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Case Report

Hybrid Ameloblastoma of the Maxilla: A Puzzling Pathology

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Abstract

Ameloblastomas are slow growing, locally invasive, benign odontogenic tumors of an epithelial origin, accounting for approximately 1% of all oral tumors. A 40-year-old man presented with a chief complaint of a swelling over the left side of his face of 4 years' duration. On examination, gross facial asymmetry was detected, and a well-defined swelling was noted intraorally involving the left maxilla medially from the mid palatal raphe and obliterating the buccal vestibule laterally. The swelling was non-tender and exhibited dual consistencies: firm in the palate and cystic in the vestibular region. Computed tomography revealed a multilocular radiolucency, which involved the left maxilla, encroached into the left maxillary sinus and the nasal complex, and caused bony erosion. Early diagnosis and treatment are the key tools in managing ameloblastomas, failure of which may lead to a significant deterioration of the prognosis and an increased recurrence rate. Uncommon variants of ameloblastomas have been gaining interest recently. To date, 25 cases of hybrid ameloblastomas have been documented in the scientific literature. We present an extremely rare hybrid type of the ameloblastoma with combined follicular, cystic, acanthomatous, and desmoplastic variants, which render it the first of its kind to have ever been reported.

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Keywords • Ameloblastoma • Maxillary sinus • Maxillary neoplasms • Odontogenic tumors

What's Known

- The most common site of involvement for a hybrid ameloblastoma is the mandible. Radiographically, it demonstrates mixed radiolucent radiopaque lesions with irregular borders and exhibits a more aggressive behavior than other variants.
- Histopathological features showing a combination of desmoplastic and conventional variants of ameloblastomas are known as the hybrid ameloblastoma.

What's New

- Our case occurred in the maxilla, which is a very rare site, and radiographically it depicted an unusual presentation with multilocular radiolucency.
- Histopathologically, it is considered the first of its kind with a rare combination of desmoplastic, follicular, cystic, and acanthomatous variants of ameloblastomas.

Introduction

Ameloblastomas are the most common neoplasms of the jaws, and they arise from the odontogenic epithelium. They begin as an asymptomatic swelling of the jaws and gradually cause the mobility, displacement, and root resorption of the involved teeth. Consequently, they expand extensively into the cortical bones, resulting in functional compromise.[1,2]

The most common histopathological types of ameloblastomas are the follicular and plexiform types, followed by the acanthomatous and granular cell types. Waldron and El-Mofty discovered a unique variant of the ameloblastoma in 1987, displaying a combined histology of desmoplastic and conventional ameloblastomas as a hybrid variant. Only a few cases have been reported worldwide, accounting for about 1.1 to 4.3% of ameloblastomas.[3] Our search of the existingliterature demonstrated that the present case is the first of its kind inasmuch as it has a unique combination of follicular, cystic, acanthomatous, and desmoplastic variants of

the ameloblastoma and involves a very unusual site (i.e., the maxilla).

Case Report

A 40-year-old man presented to the outpatient department of Drs. Sudha and Nageswara Rao Siddhartha Institute of Dental Sciences, Gannavaram, India, in May 2014, with a chief complaint of a swelling over the left side of his face of 4 years' duration. The patient revealed a history of trauma sustained at the same site 11 years previously, followed by an asymptomatic period of 4 years. He then noticed a painless swelling in the anterior region of the maxilla, for which he underwent extraction of 21 and 22. The swelling recurred after 3 years at the same site with associated dull pain on wide opening of the mouth. On extraoral examination, a well-defined swelling was seen on the left side of the face measuring 5×4 cm in size. The surface of the swelling was smooth and shiny. The obliteration of the nasolabial fold on the left side and a deviated nasal septum to the right side were noted. On palpation of the swelling, no local rise in temperature was noted and the swelling was non-tender and firm in consistency (figure 1A).

On intraoral examination, a well-defined swelling was noted involving the left maxilla and measuring 6×5 cm. The overlying surface had areas of erythema interspersed with a bluish dilated capillary vessel. On palpation, the swelling was non-tender and revealed cystic consistency in the vestibular region, although it was firm in the palatal region (figure 1B). Fine-needle aspiration was performed, which was nonproductive, and hematological investigations were within the normal limits. Panoramic radiography showed a large, irregular, osteolytic lesion demonstrating a multilocular radiolucency in the left maxilla. Wispy bone trabeculae were seen extending from 21 to 27 regions with an irregular soap-bubble appearance. The margins of the radiolucency were well-corticated and separated by thick bony septae (figure 2A).

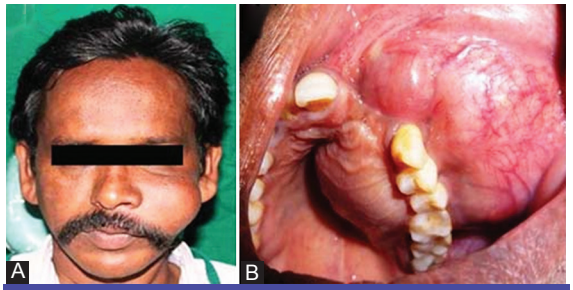

Figure 1: A) Extraoral swelling involves the left cheek. B) Intraoral swelling is seen in the left maxilla.

