Different Variants of Unicystic Ameloblastoma- A Report of Five Cases with CT Findings

Hitesh Shoor, Keerthilatha M Pai, Ankur Kaur Shergill, Monica Charlotte

Department of Oral Medicine and Radiology, Manipal College of Dental Sciences, Manipal University, Manipal, Karnataka, India

Abstract

Most ameloblastomas contain microcysts but the unicystic variant has a lining of flattened tumour cells which resemble those of non-neoplastic cysts, with nodular proliferation into the lumen without infiltration of tumour cells into the connective tissue wall. Over 80% of these cystic tumours enclose the crown of a tooth and mimic dentigerous cysts radiographically. We have reported five cases of two different variants of unicystic ameloblastoma and also highlighted their clinical, radiological and histological features.

Key Words: Unicystic ameloblastoma, Plexiform, Intraluminal, Tumor

Introducton

Ameloblastoma is a true neoplasm of odontogenic epithelium [1]. Ameloblastoma appears most commonly in the third to fifth decades but the lesion can be found in any age group including young age. Seventy percent of them develop in the posterior molar region and typically appear multilocular on radiographs. Most ameloblastomas contain microcysts but the unicystic variant has a lining of flattened tumour cells which resemble those of non-neoplastic cysts, with nodular proliferation into the lumen without infiltration of tumour cells into the connective tissue wall. Over 80% of these cystic tumours enclose the crown of a tooth and mimic dentigerous cysts radiographically. Here we report five cases of two different variants of unicystic ameloblastoma, in otherwise healthy individuals, which were not been associated with any impacted tooth. We have also highlighted their clinical, radiological, histological features and treatment modalities.

Case Reports

Case 1

A 39-year-old otherwise healthy male, reported with a complaint of swelling in relation to right side of the lower jaw with gradual increase in its size since one month. On extra oral examination, a well-defined swelling about 2 × 3 cm was seen at the right corner of the mouth, extending up to the lower border of the mandible. Swelling was non tender and firm in consistency with no local rise in temperature. On intra oral examination, a diffuse fluctuant swelling was seen in the buccal region of 44, 45, and 46 with obliteration of the sulcus. Lingual cortical expansion was seen in the region of 43, 44, 45 and mucosa over this area was erythematous but no changes of surface texture were noticed. Panoramic radiograph showed a well demarcated radiolucency corresponding to area between roots of 43, 44, 45, 46, about 4 × 4.5 cm in size with a sclerotic border. There was resorption of roots of 43, 44. CT scan showed buccolingual expansion of the cortical plate (*Figure 1*).

Figure 1. *CT scan showed buccolingual expansion of the cortical plate.*

Case 2

A 33 year old male patient reported with a swelling on lower left side of the face with gradual increase in its size since one year. There was history of associated pain and paresthesia. Root canal treatment was done in lower left back tooth region 2 years back. Extraorally, no obvious facial asymmetry was evident. Skin overlying face appeared normal, but on palpation, a 3 × 2 cm large swelling was present on left lower third of face extending from left parasymphysis region till mid of body of mandible region. Area was slightly tender on palpation and hard in consistency. Intraorally, mild swelling was evident in region of 32 to 36 on buccal and lingual aspects with obliteration of the buccal sulcus. Swelling was soft, slightly compressible but non reducible with mild tenderness. Panoramic radiograph showed a 5 × 3 cm large radiolucency extending from distal aspect of 41 to mesial aspect of 37 with well-defined corticated borders. Axial section of CT showed expansion of buccal cortical plate with extensions into the alveolar process in relation to region of incisors, premolars and 1st molar (*Figure 2*).

Corresponding author: Dr. Hitesh Shoor, M.D.S, Department of Oral Medicine and Radiology, Manipal College of Dental Sciences, Manipal University, Karnataka-576104, India, Tel: +919876772344; E-mail: hiteshshoor@gmail.com

