CASE REPORT DENTISTRY

Journal of Contemporary Medicine and Dentistry
www.jcmad.com

ISSN [P-2347-4513]
ISSN [O-2349-0799]
Year: 2021, Volume: 9
Issue: 2, p: 37 - 41

Ameloblastoma – A Diagnostic Challenge

Haris H Haridas [1], Anju Gopinathan [2], Beena Kumar I [3], Fareedi Mukaram Ali [4]

1. Asst. Prof. Department of Dentistry, PK Das Institute of medical Sciences, Ottapalam[1]
2. Reader. Department of Oral and Maxillofacial Surgery, PSM College of Dental Sciences and Research [2]
3. Professor and HOD, Department of Oral medicine and Radiology, Mar Basilios Dental college, Kothamangalam [3]
4. Asst prof, Dept of oral and maxillofacial surgery, College of Dentistry, Jazan University, Jazan [4]

Abstract

Ameloblastoma is a true neoplasm of odontogenic epithelium and is known for its local aggressive nature. There are mainly two varities of ameloblastoma –Intraosseous and Peripheral variety. The unicystic ameloblastoma especially the Plexiform variant is a rare entity that mimics the dentigerous cyst radiographically and makes it a diagnostic challenge. The treatment options for unicystic ameloblastoma consist of conservative and radical methods depending on the histopathology. A rare case of ameloblastoma is presented in this paper.

Keywords: *Unicystic Ameloblastoma (UA), aggressive, mandible*

Address for correspondence: Dr. Anju Gopinathan. T, Reader, Department of Oral & Maxillofacial Surgery, P.S.M College of Dental Sciences and Research, Bypass Road, Akkikavu P.O., Kunnamkulam, Pin: 680519 Kerala, India. E-Mail: anjgo1@gmail.com Mobile: 09497656950

Date of Acceptance: 11/08/2021

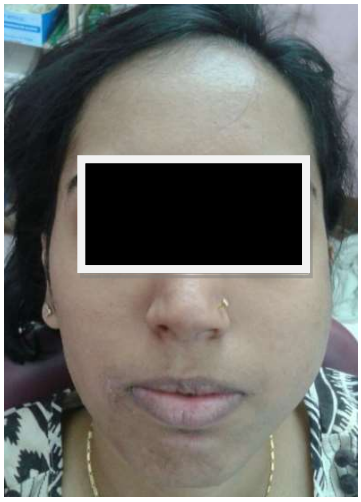
Introduction

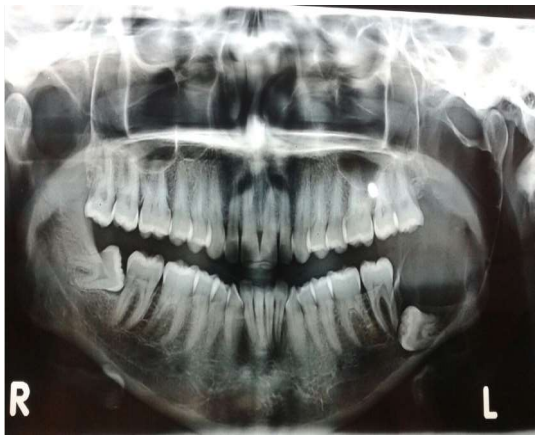
Ameloblastoma is a true neoplasm of odontogenic epithelium. It is an aggressive neoplasm that arises from the remnants of dental lamina and dental organ. Ameloblastoma may occur centrally within bone or peripherally in the soft tissues.[1] Ameloblastomas are usually first recognized between the ages of 30 and 50, being rare in children and old people. It may be slightly more common in men. About 80% form in the mandible; of these,70% develop in the posterior molar region, and often involve the ramus. Lesions are symptomless until the swelling become obtrusive. Radiographically, ameloblastoma may cast a unilocular cyst like radiolucency or a multilocular image with soap-bubble or honeycomb appearance.[2] Unicystic tumors include those that have been variously referred to as mural ameloblastomas, luminal ameloblastomas, and ameloblastomas arising in dentigerous cysts.[3] The clinical and radiologic presentation of Unicystic ameloblastoma can give a confusing picture of odontogenic cysts especially when it is seen in the interradicular or periapical area. Also, dentigerous variety may show features similar to dentigerous cyst. Hence, histopathologic examination is essential to diagnose such cases. [8] Here we present such a lesion which was initially misdiagnosed as dentigerous cyst.

