

Surgical Conundrum: Hybrid Odontogenic Tumours

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ABSTRACT

Odontogenic tumours are known for their histopathologic variations because the odontogenic tissues themselves form by a series of complex interactions among epithelial and mesenchymal components. They are delicately regulated in a timely manner. The histopathological diagnosis of odontogenic tumours can be difficult to establish due to the complex clinicopathological findings and confusion in their nomenclature. This is a case report of another combination of ameloblastoma and AOT wherein, the histopathological variant was desmoplastic ameloblastoma which is exceptionally rare with incidence rate of 4%-13% as per the available literature.

KEYWORDS

Hybrid odontogenic tumour; AOT; Ameloblastoma; Desmoplastic ameloblastoma

INTRODUCTION

Odontogenic tumours are known for their histopathologic variations because the odontogenic tissues themselves form by a series of complex interactions among epithelial and mesenchymal components [1]. They are delicately regulated in a timely manner [2-4]. The histopathological diagnosis of odontogenic tumours can be difficult to establish due to the complex clinicopathological findings and confusion in their nomenclature. Moreover, unusual combination of neoplastic components, that is hybrid odontogenic tumours are hardly seen. In fact, the occurrence of hybrid odontogenic tumours with multiple histologic types is well known and many such cases of hybrid odontogenic tumours have been mentioned in the literature. However, their commonness and combinations

are not well understood due to relatively vague histological classifications [5]. That being said, the combination of ameloblastoma and adenomatoid odontogenic tumour (AOT) appear to be very rare.

Only four cases of hybrid ameloblastoma and AOT have been reported previously in literature [6-8]. This is a case report of another combination of ameloblastoma and AOT wherein, the histopathological variant was desmoplastic ameloblastoma which is exceptionally rare with incidence rate of 4%-13% [9]. Adding to this, the desmoplastic variant was a hybrid one with a relative frequency of 4.3% [10] with osseous metaplasia. Such mixed odontogenic tumours present as a dilemma to the surgeon since the treatment modality for each odontogenic tumour differs ranging from enucleation and curettage to complex resections requiring reconstruction.

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Furthermore, it can also influence the overall prognosis of the treatment.

CASE REPORT

A 25-years old female reported to Department of Oral & Maxillofacial Surgery, KLE VKIDS, Belagavi with a chief complaint of a progressively increasing hard swelling in upper left front region of the jaw since the past 6 years. It was not associated with any underlying pain nor was there any history of trauma.



Figure 1: Frontal view.

Patient had no related medical history and gave history of extraction of the left maxillary deciduous canine following which no permanent canine erupted in the oral cavity. The individual was moderately built with normal gait. Vitals were found to be within normal limits. Facial examination showed facial asymmetry due to a diffuse swelling on left side of face with prominent flaring of the left ala of the nose along with obliteration of the nasolabial fold. The skin over the swelling appeared normal (Figure 1). Swelling was nontender on palpation.

Intra oral examination revealed a well circumscribed mass measuring about 3 cm × 2 cm in the labial vestibule extending from midline to distal aspect of 24. The overlying mucosa appeared normal. The lateral incisor was labially tipped and there was a missing canine. On palpation, the swelling was bony hard, non-tender, non-compressible, non-fluctuant and non-pulsatile. The swelling seemed to be arising from the bone. The teeth associated with the swelling were found to be vital and no mobility could be elicited. The patient had no other associated symptoms (Figure 2A and Figure 2B).



Figure 2A: Occlusal view.



Figure 2B: Lateral view.



Figure 3A: OPG showing impacted left maxillary canine associated with a mixed radiolucent and radiopaque lesion.

A clinical diagnosis of dentigerous cyst with a possible impacted canine was made with differentials of a compound odontome, adenomatoid odontogenic tumour (AOT), calcifying epithelial odontogenic tumour, odontogenic myxoma and a fibro-osseous lesion in mind.



Figure 3B: True maxillary occlusal radiograph.

Radiographs (OPG and Maxillary Occlusal Radiograph) (Figure 3A and Figure 3B) showed a diffuse, poorly defined combined radiopaque and radiolucent lesion extending from distal of left central incisor to distal of left first premolar causing distal displacement of the roots. There was an impacted tooth present with 23 on the palatal aspect which was seen on occlusal radiograph.

After clinico-radiological correlation, a tentative diagnosis of adenomatoid odontogenic tumour (AOT) or a fibro-osseous lesion of the maxilla was made. Aspiration of the lesion was non-productive, so an incisional biopsy was done under local anesthesia to come to a definitive diagnosis. Histopathology report was

suggestive of AOT (Figure 4) and the lesion was planned for enucleation.

