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CASE STUDY

NON DENTIGEROUS VARIANT UNICYSTIC AMELOBLASTOMA WITH MULTILOCULAR RADIOLUCENCY- A RARE CASE REPORT

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ABSTRACT

Unicyclic ameloblastoma is a rare, benign tumor of odontogenic epithelium. It refers to those cystic lesions that show clinical, radiographic and gross features of a mandibular cyst but on histological examination show a typical ameloblastomatous epithelium lining the cyst cavity with or without luminal and/or mural tumor growth. Most of the cases are associated with tooth impaction, the mandibular third molar being most often involved. This is the dentigerous variant. Sometimes, UA can occur in the ramus area without a relationship with teeth, the so-called 'Nondentigerous' type. As per the literature, both the dentigerous and non dentigerous variant usually demonstrates unilocular radiolucency, although multilocular features also occur. Hence, it is essential that further studies should be conducted on a large scale in order to know whether both variants i.e. dentigerous and non dentigerous with unilocular and multilocular features can alter the prognosis and treatment plan of the lesion. We present a case of unicyclic ameloblastoma – Nondentigerous type in a 38 year old patient showing multilocular radiolucency.

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INTRODUCTION

Unicyclic ameloblastoma is a rare, benign tumor of odontogenic epithelium. It refers to those cystic lesions that show clinical, radiographic and gross features of a mandibular cyst but on histological examination show a typical ameloblastomatous epithelium lining the cyst cavity with or without luminal and/or mural tumor growth (Nagalaxmi et al., 2013; Sandeep S. Gupta et al., 2011; Paikkatt et al., 2007; Nitin Gupta et al., 2011; Shambulingappa Pallagatti et al., 2003; Sudhir S Bhalerao et al., 2011; Zainab Chaudhary et al., 2011). These tumors are classified as mural ameloblastomas, luminal ameloblastomas, and ameloblastomas arising in dentigerous cysts.

(Shambulingappa Pallagatti et al., 2003; Sudhir S Bhalerao et al., 2011; Zainab Chaudhary et al., 2011; Rakesh S Ramesh et al., 2011) It appears more frequently in the younger age group with 50% of cases occurring during second or third decade (Nagalaxmi et al., 2013; Rakesh S Ramesh et al., 2011) with no sexual or racial predilection and 90% located in the mandible in association with impacted teeth especially the mandibular third molars. (Nagalaxmi et al., 2013; Paikkatt et al., 2007; Rakesh S Ramesh et al., 2011) We present a case of Unicyclic Ameloblastoma of the right posterior mandible in a 38-year-old female of the Non Dentigerous variant with multilocular radiolucency.

CASE REPORT

A 38 year old female patient presented with a history of diffuse swelling on the right side of the mandible, perceived 6 months back. The swelling was painful and had been slowly increasing

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in size for 6 months. The patient reported no significant history of trauma. The patient's personnel, family and medical history were non contributory. Extra oral examination revealed the swelling measuring about 2cm x 4cm in size on right mandible extending anterioposteriorly to about 5 cm from the symphysis region and 1cm below the lower border of the mandible. The swelling is firm in consistency and tender on palpation. Bilateral cervical lymph nodes were non palpable (Figure 1).



Fig. 1. Clinical photograph demonstrating diffuse swelling on the right side of the mandible

Intra oral examination at that time revealed the swelling extending from the mesial surface of right second premolar region to distal surface of second molar, obliterating the lingual vestibule. The regional teeth 35, 36 and 37 were non tender and not mobile (Figure 2).



Fig. 2. OPG demonstrating a well delineated multilocular radiolucency extending from the distal aspect of 33 - 37 region

Orthopantomograph revealed a well delineated multilocular radiolucency extending from the distal aspect of 33 - 37 region with root resorption of 34, but no displacement of the regional teeth. The inferior border of the mandible is intact. The cortices of mandibular canal also are intact (Figure 3). Blood investigations were requested, which rendered normal results.

Since there was no much expansion of the mandible, a provisional diagnosis of Odontogenic Keratocyst was made and an incisional biopsy was performed under local anaesthesia.

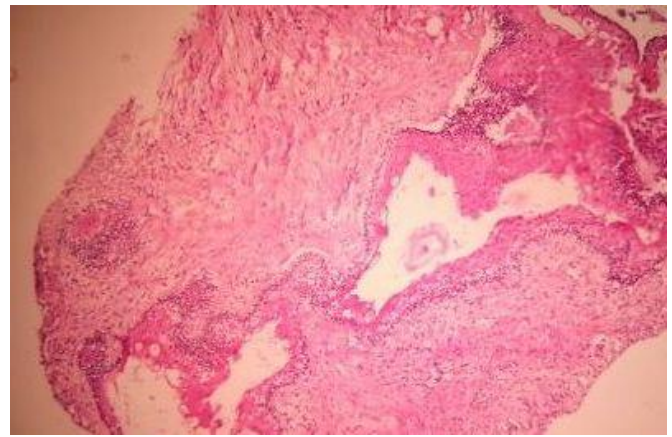


Fig. 3. H&E section demonstrating cystic cavity/lumen having an ameloblastomatous epithelium with hyperchromatic polarized basal layer

Sections from the biopsied specimen stained with Hematoxylin and Eosin revealed a cystic cavity/lumen having an ameloblastomatous epithelium with hyper chromatic polarized basal layer. The underlying connective tissue stroma revealed few islands of ameloblastic follicles with peripheral tall columnar cells and central stellate reticulum like cells (Figure 4).