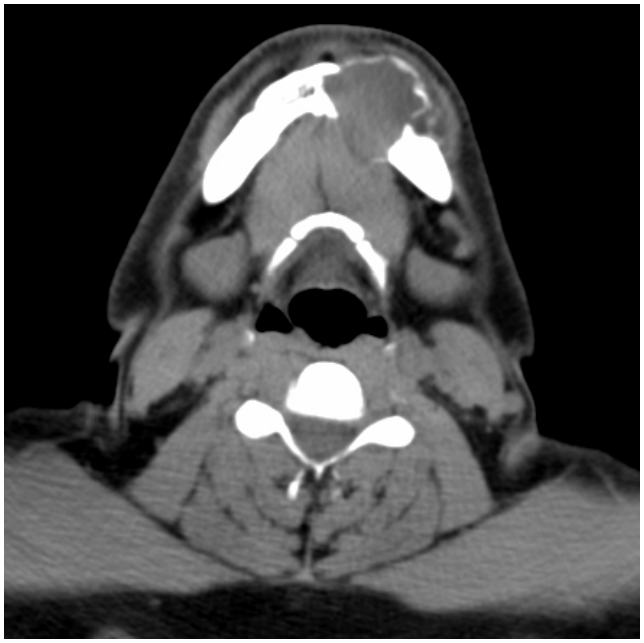

Figure 2. *Axial section of CT showed expansion of buccal cortical plate with extensions into the alveolar process.*

Case 3

A 32 year old female patient reported with history of pain, swelling and pus discharge from left lower back tooth region since 1 month. The swelling was gradually increasing in size. On extra oral examination, a diffuse swelling was seen on left side of cheek. Intraorally, there was an area of necrosis of bone visible buccal to the lower third molar with pus discharge. Panoramic radiograph showed a large unilocular radiolucency involving left angle and ramus of mandible extending till sigmoid notch region. Resorption of roots of 38 was evident. CT scan showed expansion of both buccal and lingual cortices (*Figure 3*).

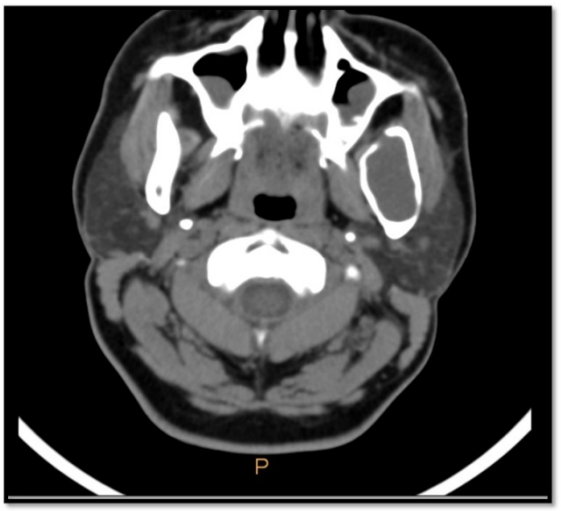

Figure 3. *CT scan showed expansion of both buccal and lingual cortices.*

Case 4

A 24 year old female patient reported with severe pain in the lower right back tooth since 2 months. There was associated slow growing swelling over the same region. On extraoral examination, a diffuse swelling was present on right lower jaw region about 1 cm posterior to right corner of mouth extending to the angle and lower border of mandible, which was soft on palpation. Intraorally, a diffuse swelling was present on the alveolar ridge distal to 47. Obliteration of buccal sulcus in the region and slight expansion of the lingual cortex were present. The swelling was soft in consistency, fluctuant and severely tender on palpation. Panoramic radiograph showed a unilocular radiolucency from the region of 47, involving the ascending ramus, with destruction of anterior and posterior borders of the ramus, but intact sigmoid notch, condyle and coronoid processes. There was floating in space appearance of 47. CT scan showed well defined expansile lesion measuring 3.8 × 2.4 cm involving ramus, angle and body of mandible on right side adjacent to 2nd molar. There was cortical erosion predominantly involving inner cortex of mandible (*Figure 4*).

Figure 4. *CT scan showed well defined expansile lesion measuring 3.8 × 2.4 cm involving ramus, angle and body of mandible on right side adjacent to 2nd molar. There was cortical erosion predominantly involving inner cortex of mandible.*

Case 5

A 58 year old male patient reported with a complaint of swelling in the front region of lower jaw since one year. Patient gave a history of trauma 2 years back due to which his lower anterior teeth were fractured and then extracted. No facial asymmetry was noticed on extra oral examination. Intraorally, a round well defined swelling was present in lower anterior region extending from 33 to 44, approximately 5 cm in diameter, involving the buccal and lingual gingiva. It was soft to firm in consistency, fluctuant and compressible, but not reducible. A well-defined unilocular radiolucency was seen in the anterior mandible on panoramic radiograph, approximately 6 × 3 cm, extending from 35 region, crossing midline to 45 region with corticated borders. CT showed an ill-defined hypoechoic expansile soft tissue density with coarse internal trabeculae noted involving body of mandible

causing erosion and displacement of adjacent lower incisor and canine roots (*Figure 5*). A follow up for 5 years in all the cases showed no signs of recurrence.

