

Unicystic Ameloblastoma of Mandible with all the Variants Mimicking a Cyst: A Case Report

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Accepted on 08.09.2012

Abstract

Unicystic ameloblastoma is a rare variant of ameloblastoma. We report a case of Unicystic ameloblastoma in a 29 year-old female with a pain and swelling in the left mandibular posterior region. Fine needle aspiration yielded no fluid. Periapical, panoramic and computer tomography scans showed well defined radiolucency present in relation to 36 and 37. Unicystic refers to those cystic lesions that show clinical, radiographic, or gross features of a cyst, but on histologic examination show a typical ameloblastomatous epithelium lining part of the cyst cavity, with or without luminal and/or mural tumor growth. Enucleation and Tumor resection was performed for the treatment. Our case report is an attempt to help the dental community in developing familiarity with the clinical presentation and at the same time advocating to develop a high index of suspicion in recognizing such cases.

Keywords: Unicystic Ameloblastoma; Odontogenic tumor; Histopathology.

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Introduction

Many benign lesions cause mandibular swellings and these can be divided into odontogenic and nonodontogenic origin. These

lesions include ameloblastoma, radicular cyst, dentigerous cyst, keratocystic odontogenic tumour, central giant cell granuloma, fibro-osseous lesions and osteomas [1].

The most common tumor of odontogenic origin is ameloblastoma which develops from epithelial cellular elements and dental tissues in their various phases of development [2].

It is a slow-growing, persistent and locally aggressive neoplasm of epithelial origin. Its peak incidence is in the 3rd to 4th decades of life and has an equal sex distribution. It is often associated with an unerupted third molar [3].

The majority of ameloblastomas arise in the mandible, and these are found at the angle and ramus region. There are three forms of ameloblastomas namely multicystic, peripheral, and unicystic tumors [2].

The unicystic ameloblastoma is a well-defined, often large monocystic cavity with a lining, focally but rarely entirely composed of odontogenic epithelium [4]. The unicystic ameloblastoma is considered as a variant of the solid or multicystic ameloblastoma, accounting for 6% to 15% of all intraosseous ameloblastomas [5]. More than 90% are located in the mandible [1]. This tumor has less aggressive biologic behavior and lower recurrence rate than the classic solid or multicystic ameloblastoma [6]. Impacted mandibular third molars are even more frequently associated with UA with figures

Figure 1: Extraoral photograph of the patient



Figure 2: Intraoral laceration with gingival swelling



ranging from 52 to 100% [7]. Unicystic tumors include those that have been variously referred to as mural ameloblastomas, luminal ameloblastomas, and ameloblastomas arising in dentigerous cysts [8]. Although the unicystic ameloblastoma is a “cystic” appearing lesion on gross examination and microscopic examination shows the presence of ameloblastoma within the cyst wall [6]. Here, we present a case of a large unicystic mandibular ameloblastoma in a 29 year old female.

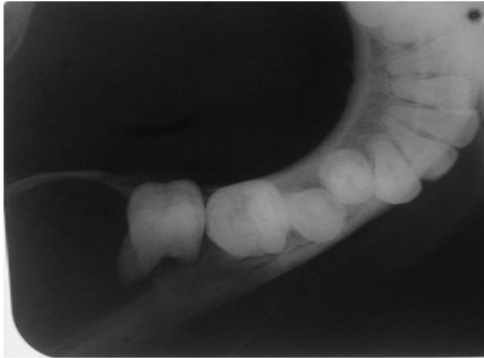
Case Report

A 29-year-old woman presented to the Department of Oral Medicine and Radiology, with pain and swelling in the lower left back region of the face since 3 months. There was

Figure 3: Panoramic radiograph showing a well defined radiolucency with corticated borders and segmented appearance



Figure 4: Mandibular occlusal radiograph showing multilocular appearance and buccal cortical expansion



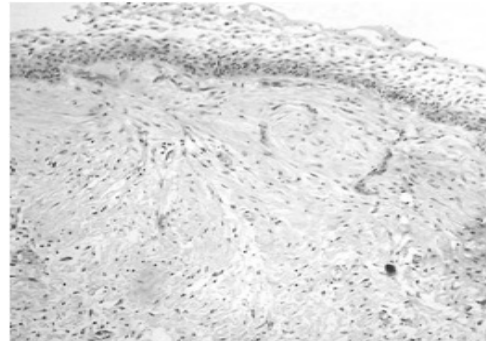
no history of trauma and her past dental/medical history was unremarkable. All vital signs were within normal limits.