Case Report

A 26 years old female came to our hospital with chief complaint of swelling of left cheek since 1 month. Associated pain or any other symptoms were not present. Her medical history was non-contributory. On examination the swelling was approximately 4x3 cm in size, extending from angle of left mandible to left corner of mouth and to lower border of left mandible superioinferiorly. Intraoral examination revealed diffuse swelling extending from distal aspect of 36 posteriorly.38 and 48 were found to be

missing. Clinical differential diagnosis included Keratocystic odontogenic tumour, Dentigerous cyst, Ameloblastoma. Panoramic radiography showed large radiolucency occupying the left side of the mandible from 37 to the neck of condylar process and coronoid process including the left ascending ramus area extending to the inferior border of the mandible. Also showed impacted 38. Incisional biopsy of the lesion was performed to establish a diagnosis. The biopsy report confirmed the diagnosis as Unicystic Ameloblastoma of Plexiform type. The patient was taken up for surgery under general anesthesia. After patient preparation under sterile conditions, surgical enucleation of the lesion followed by curettage and Carnoy's solution application and extraction of 37 ,38 was done. Follow up was performed at 6 months and 1 year post surgery and revealed no recurrence of the lesion. The patient is still under follow-up.





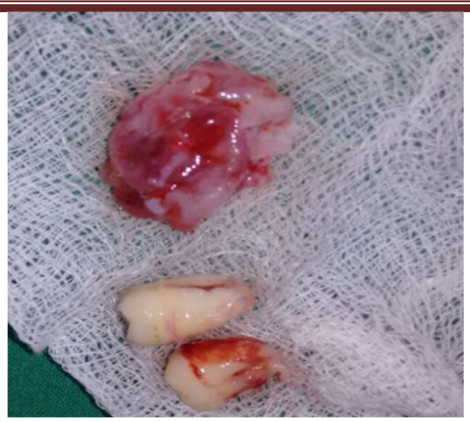


Fig 3: Intra-op

Discussion

The term meloblastoma was suggested by Churchill in 934. [2] Ameloblastomas have been categorized broadly into three biologic variants: cystic (unicystic), solid, and peripheral. This classification has a direct bearing on the pathologic behavior of these variants.[5] The most commonly occurring histological varieties of this tumor are follicular, plexiform, granular, desmoplastic, basal cell, unicystic and the peripheral variant. Unicystic ameloblastoma is a rare type of ameloblastoma, accounting for about 6% of ameloblastomas.[2] Unicystic ameloblastoma (UA) is the second and far less frequent growth pattern seen in the intraosseous ameloblastoma. Robinson and Martinez in 1977 introduced the concept of UA, which was initially called as cystic (intracystic) ameloblastoma, ameloblastoma associated with dentigerous cyst, cystogenic ameloblastoma, extensive dentigerous cyst with intra-cysticameloblastic papilloma, mural ameloblastoma, dentigerous cyst with ameloblastomatous proliferation, and ameloblastoma developing in a radicular or globulomaxillary cyst. 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.[7] About 50% of the cases occur in the second decade of life. The mandible is affected more often than the maxilla. These tumors are most commonly encountered in the posterior mandible followed by the parasymphysis region, anterior maxilla, and the posterior maxilla. Clinically and radiographically, the unicystic

