Infected Multilocular Unicystic Ameloblastoma Involving Ramus and Coronoid Process: A Rare Case Report

Abstract

Ameloblastoma is a neoplasm that originates from the odontogenic epithelium. Unicystic ameloblastoma (UA) is a rare variant of ameloblastoma occurring usually in younger population. They are characterized by slow growth and are locally aggressive, with the main site of origin being the posterior portion of the mandible. Most commonly, UA appears on radiograph as a unilocular well-demarcated radiolucency present mostly in the mandibular posterior region. Here, we report a unique case of multilocular UA in a 22-year-old male patient involving the left side of whole length of the mandibular ramus and coronoid process with impacted third molar.

Keywords: *Ameloblastoma, impacted third molar, multilocular radiolucency, unicystic ameloblastoma*

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Introduction

Ameloblastoma, the second most common tumor of odontogenic origin after odontoma,[1] is a slow-growing, persistent, and locally aggressive neoplasm developing from the remnants of the dental lamina and odontogenic epithelium.[2] It accounts for only 1% of all oral tumors.[2,3] The peak incidence of ameloblastoma is in the third to fourth decade of life, with an equal sex predilection.[3] According to the World Health Organization (WHO) Classification (2005) of Head and Neck Tumours,[4] ameloblastomas are of four types: multicystic, peripheral, desmoplastic, and unicystic ameloblastomas (UAs). UA was first described by Robinson and Martinez in 1977 as a distinct entity.[5] It accounts for about 6% of all ameloblastomas, and 50% of cases occur in the second decade of life.[2,3] It is often associated with an unerupted third molar, which may be detected during the course of routine radiography.[6] In this report, we present a rare presentation of multilocular UA in a 22-year-old male patient involving the left side mandibular angle region, extending up to the coronoid and condylar process with impacted third molar.

Case Report

A 22-year-old male patient reported to the department of oral medicine and radiology with the complaint of swelling on the left side of the face for the past 6 months and reduced mouth opening for the past 20 days. The patient's history revealed that the swelling was initially smaller in size and has increased slowly to the present size over the period of 6 months. The patient also stated that his mouth opening was approximately four fingers 20 days back, which has now reduced to approximately two fingers over a period of 20 days. His dental history revealed that he had visited a private dental clinic 10 days back for the same complaint where he was advised orthopantomogram (OPG) and magnetic resonance imaging (MRI) of the left side of the mandible and was referred to our institution for further treatment. His medical history was not significant. On extraoral examination, single diffuse swelling measuring approximately 3 cm × 4 cm was present on the left side of the mandibular angle region, extending anteroposteriorly from the level of corner of the mouth till the posterior border of the ramus of the mandible and superoinferiorly from the level of the ala-tragus line till the lower border of the mandible. The overlying skin appeared normal in color [Figure 1]. On palpation, the swelling was afebrile,

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nontender, and firm to hard in consistency. The interincisal opening was approximately 18 mm. Intraoral examination revealed inflammation in the retromolar area, with clinically missing left mandibular third molar. There was obliteration of the buccal vestibule in relation to the mandibular left second molar due to cortical expansion in the same region. There was slight tenderness in the same region. No signs of paresthesia were present. A provisional diagnosis of dentigerous cyst with the mandibular left third molar was given. Orthopantomogram (OPG) showed large well-defined multilocular radiolucency involving the left mandibular angle region and ramus, extending from the periapical region of the mesial root of the mandibular left second molar till posterosuperiorly up to the coronoid process and neck of the condyle along with distoangularly impacted third molar [Figure 2]. MRI revealed poorly marginated heterogeneous lesion in the left mandibular ramus measuring approximately 56 mm × 33 mm × 30 mm, which was predominantly hyperintense on T2 and Short-TI Inversion Recovery (STIR) and hypointense on Short-TI, suggestive of ameloblastoma of the left mandibular ramus [Figure 3]. The patient was advised biopsy from the same region. The histopathological report revealed multiple pieces of tissue showing odontogenic epithelium (luminal epithelium) overlying fibrous connective tissue stroma, with some areas of inflammatory aggregates, and in one area, numerous cholesterol clefts were also appreciated under low-power view, whereas, under higher magnification, cystic epithelium in few areas showed preameloblastic cells with dark hyperchromatic and elongated nuclei associated with star-shaped cells, suggestive of stellate reticulum-like tissue. The connective tissue stroma comprised of loose-to-dense bundles of collagen fibers with plump to spindle-shaped fibroblasts. Inflammatory infiltrate predominantly comprising lymphocytes was evident. Thus, the overall histopathological picture was suggestive of infected UA [Figure 4]. The patient was referred to the department of oral surgery for further treatment where segmental resection of the left side of the mandible was advised. The patient was posted for surgery under general anesthesia, and left segmental resection of the mandible with disarticulation was done followed by reconstruction with stainless steel condylar prosthesis [Figure 5]. The patient is under regular follow-up for more than 1½ years with no signs of recurrence.