The physical examination revealed facial asymmetry due to swelling on the lower left back region of face. On extra-oral examination, a single diffuse swelling was seen near the angle of left side of mandible. The swelling extended 3 cm from the angle of mouth to the posterior border of ramus anteroposteriorly and 4cm below the inferior border of left orbit to the inferior border of mandible superoinferiorly. The swelling was approximately 2x1 cm in size, well defined, oval in shape and had a smooth surface. The overlying skin was of same color as that of adjacent skin and was not associated with any of the secondary changes. There was no bleeding and pus discharge (Figure 1). On palpation, temperature of the overlying skin was same as that of adjacent

Figure 5: CT shows unilocular expansile lesion with the area of perforation in cortex



Figure 6. Luminal UA shows cystic wall lined by ameloblastic epithelium



skin. The swelling was diffuse, firm, tender, non compressible and non reducible over the left side of the mandible. It was fixed to the underlying structures. Submandibular lymph nodes of left side were palpable, non-tender and not fixed to underlying structures.

On intraoral examination, a single linear laceration and gingival swelling was seen on the left buccal mucosa in relation to 37 and 38 region (Figure 2). On palpation, the gingiva around 37 and 38 region was firm and non tender. The teeth in the affected area was sensitive to percussion but no mobility could be demonstrated.

The patient was then subjected to the radiographic examination. The panoramic radiograph revealed a well defined radiolucency in relation to 36 and 37 extended from distal aspect of 37 upto the retromolar area antero-posteriorly and from the crest of the alveolar ridge to the lower border of mandible superoinferiorly. It was well defined,

Figure 7: Intraluminal UA shows proliferation of epithelium into lumen

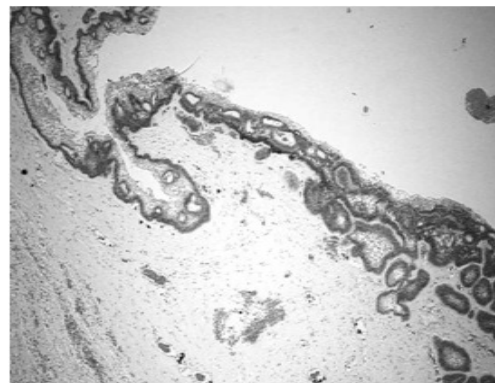
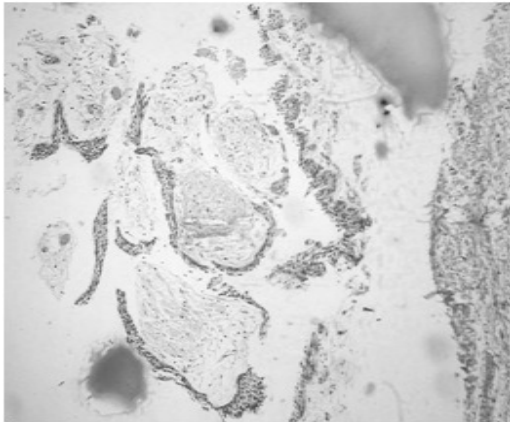


Figure 8: Mural UA shows invasive islands of ameloblastic epithelium in CT wall



corticated and had segmented appearance. Resorption of roots in relation to 36 and 37 was also seen. Impacted third molar was also seen on the left side of mandible (Figure 3). In the occlusal radiograph, the expansion of lingual cortical plate was evident. (Figure 4).

