Case Report

Odontoameloblastoma: A report of a rare case

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Abstract

Odontoameloblastoma (OA) is an uncommon mixed odontogenic tumor that contains an ameloblastomatous component and odontoma-like elements, usually seen to occur in the mandible of younger patients. Radiographically, the tumor shows central destruction of bone with extension of cortical plates and calcified structures which have the radiopacity of tooth structure. These may resemble miniature teeth similar to a compound odontoma or occur as large masses of calcified material similar to a complex odontoma. We report a case of a 17-year-old male with a hard solitary, diffuse swelling over the right lower third of the face for 8 months. Histopathological sections of tumor mass showed diverse and characteristic features of ameloblastoma along with odontogenic epithelium proliferation in unrestrained manner so as to resemble developing tooth bud in stages of morphodifferentiation, apposition and calcification. A diagnosis of OA was made. Hemimandibulectomy was performed on the patient and he remains disease free till today.

Keywords: Ameloblastic odontoma, ameloblastoma, complex odontoma, odontoameloblastoma, odontogenic tumor

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INTRODUCTION

The combination of two odontogenic tumors is seldom seen in the field of odontogenic tumors. The combined or complex odontogenic tumors are lesions characterized by the appearance of classic histological features of the two odontogenic tumors simultaneously.[1]

Odontoameloblastoma (OA) is a remarkably uncommon mixed odontogenic neoplasm with both epithelial and mesenchymal components. It has been defined by the World Health Organization (WHO) and Philipsen and Reichart as: "A neoplasm that includes odontogenic ectomesenchyme in addition to odontogenic epithelium that resembles solid multicystic ameloblastoma (SMA) in both structure and behavior. Due to the presence of odontogenic ectomesenchyme, inductive changes take place leading to the formation of dentin and enamel in parts of the tumor."[2]

OA is a combination of ameloblastoma and odontoma-like structures, occurring together.[2] Befittingly, other names for this tumor previously used were Odontoblastoma (Thoma, 1970), adamant-odontoma (Shafer *et al.*, 1983), calcified mixed odontogenic tumor (Hoffman, 1985), and ameloblastic odontoma (Hooker, 1967). The term OA was included in the 1971 WHO classification.[2,3] Kaugars and Zussmann (1991) described three histologic criteria for OA – unequivocal ameloblastoma, connective

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tissue with mature, homogenous appearance and fragments of malformed calcified dental structures.[2] From 1944, when Thoma *et al*. described the first case, till now, only around 20 cases have been reported in the medical literature, which fit into the above histologic requisites included in the WHO classification 2005 of odontogenic tumors.[2,4,5]

OA has clinical features similar to odontomas in terms of predilection for young male patients, aged 15–20 years, occurring in the posterior segments of either jaw.[4,6,7] However, unlike odontomas, it has potential to produce bone expansion, root resorption and recurrence. The biologic behavior is like the conventional ameloblastoma. The term "odontoameloblastoma" seems to be more appropriate as the tumor behaves more like ameloblastoma rather than an odontoma.[2,3]

Although the tumor is quite a rarity, it is generally thought that OA possesses unique clinical and microscopic features that allow it to be identified from the usual ameloblastomas and odontomas.[4] We present a case of OA in a 17-year-old male with a brief discussion on clinical and histologic differential diagnosis.

CASE REPORT

A 17-year-old male patient reported to the dental outpatient department with hard, solitary, diffuse swelling in the right lower one-third of the face, for 8 months. On extraoral examination, the swelling was approximately 3 cm × 2 cm in size [Figure 1a]. Intraoral examination revealed diffuse swelling extending from 47 to the right retromolar region obliterating the buccal vestibule [Figure 1b]. Orthopantomograph showed multilocular radiolucency on the right side of the mandible from 43 with displacement of 48 till the mandibular notch [Figure 2]. Intraoral periapical radiograph in relation to 46 and 47 showed root resorption. Occlusal radiograph revealed expansion of buccal and lingual cortical plates with radiopaque septae.

