UNICYSTIC ADENOID AMELOBLASTOMA: A DIAGNOSTIC RARITY

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Abstract

Adenoid ameloblastoma (AA) is a newly recognized entity among odontogenic lesions, introduced in the 5th edition of the WHO classification of odontogenic tumours. It is an epithelial odontogenic tumour characterized by a cribriform pattern with duct-like formations and dentinoid presence. Previously considered a hybrid lesion due to its varied histopathological features, the condition sparked debate regarding its nature, behaviour, and prognosis. The updated classification has clarified its diagnostic criteria, fostering a deeper understanding of its pathology and distinguishing it as a distinct entity.

This report highlights a case of a 22-year-old female presenting with swelling in the upper left front tooth region. Histopathological examination confirmed adenoid ameloblastoma. The redefinition and categorization of AA have improved diagnosis and evidence-based management. This case underscores the significance of updated literature in advancing clinical practice and enhancing the comprehension of lesions with diverse histopathological presentations.

Keywords: dentinoid material, hybrid tumor, adenomatoid odontogenic tumor, unicystic ameloblastoma, adenoid ameloblastoma

1. INTRODUCTION

A diverse category of tumors known as "odontogenic tumors" develops only in the jaw bones and/or gingival mucosa. These tumors exhibit a broad spectrum of biological behaviors that necessitate varying levels of surgical intervention, ranging from straightforward excision and enucleation to extensive resections[1]. Odontogenic tumors, like other tumor categories, are classified according to the World Health Organization (WHO) classification system[2].

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Occasionally, odontogenic tumors that are difficult to diagnose and defy classification based on current knowledge are observed, even though most odontogenic tumors can be diagnosed using the criteria provided in the WHO classification of head and neck tumors[3]. Hybrid tumors are one such entity. A hybrid odontogenic lesion (HOL) is a lesion that affects the same primary site and combines the histological features of two or more known odontogenic cysts and/or tumors. Due to their unclear clinical behavior and controversial histogenesis, these lesions pose a challenge to oral pathologists and surgeons [4]. One example is hybrid odontogenic tumors that exhibit characteristics of both ameloblastomas and adenomatoid odontogenic tumors with dentinoid, also referred to as adenoid ameloblastoma. Adenoid ameloblastoma is an odontogenic tumor with hybrid characteristics, exhibiting histopathological features of both ameloblastoma(AM) and adenomatoid odontogenic tumors (AOT) [5].

This report aims to highlight the diagnostic challenges of adenoid ameloblastoma through a case study. This report seeks to demonstrate the diagnostic difficulties of adenoid ameloblastoma through the presented cases and to review the existing literature supporting its recognition as a distinct subtype of ameloblastoma.

2. CASE REPORT

A 22-year-old female patient presented to the dental OPD with a chief complaint of swelling on the left side of the face, which gradually increased to the present size in 2-3 months. There was no relevant medical or family history.

Extraoral examination revealed a solitary swelling measuring 3 cm × 2 cm in the greatest dimension, extending from the ala of the nose to the mid-cheek region on the left side anteroposteriorly and from the left infraorbital region to the lower border of the mandible superior inferiorly. The left corner of the mouth was deviated with obliteration of the nasolabial fold. The skin over the swelling was stretched, and the overlying skin color was similar to that on the other side. Palpatory findings revealed that the swelling was firm, non-tender, non-fluctuant, and there was no localized rise in temperature.

On intraoral examination, the swelling measured 3 cm × 4 cm × 2 cm in the greatest dimension.

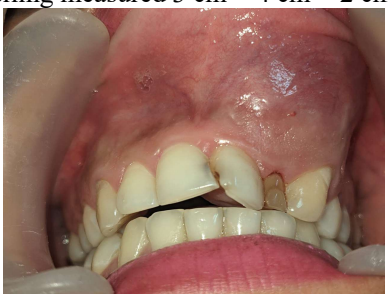

Figure-1: Intraoral Photograph Showing Localized Swelling

The swelling was expansile in nature. Retained 62 was noted. Crepitus was present on palpation in relation to 62, 23, 24, and 25. There was a clinically missing 22.

