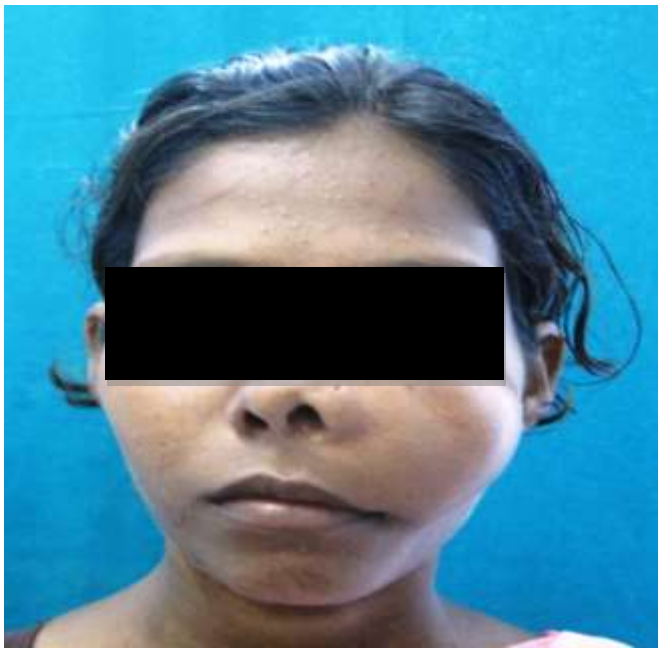

Figure I: Extraoral frontal view photograph showing well defined swelling on left side of the face causing gross asymmetry.

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Figure II: Extraoral lateral view photograph showing well defined swelling on the left side of the maxilla causing gross asymmetry obliterating left nasolabial fold.

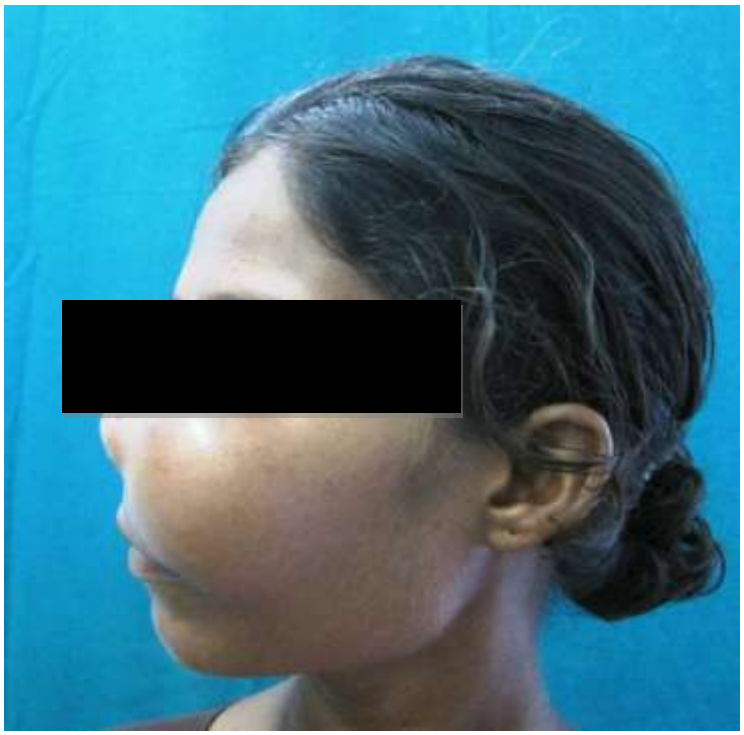

Figure 1II: Extraoral submental view photograph showing well defined swelling on left side of the maxilla displacing the nostrils and upper lip.

International Journal Dental and Medical Sciences Research
Volume 4, Issue 1, Jan-Feb 2022 pp 60-70 www.ijdmsrjournal.com ISSN: 2582-6018