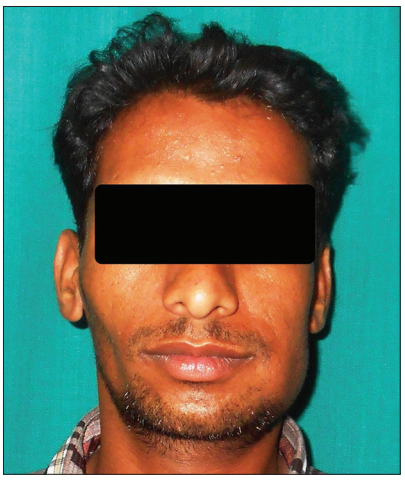

Figure 1: Facial profile of the patient showing extraoral diffuse swelling on the lower left side of the face

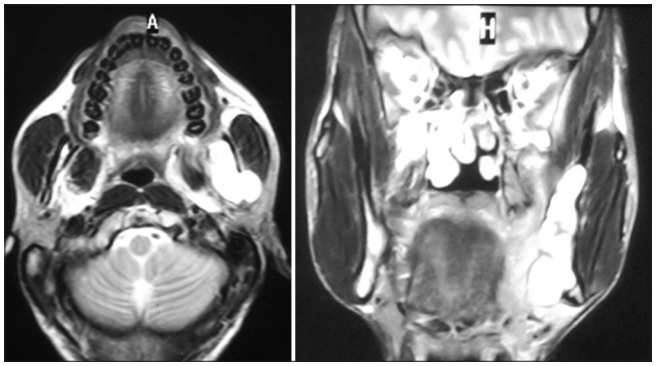

Figure 3: Axial (left) and coronal (right) sections of magnetic resonance imaging of the patient showing lesion involving the left side of the mandibular ramus

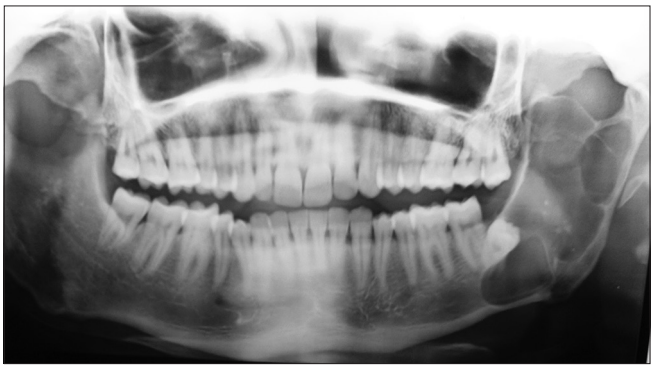

Figure 2: Orthopantomogram showing a well-defined multilocular radiolucency on the left side of the mandible with distoangularly impacted third molar

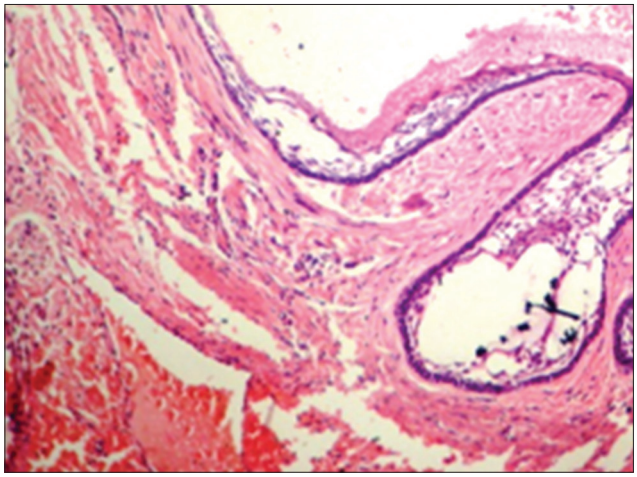

Figure 4: Histopathological picture showing preameloblastic cystic lining epithelium, with dark hyperchromatic and elongated nuclei, cholesterol clefts, and inflammatory cell infiltrate

