

CASE REPORT

An Unusual Case of Ameloblastoma arising from Dentigerous Cyst

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ABSTRACT

Ameloblastoma is a benign odontogenic neoplasm that frequently affects the mandible. The term “ameloblastoma” includes several clinicoradiological and histological types. Apart from the most commonly encountered clinicopathologic models, there are few variants, whose biological profile is unknown or not elicited. Among these types, unicystic ameloblastoma is the least encountered variant of ameloblastoma. Unicystic ameloblastoma refers to those cystic lesions that show clinical, radiographic, or gross features of a jaw cyst, but on histologic examination show a typical ameloblastomatous epithelium lining the cyst cavity with or without luminal and/or mural tumor proliferation. Ameloblastoma has been reported to arise from a dentigerous cyst on rare occasions. In this paper, a case of ameloblastoma arising from epithelium of a dentigerous cyst is presented in light of the histopathologic findings, providing evidence for ameloblastomatous change in the preexisting cyst in a 40-year-old female.

Keywords: Ameloblastoma, Benign, Dentigerous cyst, Multicystic.

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INTRODUCTION

Ameloblastoma is a rare, benign, slow-growing, locally invasive neoplasm of odontogenic origin involving the mandible (80%) and maxilla. The neoplasm was 1st discovered by Cusack in 1827. Etymologically, the name derives from the old French word “amel,” which means enamel, and the Greek word “elastos,” which means germ or bud. Overtime, this tumor has been referred to by many different names, including “cystosarcoma,” “adamantinoepithelioma,” “adamantinoma,” and “ameloblastoma.”¹ It is the most common odontogenic tumor, accounting for 10% of all such tumors. It usually occurs

in young adults 20 to 40 years old with almost equal distribution among men and women. Although ameloblastomas are defined as benign neoplasms, they are locally aggressive and infiltrative. If resection is incomplete, they may persist locally, reoccur, or are rarely metastasized.² Ameloblastomas may arise from various sources: (1) cells from the rest of enamel organ; (2) cells of the sheet of Hertwig’s epithelial cell rests of Malassez; (3) epithelium of odontogenic cyst, particularly a dentigerous cyst; (4) basal cells of the oral mucosa; and (5) heterotrophic epithelium of other parts of the body, perhaps pituitary.^{1,2} The 2005 World Health Organization (WHO) classification of ameloblastomas includes four subtypes: The solid/multicystic (91%) type followed by the unicystic (6%), extraosseous ameloblastoma (2%), and desmoplastic (1%) types. The most aggressive clinical and pathologic association is seen in the solid/multicystic type; it is also associated with the highest recurrence rate (90%).¹ Here we report a case of large ameloblastoma arising from dentigerous cyst.

CASE REPORT

A female patient aged 40 years reported to our department with chief complaints of swelling on left side of the face from 4 years and pain on the same side of the face from 3 months. The patient first noticed the swelling on the left side of the mandible 4 years ago, which was of peanut size and had gradually increased to the present size over a period of time. The patient reported associated symptoms like difficulty in chewing and trismus. She also reported recurring pain in the same region, which had sudden onset and was moderate in intensity, intermittent in nature, throbbing, and nonradiating type. No aggravating and relieving factors were reported. According to the patient, she was treated by a local physician for pain. Every time she was given an injection, following which the pain subsided but the swelling did not subside. Pain used to reoccur after 15 days, and the patient visited the physician again. Medical and family histories were noncontributory. The patient was a regular pan chewer since the last 25 years. She brushed her teeth with salt and wooden stick. General examination revealed that all the vital signs were within the normal range. Extraoral examination revealed gross asymmetry of the face. On inspection,