A provisional diagnosis of ameloblastoma was proposed on incisional biopsy, and a hemimandibulectomy was performed. Excisional biopsy consisted of hemimandibulectomy tissue specimen extending from 42 [Figure 3]. Tumor mass was extending from 43 to coronoid and condyle. The specimen was grayish-brown in color, soft to firm in consistency, with an irregular surface texture, measuring 12 cm × 7 cm × 8 cm. There was both buccal and lingual cortical plate expansion, with resorption and perforation of the lingual plate. The entire specimen was processed and sectioned.

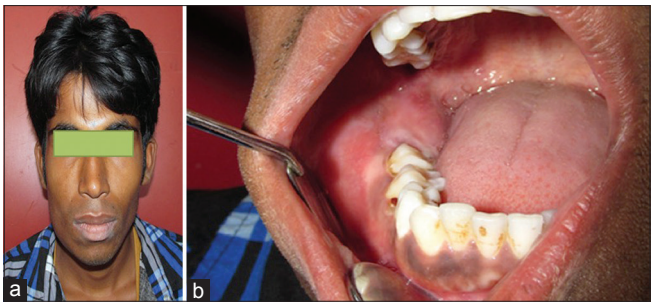

Figure 1: (a) Clinical photograph shows diffuse swelling on the right lower one-third of face (Front view). (b) Photograph showing intraoral swelling involving the right posterior mandibular region obliterating the right buccal vestibule

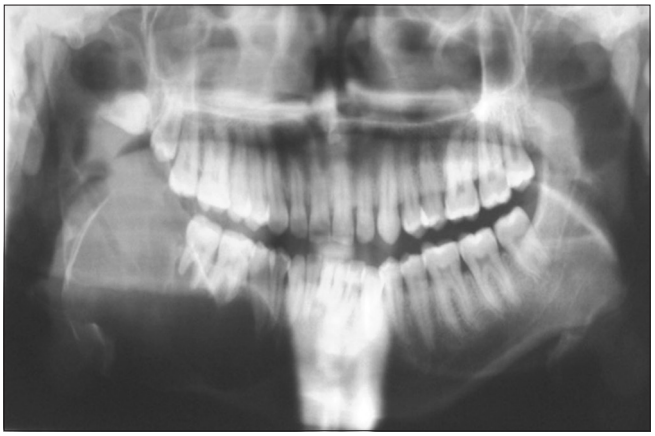

Figure 2: Orthopantomograph showing a well-defined multilocular radiolucency in the right mandibular body-ramus area with the displacement of mandibular third molar to mandibular notch area

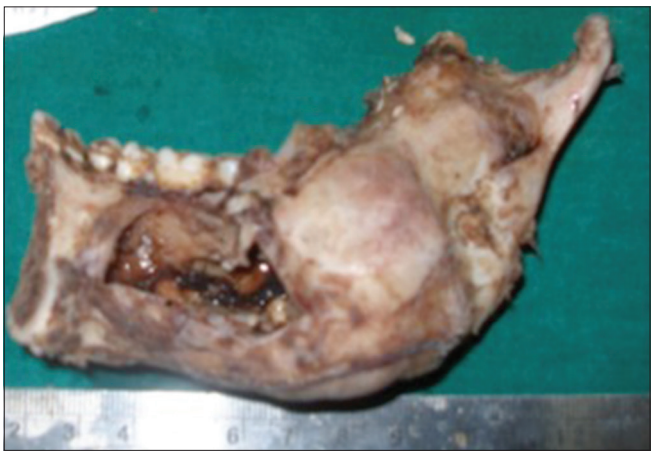

Figure 3: Photograph showing hemimandibulectomy specimen with perforation of lingual bone

