

Unicystic Ameloblastoma Presenting as Multilocular Lesion: A Case Report

Sandeep S. Gupta, Shubhangi Mhaske, *M.K. Gupta, Raju Ragavendra, Kavitha P Kamath

Department of Oral Pathology & Microbiology, *Department of Oral & Maxillofacial Surgery, People's Dental Academy, Bhanpur, Bhopal - 462037

Abstract:

Unicystic ameloblastoma (UA) refers to those cystic lesions that show clinical, radiographic, or gross features of a mandibular cyst, but on histological examination show a typical ameloblastomatous epithelium lining part of the cyst cavity, with or without luminal and/or mural tumour growth. In this case report we present a uncommon multilocular unicystic ameloblastoma encountered in a 40 year old female patient.

Key Words: Unicystic ameloblastoma, unilocular, multilocular

Introduction:

Many benign cysts and tumours involve mandible; these can be of odontogenic origin or of nonodontogenic origin. Lesions include ameloblastoma, radicular cyst, dentigerous cyst, keratocystic odontogenic tumour, central giant cell granuloma, fibroosseous lesions and osteomas (Kahairi et al, 2008). The most common tumours of odontogenic origin are ameloblastomas, which develops from epithelial cellular elements and dental tissues in their various phases of development. Much confusion still exists, when it comes to the terminology used for unicystic ameloblastomas (UAs). Some of the terms used for this lesion prior to 1977, when Robinson & Martinez

(1977) introduced the concept of UA, were cystic (intracystic) ameloblastoma, ameloblastoma associated with dentigerous cyst, cystogenic ameloblastoma, extensive dentigerous cyst with intracystic ameloblastic papilloma, mural ameloblastoma, dentigerous cyst with ameloblastomatous proliferation, and ameloblastoma developing in a radicular 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. Much confusion stems from the fact that a unicystic ameloblastoma may appear not only as a unilocular but also as a multilocular bone defect



Fig. I: Orthopentogram showing multilocular radiolucency in association with 43 to 47

Corresponding Author: Dr. Sandeep Gupta, Department of Oral Pathology & Microbiology, People's Dental Academy, People's Campus Bhanpur, Bhopal-462037,
Phone No.: +91- 7869494068
E-mail : doc.sandeepgupta@gmail.com

(Eversole et al, 1984; Li et al, 2000). We present a case of a large unicystic mandibular ameloblastoma in a 40 year old female with a relatively rare multilocular appearance.

Case report:

A 40-year-old female patient presented to the Department of Oral Pathology & Microbiology with swelling on the right side of the face for last one month. The swelling was approximately 3 X 5 cm in size, associated with dull pain on the lower right side of face. On intraoral examination, the lesion extended from 44 to 46 with obliteration of buccal sulcus. The swelling was tender on palpation. The overlying mucosa was inflamed. Right submandibular lymph nodes were palpable and tender. An orthopantomogram showed large cystic lesion in the right side of mandible, extending anteroposteriorly from 42 to 47 & superior-inferiorly from periapical region of 42 to 47 to the body of mandible. Osteolytic lesion was multilocular also showed root resorption of 45, 46, and 47 (Fig. I). Considering the site, age and it being multilocular a differential diagnosis of ameloblastoma / odontogenic

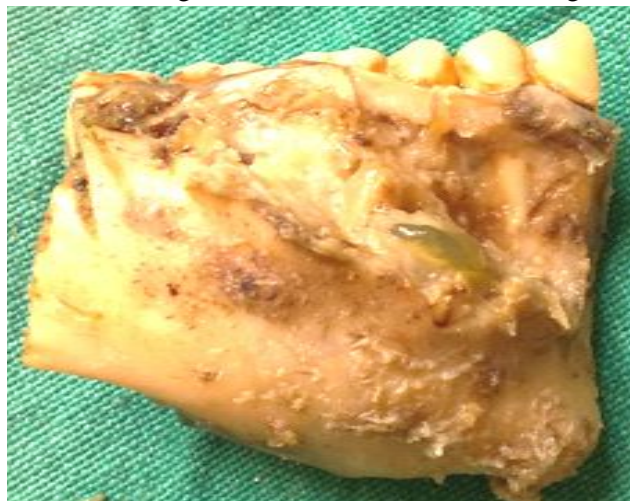


Fig. II: Gross specimen showing cortical expansion and thinning of mandible.

