

Developing complex odontoma in a 4-year-old child with active ameloblastic follicles: A case report

Harikrishnan Prasad¹, Gopal Shiva Kumar¹, Muthusamy Rajmohan¹, Korath Varkey²

¹Department of Oral Pathology and Microbiology, KSR Institute of Dental Science and Research, Tiruchengode, ²Dr Paulose Dental Clinic and Maxillofacial Centre, Salem, Tamil Nadu, India

Abstract

The World Health Organization (WHO) classification of odontogenic tumors, in its different iterations, has seen several modifications in benign mixed odontogenic tumors. Odontoma is a benign mixed odontogenic tumor that shows dental hard tissue formation to varying degrees. Although odontomas are the most common odontogenic tumors, they are mostly seen in the form of compound odontomas, where the dental hard tissues resemble tooth-like structures. Complex odontoma shows a haphazard mass of dental hard tissues and is generally identified at an older age (mean age of around 20 years). Incidence of complex odontoma in very young children is quite rare, with only around 10 cases associated with the primary dentition being reported in the literature till date. Unlike other odontomas, our case showed an active epithelial component in the form of tooth buds and ameloblastic follicles along with the hard tissues, which made it difficult to distinguish from other similar lesions like ameloblastic fibroma with hard tissue formation and ameloblastoma arising from an odontoma. This article reports the 11th case of complex odontoma associated with the primary dentition, and the youngest such case to be reported in the Indian population.

Keywords: Ameloblastic fibroma, complex odontoma, developing odontoma, mixed odontogenic tumor

Address for correspondence: Dr. Harikrishnan Prasad, Department of Oral Pathology and Microbiology, KSR Institute of Dental Science and Research, KSR Kalvi Nagar, Thokkavadi, Tiruchengode - 637 215, Tamil Nadu, India.

E-mail: prasad@ksridsr.edu.in

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INTRODUCTION

Benign mixed odontogenic lesions are a diverse group of pathologies that have been difficult to understand, mainly due to the similarity in histopathological features and resemblance to various stages of normal odontogenesis. The 2017 World Health Organization (WHO) classification of odontogenic tumors includes only ameloblastic fibroma (AF), odontoma, primordial odontogenic tumor and dentinogenic ghost cell tumor in this category.^[1] Among these, there has been considerable controversy regarding the nature and the interrelationship between AF

and odontoma. This article is a report of a case of benign mixed odontogenic tumor occurring in a 4-year-old boy, which was difficult to categorize under any one lesion.

CASE REPORT

A specimen from a 4-year-old boy was received in our department. The referring oral surgeon informed us that the child had had a swelling in the left lower jaw for a few weeks, with no other clinical signs or symptoms. The child had deciduous dentition. Radiograph of the

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affected site showed a large 3 cm × 3 cm radiopaque mass of variable intensity [Figure 1]. It was well defined and was seen in the coronal portion of the developing mandibular permanent left first molar, which was seen toward the lower border of the mandible. The entire lesion along with the developing 36 was surrounded by a rim of radiolucency and a peripheral radiopaque border. Adjacent primary teeth (74 and 75) appeared intact with no obvious pathology. Crown of developing 34 was also visible on the radiograph. The surgeon managed to excise the lesion in toto along with the partially developed 36, which could not be preserved.

Grossly, the specimen resembled a hard ball-like structure with a soft tissue attached very closely to the surface. It measured about 3 cm in diameter and was hard in consistency. The capsule was removed carefully from the calcified mass and processed separately. The calcified mass appeared yellowish to grayish in color and irregular in appearance [Figure 2a]. Ground sections and decalcified sections were made from thin slices of this calcified mass. The crown of partially developed 36 was also received. It showed development until the cervical region with intact

and undisturbed dental papilla, and the occlusal surface appeared deformed and wrinkled [Figure 2b].

