BILATERAL UNICYSTIC AMELOBLASTOMA - A CASE REPORT

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Unicystic ameloblastoma, a variant of solid or multicystic ameloblastoma is commonly reported to occur in third decade and encountered in the posterior mandible with a biologically low-grade course and with limited recurrence potential. Clinically, it frequently manifests as a painless swelling, which can be accompanied by facial deformity, malocclusion and paresthesia of the affected area.Histologically, the minimum criterion for diagnosing a lesion as unicystic ameloblastoma is the demonstration of a single cystic sac lined by odontogenic(ameloblastomatous) epithelium often seen only in focal areas whereas radiographically they represent a unilocular radiolucency.This report presents a case of 28 year old male patient with bilateral occurrence of unicystic ameloblastoma.

Key words: Unicystic ameloblastoma, luminal, intraluminal, mural ameloblastoma.

INTRODUCTION

The concept of unicystic ameloblastoma was first introduced by Robinson and Martinez in 1977[1] and this was variously termed as mural, monocystic, intracystic, cystogenic, or cystic ameloblastoma[2]. Unicystic ameloblastoma occurs at a mean age of 22 years and is located commonly in the mandible (mandible to maxilla=3 to 13:1) with female: male preponderance of approximately 1:1.3[1]. Unicystic ameloblastoma occurs in a relatively younger age group than does its conventional counterpart[3]. The simultaneous occurrence of ameloblastomas with odontogenic cysts or other non-odontogenic lesions have already been described as combined lesions[4] but the term unicystic is derived from the macro- and microscopic appearances. This lesion being in essence a well-defined, single cystic sac lined by odontogenic (ameloblastomatous) epithelium, whereas unilocular, on the other hand, is a term used in radiographic interpretation of a radiolucency having only one loculus or compartment. The subtypes of Unicystic Ameloblastoma has been named intracystic, luminal, intraluminal or mural ameloblastoma[2]. The importance of the unicystic ameloblastoma is that it possesses a much better prognosis after enucleation or curettage than does the classic intraosseous ameloblastoma[2].

We report one such case involving bilaterally premolar- molar region of mandible in a 28 year old male patient.

Case presentation

A 28 year old male patient visited the Department of Oral Pathology and Microbiology, Kothiwal Dental College and Research Centre, Moradabad with the chief complaint of pus discharge from right lower back teeth region since 10 days.

On examination, extraorally,a smooth and diffuse swelling was located in right lower cheek region causing facial asymmetry ,measuring approximately 3x 4 cm in dimensions and extending from angle of mouth to mid body region antero posteriorly and ala of nose to lower border of mandible supero inferiorly. This was associated with dull pain and was tender on palpation. (Fig1). Intra orally, the swelling was present in 45 -47 region, measured approximately 3x4 cm, was smooth surfaced and an open socket was also evident in the same region which had rolled out and erythematous borders. This was filled with yellowish necrotic material, was associated with pus discharge and foul odour. (Fig 2). An orthopantomographic evaluation revealed a unilocular radiolucency in 45-47 region with

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Fig 1. Clinical Presentation (Extra orally)

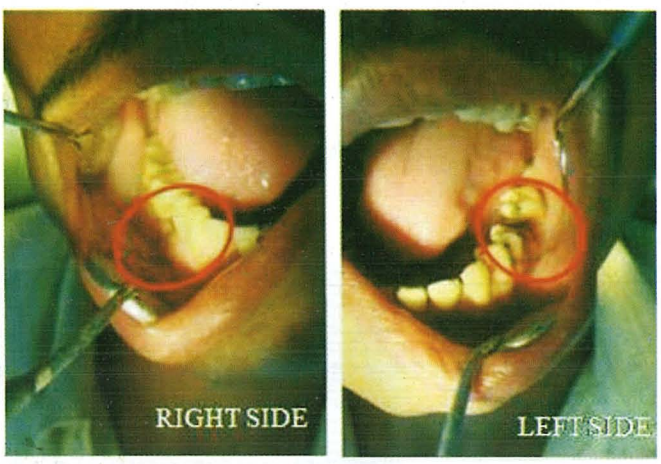


Fig 2. Clinical Presentation (Intra orally)