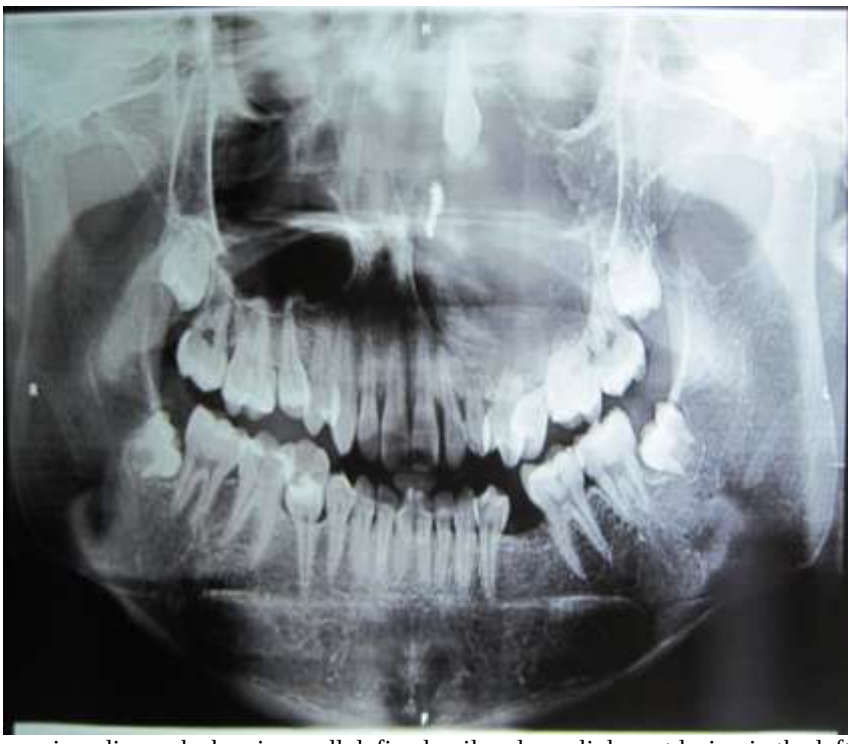

Figure IV: Panoramic radiograph showing well defined unilocular radiolucent lesion in the left maxillary sinus with displaced left maxillary canine tooth.

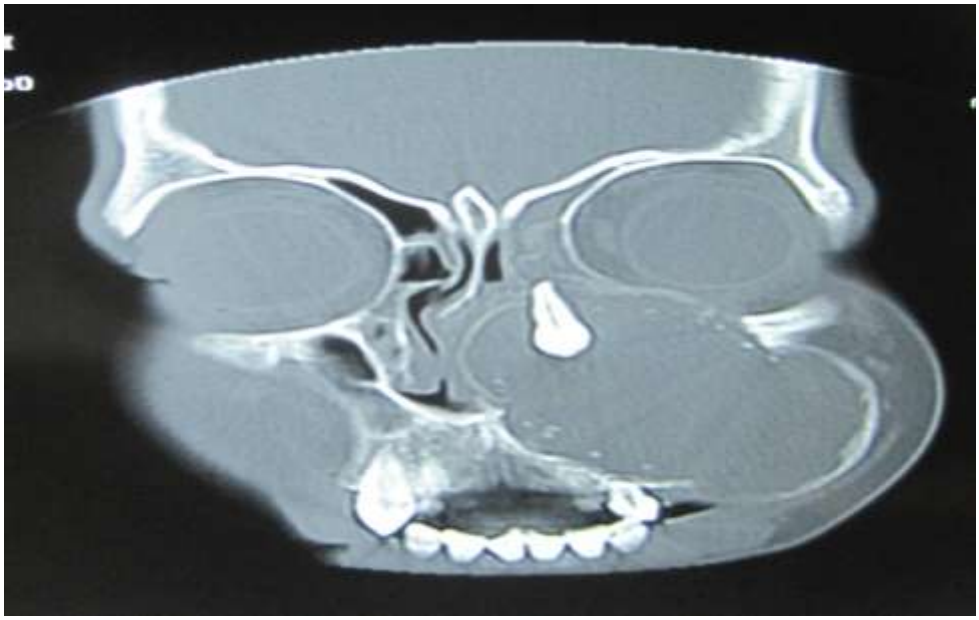

Figure V: CT coronal section of the skull showing well defined unilocular lesion in the left maxillary sinus with displaced left maxillary canine tooth.

