Plexiform Unicystic Ameloblastoma: A Case Report and Data Analysis in the Indian Population

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Abstract Unicystic ameloblastoma (UA) among the different types of ameloblastomas is least encountered. The reason for lack of perception is paucity of case reports published in the literature. The UA, first described by Robinson and Martinez in 1977, is relatively a rare type of ameloblastoma accounting for about 10–15 % of all ameloblastomas. The term, plexiform UA, refers to a pattern of one or more nodules of ameloblasts like cells projecting from the cystic lining into the lumen of the cyst. Here we present a case of swelling in the anterior mandible of a 25 year old male which on histological examination revealed plexiform UA, which was later treated by complete surgical excision. A tabular data analysis of the previous UAs occurring in the Indian population according to the cases reported by Pubmed has also been included.

Keywords Unicystic ameloblastoma (UA) · Mandible · Plexiform · Indian UAs

Introduction

Ameloblastoma is the most frequently encountered tumor arising from odontogenic epithelium, and is characterized by a benign but locally invasive behavior with a high tendency to recur [1]. Ameloblastoma appears to arise most

frequently from rest of primitive dental lamina that is located in the gingiva, alveolar bone, teeth apices, and the follicular walls of unerupted teeth.

The histological typing of odontogenic tumors by WHO classifies ameloblastomas as intra-osseous/central and extra-osseous/peripheral ameloblastomas. The unicystic ameloblastoma (UA), first described by Chana et al. [2], was reported to have a less aggressive biologic behavior and lower recurrence rate than the classic solid or multicystic ameloblastoma. Prior to the report by Robinson and Martinez, this variant has been referred to as mural or intraluminal ameloblastoma.

Leon Barnes has categorized ameloblastomas into four types based on their behavioral pattern, anatomical location, histological features and radiographic appearance; as solid (multicystic), unicystic, desmoplastic and peripheral varieties. The first three are intraosseous/central, while the peripheral is extraosseous.

Unicystic ameloblastomas are those which have been variously referred to as mural ameloblastomas, luminal ameloblastomas, and ameloblastomas arising from the lining of dentigerous cysts [3]. The six different histopathological variants of ameloblastoma are desmoplastic, granular cell, basal cell, plexiform, follicular, and acanthomatous [3]. One-third of ameloblastomas histologically are of plexiform variety.

In 1977, Robinson and Martinez first coined the term ‘unicystic ameloblastoma’. They considered unicystic variant as less aggressive and suggested simple enucleation as its treatment. UA is commonly seen in second and third decades of life. We report a case of plexiform UA, in a 25 year old male, with its clinical, radiological and histopathological presentation, along with the data analysis of the UAs occurring in the Indian population as reported by the Pubmed (Table 1).

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Table 1 Reported unicystic plexiform ameloblastomas in Pubmed from India till date

S. no.	Authors names	Age/sex	Site	Clinical features	Radiographic features	Histopathological features	Management protocol and max follow-up
1.	Sivapathasundharam and Einstein [3]	28/F	Left Posterior mandible	Diffuse non tender smooth surfaced hard swelling over left posterior aspect of jaw	Large multilocular radiolucency extending from 1st premolar to angle of mandible	Luminal and intramural plexiform epithelial proliferation with typical dentin in connective tissue capsule	Incisional biopsy performed
2.	Mahadesh et al. [4]	46/M	Right mandibular posterior region	Bony hard swelling was extending from lower right 3rd molar region to left canine region	Well defined multilocular radiolucent lesion with sclerotic borders and scalloped margins with root resorption of lower anteriors and molars	Luminal, intraluminal and mural pattern with intraluminal showing plexiform pattern and mural area showing follicular acanthomatous areas co existing with desmoplastic areas	Hemi-mandibulectomy under GA; follow-up of 2 years
3.	Yavagal et al. [5]	13/F	Left mandible	Painful swelling, fluctuant in some areas and hard in some, extending from mesial surface of 34-distal margin of 36	Well defined radiolucency from periapical region of 35–36 to inferior border of mandible	Luminal plexiform pattern with subepithelial hyalinization and odontoblastic rests were seen within fibrous capsule	Extensive enucleation with sub-periosteal dissection under GA
4.	Chacko and Kuriakose [6]	9/M	Right mandible	Pain and swelling in right mandibular area			Complete Surgical enucleation; follow up of 5 years
5.	Acharya et al. [7]	50/M	Mandible			Synchronous emergence of desmoplastic and unicystic ameloblastoma showing follicular or plexiform pattern	–
6.	Our case	25/M	Right mandible	Painless swelling (hard, non-tender), extending from distal margin of lateral incisor to the mesial margin of 1st molar	Well defined radiolucent lesion in right side of mandible	Intraluminal plexiform pattern with few areas showing subepithelial hyalinization, with mature fibrous capsule	Complete surgical excision

Case Report

A 25 year-old male reported to the Hospital with chief complaint of a slowly increasing painless growth in the right lower anterior region of the jaw, since 3 months. The swelling began as an asymptomatic enlargement of the lower jaw. It gradually increased in size to attain the

present size. There was no associated pain, difficulty in mouth opening, chewing or articulating. On physical examination, there was a hard non-tender mass, measuring 6 cm × 3 cm arising on the right side of the mandible, from the distal margin of lateral incisor to the mesial margin of first molar. Teeth associated with the lesion were vital and the overlying oral mucosa and gingiva

Fig. 1 A clinical picture shows a large expansile growth on the right anterior mandible

Fig. 2 Orthopantomogram shows a large unilocular radiolucent lesion on the right anterior mandible

Fig. 3 Gross specimen shows the cystic tissue

were normal. Cervical lymph nodes were non-palpable (Fig. 1).