Ground section under visible light and polarized light revealed irregularly arranged calcified material resembling both enamel and dentin. Enamel-like areas were deep brown in color and showed few rod-like structures. Dentin-like areas were identified by the presence of dentinal tubules, although these appeared very irregular and haphazard [Figure 3]. Decalcified sections revealed dentin-like material throughout, along with some dentinoid areas; empty spaces in between dentin, suggestive of enamel spaces, were also noticed. Loose fibrocellular



Figure 1: Orthopantomograph showing an irregular radiopaque mass covering the unerupted 36 and surrounded by a radiolucent zone

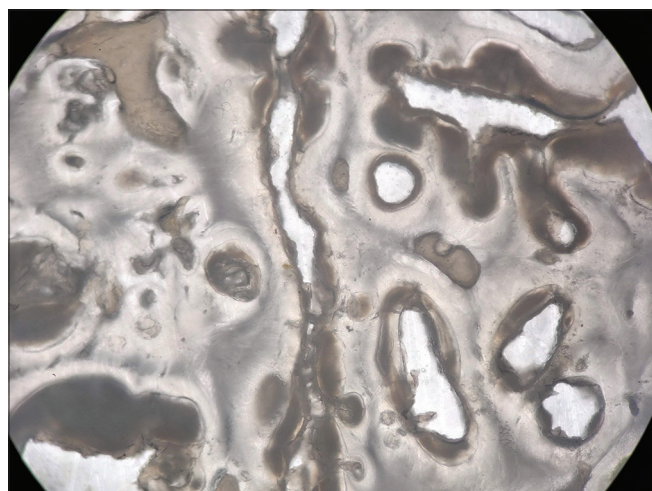


Figure 3: Ground section showing haphazardly arranged enamel- and dentin-like areas

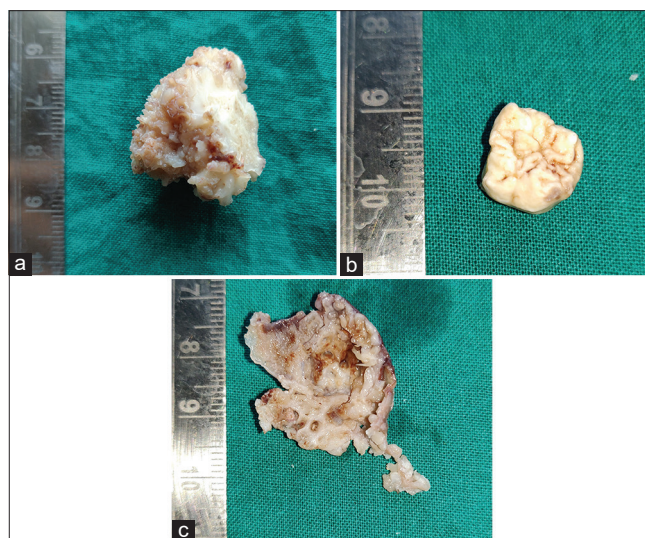


Figure 2: (a) Gross specimen showing an irregular ball-like calcified mass about 3 cm in diameter. (b) The unerupted 36 that was removed along with the lesion showed a wrinkled occlusal pattern. (c) Gross specimen showing the thin soft-tissue capsule that was separated from the calcified mass



Figure 4: Decalcified section of the hard-tissue mass showing entrapped pulp-like connective tissue, with few parallelly arranged columnar cells resembling odontoblasts (H&E stain; x200)

connective tissue was also noticed between the calcified areas. Parallely arranged columnar cells with polarized nuclei, probably suggestive of odontoblasts, were present in this connective tissue [Figure 4].

The histopathology of the soft-tissue capsule revealed dense parallely arranged collagen fibers, along with a discontinuous odontogenic epithelium on the lesional side. The odontogenic epithelium showed varied morphology. In some areas, the epithelial cells were flattened or cuboidal in shape, resembling reduced enamel epithelium. Few areas showed one to two layers of tall columnar cells with a well-polarized nucleus, probably suggestive of ameloblasts. Some areas showed proliferating epithelium resembling bud stage of tooth development, with peripheral low columnar cells and central stellate reticulum-like cells. Few large follicles of odontogenic

epithelium with peripheral columnar cells and central stellate reticulum-like cells, resembling ameloblastic follicles, were also noticed [Figures 5-8]. The connective tissue adjacent to the epithelium showed inductive changes in many places. Connective tissue also showed myxomatous changes in some areas, while few areas also showed a focal collection of plump fibroblasts.

The varied spectrum of histopathological features observed renders the diagnosis difficult in this case. It could be argued in favor of an AF or an odontoma or an ameloblastoma arising from odontoma.