International Journal Dental and Medical Sciences Research
Volume 4, Issue 1, Jan-Feb 2022 pp 60-70 www.ijdmsrjournal.com ISSN: 2582-6018

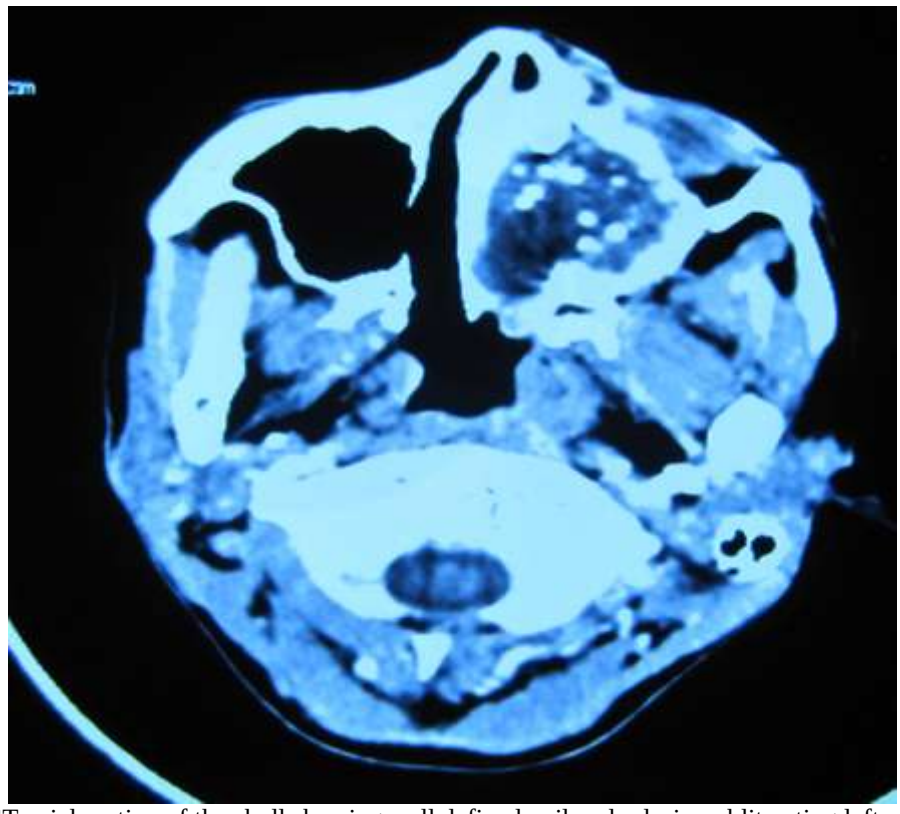

Figure VI: CT axial section of the skull showing well defined unilocular lesion obliterating left maxillary sinus with scattered calcifications.

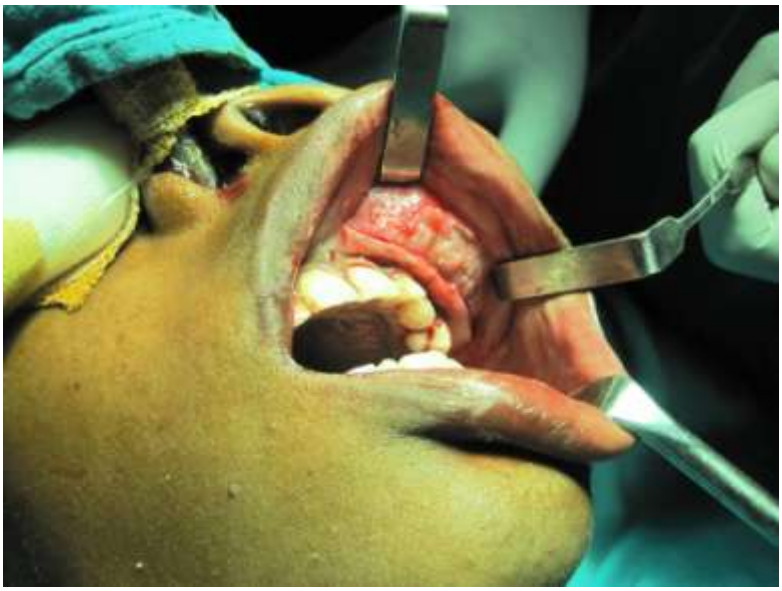

Figure VII: Intraoperative photograph showing exposed lesion using maxillary vestibular incision.

