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RESEARCH ARTICLE

UNICYSTIC AMELOBLASTOMA: A DIAGNOSTIC CONUNDRUM FOR PATHOLOGISTS.

Abelene Maria Durand and Ramesh Kumar. A.

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Abstract

Unicystic ameloblastoma is a rare variant of ameloblastoma, which usually occurs in younger age groups on comparison with conventional ameloblastoma. The aggressiveness of the tumour is reported to be related to the various histological subtypes, the most aggressive being those with luminal, intraluminal and mural proliferation. This variant of the tumour has to be treated usually with radicular resection similar to conventional ameloblastoma due to its clinical course and higher recurrence rate. The diagnosis of the tumour is usually problematic as it shares clinical and radiological features similar to other lesions. In addition to this, confirmatory diagnosis using incisional biopsy is difficult in many cases as the cystic nature of the tumour may not revealed. Here we report a case of unicystic ameloblastoma showing luminal, intraluminal and mural proliferation that was initially diagnosed as follicular ameloblastoma.

Introduction:-

Ameloblastomas are benign odontogenic epithelial tumours which are slow growing and locally aggressive. They are one of the most common odontogenic tumours affecting the posterior regions of the jaws. (Singh et al., 2015) Unicystic ameloblastoma is a rare type of ameloblastoma, accounting for about 6% of ameloblastomas. It usually occurs in a younger age group, with about 50% of the cases occurring in the second decade of life. It includes those that have been variously referred to as mural ameloblastomas, luminal ameloblastomas, and ameloblastomas arising in dentigerous cysts. (Philipsen and Reichart., 2004), (Chana et al., 2004) Patients most commonly present with swelling and facial asymmetry, pain being an occasional presenting symptom. Mucosal ulceration is rare, but may be caused by continued growth of the tumour. (Roos et al., 1994) Here we report a case which was initially diagnosed as follicular ameloblastoma with acanthomatous change and identified as unicystic ameloblastoma with luminal, intraluminal and mural proliferation following segmental mandibulectomy of the lesion.

Case presentation:-

A 43 year old female patient reported with the chief complaint of swelling and pain in the lower right side of the face for the past six months. Further history revealed that the patient had experienced dull and intermittent pain along with swelling and facial asymmetry for the past six months. An incisional biopsy was done elsewhere three months back, a diagnosis of follicular ameloblastoma with acanthomatous change was made and the patient was referred to SRM Dental College, Ramapuram, for further treatment. Patient had no relevant medical history.

Extra oral examination revealed facial asymmetry with a diffuse swelling on the lower posterior right side of the face. Intra oral examination showed the presence of a bony expansion of the jaw extending from 42 to 46. On

Corresponding Author:- Abelene Maria Durand.

palpation of the lesion, the underlying bone was thinned out and compromised in an area with tenderness and no discharge. Radiographic examination revealed a mutilocular radiolucent area extending from 42 upto 46 (Figure 1). By correlating the clinical and radiological features along with the report from the incisional biopsy, surgical resection of the mandible (region involving 42, 43, 44, 45 and 46) was done and the specimen was sent for histopathological examination (Figure 2).

The resected portion of the mandible showed buccal cortical plate expansion and perforation in an area. After creating a window on the buccal aspect, a cystic bag was evident within the bone. A part of the periosteum, the cystic bag along with tissue specimens from different sites within the lesion were processed and examined under the microscope (Figure 3, Figure 4).

Microscopic examination revealed a cystic lumen with odontogenic epithelial lining and a mature connective tissue wall. The epithelium was hyperplastic in most areas consisting of basal cells which were cuboidal or tall columnar with hyperchromatic nuclei placed away from the basement membrane and subnuclear vacuolization. Few layers of stellate reticulum – like cells were evident superficial to the basal cells in the lining. Some areas within the lumen showed epithelial proliferation in a plexiform pattern. Odontogenic epithelial islands along with some areas of the epithelium budding off into the connective tissue was evident (Figure 5, Figure 6). Based on the observed histopathological features, the lesion was diagnosed as Unicystic Ameloblastoma with luminal, intraluminal and mural proliferation.