DISCUSSION

Confusion and disagreement regarding the grouping

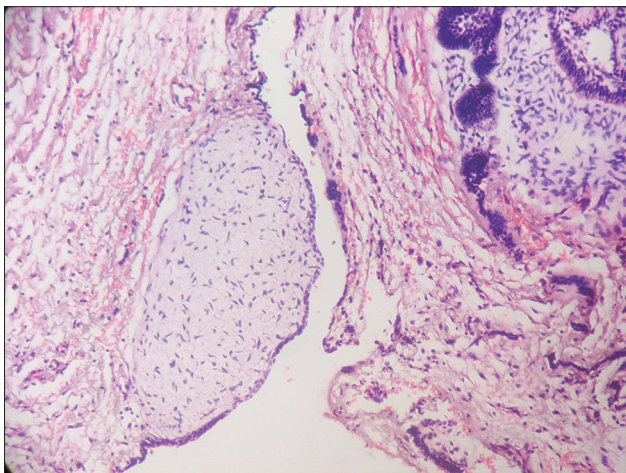


Figure 5: Reduced enamel epithelium-like lining of flattened cells seen on the lesional side of the soft-tissue capsule (H&E stain; $\times 200$)

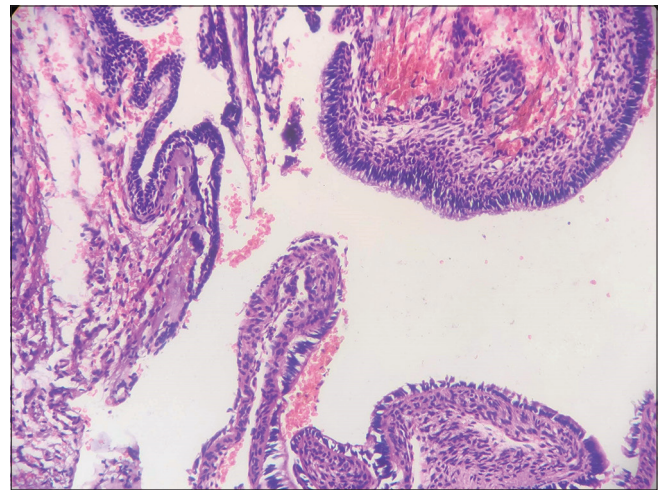


Figure 6: Polarized tall columnar cells resembling ameloblasts were seen lining the connective tissue capsule in some areas (H&E stain; $\times 400$)

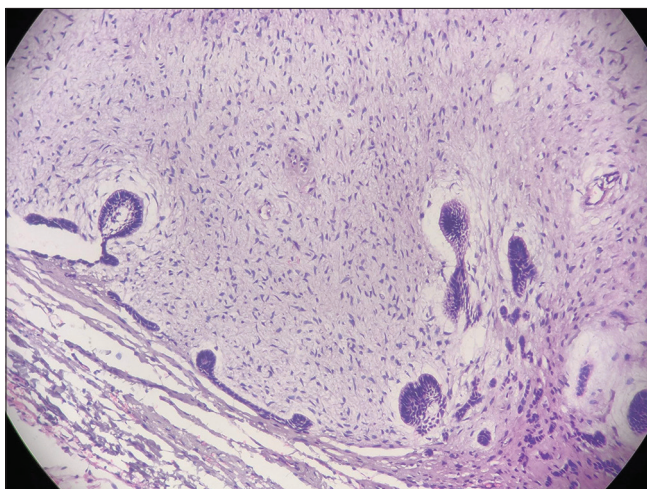


Figure 7: Proliferating epithelium resembling bud stage of tooth development was noticed in some areas of the capsule (H&E stain; $\times 200$)

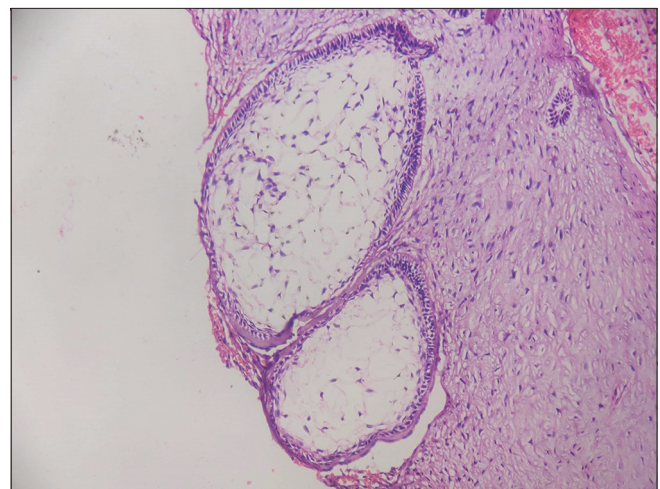


Figure 8: Few ameloblastic follicles with peripheral tall columnar cells and central stellate reticulum-like cells were noticed in the capsule (H&E stain; $\times 200$)