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solitary diffuse swelling, irregularly shaped, measuring 8 × 10 cm in size with ill-defined margins, was present on the left mandible/facial region (Fig. 1). The skin over the swelling was normal in color, stretched, and appeared smooth. Anteroposteriorly, it extended from the left parasymphysis to 2 cm beyond the left angle of mandible. Superoinferiorly, it extended from the line joining the ala of the nose to the left ear lobe till 2 cm below the lower border of the mandible. On palpation, there was no rise in temperature, the swelling was nontender, and other inspectory findings were confirmed. It had variable consistency from soft to firm and was also slightly compressible. The skin over the swelling was pinchable. Anesthesia was given in the lower lip. Intraorally, a solitary swelling was present in the left posterior region of the mandible. It measured 2.4 × 4 cm in size. Buccal vestibule was obliterated. The color of the mucosa over the swelling was normal. Margins were ill defined. The swelling extended from the lower left 2nd premolar region to the retromolar area. The swelling also extended along the anterior border of ramus, which had caused slight tilting of maxillary molar teeth, 26, 27, 28. Impingement of upper teeth was evident over the swelling. Buccal and lingual cortical

plates were expanded. 37 was displaced buccally (Fig. 2). On palpation it had variable consistency and was non-fluctuant and immovable. Based on the history and clinical examination, a clinical diagnosis of a benign odontogenic tumor undergoing cystic degeneration was considered. The differential diagnosis list included ameloblastoma, calcifying epithelial odontogenic tumor, odontogenic keratocystic tumor, and dentigerous cyst. Required radiographic examinations were conducted, which revealed large well-defined multilocular radiolucency with well-defined hyperostotic border. The multilocular radiolucency extended from 2nd molar region involving entire ramus. Additional features like thinning of lower border of mandible, expansion of the buccal and lingual cortical plates, cyst in cyst appearance, resorption of 2nd premolar and 1st molar roots, displacement of teeth, and resorption of left side of the condyle were noticed (Figs 3 to 6). The radiographic differential diagnosis included were unilocular ameloblastoma, dentigerous cyst, odontogenic keratocystic tumor, giant cell granuloma, central hemangioma, odontogenic myxoma, and ossifying fibroma. An odontogenic keratocyst may contain curved septa but usually tends to grow along the bone



Fig. 1: Extraoral swelling



Fig. 2: Intraoral swelling with buccal vestibular obliteration and buccally tilted molar

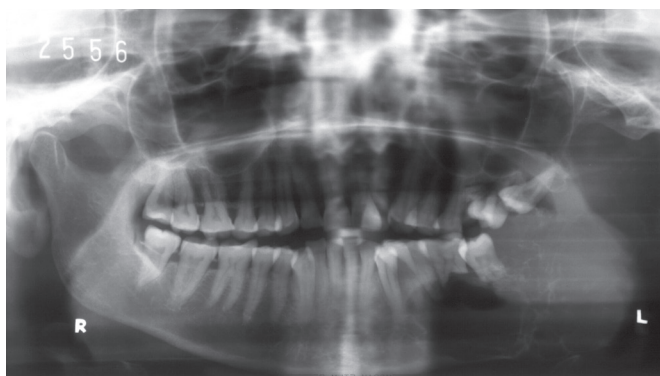


Fig. 3: A well-defined corticated multilocular radiolucency in the left posterior mandible with cortical plate expansion and displacement of teeth



Fig. 4: Mandibular occlusal radiograph showing a large multilocular radiolucency with cortical plate expansion and buccally tilted molar tooth



Fig. 5: An intraoral periapical radiograph showing a large multilocular radiolucency

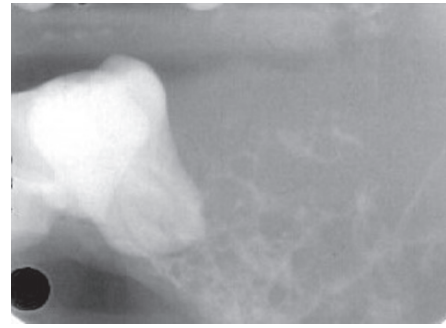


Fig. 6: An intraoral periapical radiograph showing a typical soap bubble appearance

