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Case Report

Plexiform Unicystic Ameloblastoma: A Case Report and Review

DR. Pinhaj A Sherashiya, MDS; [1]DR. Hemavathy S, MDS; [2]DR. Yogesh T L, MDS; [3]DR. Archana T S, MDS. [4]

1, Board; In- charge, Adarsh Oncopath Lab, Palanpur, Gujarat- 385001, INDIA
2, Professor and Head,
3, Assistant Professor,
4, Post graduate student,
2,3,4, Department of Oral and Maxillofacial Surgery, Sri Rajiv Gandhi College of Dental Sciences and Hospital, Bengaluru -32, Karnataka, INDIA

Correspondence: Dr Pinhaj A Sherashiya, MDS; Adarsh Oncopath Lab, Near Gathaman Gate, Palanpur, Gujarat-385001, INDIA. hi-pinhaz@gmail.com

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ABSTRACT

Background and Setting: Unicystic ameloblastoma is a unique variant of ameloblastomas that are commonly seen in younger populations. They show a slow growth rate and yet exhibit a relatively local aggression; with the predominant site of origin being the posterior portion of the mandible. Recurrence, following surgical management, is common, depending on the site of involvement, the histological type and the primary method of treatment range from en-bloc excision to curettage.

The case presented here is of a 16-year-old female patient, who reported to the Department of Oral and Maxillofacial Surgery, with a unilocular radiolucency involving 48 region that was discovered during a routine radiographic examination. The radiolucency measured around 2 x 2 centimetres in size and surrounded the unerupted crown of 48. The lesion was asymptomatic, and a provisional diagnosis of the dentigerous cyst was made. Enucleation of the cyst was done after extraction of 48 under local anaesthesia. On histological examination, the lesional tissue showed cystic areas lined by ameloblastic epithelium and a thin overlying layer of stellate reticulum like cells. The cystic lumen showed large nodular masses of plexiform ameloblastoma confined to the lumen, and an infiltration of ameloblastic epithelium into the connective tissue wall. A confirmatory histopathological diagnosis of unicystic ameloblastoma–mural variant of plexiform type was made. The patient was under observation for 6 months, and there has been no reported recurrence of the lesion.

INTRODUCTION

Unicystic ameloblastoma, a type of ameloblastoma, was first described by Robinson and Martinez. It accounted for 10-15% of all extraosseous ameloblastomas. The origin of the neoplasm was controversial; it was said to originate de-novo or as a result of the ameloblastomatous transformation of the cyst epithelium.

The neoplasm is often seen in young patients, with about 50% of such tumors occurring during the second decade of life. More than 90% of unicystic ameloblastomas are found in the mandible, usually in the posterior region. The lesion is usually asymptomatic, but if large, it may result in a painless swelling of the jaws. [1]

Ackerman has described 3 histopathologic variants of unicystic ameloblastoma: [2]

1. In the **luminal unicystic ameloblastoma**: the tumor is located on the luminal surface of the cyst; the lesion also consists of fibrous cyst wall that is partially or totally lined by an ameloblastic epithelium. The epithelium

comprises of a basal layer of columnar or cuboidal cells with cytoplasmic vacuolisation, and hyperchromatic nuclei that exhibit reversal of polarity. The overlying epithelial cells resemble stellate reticulum and are loosely cohesive. This was said occur due to an inflammatory edema.

2. In the intraluminal unicystic ameloblastoma, one or more ameloblastomatous islands project from cystic lining into the lumen. These islands may be small or may be large enough to fill the cystic lumen. In some cases, the islands of tumor that project into lumen exhibit an edematous, plexiform pattern simulating the plexiform pattern seen in conventional ameloblastomas. These lesions are referred to as plexiform unicystic ameloblastomas. [1]The intraluminal plexiform UA has peripheral epithelial cells that are neither columnar nor hyperchromatic; or even polarised in a reverse manner, and central follicle is made up of squamous or solid cells instead.
3. In the mural unicystic ameloblastoma (MUA) the cystic fibrous wall is infiltrated by typical plexiform or follicular ameloblastomatous islands, whose extent and depth may vary. To confirm a case of MUA multiple sections of the specimen are necessary to rule out the possibility of an ameloblastoma. The diagnosis of unicystic ameloblastoma is based on clinical, histopathologic and radiographic features. Unicystic ameloblastoma has a high chance of recurrence, especially of the typical stellate reticulum cells. This type is said to have a higher recurrence rate and is said to be more invasive, [3]in cases where the ameloblastic islands invade the adjacent tissue. [1]