and classification of benign mixed odontogenic tumors still exist.^[2] Although the first classification of odontogenic tumors was published by the WHO in 1971, a separate group labeled benign odontogenic tumor of mixed origin was created only in the second WHO classification in 1992. The recent 2017 classification of the WHO simplified this group and deleted lesions such as ameloblastic fibrodentinoma (AFD), ameloblastic fibro-odontoma (AFO) and odontoameloblastoma while incorporating a new entity called primordial odontogenic tumor.^[1] Much of the confusion in this group stems from the overlapping histopathological features among these lesions.

Odontogenic tumors show inductive interactions between the odontogenic epithelium and the ectomesenchyme. This results in the formation of either abortive or normal-appearing dental hard tissues to different extents. While some evidence suggests that the odontogenic epithelial proliferation and associated hard-tissue formation represents a hamartoma, others suggest that a true neoplastic proliferation cannot be ruled out. However, conclusively identifying whether the lesion is a hamartoma or a neoplasia is almost impossible.

The maturation theory proposed by Cahn and Blum favors the hamartoma line and suggests that an AF will develop in time into an AFD, mature into an AFO and eventually turn into an odontoma.^[3] It has been suggested that once dental hard tissues are formed in an AF, it is programmed to develop into an odontoma in due course. Therefore, AFD and AFO are now considered to be stages in the formation of an odontoma, rather than unique pathologies. Many cases have also been reported in the past, in which recurrent lesions showed no maturational changes, and were compatible with a diagnosis of AF even after several months or years.^[4] This suggests the possible existence of a truly neoplastic AF that does not undergo maturation into an odontoma. Trodahl suggested that the truly neoplastic AF may appear to be somewhat more aggressive and have greater potential for recurrence. However, during the period of odontogenesis (first two decades of life), a lesion diagnosed as AF can either belong to the neoplastic line or the hamartomatous line.

Odontomas are considered to be benign lesions in which all odontogenic tissues are noticed in varying proportions and different degrees of development.^[5] A complex odontoma comprises a well-delineated, roughly spherical mass of haphazard conglomerate of mature dental hard tissues.^[2] Islands of pulp tissue with buds and cords of odontogenic epithelium may be found in the periphery of

Table 1: Reported cases of complex odontoma associated with the deciduous dentition^[6]

Reported by	Patient age	Sex	Location
Hitchin and White (1955)	4 years	Male	Left mandible
Ohtake <i>et al.</i> (1993)	10 years	Male	Right mandible
Motokawa <i>et al.</i> (1998)	3 years	Female	Left maxilla
Hisatomi <i>et al.</i> (2002)	3 years 7 months	Female	Right mandible
Sheehy <i>et al.</i> (2004)	4 years 8 months	Female	Right maxilla
Ozec <i>et al.</i> (2007)	5 years 2 months	Male	Left mandible
Johnson <i>et al.</i> (2007)	7 years	Male	Right maxilla
Losso (2008)	4 years	Female	Left maxilla
Matsuo <i>et al.</i> (2013)	3 years	Male	Left mandible
Gill and Yadav (2014)	7 years	Male	Right mandible
Present case	4 years	Male	Left mandible

the lesion. However, the epithelial component in a complex odontoma is usually inactive at diagnosis. Philipsen noted that when the epithelial component appears to be active and abundant, it might represent a developing complex odontoma or an AF.

In our present case, the hard-tissue component was exceedingly large than the soft-tissue component. The epithelial component in our case was active and arranged in the form of follicles and buds. Only focal areas in the connective tissue showed plump fibroblastic proliferation, while most areas resembled ectomesenchyme-like tissue. These features were against a diagnosis of AF. On the other hand, the exuberant amount of hard-tissue component noticed at such an early age (4 years) makes us question the diagnosis of a complex odontoma. Although odontomas have been reported in a wide age range, very few cases of complex odontoma have been reported in children <5 years of age [Table 1].^[6] Exuberant hard tissue formation (3cm x 3cm size) at this young age (4 years) is extremely rare. This suggests an aggressive or accelerated activity of the odontogenic epithelial tissue in this case.