Figure 5. *CT showed an ill-defined hypoechoic expansile soft tissue density with coarse internal trabeculae noted involving body of mandible causing erosion and displacement of adjacent lower incisor and canine roots.*

Histopathological examination

Histopathological examination of incisional biopsy specimen of case 1,2 revealed an irregular cystic space lined by disrupted epithelial lining, with lumen showing interlacing strands and cords of odontogenic epithelial cells. There were peripheral palisaded cuboidal/columnar cells and central loosely arranged stellate reticulum like cells with squamous metaplasia, suggestive of intraluminal/plexiform Unicystic Ameloblastoma (*Figure 6*). The lesion was excised under general anesthesia along with removal of 43, 44, 45.

Histopathological examination of biopsy specimen of case 3,4,5 revealed a cystic lining of varied thickness resembling odontogenic epithelium. The basal cells appeared columnar to cuboidal with nucleus exhibiting hyperchromatasia, palisading and reverse polarity the suprabasal cells are flattened with few areas resembling stellate reticulum. Areas of hylanization are seen adjacent to the cystic lining. The underlying cells showed small nests of tumor cells of varying size and shape. The follicle showed peripheral columnar cells with reversal of polarity and central stellate reticulum like cells, with areas of squamous metaplasia. Areas of hemorrhage and peripheral new bone formation was also evident which all were suggestive of mural variant of unicystic ameloblastoma (*Figure 7*).

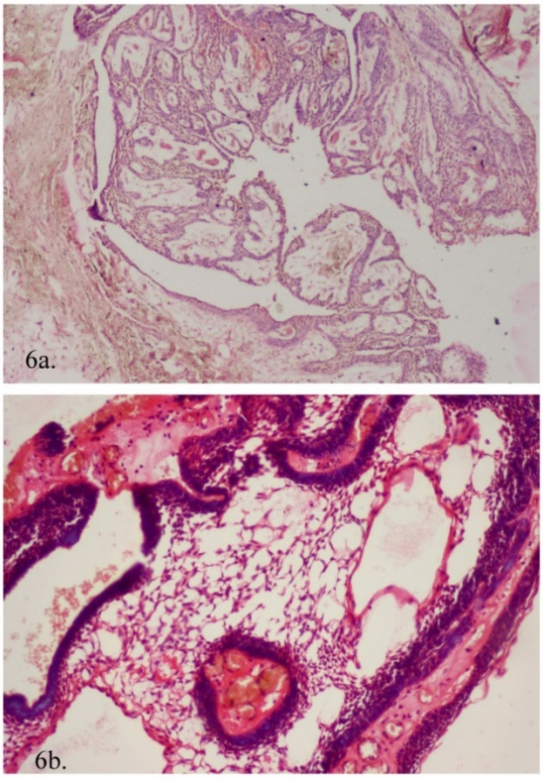

Figure 6. *Intraluminal/plexiform unicystic ameloblastoma.*

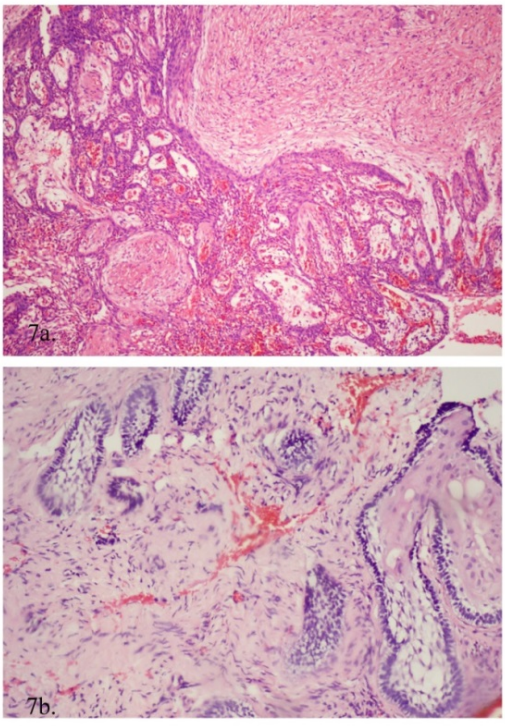

Figure 7. *Mural variant of unicystic ameloblastoma.*

Discussion

Ameloblastoma is a benign epithelial odontogenic tumor which is often aggressive and destructive, with the capacity to attain great size, erode bone and invade adjacent structures