Philipsen and Reichart have described the following forms of UA:

- Subgroup 1. Luminal UA;
- Subgroup 1.2. Luminal and intraluminal;
- Subgroup 1.2.3. Luminal, intraluminal and intramural;
- Subgroup 1.3. Luminal and intramural. [4]

CASE REPORT

A case report of a 16-year-old female patient, who reported to Department of Oral and Maxillofacial Surgery, for a general check-up of her teeth, has been presented here. Incidentally, a unilocular radiolucency involving 48 regions was identified, during the routine radiographic examination (Figure 1).

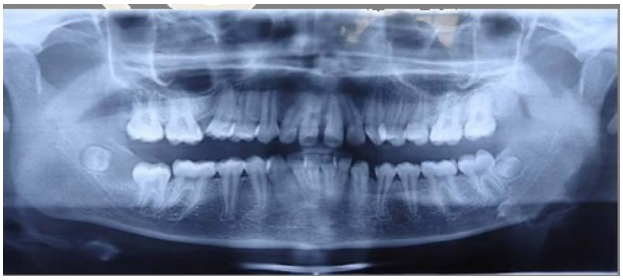

FIGURE 1: *OPG shows a unilocular radiolucency with 48*

Radiolucency was about 2 x 2 centimetres in size and surrounded the crown of the unerupted 48. The patient complained of no associated pain or paraesthesia. A provisional diagnosis of the dentigerous cyst was made. Enucleation of the cyst was done after extraction of 48 under local anaesthesia (Figure 2).

FIGURE 2: *Enucleation site of the lesion seen in relation to 48 regions*

On microscopic examination, hematoxylin and eosin stained section showed lesional tissue composed of cystic areas lined by ameloblastic epithelium, with a basal cell layer composed of columnar cells that were hyper chromatic with a palisaded nuclei. Reversal of polarity of nuclei was seen, and a sub-nuclear vacuole was noted between the basement membrane and nucleus. (Figure3). Within cystic lumen, the proliferation of islands of plexiform ameloblastoma (Figure4); and an infiltration of ameloblastic epithelium into connective

tissue wall as separate islands, which was not in direct continuity with the epithelial lining; was observed. (Figure5). The histopathological impression confirmed a unicystic ameloblastoma – mural variant of plexiform type. The patient was monitored for recurrence during the periodic visits. The lesion showed no recurrence 1 year after removal.

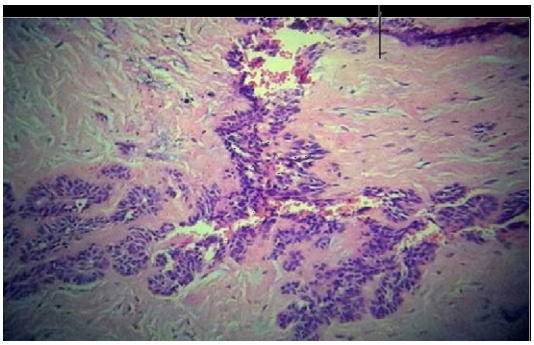

FIGURE 3: *Cystic lining composed of odontogenic epithelium of variable thickness proliferating into the lumen in a plexiform pattern*