3. INVESTIGATIONS

Fine Needle Aspiration Cytology (FNAC) yielded 3 ml of hemorrhagic fluid, which on Papanicolaou (PAP), Giemsa, and Leishman staining showed neutrophils, lymphocytes, and cystic macrophages in a proteinaceous background. The cytomorphological features were suggestive of an inflammatory pathology.

Radiographic examinations using orthopantomography (OPG) revealed a well-circumscribed unilocular radiolucency with a sclerotic border circumscribing the impacted 22, pushing the floor of the maxillary sinus and deviating from the midline. Radiolucency was extending from 11 to 25, causing root displacement of 21, 23 and 24 along with resorption of 25, with bicortical expansion ranging from 21 to 25

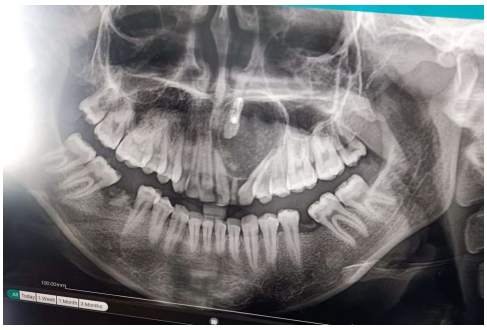

Figure 2: OPG Revealing a Unilocular Lesion Involving Impacted 22 causing Displacement of 21, 23, 24.

In addition, 3D imaging of Cone Beam Computed Tomography (CBCT) revealed a well-defined mixed radiolucent lesion involving 11, 12, 21 to 26, and 62

Figure 3: 3D Imaging(CBCT) of the Lesion.

The lesion was 29.36 mm buccolingually and 28.22 mm mesiodistally. A Breach in the cortical plates and displacement of roots 11, 12, and 21, and poorly defined borders and impacted 22 noted in the lesional area. An excisional biopsy was performed under local anesthesia after obtaining consent from the patient. Cystic lining was removed in toto along with extraction of impacted 22 and 62, 23, 24 & 25. Gross examination revealed multiple soft and hard tissue specimens and bony flakes.

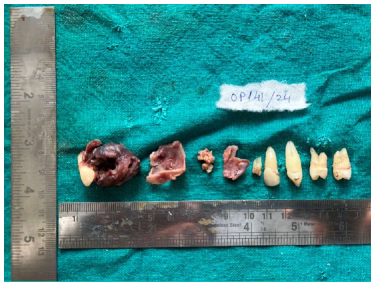

Figure 4: Gross Image of the Specimen.

The specimen measured 4x2x2.5 cm. It was firm in consistency with a cystic lining attached to 22 at the cemento-enamel Junction (CEJ), brownish in color, and a cystic cavity showing brownish material. The tissue was then subjected to routine processing. Histopathological examination revealed cystic lining epithelium and features of adenoid ameloblastoma. The histopathological features were suggestive of Unicystic Adenoid Ameloblastoma.

4. DIFFERENTIAL DIAGNOSIS

Differential diagnosis was challenging because this is a new entity. However, some differentials that could be possible were AOT, Unicystic Ameloblastoma, hybrid tumor, and Hemangiomatous Ameloblastoma. They were ruled out based on histopathological findings such as the presence of dentinoid, duct-like structures, and stellate reticulum-like areas along with thin cystic lining.

5. TREATMENT

Enucleation of the cystic lesion was done, and whole tissue was excised along with 21,62, 23, 24 & 25. In addition, the cystic lining was removed in toto, along with the impacted 22. There was no evidence of recurrence till the patient last reported for a follow-up after 6 months.

6. OUTCOME & FOLLOW-UP

The resected tissue was sent for further evaluation to the department of oral and maxillofacial pathology. On meticulous examination, a thin cystic lining around 2-3 cell layered thick resembling reduced enamel epithelium was noted.

Figure 4: 4x Photomicrograph of H&E Stained Section Showing Thin Epithelial Lining & Connective Tissue Wall.

In one focal area, cells resembling the stellate reticulum were observed in the cystic epithelium. In a few focal areas, the connective tissue capsule revealed a pattern resembling AOT-like areas, including

cystic spaces and glandular/ductal structures lined by cuboidal cells. Areas of rosette, whorled, and cribriform patterns were noted.