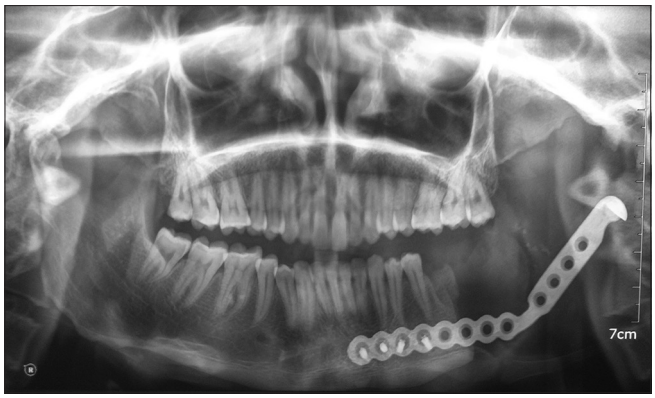

Figure 5: Postoperative orthopantomogram showing stainless steel reconstruction plate on the left side of the mandible

Discussion

Ameloblastoma was first described by Cusack in 1827.[7] Later on, in 1885, Malassez introduced the term "adamantinoma," which is currently used for a rare form of bone cancer described in 1913 by Fisher.[8] Although first described in detail by Falkson in 1879, the term ameloblastoma was given by Ivey and Churchill in 1930.[9] It is a benign odontogenic tumor developing from the epithelial rests of Malassez, which is responsible for 1% of all the oral tumors and approximately 9%–11% of odontogenic tumors.[10] As per the 2005 WHO Classification,[4] ameloblastomas are of four types: multicystic, peripheral, desmoplastic, and UAs, whereas as per the 2017 updates, the classification has been narrowed down to three types: conventional ameloblastoma, UA, and extraosseous/peripheral types.[11]

UA is a distinguishable entity of ameloblastoma which refers to horse cystic lesions, which has clinical and radiographic features similar to that of a cyst; however, on histopathological examination, it shows a part of ameloblastomatous lining epithelium of the cystic cavity, with or without luminal and/or mural tumor growth.[1] It accounts for 15% of all intraosseous ameloblastomas.[6]

Various theories reported in the literature state that UAs may arise from reduced enamel epithelium associated with developing tooth or it may develop in a preexisting dentigerous cyst or other types of odontogenic cyst or solid ameloblastoma may undergo cystic degeneration, resulting in UA. However, it is difficult to produce sufficient evidence to prove these theories. Others are of opinion that UAs arise *de novo* as cystic neoplasms.[12] In contrast to conventional ameloblastoma, UA usually occurs in a younger age group with more than 50% of cases occurring in the second decade of life, and it has a slight male predilection.[1,6] UA occurs more frequently in the mandible than the maxilla, with the mandible:maxilla ratio of 13:1, and more than 75% of cases are located in the molar ramus region.[1] The present case was in agreement with all these findings. Usually, the lesion presents as local asymptomatic swelling with facial asymmetry, pain, and less often, lip numbness. The discharge or drainage is seen only when it is secondarily infected as was reported in one of the cases in 2015.[12] In the present case, the patient reported with the chief complaint of reduced mouth opening and swelling on the left side of the mandibular angle region. There were no signs of drainage or discharge from the lesion, but the histopathological report was suggestive of infected UA which is again rarely seen. The lesion is most commonly observed in the mandibular molar–ramus region as was seen in the present case, but it can also be seen in interradicular, periapical, and edentulous regions of the jaw.[2]