International Journal Dental and Medical Sciences Research
Volume 4, Issue 1, Jan-Feb 2022 pp 60-70 www.ijdmsrjournal.com ISSN: 2582-6018

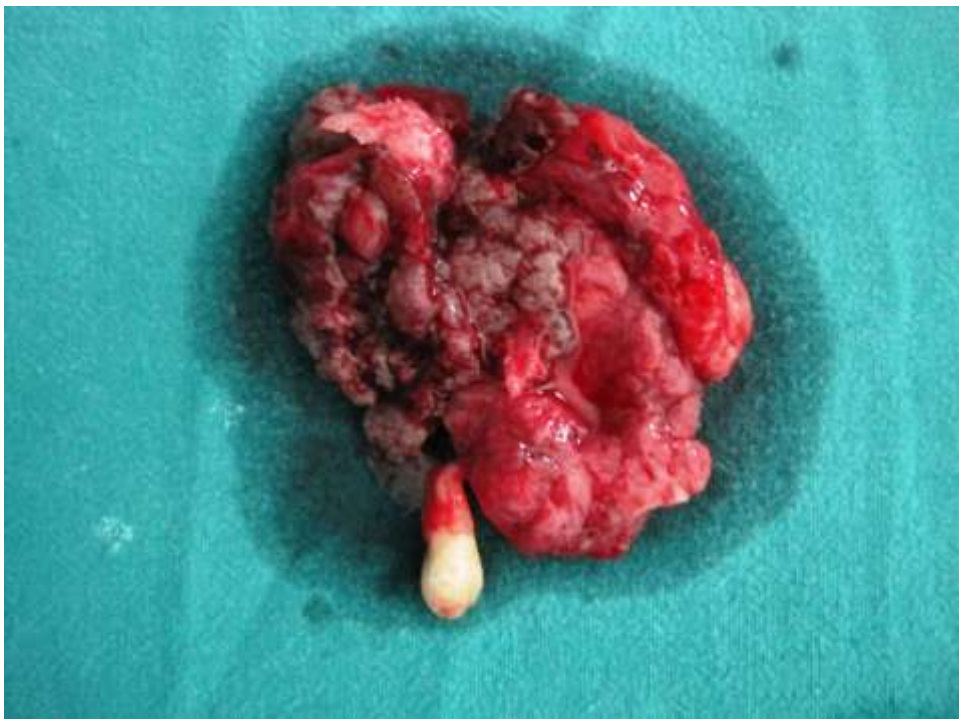

Figure VIII: Excised specimen of the lesion showing a variegated appearance with maxillary left canine tooth.

International Journal Dental and Medical Sciences Research
Volume 4, Issue 1, Jan-Feb 2022 pp 60-70 www.ijdmsrjournal.com ISSN: 2582-6018