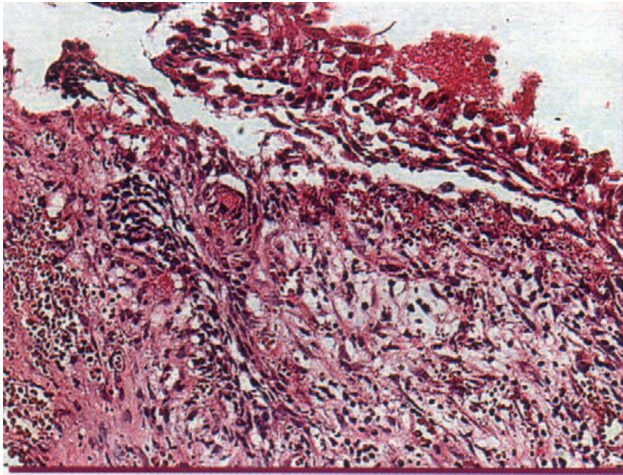


Fig. 7: Epithelial lining resembling dentigerous cyst

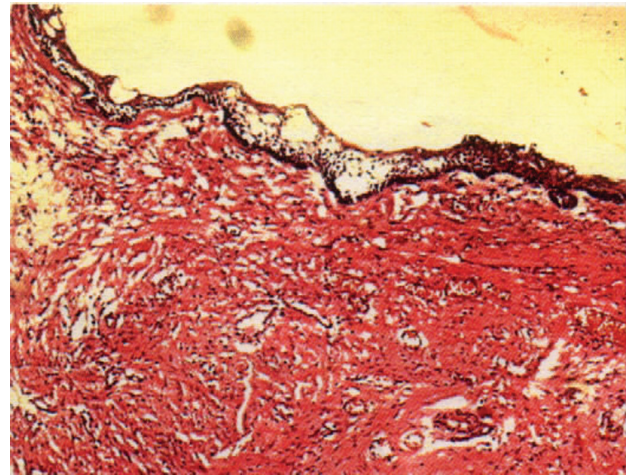


Fig. 8: Islands of odontogenic epithelium displaying peripherally ameloblast-like cells, centrally stellate reticulum-like cells, with areas of squamous metaplasia

without marked expansion as in ameloblastoma. Giant cell granulomas occur in younger age group and have more granular or wispy ill-defined septa. Odontogenic myxoma has one or two thin sharp, straight septa resembling a tennis racket. Septa in ossifying fibroma are usually wide, granular, and ill defined and often have small, irregular trabeculae. Fine needle aspiration cytology and incisional biopsies were performed along with complete blood examination. Fine needle aspiration cytology yielded straw-colored fluid. Incisional biopsy report revealed thin discontinuous epithelial lining resembling that of a dentigerous cyst. Islands of odontogenic epithelium comprising peripherally ameloblast-like cells, centrally stellate reticulum-like cells, along with areas of squamous metaplasia were found. Also foci of isolated tissue showed inflammatory cell infiltrate, blood capillaries, and extravasated RBSs. These features suggested dentigerous cyst with ameloblastic changes (Fig. 7). The histopathologic examination of excised specimen revealed discontinuous epithelial lining having a basal layer of ameloblast-like cells and stellate reticulum-like cells. Islands of odontogenic epithelium displayed peripherally ameloblast-like cells, centrally stellate reticulum-like cells, along with areas of squamous metaplasia. These features

were suggestive of follicular ameloblastoma arising from an odontogenic cyst (Fig. 8). In the end, based on the history and clinical, radiographic, and histopathological examinations, a final diagnosis of follicular ameloblastoma arising from a dentigerous cyst was made. The patient was further referred to the Department of Oral Surgery, and hemi-mandibulectomy was performed and graft was placed; the patient was called again after 10 days. However, the patient did not maintain a proper oral hygiene and the graft became infected. Subsequently, regrafting was done. The patient was called again after few days, and she was found to be doing well.