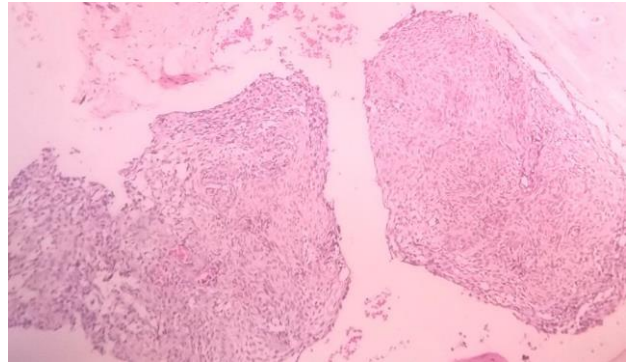


Figure 4: Cellular odontogenic tumour islands showing cuboidal, columnar and spindle cells arranged in sheets and whorls.



Figure 5: Tumour exposed.



Figure 6: Lesion enucleated in tooth with the impacted canine.

Under general anesthesia, after raising a mucoperiosteal flap buccally, the underlying tumour was exposed. A firm, well-circumscribed and nonencapsulated intraosseous mass was seen (Figure 5). The expanded buccal bone was inseparable from the tumour so an osteotomy was performed leaving 5 mm of surrounding normal bone after which the tumour was excised along

with the impacted canine with the help of a chisel and mallet (Figure 6). The underlying bone showed a honey-comb appearance (Figure 7) indicating infiltration into the bone which does not go in favor of adenomatoid odontogenic tumour (AOT). Hence, a peripheral osteotomy was performed with a large round carbide bur till normal bone was seen. After the procedure, the nasal floor was found to be exposed whereas the maxillary sinus and palatal tissues were intact. Since the tumour and the underlying bone did not resemble an AOT clinically, after performing peripheral osteotomy, the cavity was packed with carnoy's solution for 5 minutes to ensure control of micro spread of the tumour. The surgical field was irrigated with copious saline following which a water tight closure was done using 3-0 vicryl.



Figure 7: Intra-operative picture showing honey comb appearance of bone.

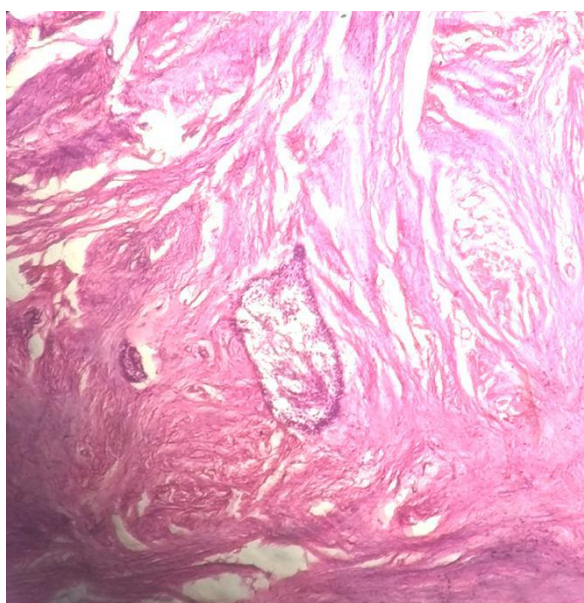


Figure 8A: Ameloblastic follicles in dense collagen stroma.

The excised mass was sent for histopathological examination which was suggestive of hybrid desmoplastic ameloblastoma with osseous metaplasia (Figure 8A - Figure 8C) with adenomatoid odontogenic tumour (AOT).

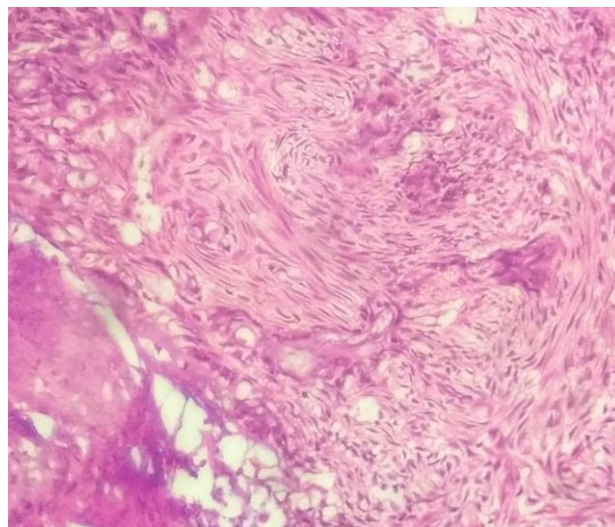


Figure 8B: Hypercellular spindle cells.