Figure 1:- OPG showing multilocular radiolucency

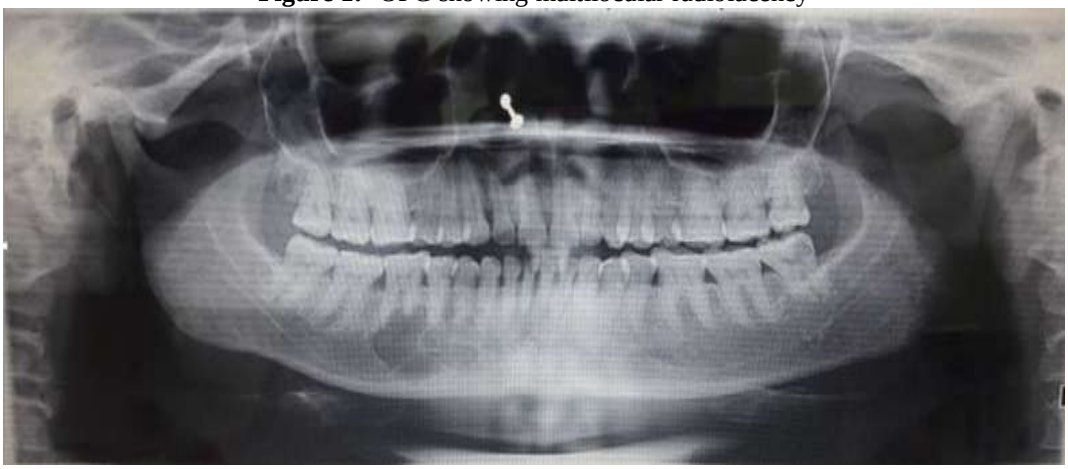


Figure 2:- Resection of the lesional portion of the jaw

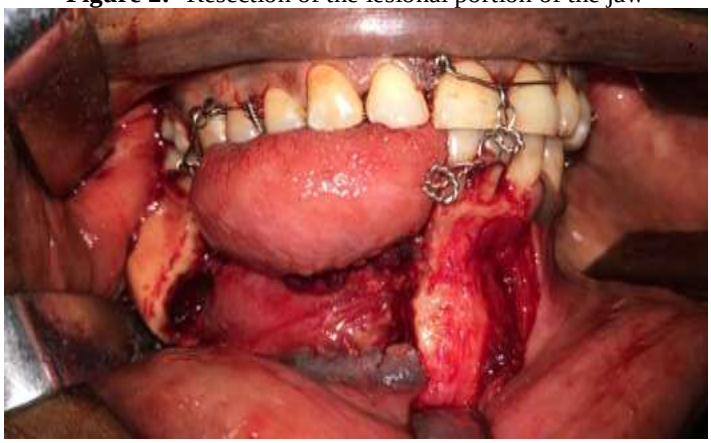


Figure 3:- Buccal cortical expansion

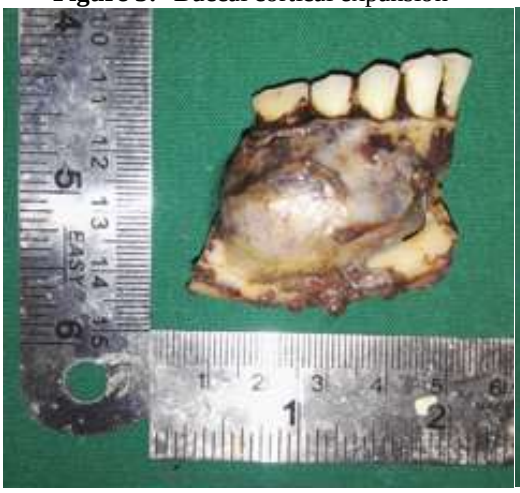