Computed tomography of the lesion showed a large unilocular expansile lesion of size 3.5x2.5x4.5cm was seen in relation to alveolar process of left side of mandible. Both buccal and lingual plates were thinned out with the area of perforation in cortex. Impacted third molar was also seen within the lesion (Figure 5).

Based on the clinical and radiological appearance, a provisional diagnosis of dentigerous cyst was made. Aspiration of the lesion was non-productive and a complete hemogram showed all the values within the normal range. An incisional biopsy was performed under local anaesthesia to establish a definitive diagnosis.

Histologically, it showed cystic wall lined by ameloblastic epithelium which in areas shows columnar basal cells with hyperchromatic nuclei, nuclear palisading with polarization and cytoplasmic vacuolation with intercellular spacing, thin layer of stellate reticulum like cells (Figure 6). The ameloblastic epithelium which is thickened at spaces showed papillary projections (Plexiform), extending into the lumen (Figure 7). The cystic stroma in one area shows an ameloblastic follicle with central

acanthomatous change (Figure 8). All these features were consistent with unicystic ameloblastoma. Patient had subjected to enucleation and tumor resection was done.

Discussion

The term Ameloblastoma was suggested by Churchill in 1934 [4]. There are almost upto fifteen different types of this tumor recorded till date. The most commonly occurring varieties of this tumour histologically are follicular, plexiform, granular, desmoplastic, basal cell, unicystic and the lesser occurring peripheral variant [4].

Unicystic ameloblastoma is a rare type of ameloblastoma, accounting for about 6% of ameloblastomas [2]. The concept of this tumor was first introduced by Robinson and Martinez in 1977 [9]. About 50% of the cases occur in the second decade of life [1]. Mandible is affected more than maxilla. They are most commonly encountered in the posterior mandible followed by the parasymphysis region, anterior maxilla, and the posterior maxilla [10]. Between 50 and 80% of cases are associated with tooth impaction, the mandibular third molar being most often involved [1]. Clinically and radiographically, the unicystic ameloblastoma often has the appearance of a dentigerous cyst [5]. The radiographic appearance is peculiar with the association of a circumscribed radiolucency with the crown of a tooth. The margins are well delineated, with well decorticated margins present in most of the cases [5]. In our case, tumor was associated with impacted mandibular third molar and consistent with all findings in the literature. A confirmatory diagnosis of unicystic ameloblastoma is made by histopathological evaluation of biopsy specimens. The following features are usually observed during microscopic examination.

Ackermann *et al* classified this entity into 3 histologic groups:

Group 1- Luminal UA lesions consist of a unilocular cyst lined by epithelium that in areas

shows ameloblastic transformation without infiltration into the connective tissue wall.

Group 2- Intraluminal/plexiform UA lesions consist of a unilocular cyst with the lining epithelium showing a nodular proliferation of plexiform ameloblastoma into the lumen without infiltration of tumor cells into the connective tissue wall.

Group 3- Mural UA lesions have invasive islands of ameloblastomatous epithelium in the connective tissue wall, that may or may not be connected to the cyst lining epithelium [5]. Our case was consistent with all the common features reported in the literature.

Odontogenic keratocyst, residual cysts, adenomatoid odontogenic tumor, giant cell lesions and sometimes solid ameloblastoma can be the possible differential diagnoses for a unilocular lesion with or without a 'dentigerous' relationship occurring within the jaws [10].

The treatment is decided by the clinical behavior and which in turn is dictated by the histological pattern of the ameloblastoma [10]. In cases of the luminal, intraluminal or plexiform pattern, enucleation generally suffices but if there is a mural component, bony resection is necessary to ensure adequate removal [1].

Conclusion

The unicystic ameloblastoma is characterized by specific clinical, imaging, and histological features. For proper understanding of such cases, more in depth analysis and long term follow up is required. The clinician has to be alert regarding the unusual presentation of this neoplasm and should include unicystic ameloblastoma as differential diagnosis in any lesion ranging from simple abscess to any fibro-osseous lesions/neoplastic growth presenting in posterior mandible. The definite diagnosis requires histopathological examination. Also

with the potential for recurrence, such cases should always be treated by complete resection.

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