[2,3]. Although the term 'ameloblastoma' was coined by Churchill in 1933, the first detailed description of this lesion was by Falkson in 1879 [4]. It represents about 1% of all oral ectodermal tumors and 9% of odontogenic tumors [5]. It is an aggressive neoplasm that arises from remnants of the dental lamina and dental organ (odontogenic epithelium) [6]. Its histological appearance is similar to that of the early cap-stage ameloblastic elements of developing (tooth) without complete differentiation to the stage of enamel formation [1,7]. The WHO classified ameloblastoma into four variants which differ in their relation to age, distribution, localization, imaging features and prognosis. These are the solid/multicystic ameloblastoma, extraosseous/peripheral ameloblastoma, desmoplastic ameloblastoma and unicystic ameloblastoma.

Diverse histological patterns have been described in the literature, including those with follicular, plexiform, acanthomatous, papilliferous keratotic, desmoplastic, granular cells, vascular and with dentinoid induction [4]. The term "plexiform" refers to the appearance of anastomosing islands of odontogenic epithelium in contrast to a follicular pattern [8]. In two of our cases, a large plexiform ameloblastoma found in the mandible and it was not associated with a non-erupted tooth. In other two cases it was a mural variant with occurring unicystic ameloblastomas, differ from the solid types in that they can be enucleated with a low risk of recurrence and has better prognosis. A 10% recurrence rate 10 years after enucleation can be expected. However, in some unicystic ameloblastomas tumour tissue extends from the cyst wall into the surrounding tissues. The diagnosis of unicystic ameloblastoma therefore cannot be made on a unilocular radiographic appearance or a single incisional biopsy. Multiple areas need to be evaluated to confirm that the tumor does not extend beyond the cyst wall [4]. The most common treatment modality is enucleation followed by chemical cauterization. However larger, more aggressive lesions require more radical treatment modalities including marginal, segmental or composite resection. Unicystic variants have a high risk of recurrence after enucleation. The most common sites of metastases are the lungs followed by regional lymph nodes, pleura, vertebrae, skull, diaphragm, liver, parotid and small intestine [4]. Mechanisms of distant spread are debated and include aspiration, haematogenous spread, lymphatic spread and malignant activation of the developmental rests of epithelial tissue.

It is important for oral diagnosticians to keep the salient features of this unique entity in mind while diagnosing the condition, as it has very distinct radiological and histological features. The importance of knowing the different variants of ameloblastoma lies in considering the possible differential diagnosis and prognosis, and after confirmation, applying the appropriate therapy.

References

1. White SC, Pharoah MJ. Oral radiology principles and interpretation (4th edition). *Mosby.* 2000: 386-390.

2. Chen WL, Li J, Yang ZH, Wang JG, Zhang B. Recurrent ameloblastoma of the anterior skull base: Three cases treated by radical resections. *Journal of Cranio-Maxillo-Facial Surgery.* 2006; **34**: 412-414.

3. Gümgüm S, Hosgören B. Clinical and radiologic behaviour of ameloblastoma in 4 cases. *Journal of Canadian Dental Association.* 2005; **71**: 481-484.

4. Iordanidis S, Makos C, Dimitrakopoulos J, Kariki H. Ameloblastoma of the maxilla. Case report. *Australian Dental Journal.* 1999; **44**: 51-55.

5. Torres-Lagares D, Infante-Cossío P, Hernández-Guisado JM, Gutiérrez-Pérez JL. Mandibular ameloblastoma. A review of the literature and presentation of six cases. *Medicina Oral Patologia Oral y Cirugia Bucal.* 2005; **10**: 231-238.

6. Adebiyi KE, Ugboko VI, Omoniyi-Esan GO, Ndukwe KC, Oginni FO. Clinicopathological analysis of histological variants of ameloblastoma in a suburban Nigerian population. *Head & Face Medicine.* 2006; **2**: 42-50.

7. Artes-Martínez MJ, Prieto-Rodríguez M, Navarro-Hervás M, Peñas-Pardo L, Camañas-Sanz A, et al. Ameloblastoma. Diagnosis by means of FNAB. Report of two cases. *Medicina oral, patología oral y cirugía bucal.* 2005; **10**: 205-209.

8. Kovács A, Wagner M, Ghahremani M. Considerations on a long-term course of a plexiform ameloblastoma withn a recurrence in the soft tissue. *Revista Médica del Hospital General de México.* 1999; **62**: 48-53.