Figure 4:- Cystic bag within the bone

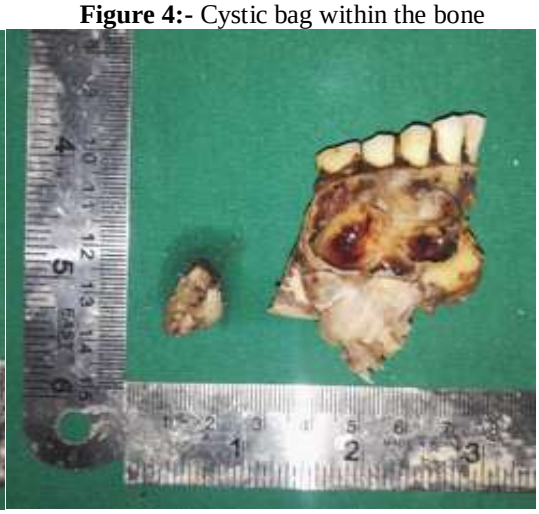


Figure 5:- Odontogenic epithelial lining showing intraluminal and mural proliferation (H&E, 4x)

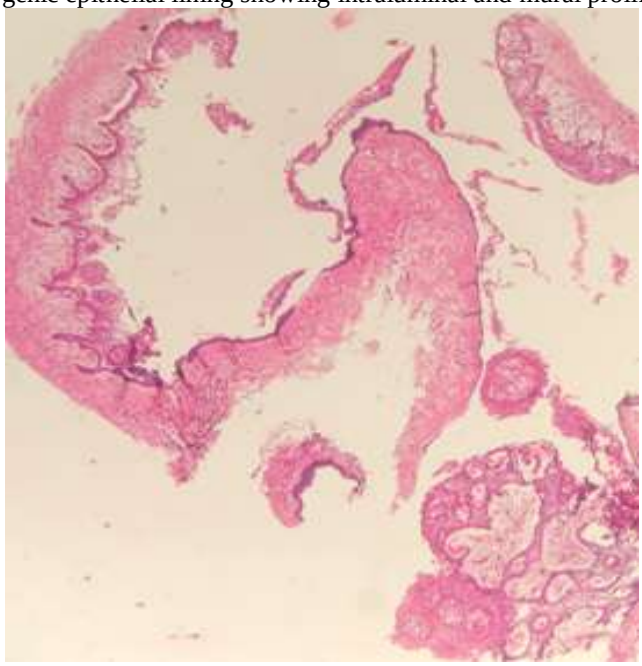
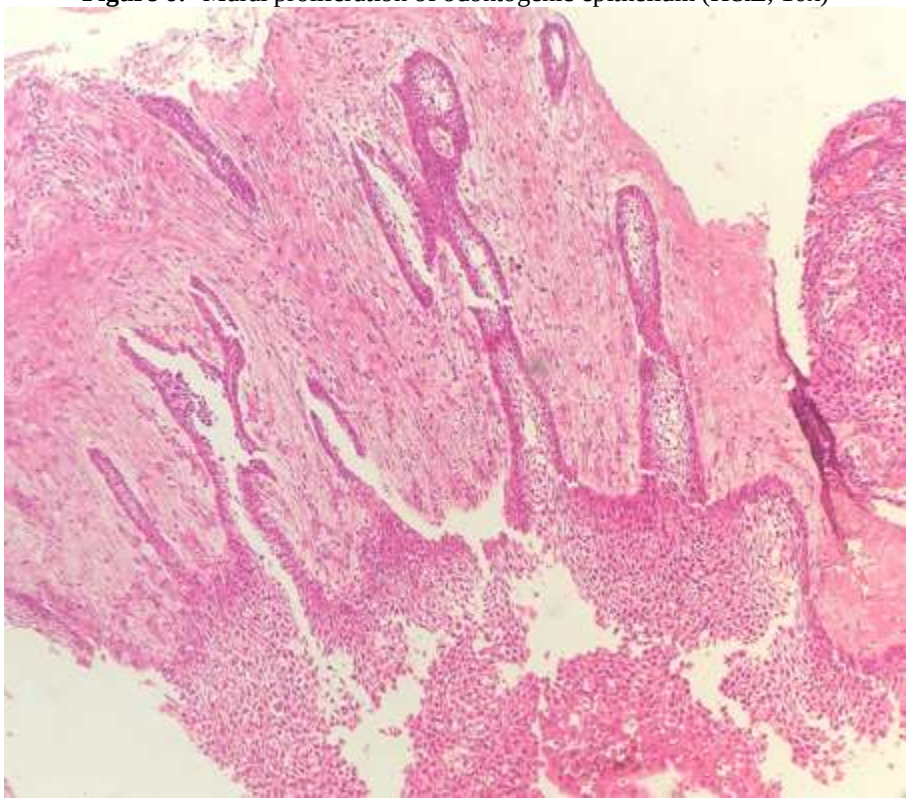