Basically, there are two main radiographical patterns of UA: unilocular and multilocular.[2] However, according to the literature, various radiographic patterns of UA can be observed such as pericoronal unilocular, extensive pericoronal unilocular, pericoronal scalloped, and periapical unilocular, inter-radicular, and even multilocular, but with a clear preponderance for the unilocular pattern.[13] However, pericoronal multilocular radiographic pattern involving impacted third molar is rare, which was seen in the present case. According to the world literature review published by Meshram *et al*.[14] in 2017, only 74 cases of UA were reported in the literature, among which 67 were in the mandible and 7 were in the maxilla. Of the 67 cases, in 64 cases, the lesion was unilocular in appearance and only three cases showed multilocular radiographic appearance. In the present case also, the lesion was multilocular in appearance. Tooth impaction is associated in approximately 50%–80% of cases, with the mandibular third molar being the most commonly involved tooth,[6] as was seen in the present case where the impacted mandibular left third molar tooth was involved. Pericoronal unilocular type of UA with the impacted third molar shows similarity to a dentigerous cyst, and hence, it is categorized as dentigerous variant (with impacted tooth) of ameloblastoma.[13] In majority of such cases, a radiographic diagnosis of dentigerous cyst is made considering the age of the patient. Therefore, biopsy of the lesion becomes mandatory for accurate diagnosis and appropriate treatment plan, which was followed in the present case as well. According to the literature, the "dentigerous" type occurs earlier than the "nondentigerous" variant by approximately 8 years, and the mean age for unilocular, impaction-associated UAs is 22 years, whereas the mean age for the multilocular lesion unrelated to an impacted tooth is 33 years.[1,3] In contrast to this, UA in the present case was multilocular in appearance associated with an impacted mandibular third molar, and the age of the patient was 22 years.

Based on histological examination, to diagnose a lesion as UA, the minimum criteria are the demonstration of the presence of a single cystic sac lined by odontogenic ameloblastomatous epithelium, which was seen in the present case.[15] Ackerman in 1988 classified UA

into following three histologic groups based on his clinicopathological study of 57 cases:[16]

i. Luminal UA (tumor confined to the luminal surface of the cyst)
ii. Intraluminal/plexiform UA (nodular proliferation into the lumen without infiltration of tumor cells into the connective tissue wall)
iii. Mural UA (invasive islands of ameloblastomatous epithelium in the connective tissue wall not involving the entire epithelium).

Philipsen and Reichart have described another histologic subgrouping as follows:[17]

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

As per the published literature, UA diagnosed as subgroups 1 and 1.2 can be treated conservatively only with careful evaluation, and the subgroups that show intramural growths (subgroups 1.2.3 and 1.3) should be treated with radical resection.[15] Since UA is considered to be less aggressive in nature, it can be treated successfully by simple enucleation or other less aggressive surgical treatments. Stoelinga and Bronkhorst[18] in 1988 suggested the use of Carnoy's solution to decrease the risk of recurrence after conservative surgical management of UAs. Furthermore, it is stated that vigorous curettage of the bone should be avoided since it may result in the implantation of foci of ameloblastoma deeply into the bone. The response of UA to enucleation or curettage is more favorable than the solid or multicystic ameloblastomas.[2] The most commonly advocated treatment for the management of solid or multicystic ameloblastoma is wide surgical excision to prevent the recurrence. More than 1 cm of the normal margin of the mandible and overlying periosteum, if cortical perforation had occurred, has to be excised. More conservative treatment modalities such as curettage, cryotherapy, or enucleation have resulted in recurrence rates of approximately 75%–90%.[19]

In case of UA, a recurrence rate of approximately 7%–25% has been reported after treatment which is said to be dependent on its histological type, site of origin, and initial treatment modality.[6] Among the three subtypes described by Ackerman *et al*.,[16] the mural type has been reported to have the highest recurrence rate. Li *et al*.[20] have reported a higher recurrence rate of 35.7% for the mural type and a lower recurrence rate of 6.7% for the other two types. Hence, for the mural type of UA, radical resection appears to be more appropriate management protocol. UAs occurring in the mandible are usually treated by conservative interventions, but the same is not advocated for the lesions occurring in the maxilla, the reason being the spongy bone architecture of the maxilla, which facilitates spread of the tumor and its proximity to vital structures, such as the orbit, pterygomaxillary fossa, and cranium. Therefore, more aggressive treatment in the form of resection is suggested to eliminate the risk of recurrence.[6] Lau and Samman[21] reported recurrence rates of 3.6% for resection, 30.5% for enucleation alone, 16% for enucleation followed by Carnoy's solution application, and 18% by marsupialization followed by enucleation, where the lesion has reduced in size. Therefore, in the present case, segmental resection of the left mandible involving the whole lesion with safe margin was done, followed by reconstruction.

Conclusion

UA is a tumor with a strong propensity of recurrence, especially when the ameloblastic focus penetrates the adjacent tissues from the wall of the cystic cavity. Radiographically, most of the conventional ameloblastomas show multilocularity, whereas UAs show a single large unilocular radiolucency. Very rarely, we come across a case with the presentation of pericoronal multilocular radiographic pattern involving impacted third molar as was reported in the present case which makes it unique.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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