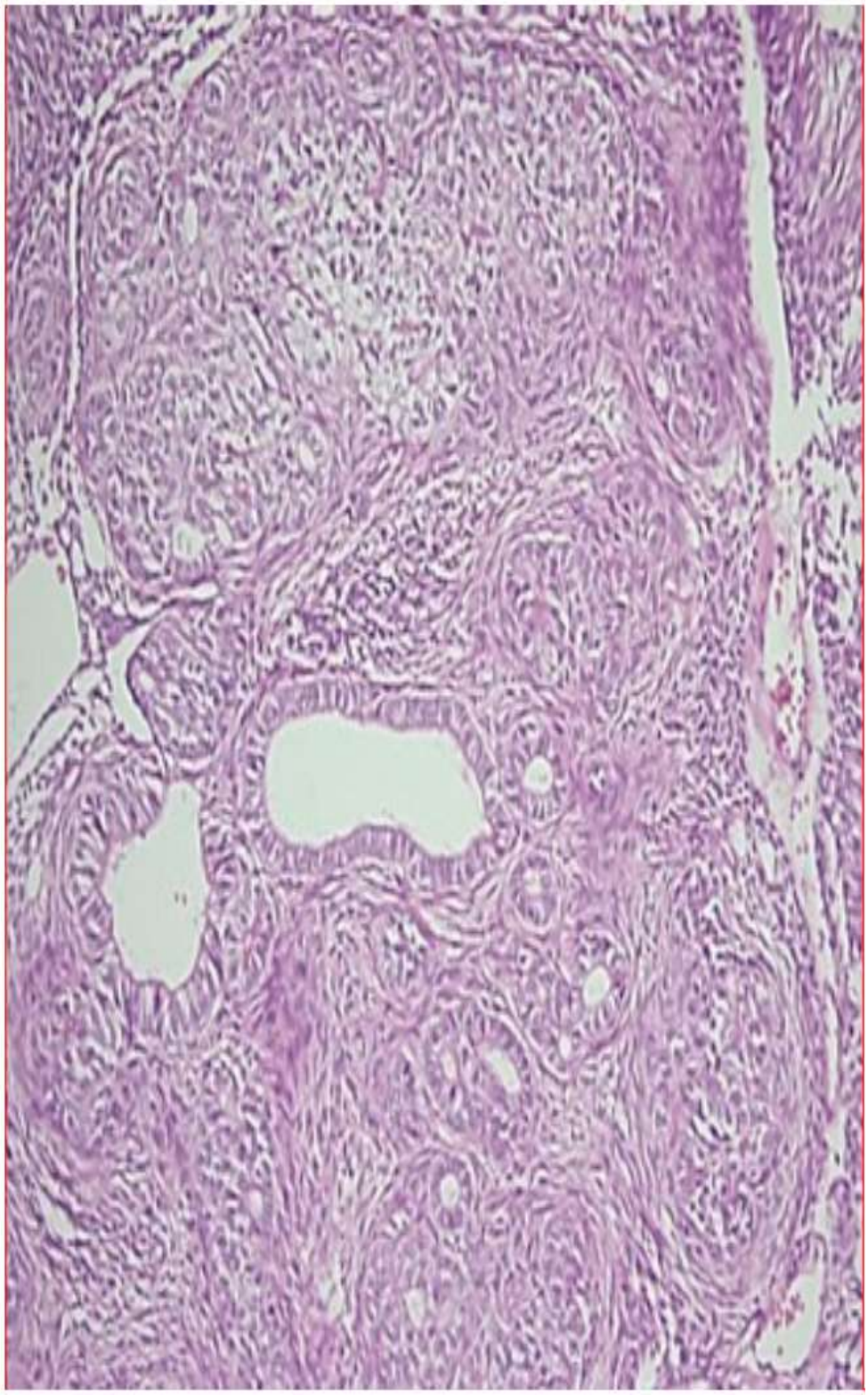

Figure IX: Histopathological picture showing well-known sheets and strands of polyhedral, polymorphous epithelial cells with clear intercellular bridges.

International Journal Dental and Medical Sciences Research
Volume 4, Issue 1, Jan-Feb 2022 pp 60-70 www.ijdmsrjournal.com ISSN: 2582-6018