Figure 6:- Mural proliferation of odontogenic epithelium (H&E, 10x)



Discussion:-

Based on the results obtained by studying 57 cases of unicystic ameloblastoma, Ackerman has classified the tumour into 3 histological subtypes.

1. **Luminal Unicystic Ameloblastoma**, where, tumor is confined to the luminal surface of the cyst.
2. **Intraluminal Unicystic Ameloblastoma**, where there is nodular proliferation into the lumen without infiltration of tumour cells into the connective tissue wall.
3. **Mural Unicystic Ameloblastoma**, where there are invasive islands of ameloblastomatous epithelium in the connective tissue wall not involving the entire epithelium. (Philipsen and Reichart., 2004)

Philipsen and Reichart (Philipsen and Reichart., 2004) have also classified unicystic ameloblastoma into the following histological groups, namely,

1. Subgroup 1: Luminal (treated conservatively)
2. Subgroup 1.2: Luminal and intraluminal (treated conservatively)
3. Subgroup 1.2.3: Luminal, intraluminal and intramural (radical resection)
4. Subgroup 1.3: Luminal and intramural (radical resection)

Philipsen and Reichart reviewed 193 cases of unicystic ameloblastoma and divided them into two categories:

1. **Dentigerous variant:-** Unicystic Ameloblastomas associated with an unerupted tooth
2. **Non dentigerous variant:-** Unicystic ameloblastomas lacking an association with an unerupted tooth. (Philipsen and Reichart., 1998)

The non dentigerous variant is more common in older individuals (mean 35.2 years) and also has a female predilection. (Philipsen and Reichart., 1998) Radiologically, unicystic ameloblastomas may be unilocular or multilocular, with the mutlilocular unicystic ameloblastomas being more common in the non dentigerous variant (Eversole et al., 1984) The case discussed here was not associated with any unerupted tooth, thereby deeming it as a non dentigerous variant. The occurrence of the tumour in a female patient as well as a radiologically multilocular lesion reported in this case was in accordance with the findings observed by Philipsen and Reichart. (Philipsen and Reichart., 1998)

The recurrence rate for unicystic ameloblastomas showing luminal, intraluminal and mural proliferation is high and hence has to be treated aggressively. Diagnosing a unicystic ameloblastoma by an incisional biopsy has proved to be problematic in cases where the cystic nature of the lesion is not clearly evident. There have been previous reports where unicystic ameloblastomas have been misdiagnosed as other lesions. (Chaudhary et al., 2011) The tumour reported here was initially diagnosed as follicular ameloblastoma with acanthomatous change and following treatment was identified as unicystic ameloblastoma with intraluminal and mural change. The reason for this possible variation in diagnosis could be attributed to the possibility that the section of tissue which was studied could have been from a superficial area, presumably the connective tissue or it could have been from a proliferation of the ameloblastomatous epithelium into the lumen, thereby eliminating the lining epithelium needed for diagnosis of a unicystic ameloblastoma.

Conclusion:-

Diagnosing a unicystic ameloblastoma preoperatively proves to be difficult in most cases, as it shares clinical and radiological features with other tumours and cysts. An incisional biopsy does not always guarantee a reflection of the true nature of the disease as was seen in this case and can be diagnosed as any other odontogentic tumour or cyst. As the final diagnosis of the case was revealed to be unicystic ameloblastoma with mural, luminal and intraluminal proliferation, surgical resection as a treatment option is justified. However, if the final diagnosis had revealed only a luminal or intraluminal proliferation, the need for resection would have been questionable. Hence, it is strongly recommended that multiple or serial sections are made in order to minimize this error in diagnosis.

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