DISCUSSION

Ameloblastoma is a benign, locally aggressive odontogenic neoplasm with variable clinical expression and accounts for 1% of all cysts/tumors of jaws and 18% of all odontogenic neoplasms. It rarely metastasizes but has a high recurrence (55–90%) if not removed adequately.³ Its peak incidence is in the 3rd and 4th decades of life and has an equal sex predilection. It is often associated with an unerupted 3rd molar. A vast majority of ameloblastomas arise in the mandible, and the majority of these are found in the angle and ramus region.⁴ Global incidence has been

estimated at 0.5 cases per million persons per year.¹ As per the 2005 WHO classification, ameloblastoma is classified based on the difference in biologic behavior, treatment plan, and recurrence rate as follows: Classic solid/multicystic, unicystic, peripheral, and desmoplastic including so-called hybrid lesions and malignant types. It has been reported that the epithelium of the odontogenic cysts may be transformed into benign odontogenic tumors like ameloblastoma.⁵ As in our case it occurred due to ameloblastic transformation of reduced enamel epithelium associated with a developing tooth with subsequent cystic development. Ameloblastomas arising in dentigerous cyst or in others in which neoplastic ameloblastic epithelium is preceded temporarily by a non-neoplastic stratified squamous epithelial lining or a solid ameloblastoma undergo cystic degeneration of the ameloblastic islands, with subsequent fusion of multiple microcysts and develop into unicystic lesions.³ Thus, the epithelium of odontogenic cysts may be transformed into odontogenic tumors like ameloblastoma and odontogenic adenomatoid tumor or into non-odontogenic malignant tumors like epidermoid and mucoepidermoid carcinomas. Ameloblastoma may arise from the remnants of the dental lamina and the enamel organ, or from the basal layer of the oral mucosa as well as the epithelium of dentigerous cyst.^{6,7} Some researchers believe that 5 to 30% of ameloblastomas originate from dental cysts. The walls of dentigerous cysts may present with a proliferation similar to an ameloblastoma, but lack the ameloblastoma-like appearance of the peripheral cells of the true ameloblastoma follicles. On the contrary, in some cases, small mural thickenings of the cyst wall of dentigerous cysts may contain proliferating odontogenic epithelium, and these strands also may show follicular enlargements similar to an ameloblastoma.⁸ Various etiologic factors have been proposed for the ameloblastomas arising from odontogenic cysts, including nonspecific irritational factors (extraction, trauma, infection, inflammation, unerupted tooth), nutritional deficit disorders, and viral infection.⁵ Several reports in the literature have also documented the combined microscopic features of calcifying odontogenic cyst and ameloblastoma merging from one to the other.⁹ Unicystic ameloblastomas (UA) and dentigerous cysts have identical clinical and radiographic appearances. Some subtypes of UA have a better prognosis than that of solid or multicystic ameloblastomas. Unicystic ameloblastomas with small islands of ameloblastomatous epithelium may be misdiagnosed as a dentigerous cyst or keratocyst as in our case.¹⁰ The immune-histochemical data on Ki-67 expression in ameloblastomas, which arise from dentigerous cysts, confirm the hypothesis that ameloblastomas that arise from dentigerous cysts have a biologic behavior, which is similar

to that of UAs.¹¹ The most common presentation for ameloblastoma is a painless swelling in mandible or maxilla with pain being an uncommon feature but can occur because of hemorrhage, especially following a fine needle aspiration. Pain with rapid growth may represent rare malignant ameloblastoma. Tooth displacement and root resorption are infrequent and paresthesia are uncommon with rare cases of perineural invasion have been reported.¹ Differential diagnosis includes keratocystic odontogenic tumor, odontogenic myxoma, nonodontogenic lesions, such as mucoepidermoid carcinoma, and traumatic bone cyst.²

Preoperative diagnostic evaluation includes imaging and possible biopsy. Pantomographic or plain X-ray film examination reveals usually a lytic lesion with scalloped margins, resorption of tooth roots, and impacted molars. The classic "soap bubble" appearance is seen with the most common ameloblastoma, the multilocular/solid type. Although sometimes adequate for complete evaluation, plain X-rays lack sensitivity and specificity for the extent of bone and soft tissue invasion. Computed tomography (CT) is the most useful diagnostic imaging modality, typically demonstrating well-defined radiolucent uni/multilocular expansile lesions. It is also useful for the evaluation of cortical destruction and soft tissue extension identifying the full extent of tumor to support surgical planning. Magnetic resonance imaging (MRI) provides potentially more complete information than CT about soft tissue extension beyond the lytic bone cavity. Magnetic resonance imaging is particularly useful for ameloblastomas arising from the maxilla, as it characterizes extension to orbit, paranasal sinuses, and skull base. Imaging findings are characteristic but not pathognomonic, and the diagnosis is classically established by histology. Histopathologically, ameloblastoma resembles normal odontogenic/enamel epithelium and ectomesenchyme. Microscopic patterns of ameloblastoma include follicular, plexiform, acanthomatous, spindle, basal cell-like, desmoplastic, and granular cell.^{1,2} Surgery is the standard treatment for ameloblastomas. The extent of resection has been controversial, comprising of two surgical options – conservative and radical. The former involves enucleation/curettage of the bony cavity, while the latter involves a radical operation with appropriate margins.¹