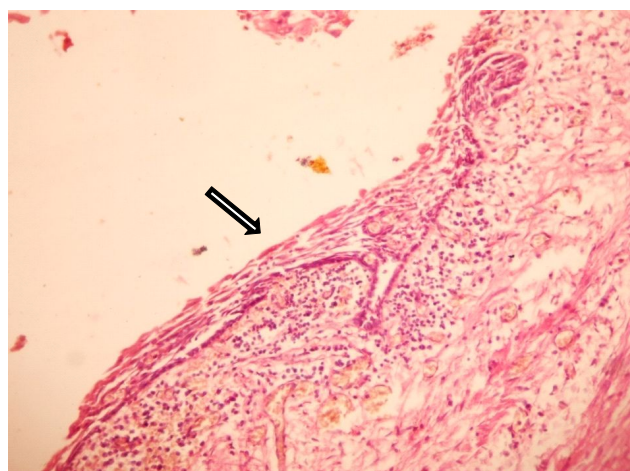


Fig. III: Histological section showing luminal unicyclic lining (H&E stain 10X)

keratocyst was considered. Segmental mandibulectomy was done under general anaesthesia and the specimen was subjected to histopathological examination. The gross specimen (Fig. II) revealed buccal cortical expansion with thinning of cortical plates. No expansion of the lingual cortical plate was noted.

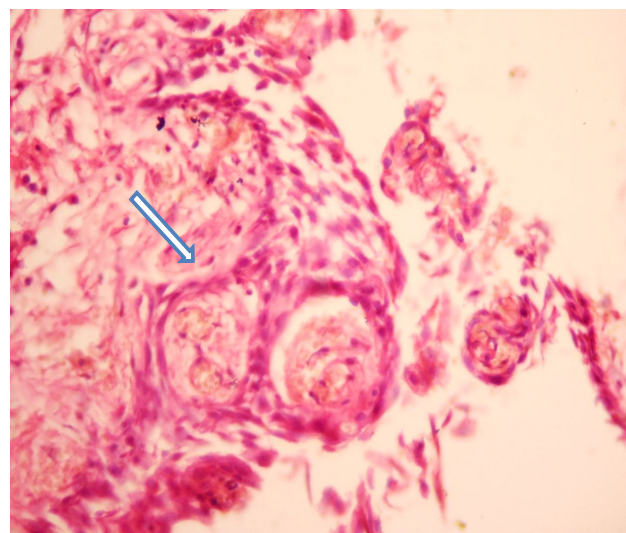


Fig. IV: Histological section showing intraluminal unicyclic lining (H&E stain 10X)

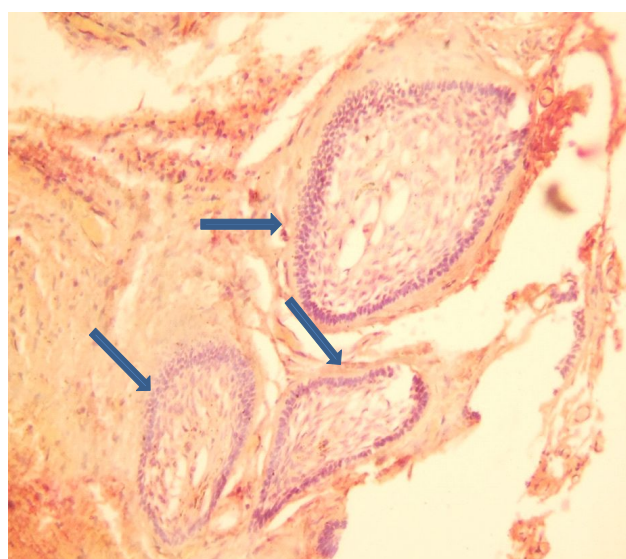


Fig. V: Histological section showing mural follicles in unicyclic ameloblastoma (H&E stain 10X).

Histopathological examination revealed presence of an odontogenic epithelium showing Vickers & Gorlins criteria (Vickers & Gorlin 1970) with the intraluminal and mural proliferation of the odontogenic islands and stands. Inflammatory infiltration was also observed (Fig. III, IV, & V). Based on the histopathological findings, diagnosis of unicyclic ameloblastoma showing intraluminal and mural proliferation was given.