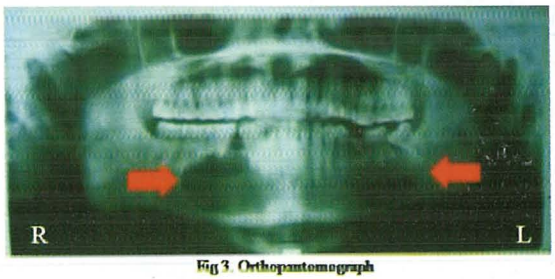


Fig 3. Orthopantomograph

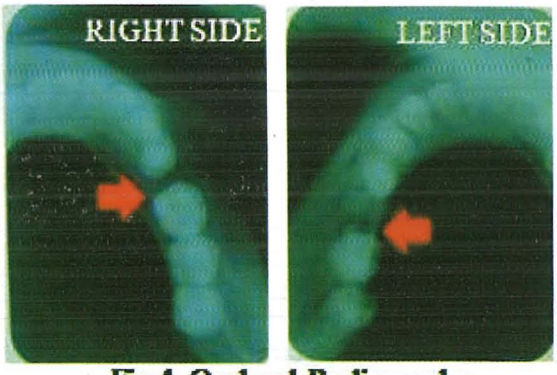


Fig 4. Occlusal Radiographs

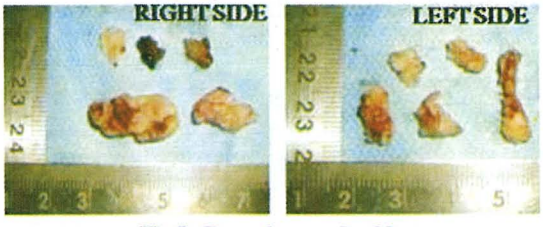


Fig 5. Gross tissue after biopsy

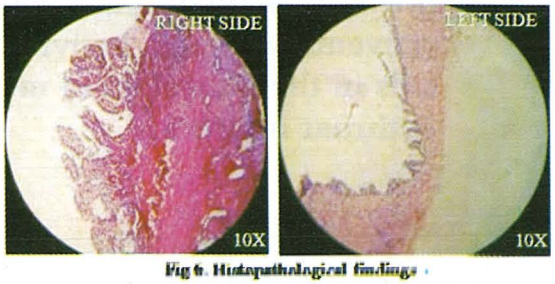
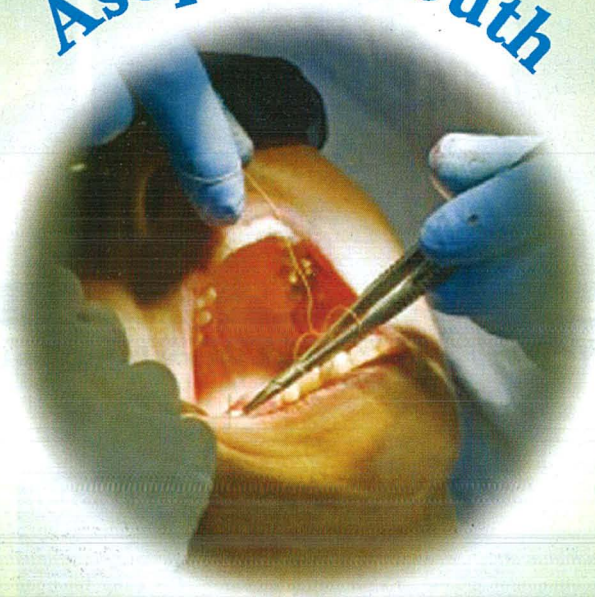


Fig 6. Histopathological findings

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slightly scalloped radiopaque margins.However, a concomitant unilocular radiolucency was also present in 35-37 region measuring roughly 3x3 cm radiographically (Fig 3). The occlusal radiographs revealed expansion of buccal cortical plate on right side only (Fig 4). Based on the above mentioned findings,a differential diagnosis of bilateral odontogenic keratocyst, radicular cyst and ameloblastoma were made. The aspirate from both the sides were taken and micro protein estimation values using semiautomatic analyser for right and left sides were 8.6 and 11.08 gm% respectively which ruled out the possibility of odontogenic keratocyst . Following this, enucleation on either side was performed in the Department of Oral and Maxillofacial Surgery, the specimens were labeled, fixed in 10% neutral buffered formalin and appeared as multiple soft tissue bits which were yellowish white in colour and variable in dimensions. (Fig 5). The tissues were processed for routine Haematoxylin and Eosin staining and the stained sections on right side revealed the presence of 2-3 layered epithelium overlying highly inflamed fibrocellular connective tissue stroma.The epithelial lining showed papillomatous projections of odontogenic (ameloblastomatous) epithelium at focal areas which extended into the lumen extensively. The basal layer was composed of low columnar cells displaying hyperchromatic, palisaded nuclei, Suprabasal cells revealed reduced numbers of intercellular contacts and appeared edematous whereas the surface cells had a pulled up appearance with partial separation from the outer layers. The connective tissue stroma appeared fibrous with abundant fibroblasts, diffusely infiltrated odontogenic epithelial strands with increased proliferative activity, chronic inflammatory cells, a few foamy macrophages and endothelium lined blood capillaries with extravasated RBCs.The overall histopathological features were suggestive of ***inflamed intraluminal unicystic ameloblastoma on right side.*** On the left side the H & E stained sections revealed the presence of a large cystic cavity with a lining composed focally of ameloblastomatous cells.The cells of these luminal proliferations appeared heaped upto 7-8 layers, swollen and were loosely cohesive.The connective tissue had similar findings as on the right side except for negligible inflammatory response.The overall histopathological features were suggestive of ***intraluminal unicystic ameloblastoma on left side.***