Histopathological examination of the hematoxylin- and eosin-stained sections of the specimen revealed proliferating odontogenic epithelium arranged in islands, cords, follicles and interconnecting strands in mature fibrous connective tissue stroma, characteristic of a SMA. Plexiform pattern with reversal of polarity and cystic degeneration of stroma was predominantly seen [Figure 4a]. Odontogenic

follicles are also seen with peripheral tall columnar palisaded ameloblast-like cells showing a reversal of polarity and central stellate reticulum-like areas [Figure 4b]. Numerous developing enamel organ-like structures are seen [Figure 5a]. Plenty of dental papilla-like areas with extravasated red blood cells, areas of hemorrhage and macrophages are seen [Figure 5b]. Basophilic areas resembling enamel are seen [Figure 6a]. It resembles the form of crown [Figures 6b-d]. Large masses of dentin [Figure 7a], dentinoid material [Figure 7b] and enamel/cementum [Figure 7c and d] are seen. Areas showing eosinophilic and basophilic material between tall columnar and mesenchymal cells are also evident [Figure 8]. Calcified tooth-like structure (compound odontoma) composed of dentin with pulp tissue is seen in association with proliferating columnar cells at the periphery [Figure 9]. Osteodentin, Ghost cells [Figure 10] and squamous metaplasia [Figure 11] are also evident. All the features are suggestive of OA.

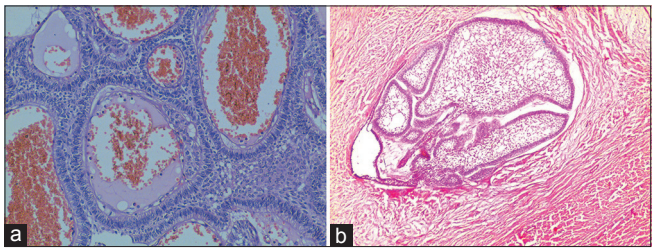

Figure 4: (a) Histopathological image shows odontogenic epithelium in plexiform pattern with reversal of polarity and cystic degeneration of adjacent stroma (H&E, ×40). (b) Histopathological image shows ameloblastic follicles with reversal of polarity of peripheral tall columnar cells and central stellate reticulum-like areas (H&E, ×40)

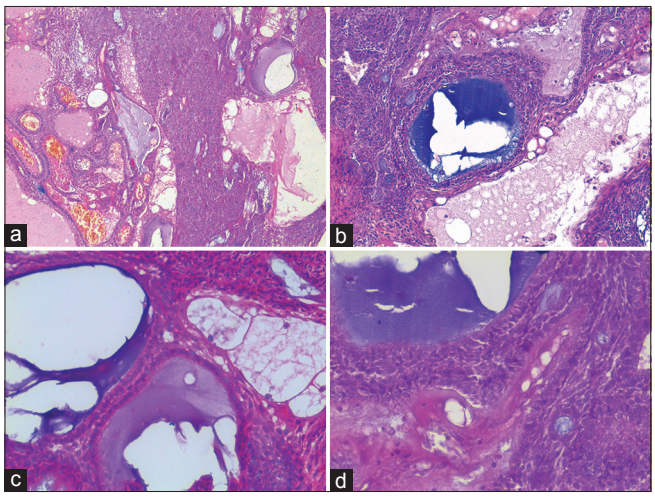

Figure 6: (a) Histopathological image shows enamel matrix and dentin-like material (H&E, ×40). (b) Histopathological image shows enamel matrix in shape of crown (H&E, ×100). (c) Histopathological image shows enamel matrix and dentin in shape of crown (H&E, ×200). (d) Histopathological image shows enamel matrix in shape of crown (H&E, ×400)

DISCUSSION

OA is a mixed odontogenic neoplasm, destructive in its behavior and characterized by simultaneous microscopic presentation of ameloblastoma and odontoma.[3] Its clinical features similar to odontoma in terms of predilection for young age, occurrence in either jaw and tendency to cause bone expansion like ameloblastoma, may aid in clinical differentiation.[4] OA or ameloblastic odontoma has created much debate in literature, as supported by reporting of many lesions with diverse and varied behavior, (ameloblastic fibro-odontomas [AFOs], developing odontomas and OAs) all under the terms of ameloblastic odontoma. To clarify this confused reporting of cases, the WHO in 2005 subdivided the category into AFO, ameloblastic fibro-dentinoma (AFD) and OA.[8,9]

There is difference between the clinical behavior of AFO, AFD and OA. AFO and AFD are usually, asymptomatic, slow-growing lesions with no propensity for bony expansion, mostly associated with an unerupted tooth