An orthopantomogram (OPG) revealed large well defined unilocular lesion in the right side of mandible (Fig. 2). The patient was taken up for surgery under local anaesthesia and

complete surgical excision of the lesion was done. The excised mass was then sent for histopathological examination (Fig. 3). Microscopically, sections revealed cystic lumen lined by 3–4 cell layer thick odontogenic epithelium proliferating in the form of plexus and interlacing bundles with columnar basal cells having hyperchromatic nuclei, reverse polarity and cytoplasmic vacuolization. Few areas of focus showed subepithelial hyalinisation. The underlying stroma also revealed endothelial lined blood vessels and extravasated RBCs at few areas of focus. Mature fibrous capsule was also seen (Fig. 4A–C). Histologically the lesion was diagnosed as plexiform UA. The post-operative healing was uneventful. The patient was scheduled for dental prosthesis after 3 months. Patient did not turn up in the department after collection of the histopathology report.

Discussion

The term plexiform UA refers to a pattern of epithelial proliferation arising from cystic lesion of the jaws. It does not exhibit the histological criteria for ameloblastoma published by Vickers and Gorlin, and has therefore been considered by some pathologists to be a hyperplastic epithelial proliferation of the cystic lining rather than true ameloblastoma [8].

Unicystic ameloblastoma (UA) is relatively a rare type of ameloblastoma, accounting for about 10–15 % of all ameloblastomas. UAs are known to be lined by a variable epithelium ranging from typical ameloblastic characteristics to metaplastic which appears completely nondescript consisting of several layers of nonkeratinizing squamous cells. In such cases the differentiation of odontogenic cysts, such as dentigerous cysts from the UA can be challenging [9]. UAs and dentigerous cysts have an identical clinical and radiographic appearance.

Ackerman et al. [10] reviewed 57 cases of UA and found that nearly half of the patients were <20 years of age. Compared to multicystic ameloblastoma, unicystic variety occurs more commonly at younger age as in our patient. In our case the lesion occurred in the mandible body area. UAs have a marked predilection for occurrence in the mandible. In a report by Gardner et al.'s, 46 UA, referred to as plexiform UAs occurred in mandible and not even a single lesion occurred in the maxilla [8]. The ratio of the mandible: maxilla being 13:1. Most of the time ameloblastomas are asymptomatic and are first seen on a routine radiograph. As the lesion slowly enlarges the patient seldom notices it until it becomes obviously disfiguring [10]. Hence most of the patients report at later stage of the disease as in our case.

There are various subtypes of UA depending on the character and extent of ameloblastic proliferation within

Fig. 4 A–C Photomicrographs showing a plexiform unicystic ameloblastoma ($\times 4$, $\times 20$, H/E)

the cyst wall. In a clinicopathologic study of 57 cases of UA, Ackerman et al. [10] classified this entity into the following three histologic groups:

- 1: Luminal UA
- 1.2: Luminal and intraluminal UA
- 1.2.3: Luminal, intraluminal and intramural UA
- 1.3: Luminal and intramural UA

Radiographically, UAs have been divided into two main patterns: unilocular and multilocular. UAs have clear preponderance for the unilocular pattern which was well appreciated in our case. As per the Pubmed records of the published cases of UA and based on our findings in the Indian population the data revealed that UA showed a marked predilection for males as compared to females with the ratio being 4:2. UA has been reported to occur more often in the younger population (3rd decade) [9]. Few cases of UAs in pediatric patients have also been reported [5, 6]. However, based on our findings UA showed a varied age distribution (1st–5th decade). The most frequent site was found to be almost exclusively the mandible, which was consistent with Gardner et al.'s study of 46 examples of histological variants of UA with not one occurring in the maxilla [8]. The clinical findings ranged from a painless hard swelling to painful fluctuant swellings. The radiographic features were consistent in all the cases having a well defined radiolucency. Mahadesh et al. [4] described, in their case, a mixed radiolucent and radio opaque lesion in front of a clearly defined radiolucent lesion which later

turned out to be a hybrid ameloblastoma. The histopathological findings conversely showed a synchronous pattern in which more than one histological variant occurred simultaneously. Varied histopathologies have also been reported. Sivapathasundharam and Einstein [3] reported the presence of hard tissue (dentin) in their reported case of UA. Mahadesh et al. [4] and Acharya et al. [7] reported histopathological diagnosis of UAs alongwith desmoplastic component. The management of UA remains controversial. Some surgeons believe that local resection of the area is indicated as a prophylactic measure whereas others prefer to do a conservative removal of the lesion and keep the patient under close radiographic observation. Mandibular reconstruction and careful prosthetic planning with placement of osseointegrated implants could achieve the successful rehabilitation of patients with enucleation [9]. A recurrence rate of 53 % has been reported in UAs during the first 5 years of the surgery [4].

Conclusion

The basic objective of reporting such a case was to discuss the clinical features, radiographic features and histological features in plexiform UA with an added emphasis on review of literature. According to recent literature these lesions are more aggressive than previously thought. Most of the times the lesion is diagnosed as a dentigerous cyst both clinically as well as radiographically; in our case too, the preoperative

provisional diagnosis of dentigerous cyst was ruled out after the histopathological diagnosis. Hence, it is imperative that examination of the entire lesion histopathologically is mandatory for securing the final diagnosis and consideration should be given to plan a proper treatment.

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