Today, one of the points of controversy about this tumor is the differential diagnosis between ameloblastoma showing cystic degeneration and ameloblastomas arising from an odontogenic cyst or the ameloblastomatous hyperplasia in the odontogenic cysts. Many authors suggest that most of the ameloblastomas may arise from the dentigerous cysts and the most likely explanation for the pathogenesis is the ameloblastic transformation of the ordinary dentigerous cyst linings.⁶

CONCLUSION

Odontogenic tumors of the jaws arising from the tooth-forming tissues are uncommon. Although ameloblastoma is the most common of the epithelial odontogenic tumors, it is still as rare as 1% of the tumors and cysts of the jaws, and whether the ameloblastoma arises from the dentigerous cyst or not is still controversial. Thus, a histological examination is the most sensitive tool that can be used for differentiation of dentigerous cysts from unicystic ameloblastomas. Clinical and radiologic findings contribute to the diagnosis. Thus, lesions that clinically and radiographically appear to be odontogenic cysts may prove to be ameloblastomas. Our present case substantiated all the above features.

REFERENCES

1. McClary AC, West RB, McClary AC, Pollack JR, Fischbein NJ, Holsinger CF, Sunwoo J, Colevas AD, Sirjani D. Ameloblastoma: a clinical review and trends in management. *Eur Arch Otorhinolaryngol* 2015 Apr 30.
2. Cankurtaran CZ, Branstetter BF, Chiosea SI, Barnes EL Jr. Ameloblastoma and dentigerous cyst associated with impacted mandibular third molar tooth. *Radiographics* 2010 Sep;30(5):1415-1420.
3. Nagalaxmi V, Sangmesh M, Maloth KN, Kodangal S, Chappidi V, Goyal S. Unicystic mural ameloblastoma: an unusual case report. *Case Rep Dent* 2013;2013:1-6.
4. Goyal N, Pillai AK, Moghe S, Vishnu V, Nahar S. An dentigerous variant of ameloblastoma in mandible – an unusual case report and review of literature. *J Dent Med Sci* 2014 May; 13(5):32-36.
5. Lawal AO, Adisa AO, Olajide MA. Cystic ameloblastoma: a clinicopathologic review. *Ann Ib Postgrad Med* 2014 Jun;12(1):49-53.
6. Ahyan S, Oygur T, Çenetoglu S. Ameloblastoma arising from a dentigerous cyst: a case report. *Gazi Med J* 1998; 9(2):92-95.
7. Savithri V, Janardhanan M, Rakesh S. Unicystic ameloblastoma as a differential diagnosis for odontogenic cysts. *Oral Maxillofac Pathol J* 2014;5(1):466-469.
8. Piattelli A, Iezzi G, Fioroni M, Santinelli A, Rubini C. Ki-67 expression in dentigerous cysts, unicystic ameloblastomas and ameloblastoma arising from dental cysts. *J Endod* 2002 Feb;28(2):55-58.
9. Birdar VG. Ameloblastoma arising in calcifying odontogenic cyst: a rare histologic variant. *Int J Head Neck Surg* 2012; 3(2):106-107.
10. Masthan KMK, Rajkumari S, Deepasree M, Babu NA, Sankari SL. Neoplasms associated with odontogenic cysts. *J Dent Oral Hyg* 2011 Nov 18;3(10):123-130.
11. Bhushan NS, Rao NM, Navatha M, Kumar BK. Ameloblastoma arising from a dentigerous cyst – a rare case report. *J Clin Diagn Res* 2014 May;8(5):23-25.