Axial computed tomography images demonstrated a large, well-defined, multilocular, expansile lesion with thin, corticated, bony walls. The internal structure of the lesion revealed multiple thin septae in the left maxilla extending superiorly from the infraorbital margin and inferiorly involving the alveolar ridge. The lesion extended medially to the lateral border of the nasal septum and the palatal process of the maxilla as far as the mid palatine raphe, encroaching the entire left maxillary sinus and laterally expanding into the buccal cortical plate (figure 2B). The lesion showed a bulge into the oral cavity involving the maxillary sinus and roots of all the teeth on the affected side with a deviated nasal septum to the right side. The osteomeatal complex and frontal recesses were blocked on both sides. Radiological differential diagnoses included ameloblastoma, odontogenic myxoma, and central salivary gland tumor.

After a complete examination of the patient, the surgical resection of the tumor and hemimaxillectomy were performed under general anesthesia. The excision was done with a wide surgical margin, preserving the orbital floor (figure 3A). A split thickness graft was placed to close the surgical defect, and an immediate obturator was placed over the graft (figure 3B).

The excised specimen was sent for histopathological examination (H & E 20X), which revealed islands of cells in which the peripheral cells were tall and columnar with subnuclear vacuolations and a peripheral prominent palisading appearance of basal cell

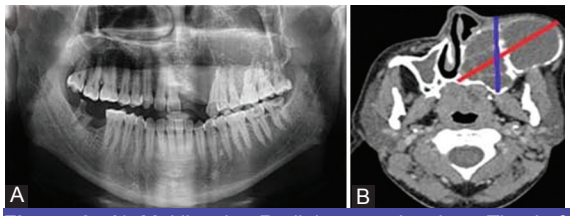

Figure 2: A) Multilocular Radiolucency Involves The Left Maxilla And The Adjoining Sinus, Exhibiting A Soap-Bubble Pattern. B) Axial Computed Tomography Image Shows A Multilocular Expansile Lesion Encroaching Into The Sinuses And The Nasal Cavity.

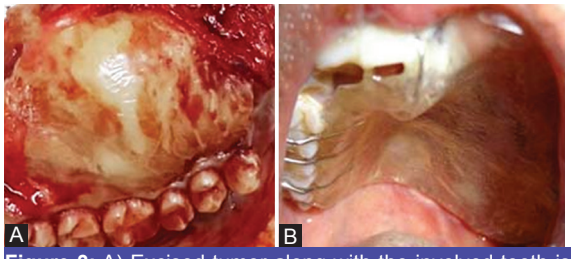

Figure 3: A) Excised tumor along with the involved teeth is depicted here. B) Placement of the obturator to close the surgical defect is illustrated here.

nuclei with a reverse polarization (black and green arrow). The central cells appeared to be stellate reticulum-like cells with a few islands showing central necrosis, and there were large areas of cystic degeneration between the cells (red arrow). The tumor showed foci of squamous metaplasia (blue arrow) infiltrating the soft tissues and areas of dense hyalinization with focal desmoplasia of the connective tissue (yellow arrow), suggestive of an ameloblastoma with mixed subtypes consisting of follicular, cystic, acanthomatous, and desmoplastic variants (figures 4A, B, and C). The final histopathology picture was consistent with the hybrid type of the ameloblastoma. Written informed consent was obtained from the patient regarding the publication of the current case report along with the accompanying images.

Discussion

Ameloblastomas are common benign but locally destructive neoplasms. The average age at the occurrence of hybrid ameloblastomas is similar to that of conventional ameloblastomas and it varies between the first and the seventh decades of life with a mean in the fourth decade, showing a male predilection.[4] Our patient was male and was in the fourth decade of his life.

According to the literature, the most common site of involvement for hybrid ameloblastomas is the mandible at a ratio of 5:3 when compared with the maxilla.[1] When the tumor occurs in the maxilla, the posterior region is the most affected, which may often compromise the maxillary sinus and the orbit.[5] In the present case, the maxilla was involved, which is a very rare anatomical location for the occurrence of an ameloblastoma. Maxillary tumors spread very quickly compared to mandibular neoplasms due to the thin cortical bone that forms the weak barrier for the spread of the tumor. In addition, their abundant blood supply provides another possible mode of spread. Invasive maxillary ameloblastomas have grave prognoses owing to their extension into the orbit, frontal sinus, skull base, middle cranial fossa, and petrous apex.[6,7]