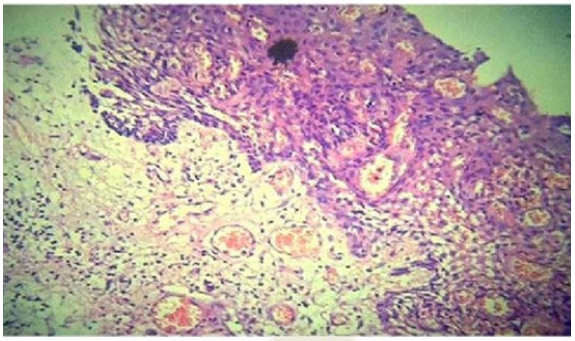

FIGURE 4: *Lining epithelium showing typical ameloblastomatous features*

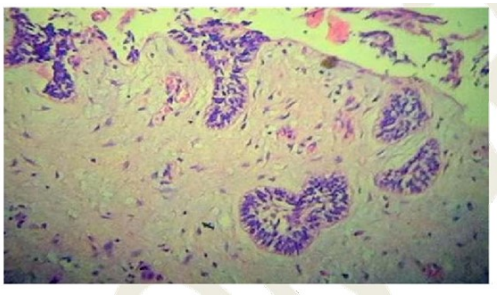

FIGURE 5: *Ameloblastomatous islands in the connective tissue capsule*

DISCUSSION

Ameloblastoma is a considered to be a locally invasive tumor that originates from

- Remnants of the dental lamina and
- Odontogenic epithelium

They comprise only 1 % of all oral tumors. World Health Organization (WHO)'s classification of head and neck tumours stated four forms of ameloblastomas: multicystic, peripheral, desmoplastic and unicystic ameloblastomas.

In 1970, Vickers and Gorlin described Unicystic ameloblastoma, as a lesion that showed alterations in the epithelium. They included:

- Palisading and polarisation of basal cell nuclei of epithelium lining the cystic cavity
- Hyperchromatism of basal nuclei of epithelium
- Cytoplasmic vacuolisation of the basal cells lining the lumen

UA makes up about 6 % of ameloblastomas, and nearly 50 % of the cases were seen in the second decade of life. It was found to affect the mandible more than the maxilla, with a ratio of 13:1. The mandibular-ramus region was predominantly affected, while the lesions in the posterior region of the maxilla, was considered to be rare and atypical. The lesion was seen in inter-radicular, periapical and edentulous regions as well. The response of UA to enucleation or curettage is more favourable than the solid or multicystic types. Robinson and Martinez (1977) and Ackermann et al. (1988), hypothesised that; as the ancestry of the ameloblastomas and odontogenic cysts are common, their epithelium may become neoplastic from a non-neoplastic (14-33%) [one, 3] this is a possibility; though infrequent. [2] But this theory is not applicable; as the patient ages to more than 30 years; the dentigerous cyst epithelium becomes squamous. Histologically the lesion is said to mimic a Keratocystic odontogenic tumor, dentigerous cyst or even a radicular/residual cyst.

Treatment: As the UA is considered to be a less aggressive form of ameloblastoma, a more conservative surgical approach by simple enucleation or any other less aggressive surgical methods, has been recommended.

Stoelinga and Bronkhorst in 1988 suggested the use of Carnoy's solution to minimise the risk of recurrence, after conservative surgical treatment. It has also been advised to avoid vigorous curettage of the bone, as it may plant foci of the lesion, deeper into bone. After the conservative surgical treatment (enucleation or curettage) the recurrence

rate for UA's is reported at 10–20 % and on average, <25 %; much lesser than 50–90 % recurrence rates that are observed after the conventional curettage of solid or multicystic ameloblastomas. Lau and Samman reported a recurrence of 3.6 % for resection and 30.5 % for enucleation; 16 % for Carnoy's solution application after enucleation, and 18 % where treatment was enucleation that followed marsupialisation; in which case the size reduced, after the conventional curettage of solid or multicystic ameloblastomas.

Two main radiographic patterns have been recognised:

- Unilocular- which is similar to a dentigerous cyst and
- Multilocular [2]

According to Eversole and co-workers, there are 6 radiographic presentations:

- Small, pericoronal, unilocular radiolucency- unicystic (>2 cm)
- Large, expansile, unilocular radiolucency extending to the coronoid process
- An expansile radiolucency with scalloped margins
- Large, expansile, periapical radiolucency with root resorption
- Pear shaped radiolucency invaginated between 2 teeth causing root divergence
- Periapical, multilocular radiolucency with root resorption [3]
-

CONCLUSION

As UA has close similarities: both clinically and radiologically; with many odontogenic cysts and tumors, all unilocular radiolucencies of the jaws should be closely observed and examined. The incisional biopsy or the aspiration cytology may fail to reflect the true nature of the lesion, and a long-term follow-up is essential.

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