The clinicopathological correlation of the findings were conclusive of a final diagnosis of ***Bilateral Unicystic Ameloblastoma*** of mandibular body region.The patient was kept on periodic follow up for survillience of recurrences.

DISCUSSION

Philipsen & Ricchart (1998) concluded that ameloblastic epithelium was considered to be present in unicystic ameloblastoma when any of the following, either singly or in combination, was present (1) a lining epithelium in which the basal cells were clearly columnar, with hyperchromatic nuclei and the overlying cells were only loosely textured with the absence of 'cohesiveness'. This separation of the suprabasilar cells could not be explained on the basis of inflammatory oedema. These criteria coincide very closely with those selected some years earlier by Vickers and Gorlin as representative of early ameloblastoma change. (2) Downgrowth of ameloblastic epithelium described into the connective tissue portion of the cyst wall. (3) The presence within the connective tissue portion of the cyst wall of islands composed of a periphery of columnar epithelial cells and a centre identical to stellate reticulum.(4) Intraluminal nod ules composed of anastomosing cords and islands of epithelium[1]. Leider et al has proposed three pathogenic mechanisms for its evolution :(1) the reduced enamel epithelium associated with a developing tooth undergoes ameloblastic transformation with subsequent cystic development;(2) ameloblastomas may arise in a dentigerous or other type of dental cyst in which the neoplastic ameloblastic epithelium is preceded temporarily by a non neoplastic stratified squamous epithelial lining; (3) a solid ameloblastoma undergoes cystic degeneration of

ameloblastic islands with subsequent fusion of multiple microcysts to develop a unicystic lesion[1]. The second hypothesis is most accepted and favours for the case.

Unicystic ameloblastoma most commonly manifest facial disfigurement with noticeable swelling which does not corroborate with the clinical presentation of the lesion on left side in our case. However, on right side clinically significant symptomatic presentation of the lesion coincides with the findings explained by Philipsen & Riechart (1998) along with the male preponderance as reported by Olaitan et al (1997).

The most common site for the occurrence of unicystic ameloblastoma in the mandible is the third molar area but Olaitan et al (1997) in the study on unicystic ameloblastoma confirmed that mandibular body region is not an uncommon site for its occurrence and this strengthened the findings of our case.However, its presence BILATERALLY could not be linked to any such case mentioned in English literature to the best of our knowledge.

Ronsenstein et al (2001) reported that the histology of cystic ameloblastomas is deceptively bland with basal palisading and intercellular edema[5] which is very similar to the findings reported.

Yunus et al (2009) reveals that although cystic ameloblastomas follow a biologically low-grade course, recent evidence suggests that they may often behave clinically as biologically aggressive tumors. This is supported by the high incidence of cortical perforation, tooth resorption, lesion size, bony destruction, and a high rate of recurrence after simple enucleation[6].

To conclude, the diagnosis of unicystic ameloblastoma is based on two features. First, the lesion must be unilocular, clinically and radiologically. Second, on microscopic examination it must appear as a single cystic lesion with the epithelial lining consisting of ameloblastoma[2]. Unicystic-type ameloblastoma are generally removed as a dentigerous cyst without preoperative biopsy, and all of the tissue must be included for a proper diagnosis[7]. Its recurrence rate after these procedures is around 15%, whereas recurrence is much higher after curettage of classic intraosseous ameloblastoma[2]. In case of a unilocular cystic lesion noted within maxilla or mandibular body, aspiration is followed by enucleation and excision, but if mural type is reported, partial maxillectomy or marginal mandibulectomy should be performed[8].

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