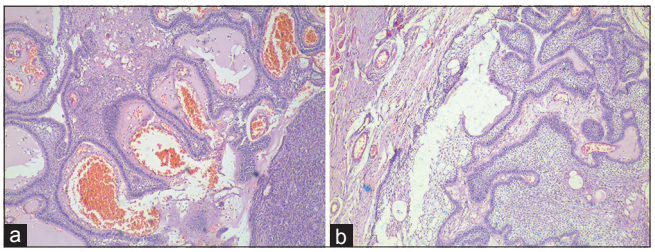

Figure 5: (a) Histopathological image shows enamel organ-like structures (H&E, ×40). (b) Histopathological image shows dental papilla-like areas (H&E, ×40)

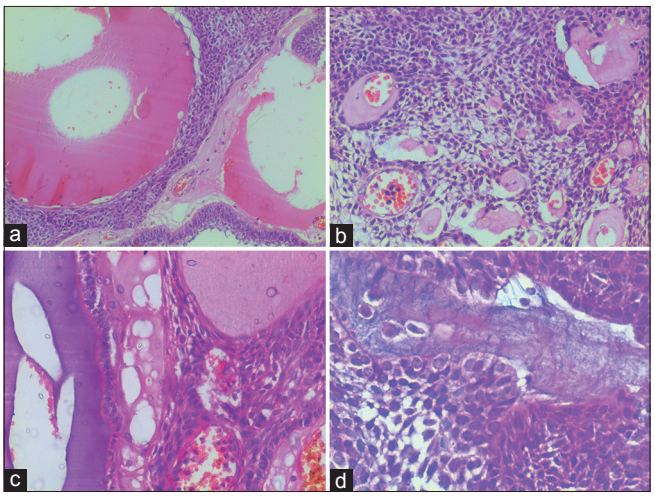

Figure 7: (a) Histopathological image shows dysplastic masses of dentin (H&E, ×200). (b) Histopathological image shows masses of dentinoid-like eosinophilic material (H&E, ×100). (c) Histopathological image shows dysplastic masses of dentin and enamel along with tall columnar cells (H&E, ×200). (d) Histopathological image shows cellular cementum-like material (H&E, ×200)

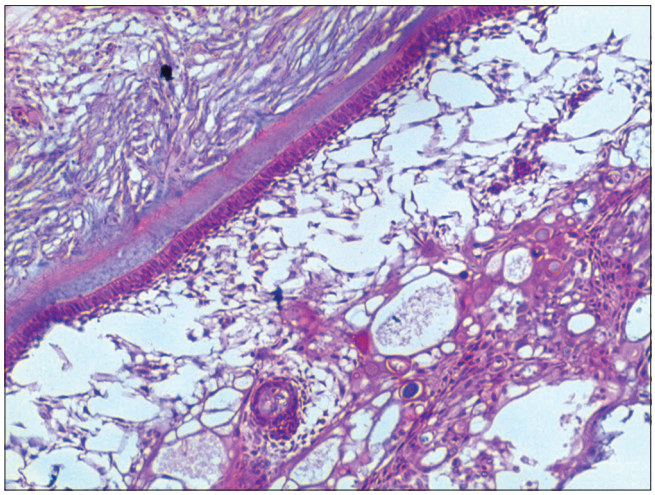

Figure 8: Histopathological image shows eosinophilic and basophilic material adjacent to tall columnar and mesenchymal cells (H&E, ×200)

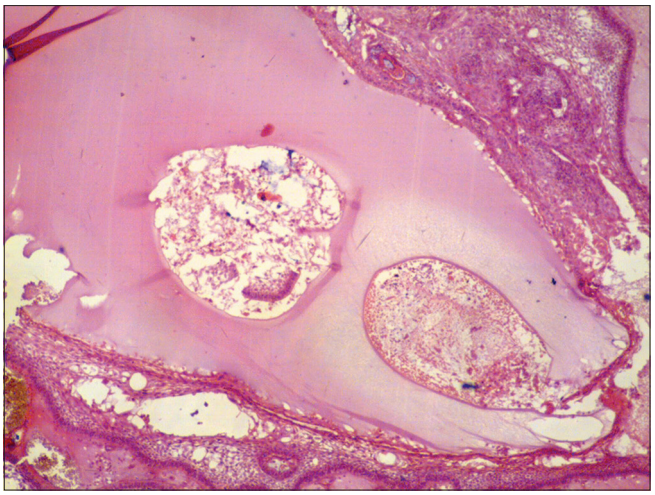

Figure 9: Histopathological image shows a tooth-like structure with dentin and pulp tissue evident (H&E, ×100)