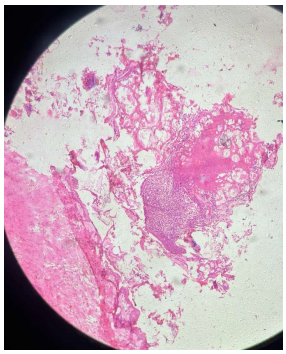

Figure 5: Photomicrograph reveals A. Stellate Reticulum Like Areas, B. Cystic and Glandular Structure Resembling AOT.

However, one area showed a loose arrangement of cells near the stellate reticulum. Areas showing eosinophilic material were also noted, consistent with dentinoid structures.

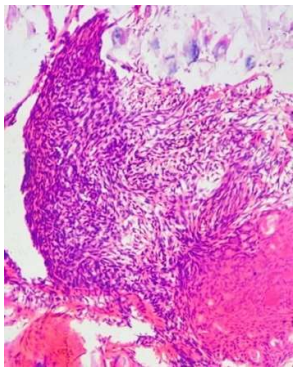

Figure 6: Stellate Reticulum Like Cells with AOT Like Areas.

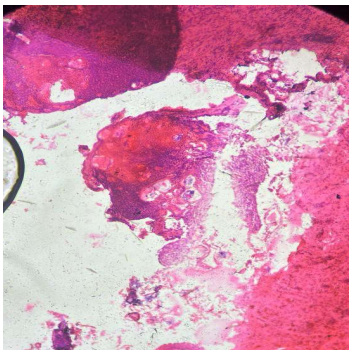

Figure 7: Photomicrograph Showing Cystic Spaces with Eosinophilic Coagulum.

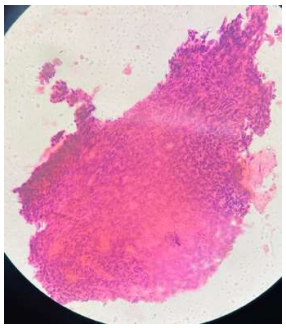

Figure 8: Photomicrograph Showing Arrangement of Cells in Rosette Pattern.

Therefore, it was considered a hybrid tumor, as sufficient data are not available on this type of lesion. However, in 2022, the WHO introduced this entity with the abovementioned features, which were present in this case as Adenoid Ameloblastoma. Hence, the final diagnosis of Unicystic Adenoid Ameloblastoma was made based on the presence of both cystic lining and ductal structures.

7. DISCUSSION

Waldron's 1959 identification of the adenoid characteristics of ameloblastoma (AM) laid the groundwork for subsequent findings. Slabbet et al. first used the name "dentino ameloblastoma" in 1992. In 1994, Brannon coined the term “adenoid ameloblastoma with dentinoid” at the Armed Forces Institute of Pathology [6].

In a single lesion, two or more different tumor types may coexist, known as hybrid tumors. These lesions display a mix of histological characteristics from two or more previously identified tumors and/or cysts that fall into various categories, according to Ide et al. [7]. However, these tumors were excluded from the 2017 WHO classification of odontogenic neoplasms because of the small number of reported instances [8].

A hybrid odontogenic tumor can exhibit both AM and AOT histological traits. This specific hybrid tumor was initially identified as a dentinoameloblastoma by Slabbert et al. in 1992 [9].

Since its first description, these lesions have been referred to by several names, such as hybrid ameloblastoma and AOT arising within a unicystic ameloblastoma, atypical plexiform ameloblastoma with dentinoid, ameloblastoma with dentinoid features, and atypical adenoid ameloblastoma (AAD). A retrospective analysis by Loyola et al. highlighted the possibility that AAD may be disregarded [10], wherein four of the forty-five cases that were first diagnosed as AM were reassessed and reclassified as AAD. It implied that the literature may greatly underreport the actual incidence of these cases.

Accurate diagnosis can be difficult because AM and atypical adenoid ameloblastoma (AAD) share several common histological characteristics. The aggressive character and broad development potential of this tumor are shown in the seven documented cases in which the lesion crossed the midline and affected both sides of the entire dental arch [6]. Our case is unique because it is unilateral and does not cross the midline.

Only two of eight cases had multilocularity, according to Adorno-Farias et al. [11]; however, our case stood out from previously reported cases because of its unique unilocular radiolucency. Only one documented instance, as described by Salehinejad et al., exhibited characteristics of desmoplastic ameloblastoma (AM) [12]. In contrast, squamous metaplasia was consistently observed in each of the eight cases reported by Adorno-Farias et al. [11].