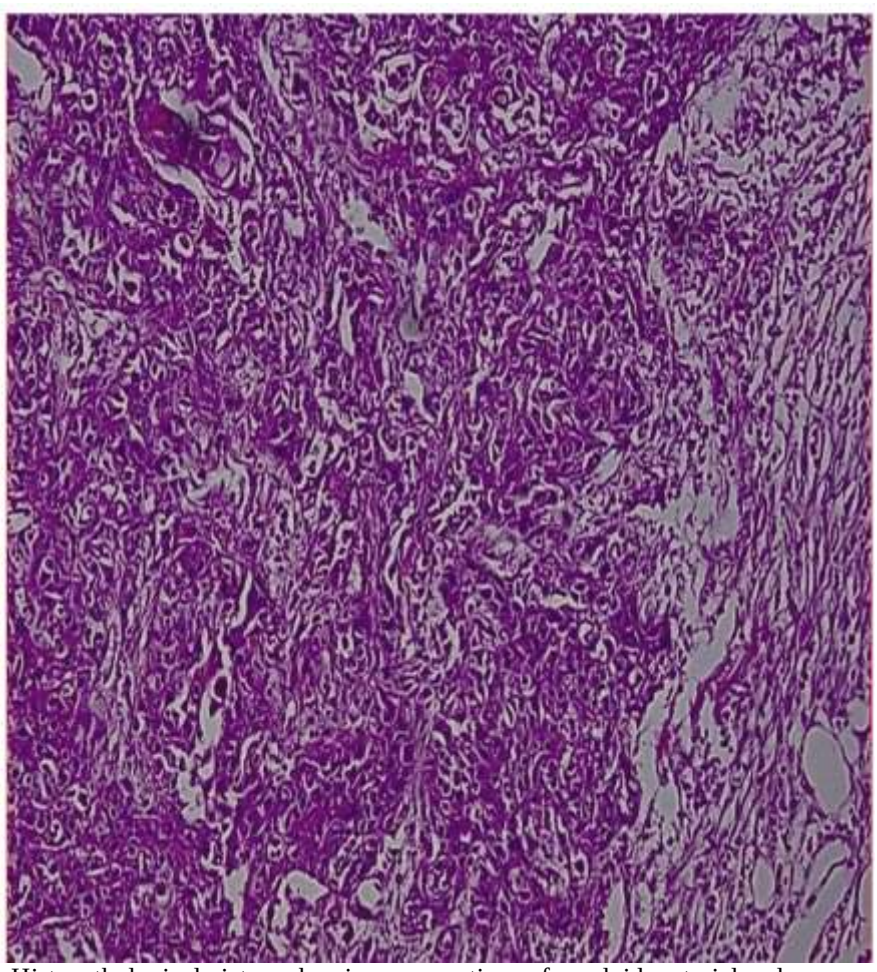

Figure X: Histopathological picture showing aggregations of amyloid material and very small foci of calcification were observed with closely packed tumour cells exhibit variation in nuclear size and shape along with stromal and intraepithelial aggregates of amyloid deposition.

International Journal Dental and Medical Sciences Research
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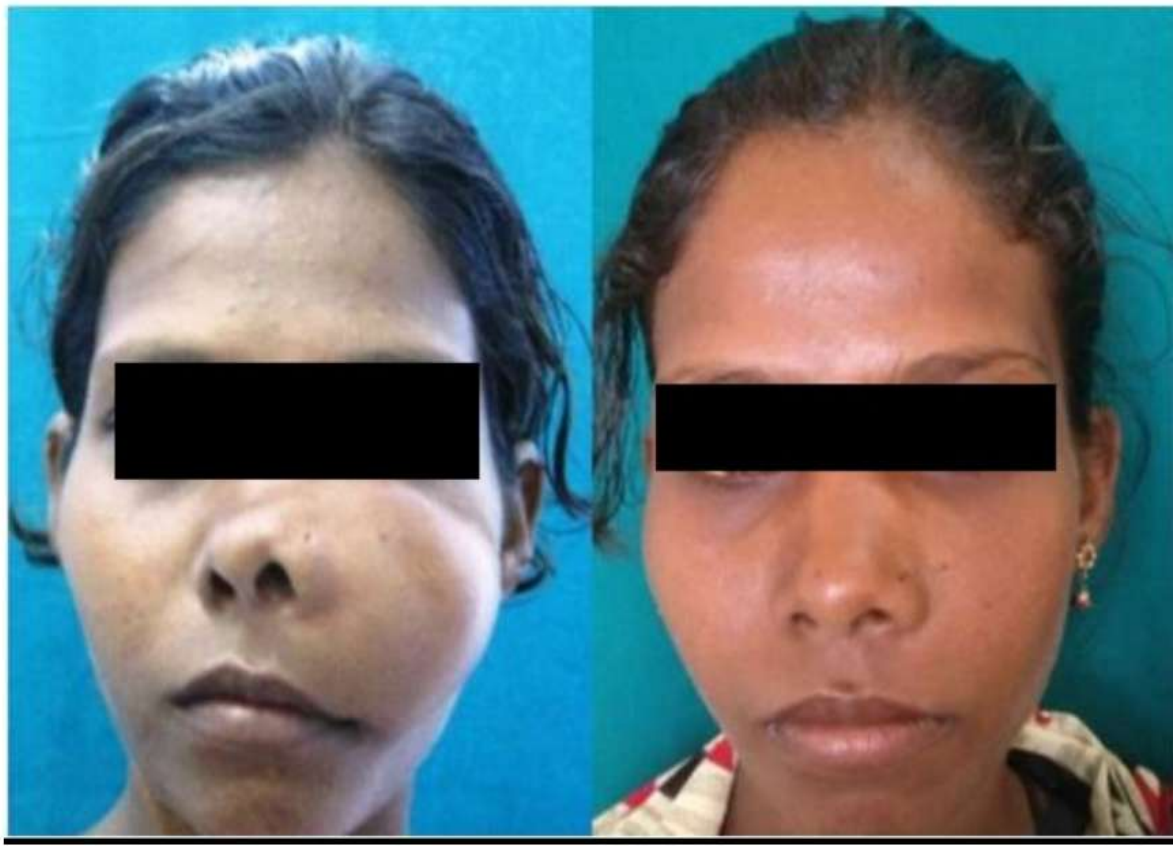

Figure XI: Pre operative and postoperative photograph after one year.