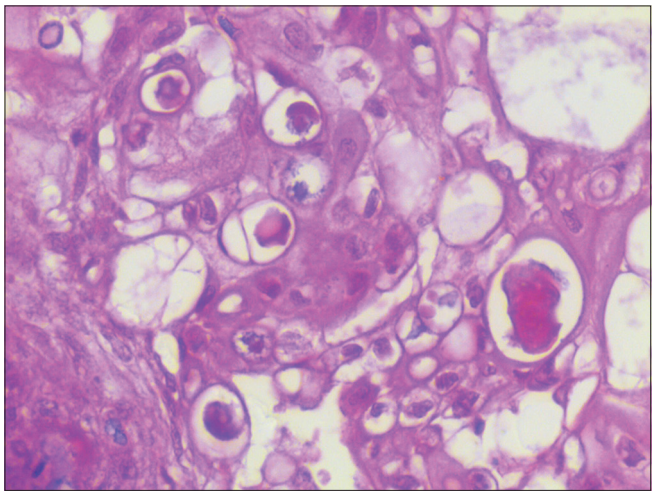

Figure 10: Histopathological image shows ghost cells (H&E, ×400)

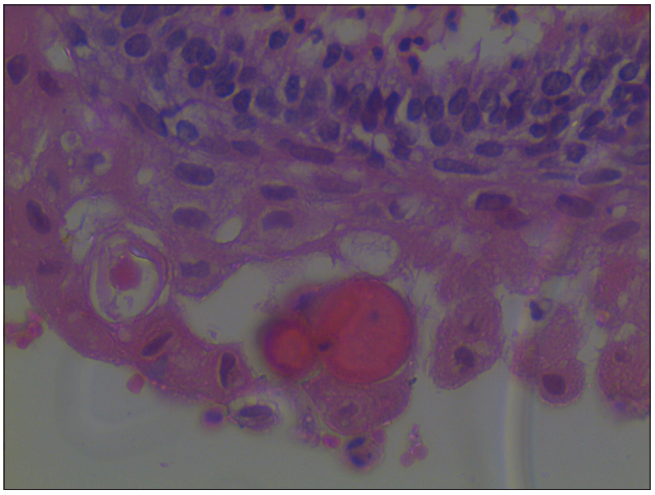

Figure 11: Histopathological image shows squamous metaplasia of the odontogenic epithelial cells (H&E, ×400)

and can be treated with enucleation without much danger of recurrence.[2,10-12] However, OA is a locally invasive, aggressive odontogenic tumor, which spreads by infiltration between the bony trabeculae. It might cause divergence and resorption of the roots.[6,9-11] Sometimes, there might be dull or intermittent pain.[12]

Radiographic features of OA include well-defined unilocular or multilocular radiolucencies with radiopaque calcified masses, which may or may not resemble a tooth. Hence, the clinical differential diagnosis should include lesions with mixed radiographic patterns such as AFO, AFD, calcifying epithelial odontogenic tumor, adenomatoid odontogenic tumor, calcifying odontogenic cyst (COC), odontoma and central ossifying fibroma.[1,6,9]

The histopathology of the OA consists of solid and cystic areas of ameloblastoma showing follicular or plexiform pattern. These are present in mature connective tissue along with odontogenic epithelial proliferation in the form of cords, islands and nests. Unlike conventional ameloblastoma, there is the inductive production of dentin, cementum and enamel. These dental tissues might be haphazardly arranged as in complex odontoma or may resemble rudimentary teeth as in compound odontoma. Admixed with these, there might be the presence of structures representing various stages of odontogenesis such as enamel organ, dental papilla and pulpal tissue; sometimes, osteodentin, dentinoid and bone might be present.[3,6,8-10,13]