ameloblastoma often has the appearance of a dentigerous cyst. [2] In our case, the lesion showed circumscribed radiolucency associated with impacted third molar and was provisionally diagnosed as dentigerous cyst. However, on histopathological evaluation, confirmatory diagnosis of Plexiform Unicystic ameloblstoma was made based on ameloblastomatous lining of the cystic cavity. On removal of the UA, it is important to examine both the interior and exterior of the cyst sac. Careful macroscopic inspection of the specimen may reveal important diagnostic clues. The inner surface of the cyst (facing the lumen) may show one or several polypoid or papillomatous, pedunculated, exophytic masses, filling the cyst lumen. This subtype of UA has been called intracystic, luminal, or intraluminal ameloblastoma and corresponds to the plexiform UA. In addition to the intraluminal excrescences, the cyst capsule may show one or several rounded and slightly protruding nodules that may also be seen macroscopically when viewing the cyst wall from the outside. These formations have been named mural or intra-mural nodules. UA arises from pre-existing odontogenic cysts, in particular a dentigerous cyst, while others maintain that it arises de novo. Leider et al., proposed three pathogenic mechanisms for the evolution of UA: 1. The reduced enamel epithelium associated with a developing tooth undergoes ameloblastic transformation with subsequent cystic development; 2. Ameloblastomas arise in dentigerous or other types of odontogenic cysts in which the neoplastic ameloblastic epithelium is preceded temporarily by a non-neoplastic stratified squamous epithelial lining; and 3. A solid amelo- blastoma undergoes cystic degeneration of ameloblastic islands with subsequent fusion of multiple microcysts and develops into a unicystic lesion. It is difficult to produce convincing evidence for any of the theories presented.[1]

Ackermann et al [2] classified this entity into 3 histologic groups:

Group 1 - Luminal unicystic ameloblastoma lesions consist of a unilocular cyst lined by epithelium that in some areas shows ameloblastic transformation without infiltration into the connective tissue wall.

Group 2- Intraluminal/plexiform unicystic ameloblastoma 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 unicystic ameloblastoma lesions have invasive islands of ameloblastomatous epithelium in the connective tissue wall that may or may not be connected to the cyst lining epithelium.

Many studies have been done on the various methods which could be used to differentiate UA from odontogenic cysts. The earliest attempts were on the expression of blood cell carbohydrates. Though it was found useful initially, it was later disproved. Imaging studies like the use of contrast enhanced MRI have also been documented. It has been found to be useful as there was thick enhancement in the walls of UA. Histoenzymological studies were done to find out the variations in levels of activities of oxidative enzymes, diaphorases, acid phosphatases and naphthol esters. Odontogenic cysts showed nonspecific activity whereas in ameloblastoma there were uniformly low oxidative enzymatic activities in the epithelium and widespread activity of alkaline phosphatase in the stroma. Thus, alkaline phosphatase activity may be useful in distinguishing the cystic ameloblastomas. A definitive diagnosis of UA can be made only after examining the whole specimen. Hence, incision biopsy may not always be correct as the epithelium shows variation. Thus, multiple sections from the whole specimen should be examined for a final diagnosis.[8] Two types of treatment options have been proposed for the management of ameloblastoma, ranging from curettage to a combination of surgery and radiation therapy. However, curettage is the least desirable of all methods due to its association with a high recurrence rate, and radiation therapy is usually not warranted as the lesion is radioresistant. Diagnosis of unicystic ameloblastoma plays a pivotal role in planning the treatment for a patient. This is owed to the fact that the recurrence rate of this lesion is distinctly lower indicating a less aggressive nature of this variant compared to the characteristic ameloblastoma. Thus, the overall prognosis for unicystic