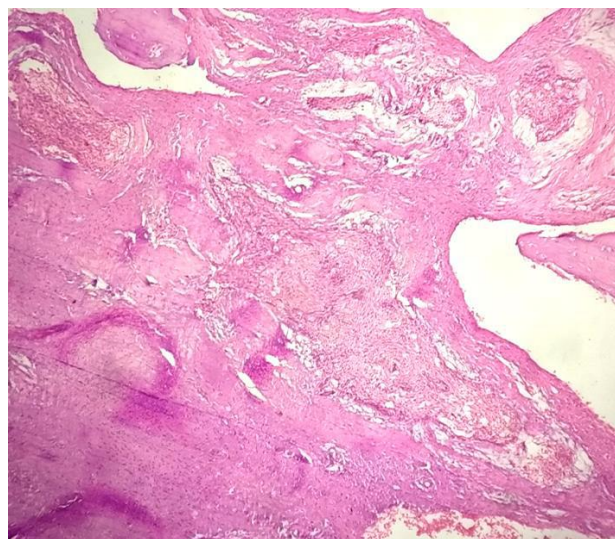


Figure 8C: Irregular stroma with tumour islands and osseous metaplasia.

The patient's post-operative course was uneventful with no recurrence at follow-up period of 3 years (Figure 9).

DISCUSSION

Ameloblastoma is an uncommon tumour of odontogenic origin that accounts for about 1% of all jaw tumours and cysts [9]. Conventional/solid, unicystic, and peripheral/

extraosseous clinico-radiographic subtypes of ameloblastoma are recognized [10], along with the histologic subtypes of which the follicular and retiform patterns are noticed in vast majority of cases.

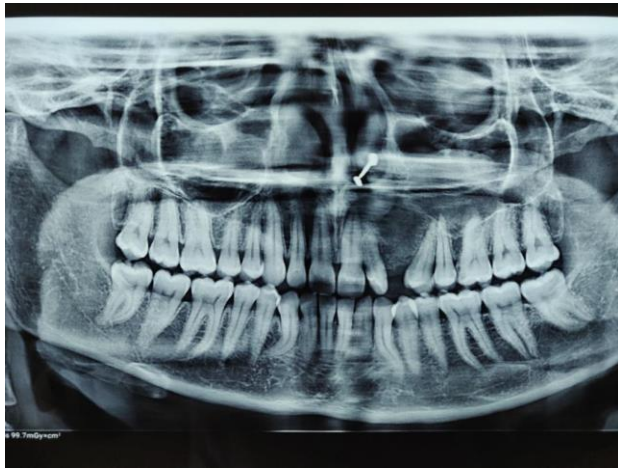


Figure 9: OPG after a period of 3 years showing no signs of recurrence.

The desmoplastic variant is among the recently identified patterns which was introduced in 1984 by Eversole et al. [11]. It is confirmed from the literature that desmoplastic ameloblastoma is the least frequent tumour originating from odontogenic tissues. Amidst the 92 cases of ameloblastoma studied by Takata et al. [12], seven cases were diagnosed as desmoplastic ameloblastoma and only one case as “hybrid lesion”. Waldron and El-Mofty described hybrid ameloblastoma as a tumour variant in which, areas of follicular and retiform ameloblastoma exists simultaneously with typical areas of desmoplastic ameloblastoma [13]. In the analysis by Waldron and El-Mofty, the relative occurrence of hybrid lesions was recorded as 4.3% [13] while in studies done by Higuchi et al. [14] and Takata et al. [12], it was 3.4% and 11% respectively. Philipsen et al. [15] analysed the clinical characteristics of hybrid ameloblastoma lesions. Iwase et al. reviewed 36 cases of hybrid ameloblastoma lesions of desmoplastic ameloblastoma and conventional ameloblastoma that have been recorded in the literature [16]. They hypothesized that secondary desmoplastic alterations take place in the stroma of conventional ameloblastoma or, areas of primary desmoplastic

ameloblastoma transforms giving rise to a conventional ameloblastoma [16]. In Japan, desmoplastic ameloblastoma was found in only 5.3% of all intraosseous ameloblastomas reported over a period of 27 years [17,18]. Ameloblastoma contributed for 1.18% of the total 7,700 tissue samples received over a 25-years period, with desmoplastic ameloblastoma accountable for just 2.2% of all histological forms of ameloblastoma, according to an Indian study. No cases of hybrid ameloblastoma were found [19]. The hybrid desmoplastic ameloblastomas showed a nearly identical sex predilection and surfaced in the age range of 17 years - 82 years. They have been seen in Asians in particular with a fairly high predilection towards mandible [17].