Microscopically, there is a subtle distinction between OA, AFO and AFD.[8] Unlike OA which has a mature connective tissue, both AFO and AFD are composed of loose connective tissue resembling primitive dental papilla intermingled with cords, nests, islands of odontogenic cells and foci of calcified masses. When the calcified masses

are similar to dentin and enamel, then term AFO is used. Whereas AFD consists of dentin matrix or dentinoid material as calcified mass.[10,12]

The presence of ghost cells has been reported in OA; however, it is not difficult to differentiate it from ghost cell containing lesions such as COC and odontogenic ghost cell tumors, which have characteristic phenotype.[2,6] Some of OAs have shown squamous differentiation similar to those observed in keratocyst and COC.[3] Takeda *et al*. have described melanocytic differentiation in the epithelial cells of OA.[7] Peripheral OA has been reported in literature, with identical microscopic features to the central OA.[4]

According to literature, OA, a mixed tumor, should show positivity for immunohistochemical (IHC) markers CK14, CK19 and amelogenin, for odontogenic epithelium. Similarly, ectomesenchymal component with its inductive function should show positivity for ameloblastin (unlike ameloblastoma, OA has secretory phase ameloblast-like cells), bone morphogenic protein, nestin and tenascin.[14] However, there are not many IHC studies on evaluation of tumor cells in OA except for analysis of extracellular matrix proteins of the basement membrane by Yamamoto *et al*. They demonstrated high proliferation potential of tumor cells, by positive expression of proliferating cell nuclear antigen and tenascin in the basement membrane of the odontogenic epithelium and hypothesized it to high recurrence rate.[5]

Apart from humans, OA has also been reported in animals such as monkeys, sheep, cats and rats.[7] More elaborate IHC evaluation of tumor cells of OA has been done in Japanese monkeys. The tumor cells showed consistent positive reaction for keratin, cytokeratin AE1, cytokeratin AE3 and epithelial membrane antigen. They were negative for carcinoembryonic antigen, S-100 protein, neuron-specific enolase and glial fibrillary acid protein.[15]

The pathogenesis of OA still remains unknown due to paucity of studies, as most of the OA has been reported as case reports. However, theories have been put forth by researchers. According to Thompson *et al*., the mineralized dental tissues are formed as a hamartomatous proliferation in response to inductive stimuli produced by the proliferating epithelium over the mesenchymal tissue, as seen in a dental follicle.[5]

Choukus and Tots (1964) suggested that OA is a true collision tumor produced by the ameloblastoma and the odontoma, which develop separately but simultaneously at the same topographical area. However, they become enclosed, due to the invasive growth of the odontoma and aggressive nature of ameloblastoma.[11] This hypothesis was discarded by Mosqueda-Taylor *et al*. on the basis of clinical and microscopic features of OA, which show intimate relationship between ameloblastoma and odontoma unlike a true collision tumor.[6]

The latest WHO 2017 classification does not group OA as a separate entity, as there is no sufficient evidence regarding its histogenesis. As association of ameloblastoma and odontoma is well documented and accepted, OA is now described as ameloblastoma arising from primitive ectoderm present in odontoma. Hence, OA is similar to the conventional ameloblastomas arising from primitive ectoderm involved in tooth development. AFO and AFD have been regrouped under developing odontomas rather than as separate neoplastic entities.[16]

OA is unusual in that a relatively undifferentiated neoplastic tissue is associated with highly differentiated tissue, both of which show recurrence after adequate removal. For these reasons, OA should be aggressively treated like a conventional ameloblastoma, with wide surgical excision and close follow-up for at least 5 years.[8-11]

CONCLUSION

We report a case of a 17-year-old male with a hard, solitary, diffuse swelling over the right lower third of the face for 8 months. Histopathology revealed diverse microscopic features characteristic of OA which was treated like conventional ameloblastoma, with hemimandibulectomy. This may be the treatment of choice as the patient remains disease free, without any recurrence till today.

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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