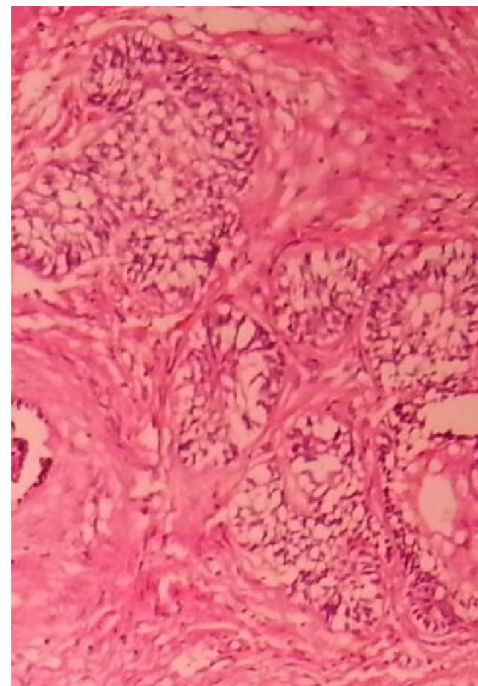


Fig. 4. Connective tissue stroma reveals few islands of ameloblastic follicles with peripheral tall columnar cells and central stellate reticulum like cells

On higher magnification, ameloblastic lining composed of columnar basal cells in palisading pattern with vacuolated cytoplasm and hyper chromatic nuclei polarized away from the basement membrane was evident, hence consistent with the classic Vicker's Gorlin (V & G) criteria (Figure 5), thus leading to the diagnosis of Unicystic Ameloblastoma Subgroup 1.3 (luminal and intramural).

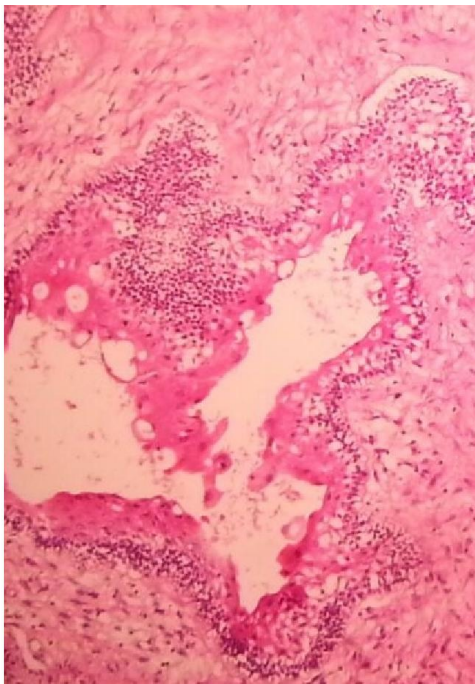


Fig. 5. Higher magnification demonstrating Vickers Gorlin criteria with ameloblastic lining composed of columnar basal cells in palisading pattern with vacuolated cytoplasm and hyperchromatic nuclei polarized away from the basement membrane was evident

DISCUSSION

Ameloblastoma is a benign, locally aggressive, slow growing, persistent odontogenic neoplasm of epithelial origin that is the second most common odontogenic tumor, the first being odontoma (Sudhir S Bhlerao *et al.*, 2011; Zainab Chaudhary *et al.*, 2011). According to the World Health Organization 2003, ameloblastomas are classified based on differences in biologic behaviour, treatment plan and recurrence rate into the following types: conventional - solid / multicystic, unicystic, and peripheral and desmoplastic including the so called hybrid lesions (Nagalaxmi *et al.*, 2013).

Unicystic ameloblastoma, first introduced by Robinson and Martinez in 1977 (Sandeep S. Gupta *et al.*, 2011; Zainab Chaudhary *et al.*, 2011; Mohammad Yunus *et al.*, 2009) is one of the least encountered variant of the ameloblastoma accounting for about 10-15% of all ameloblastomas. (Paikkatt *et al.*, 2007; Zainab Chaudhary *et al.*, 2011; Mohammad Yunus *et al.*, 2009) The term unicystic is derived from the macro and microscopic appearance, the lesion being essentially a well defined, often large monocystic cavity with a lining, focally but rarely entirely composed of odontogenic ameloblastomatous epithelium. (Sandeep S. Gupta *et al.*, 2011; Peter A Reichart and Hans P Philipsen, 2004) The most common age of occurrence of this cyst is the second or third decade with no sexual or racial predilection. It is almost exclusively encountered asymptotically in the posterior mandible (Nagalaxmi *et al.*, 2013; Paikkatt *et al.*, 2007; Zainab Chaudhary *et al.*, 2011; Nadendla, 2012) It is believed to be less aggressive and hence can be treated by conservative surgery than the solid or multicystic ameloblastomas. (Nagalaxmi *et al.*, 2013; Zainab Chaudhary