The present case was a 25-years old female patient in whom the lesion was found in the maxillary anterior region. There are two histologic variants of desmoplastic ameloblastoma; the predominant simple desmoplastic ameloblastoma (88.0%) and the rare desmoplastic ameloblastoma with osteoplasia (12.0%) [20,21]. Philipsen et al. in 1992, narrated that when a tumour cell stimuli to stromal fibroblast causes desmoplasia, it also affects osteoblast (another type of mesenchymal cell) giving rise to new bone called osteoplasia [22]. Prominent new bone in the tumour tissue noticed in our case has only been recorded in eight other desmoplastic ameloblastomas till date. [23]. The osteocyte rich metaplastic trabeculae of bone lined by healthy functional osteoblasts showed variation from woven bone to mature bone. Non-neoplastic bone remnants often tend to linger in the tumour tissue. Hybrid neoplasms consisting of two or more distinct histologic forms seldom occur, but among odontogenic tumours, their incidence have been fairly well recognized. A hybrid odontogenic tumour is defined as: “A lesion showing the combined histopathological characteristics of two or more previously recognized tumours and/or cysts of different categories.” [5]. It is exceptionally rare to see a combination of ameloblastoma and adenomatoid

odontogenic tumour (AOT). Literature shows only four cases of hybrid ameloblastoma and adenomatoid odontogenic tumour (AOT) reported previously and this is the second case consisting of solid multicystic ameloblastoma and AOT [6-8]. The present case is a hybrid ameloblastoma composed of areas of desmoplastic ameloblastoma and follicular ameloblastoma along with osteoplasia and adenomatoid odontogenic tumour (AOT). The incisional biopsy specimen showed highly cellular sparse lesional tissue surrounded by compact bone comprising of cuboidal, columnar and spindle cells arranged in sheets and whorls. Focal areas of eosinophilic material were also noted, suggestive of adenomatoid odontogenic tumour (AOT). However, after enucleation, the specimen showed large, irregular odontogenic tumour islands, cords and follicles widely separated with dense fibrous and moderate cellular stroma. The tumour islands show peripheral cuboidal cells with hyperchromatic nucleus and few odontogenic islands show columnar cells with reversal of polarity. The center of odontogenic islands show hypercellular spindle shaped cells with few islands present with eosinophilic amorphous deposits, also few islands show cystic degeneration in the center. The stroma shows moderate cellular and fibrous appearance. The collagen fibers are thick and seem to be compressed by epithelial islands in some areas. Metaplastic bony trabeculae surrounded by functional osteoblasts and lacunae showing osteocytes along with resting and reversal lines are noted. The final histopathological appearance was suggestive of Hybrid desmoplastic ameloblastoma with osseous metaplasia and adenomatoid odontogenic tumour (AOT). Marx et al. opined in the text book of Oral Pathology that desmoplastic ameloblastoma should be treated in the same manner as solid multicystic ameloblastoma (SMA) [24]. Sun et al. [25] opined that more radical approach is required for the ill-defined borders of desmoplastic ameloblastoma. Usually, this non-encapsulated tumour infests in cancellous bone

trabeculae (masking under the healthy-looking cortical bone), which results in high tendency to recur. So, curettage is never an appropriate treatment for desmoplastic ameloblastoma. The preferred mode of treatment is wide surgical excision as opined by Yoshimura et al, in 2009 [26]. Regarding the clinical behavior of desmoplastic ameloblastoma, WHO classification of odontogenic tumours says that desmoplastic ameloblastoma, like unicystic ameloblastoma and peripheral ameloblastomas can have a lower rate of recurrence than other forms of ameloblastomas [27]. In contrast to this, a review of the literature revealed that desmoplastic ameloblastoma has same recurrence rate (15.9%) as that of other forms of ameloblastomas [25]. Keszler et al. highlighted a higher recurrence rate of desmoplastic ameloblastoma (21.4%) as compared to the non-desmoplastic variants of ameloblastoma (10.1%) [28]. The justification for this may be the fact that radiographically, desmoplastic ameloblastomas are easily confused with fibro-osseous lesions.

CONCLUSION

The precise diagnosis of a desmoplastic ameloblastoma can be difficult to establish before the operation. Moreover, desmoplastic ameloblastoma frequently exhibits ill-defined border making the exact attachment of the lesion with normal bone difficult to investigate. Also, the more common location in the maxilla may cause an early invasion of the adjacent structures. So as was rightly said by Marx and Stern that “The best chemotherapy for odontogenic tumours is a jar of formalin” it applies for this infrequent variant as well. As the clinical nature, radiographic features and histological characteristics of hybrid lesions are not clear till date, the lesion requires more detailed tumour analysis, follow-up and reporting in literature.

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None

**CONFLICT OF INTEREST & FINANCIAL
DISCLOSURE**

None

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