ameloblastoma is considerably better than the other variants.[3] Considering the characteristics of ameloblastoma as a locally invasive but slow-growing and extremely rare metastasizing benign tumor, the priority of the treatment method should be discussed from the points of morbidity and quality of life of the patients, noting that the recurrence rate is not always the primary factor. Wide resection of the jaw is usually the recommended treatment for ameloblastoma, should priority be given to the recurrence rate. However, radical surgery often means that the patients have serious complications including facial deformity, masticatory dysfunction, and abnormal jaw movement. Two therapy strategies are mentioned in literature: a conservative way of treatment and radical procedures. Non-radical surgical procedures like enucleation and curettage, combined with liquid nitrogen spray cryosurgery, or just drilling of the perilesional bone are mentioned to be useful in unicystic ameloblastomas, especially in children and young patients. Other authors show high rates of recurrence of ameloblastoma after conservative treatment protocols and therefore recommend radical surgical treatment. Authors suggests a "rational radical conservative" resection of the mandible with preservation of the lower border of the mandible to maintain the continuity of the lower jaw and the facial contours. In the previous reports, conservative treatments for ameloblastoma appeared to have failed to control local recurrences. Sehdev *et al*, reported recurrence after the conservative approach (curettage) in more than 90% of 92 ameloblastomas. Shatkin and Hoffmeister reported that 86% of 20 mandibular ameloblastomas recurred after curettage compared with a 14% recurrence rate after en bloc resection. Other authors have reported a series of 84 ameloblastomas in which they found a 52% rate of recurrence in patients treated conservatively and a 25% rate of recurrence in patients with primary tumor treated by the radical approach. However, extensive tumors require a more radical approach. The amount of resection is variable and depends on the site and extension of the tumor. When planning the treatment of ameloblastoma, it is important to understand the growth characteristics and to remove the full extent of the tumor, including the surrounding tissues. Otherwise, the remaining tumor cells may lead to multiple morbidities of recurrence. Recent advancements in the understanding of the biological behaviors of ameloblastoma have revealed that unicystic lesions are well-localized by the fibrous capsule of the cyst, with few tumors broaching peripheral tissues, whereas multicystic and solid lesions are characterized by an aggressive infiltration to adjacent tissue. Gardner discussed the treatment of ameloblastoma on the basis of pathological and anatomical considerations. He stated that the recommended treatment for solid and multicystic ameloblastoma was radical treatment, whereas unicystic ameloblastoma was usually cured by curettage. [7] Chemical cauterization with Carnoy's solution is advocated for luminal and intraluminal variety.Carnoy's solution a powerful fixative penetrates the cancellous spaces and thus fixes the remaining tumour cells. Usually, Carnoy's solution is applied for 3-5 min. However, Frerich et al, suggested that the application of Carnoy's solution should not exceed by 3 min and should not be directly applied over the nerve as it could lead to nerve impairment.[4] Due to technical and financial constraints, and because the lesion was histologically diagnosed as Peripheral Unicystic Ameloblastoma,we performed surgical enucleation of the lesion followed by curettage and application of Carnoy's solution. In our follow-up regime, patient was scheduled for clinical and radiological examination twice a year for the first 5 years and after that only once a year. No recurrence has been noted 1 year post surgery.

Conclusion

Ameloblastoma, though a benign odontogenic tumour requires thorough clinical and radiographic evaluation and belligerent management due to its local aggressive behavior. Unicystic ameloblastoma is a rare entity that requires special management. A long follow-up period for at least 10 years is suggested as recurrence may also appear years after primary surgery.

***Conflict of Interest*: None**
***Source of support*: Nil**
***Ethical Permission*: Obtained**

References

1. Gabhane et al. Unicystic Ameloblastoma of Mandible: A Case Report. Indian J Stomatol 2011;2(4):273-76.
2. Pallagatti S et al. Unicystic Ameloblastoma of the Mandible with all Variants. JPMS 2013;3(2):106-109.
3. Kanneppady S et al. Unicystic Ameloblastoma in Young Female-A Case report and Review of Literature. World Journal of Dentistry. 2011;2(3):263-67.
4. Ramesh et al. Unicystic ameloblastoma of the mandible – an unusual case report and review of literature. Head Neck Oncology 2010, 2:1.
5. Malheiro P et al. Unicystic Plexiform Ameloblastoma – Case Report. Revista De Saude Amato Lusitano 2013; 32:52-54.
6. Ricci M et al. An unusual case of intramural unicystic ameloblastoma and review of literature. Contemp Clin Dent. 2012; 3(Suppl 2): S233–39.
7. Dandriyal R et al. Surgical management of ameloblastoma: Conservative or radical approach. Natl J Maxillofac Surg. 2011; 2(1): 22–27.
8. Savithri V et al. Unicystic Ameloblastoma as a Differential Diagnosis for Odontogenic Cysts. Oral and Maxillofacial Pathology Journal. 2014;5(1):466-469.