et al., 2011) Between 50 and 80% of cases are associated with tooth impaction, the mandibular third molar being most often involved. This is the dentigerous variant. Sometimes, unicystic ameloblastomas can occur in the ramus area without a relationship with teeth, the so-called 'Nondentigerous' type. In our case, it demonstrated a multilocular honeycomb type of radiolucency without an association with impacted teeth – non dentigerous type. Since the lesion slowly enlarges, a slight, nontender swelling becomes evident on clinical examination. This swelling is the result of an expansion of the cortical plates of the jaw. It can be identified by palpation as hard and bony. (Mohammad Yunus *et al.*, 2009) In the present case, there was no much bony expansion and no involvement of the impacted third molar, hence classified as Non dentigerous type.

The pathogenesis of this tumor and cystic degeneration are enigmatic to understand. However, it has been suggested that the reason why some ameloblastomas become completely cystic may be due to:- epithelial dysadhesion (e.g. defective desmosomes) or the intrinsic production of proteinases (e.g. metalloproteinases, serine proteinases), or enzymes that normally degrade the central zone of the enamel organ during odontogenesis. (Mohammad Yunus *et al.*, 2009) In 1985, Leider *et al.* proposed three pathogenic mechanisms for the occurrence of UA. Firstly, the reduced enamel epithelium which is associated with a developing tooth undergoes ameloblastic transformation with subsequent cystic development.

Secondly, ameloblastomas arise in dentigerous cysts or in others in which the neoplastic ameloblastic epithelium is preceded temporarily by a nonneoplastic stratified squamous epithelial lining. Finally, a solid ameloblastoma undergoes cystic degeneration of the ameloblastic islands, with subsequent fusion of multiple microcysts and develops into unicystic lesions. (Nagalaxmi *et al.*, 2013; Peter A Reichart and Hans P Philipsen, 2004) The radiographic feature of unicystic ameloblastoma is usually unilocular with well circumscribed area of radiolucency. Though, the fact that the term unicystic would imply a unilocular radiographic appearance, the lesion can sometimes have a multilocular radiographic appearance (Mohammad Yunus *et al.*, 2009) as in our case. Considering these criteria, Eversole and Paikkatt *et al.* (2007) identified six radiographic patterns for this neoplasm- Unilocular, scalloped, macromultilocular, pericoronar, interdicular, or periapical expansile radiolucencies. (Nadendla, 2012) In our case it is macromultilocular with smooth borders.

Histologically, the minimum criteria for diagnosing a lesion as unicystic ameloblastoma is the demonstration of a single cystic cavity lined by odontogenic (ameloblastomatous) epithelium often seen only in focal areas. It was classified into three histologic groups as follows:- (Zainab Chaudhary *et al.*, 2011; Rakesh S Ramesh *et al.*, 2011; Stručni Rad, 2012; Badal, 2011)

- Group I: Luminal UA (tumor confined to the luminal surface of the cyst)
- Group II: Intraluminal/plexiform UA (nodular proliferation into the lumen without infiltration of tumor cells into the connective tissue wall), and

Group III: Mural UA (invasive islands of ameloblastomatous epithelium in the connective tissue wall not involving the entire epithelium)

Another histologic subgrouping by Philipsen and Reichart (**Rakesh S Ramesh et al., 2011**) has also been described:

Subgroup 1: Luminal UA

Subgroup 1.2: Luminal and intraluminal

Subgroup 1.2.3: Luminal, intraluminal and intramural

Subgroup 1.3: Luminal and intramural

Based on both these classification, tumors diagnosed as subgroups 1 and 1.2 can be treated conservatively whereas subgroups 1.2.3 and 1.3 showing intramural growths require aggressive treatment with radical resection. In the present case, as it showed intramural growth with ameloblastic follicles into the connective tissue, it was classified as group 3 with subgroup 1.3. The cystic epithelium also demonstrated the classic Vicker's Gorlin criteria demonstrating ameloblastic lining composed of columnar basal cells in palisading pattern with vacuolated cytoplasm and hyperchromatic nuclei polarized away from the basement membrane. Hemimandibulectomy was performed along with bone grafting. The patient is being followed up at regular intervals to check for any recurrences.

In conclusion, unicystic ameloblastoma, presents with a variety of clinical, radiological and histopathological features. Hence, it presents as a challenge both for its diagnosis and treatment. As per the literature, both the dentigerous and non dentigerous variant usually demonstrates unilocular radiolucency, although multilocular features also occur. Hence, it is essential that further studies should be conducted on a large scale in order to know whether both variants i.e. dentigerous and non dentigerous with unilocular and multilocular features can alter the prognosis and treatment plan of the lesion.

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