The other element frequently linked to ameloblastoma (AM) in cases of atypical adenoid ameloblastoma (AAD) is the adenomatoid odontogenic tumor (AOT). The biological process that allows these two different tumor forms to coexist is unknown. The conversion of one lesion into another is a tenable pathogenic explanation; however, further research is needed to support this notion [13].

Alongside the ameloblastoma (AM) and adenomatoid odontogenic tumor (AOT) components, an extracellular homogenous dentinoid material of varying quantities is regularly observed in atypical adenoid ameloblastoma (AAD) [14]. In 1992, Slabbert et al. made the first suggestion that the eosinophilic material in atypical adenoid ameloblastoma (AAD) was dentinoid [9]. A similar dentinoid substance was observed in the present case. Using Van Gieson and Masson's trichrome stains, the study showed that the eosinophilic material stains favorably for collagen. However, Congo Red and formic acid-Alcian blue staining yielded negative results for amyloid and keratin, respectively. Sonone et al. further verified the collagenous nature of the dentinoid in atypical adenoid ameloblastoma (AAD), noting negative Congo Red findings and positive Van Gieson staining [15].

It has been hypothesized that the conversion and co-expression of the mesenchymal phenotype allow odontogenic epithelial cells devoted to ameloblastic development, including neoplasms, to form dentinoid material [16].

According to Khalele and Al-Shiaty's description of atypical adenoid ameloblastoma (AAD), there has only been one documented instance of cellular atypia and aberrant mitosis [4]. It becomes more difficult to accurately determine the actual frequency of AAD in reported cases when these malignant symptoms are present since there is a chance that some instances of AAD with these characteristics could be misinterpreted as ameloblastic carcinoma.

It was recently shown that AA does not contain BRAF and KRAS mutations, which are frequently found in histologic mimics of AA, such as adenomatoid odontogenic tumors and ameloblastomas. Instead, nuclear accumulation of β-catenin and CTNNB1 hotspot mutations have been found in patients with AA. A characteristic of WNT pathway activation is the nuclear translocation of β-catenin, which stimulates transcription and several cellular functions. Similarly, subgroups of WNT-activated odontogenic lesions, such as AA, have been shown to lack CTNNB1 hotspot mutations [2].

Alcian blue and mucicarmine showed positive staining in part of the cystic or duct-like areas in atypical adenoid ameloblastoma (AAD), indicating the presence of mucin, according to Matsumoto et al.[15]. Furthermore, in six out of eight instances, Adorno-Farias et al. observed high immunopositive staining for CK14 in several tumor island central and peripheral cells, as well as in the surrounding cells and adenoid structures [11].

Furthermore, according to Loyola et al., CK8/18 demonstrated localized positivity in three of five cases of atypical adenoid ameloblastoma (AAD). Although this varied across AAD cases, most reports showed that the proliferation marker Ki-67 expression was borderline to low. To precisely determine the type of lesion and forecast its prognosis, an immunohistochemistry (IHC) panel containing proliferative markers was used. Surgeons can use this information to decide on the best course of treatment. It is interesting to note that even after surgical treatment and follow-up, more than 50% of AAD cases experienced repeated recurrences despite the typically low proliferative indices seen in IHC analyses [10].

8. CONCLUSION

Adenoid ameloblastoma shares demographic characteristics with conventional ameloblastoma but differs in histopathological features and is associated with a higher recurrence rate. Molecular findings further support its classification as a distinct entity, separate from ameloblastoma and adenomatoid odontogenic tumors (AOT). However, its diagnosis poses a challenge for oral pathologists because of the presence of duct-like structures and dentinoid formation, which may mimic AOT. In some cases, the predominance of AOT-like areas can overshadow ameloblastomatous components, leading to potential underdiagnosis or misdiagnosis. Therefore, a meticulous examination of histological slides is crucial. Accurate diagnosis is essential, and future research incorporating genetic analysis is warranted to better understand the biological behavior of these tumors. Given the evolving nature of terminology and classification of odontogenic tumors, the use of immunohistochemical (IHC) markers is imperative, especially in cases exhibiting overlapping histological features.

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