Factors favoring a developing complex odontoma are a regular oval- or ball-shaped lesion and the relation to the occlusal surface of an impacted tooth.^[7] Although the features are in favor of a diagnosis of AFD or AFO, these lesions are no longer recognized entities, and hence, it is difficult to come to such a conclusion.^[8] Therefore, we are of the opinion that this case could represent an immature form of a complex odontoma that is still in the active developmental stage. Odontogenic epithelium in odontogenic tumors generally has a propensity to proliferate until the end of odontogenesis (around 22 years of age) and tries to recapitulate normal tooth formation. An aberrancy in this process leads to abnormal proliferation of the epithelium, which can induce the underlying ectomesenchyme to produce masses of dental hard tissue in the form of tooth-like structures (compound odontoma)

Table 2: Features in our case that were in favor of or against the diagnosis of the lesions considered

Diagnosis	Factors in favor of	Factors against
AF (turning into an odontoma)	Age (AF usually seen in the first and second decades, although the mean age is around 15 years) Epithelial component resembles ameloblastoma Connective tissue showing inductive changes	Hard-tissue component far exceeding the soft-tissue component, especially considering the duration of the lesion Connective tissue did not appear cell-rich; neoplastic soft-tissue component not noticed
Complex odontoma	Radiographic picture showing a large radiopaque mass with thin radiolucent periphery An insignificant connective tissue component resembling ectomesenchyme Disordered mix of dental hard tissues noticed in the demineralized sections	Age (complex odontomas are usually seen in 2 nd -3 rd decades with a mean age of around 20 years) Epithelial component appears active, with numerous bud-like structures and few ameloblastic follicles
Ameloblastoma arising from odontoma	Epithelial component showing ameloblastic follicles Radiographic and histopathologic evidence of complex odontoma, along with the above	Age (ameloblastoma arising from odontoma tends to arise in 2 nd and 3 rd decades) Clinical presentation of a benign lesion (ameloblastoma arising from odontoma is a locally invasive, aggressive tumor)

AF: Ameloblastic fibroma

or conglomerate masses (complex odontoma). In our case, if the lesion were not diagnosed or intervened at this age, it could probably have progressed into a much larger lesion.

We also considered the possibility of this lesion being an ameloblastoma arising from an odontoma, due to the presence of few discrete ameloblastic follicles with peripheral columnar cells showing polarized nuclei. Ameloblastoma arising from an odontoma, previously referred to as odontoameloblastoma, is a locally invasive, aggressive tumor with a tendency to spread by infiltration between bony trabeculae, and might cause resorption or divergence of tooth roots, with occasional symptoms like pain.^[9] However, none of these features were noticed in our patient. A summary of features in favor of and against the various diagnosis considered is presented in Table 2.

As mentioned earlier, very few cases of complex odontoma associated with the deciduous dentition have been reported till date. Interestingly, Matsuo *et al.*^[10] and Ohtake *et al.*^[11] have reported multiple complex odontomas in young children (a 3-year-old boy and a 10-year-old boy, respectively), with the case reported by Matsuo *et al.* even showing recurrence after incomplete removal. This lends credence to the assumption that the epithelium in an odontoma may sometimes be functionally active at diagnosis, and such cases should be diligently followed up, since they might grow very large if untreated.

As the evidence suggests, the epithelium in our case was active and arranged rarely in follicles, and the connective tissue was predominantly ectomesenchyme-like tissue with abundant haphazard dental hard-tissue formation. Therefore, taking into consideration the age of the child, the clinical features, radiological features and the diverse histopathological features, we arrived at a final diagnosis of a developing complex odontoma with active odontogenic

epithelium resembling ameloblastic follicles. Ours is the youngest case report of complex odontoma associated with the deciduous dentition to be reported in the Indian population.

CONCLUSION

Mixed odontogenic tumors are a set of lesions that have overlapping histopathological features and are likely to be misdiagnosed easily due to their close resemblance to normal odontogenesis. Trodahl in 1972 remarked that it was difficult to identify whether an AF might be hamartomatous or neoplastic and suggested that future research might be able to help. However, almost 50 years later, we are still unable to unravel the mysteries behind this diverse group of mixed odontogenic tumors. This case report of a rapidly growing immature complex odontoma proves this. Incidence at a very young age of 4 years (the youngest reported in Indian literature), the large mass of dental hard tissues, a relatively sparse soft-tissue component and the presence of abundant active odontogenic epithelium histopathologically are some of the features that make this case 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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Nil.

Conflicts of interest

There are no conflicts of interest.

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