Discussion:

Ameloblastomas are benign tumours whose important lies in its potential to grow to enormous size with resulting bone deformity. Ameloblastomas are typically differentiated histologically into unicystic intraosseous, multicystic, and solid. The relative frequency of occurrence of unicystic ameloblastoma has been reported as between 5% to 22% of all types of ameloblastomas (Reichart et al, 1995). The mean age at the time of diagnosis differs considerably according to the UA variants. Those diagnosed as dentigerous, occurred in much younger patients with mean age of 16.5 years, 78.3% occurring in the 1st and 2nd decades while for nondentigerous the mean age was 35.2 years with age ranging from 40 to 70 years (Reichart & Philipsen, 2004). In the present case patient was 40-year-old which is in accordance to the literature (Reichart & Philipsen, 2004). The location of the UA within the jawbones shows a marked predominance for the mandible irrespective of the variant. The maxilla: mandible ratio being 1:7 for the dentigerous type and 1:4.7 for the nondentigerous type (Philipsen & Reichart, 1998) as seen in the present case. The radiographic appearance of all UAs is divided into two main patterns, unilocular and multilocular, there is a clear predominance of the unilocular configuration in all studies. This predominance was exceptionally marked for the dentigerous variant where the unilocular: multilocular ratio was 4.3:1.2. For the nondentigerous type this ratio was 1.1:1 (Eversole et al, 1984). The various histological subtypes of unicystic ameloblastoma are determined by the pattern and extent of ameloblastomatous proliferation in relation to the cyst wall. In a clinicopathologic study of 57 cases of unicystic ameloblastoma, Ackermann et al (1988) classified this entity into the following three histologic groups:

- Group I: Luminal UA (tumor confined to the luminal surface of the cyst);
- Group II: Intraluminal/plexiform UA (nodular proliferation into the lumen without infiltration of tumor cells into the connective tissue wall);
- Group III: Mural UA (invasive islands of ameloblastomatous epithelium in the connective tissue wall not involving the entire epithelium)

On histopathology of the present case, luminal, intra luminal and mural type of ameloblastomatous proliferation was present. This type of UA can be better classified as subgroup 1.2.3 according to Philipsen & Reichart (1998) who have categorized UA into:

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

In the present case the luminal areas of the tumour satisfied Vickers & Gorlins criteria (1970) with presence of prominent subepithelial hyalinization. The intraluminal ameloblastomatous proliferation resembled plexiform pattern. The intramural ameloblastoma tissue was seen as an infiltration from the cyst lining or as free islands of follicular solid multicystic ameloblastoma (SMA), often with central cystic degeneration.

Due to the presence of mural proliferations tumour should be treated aggressively in the same manner as the classic SMA (Stoelinga & Bronkhorst 1988).

Bibliography:

- Ackermann GL, Altini M, Shear M: The unicystic ameloblastoma: A clinicopathologic study of 57 cases. *Journal of Oral Pathology & Medicine*, 1988; 17(9-10):541-546.
- Eversole LR, Leider AS, Strub D: Radiographic characteristics of cystogenic ameloblastoma. *Oral Surgery Oral Medicine & Oral Pathology*, 1984; 57(5):572-577.
- Kahairi A, Ahmad RL, Wan Islah L, Norra H: Management of large mandibular ameloblastoma - A case report and literature reviews. *Archives of Orofacial Sciences*, 2008; 3(2):52-55.
- Li TJ, Wu YT, Yu SF, Yu GY: Unicystic ameloblastoma: A clinicopathological study of 33 chinese patients. *The American Journal of Surgical Pathology*, 2000; 24(10):1385-1392.
- Philipsen HP, Reichart PA: Unicystic ameloblastoma: A review of 193 cases from the literature. *Oral Oncology*, 1998; 34(5):317-325.
- Reichart PA, Philipsen HP, Sonner S: Ameloblastoma: Biological profile of 3677 cases. *European Journal of Cancer (Part B: Oral Oncology)*, 1995; 31B(2): 86 - 99.
- Reichart PA, Philipsen HP: Unicystic ameloblastoma. Odontogenic tumors and allied lesions. 1st Edn.; Quintessence Publication. Co. Ltd London, 2004; pp77-86.
- Robinson L, Martinez MG: Unicystic ameloblastoma: A prognostically distinct entity. *Cancer*, 1977; 40(5): 2278-2285.
- Stoelinga PJ, Bronkhorst FB: The incidence, multiple presentation and recurrence of aggressive cysts of the jaws. *Journal of Craniomaxillofacial Surgery*, 1988; 16(4):184-189.
- Vickers RA, Gorlin RJ: Ameloblastoma: Delineation of early histopathologic features of neoplasia. *Cancer*, 1970; 26(3): 699-710.