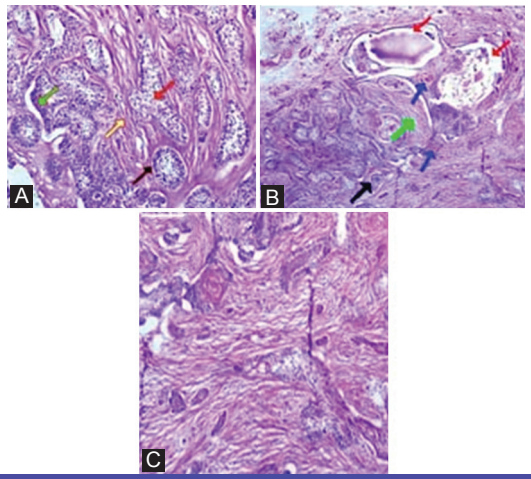

Figure 4: A) H & E 20x section shows follicles (black arrow) and peripheral hyperchromatic palisading ameloblast-like cells (green arrow) with stellate reticulum-like cells (red arrow) in the center intervening with a dense hyalinized desmoplasia connective tissue (yellow arrow). B) H & E 20x section presents follicles with peripheral hyperchromatic palisading-like cells (black arrow) with squamous metaplasia (blue arrow) and a few cells undergoing cystic degeneration (red arrow) with dense hyalinized desmoplasia (green arrow) like connective tissue stroma. C) H & E 20x section depicts stromal desmoplasia.

Philipsen and coworkers[8] proposed that a hybrid variant was a “transitional” form of the desmoplastic type, comprising the microscopic features of both desmoplastic and “classic” follicular or plexiform variants.The hybrid variant demonstrates varied radiographical appearances such as mixed radiolucent and radiopaque lesions with irregular borders similar to the common radiological pattern observed in the desmoplastic variant (with osteoplasia) or fibro-osseous lesions or malignant tumors owing to its high infiltrative nature, while few cases of hybrid ameloblastomas exhibited a multilocular radiolucent pattern, which is similar to that of conventional ameloblastomas,[6,8] as was seen in our patient. The clinicopathological and radiographical features of a few reported cases of hybrid ameloblastomas are illustrated in table 1. However when the ameloblastoma shows a typical expansile multilocular aspect, the differential diagnosis can include a variety of odontogenic or non-odontogenic lesions with similar characteristics like odontogenic keratocysts, aneurysmal bone cysts, adenomatoid odontogenic tumors, odontogenic myxomas, and giant cell central lesions.[5]

Follicular and plexiform ameloblastomas are the most common histological variants and account for 32.5% and 28.2%, respectively, followed by the acanthomatous subtype with 12.1%. However, the desmoplastic variant is extremely rare, with an incidence ranging from 4 to 13%.[3] The synchronized occurrence of the desmoplastic variant with another variant in the hybrid lesion is mysterious. It is considered a puzzling pathology in that it is doubtful whether a part of the primary desmoplastic variant transforms into the conventional variant, whether the desmoplastic change occurs secondarily in the stroma of a pre-existing solid multicystic ameloblastoma, or whether it is a collision tumor.[3,15] Two or more tumors that arise from independent topographic sites are regarded

Table 1: Clinicopathological and radiographical features of a few reported cases of hybrid ameloblastomas

Author & years	Age and gender	Location	Radiological Features	Histopathological features
Wakoh et al., 2002[9]	35/female	Anterior mandible	Mixed radiolucent/radiopaque with adjacent cystic radiolucent areas	Follicular and desmoplastic
Hirota et al., 2005[10]	17/female	Posterior maxilla	Well-defined radiolucent lesion	Follicular, plexiform, and desmoplastic
Desai et al., 2006[11]	32/male	Posterior mandible	Well-defined unilocular radiolucency	Desmoplastic and follicular
Yazdi et al., 2009[12]	48/female	Anterior mandible	Mixed radiolucent/radiopaque	Desmoplastic and follicular
Brooks et al., 2010[13]	66/female	Anterior mandible	Mixed radiolucent/radiopaque	Desmoplastic and acanthomatous
Gupta et al., 2011[1]	35/female	Anterior mandible	Mixed radiolucent/radiopaque	Desmoplastic and follicular
Angadi et al., 2011[14]	64/female	Posterior maxilla	Mixed radiolucent/radiopaque	Desmoplastic and follicular
Effiom et al., 2013[3]	50/female	Anterior mandible	Mixed radiolucent/radiopaque	Desmoplastic and follicular
Rai et al., 2014[15]	50/female	Anterior and posterior maxilla	Mixed radiolucent/radiopaque	Desmoplastic and plexiform
Present case	40/male	Anterior and posterior maxilla	Multilocular radiolucency	Acanthomatous, follicular, and desmoplastic

as collision tumors. The desmoplastic and conventional variants of hybrid ameloblastomas may develop simultaneously, which supports the collision concept.[3]

Histopathologically, hybrid ameloblastomas show areas of extensive stromal collagenization or desmoplasia surrounding compressed islands of the odontogenic epithelium.[8,16] The present case was diagnosed as a hybrid ameloblastoma according to the diagnostic criteria established by Waldron and El-Mofty[10] as it showed histopathological features of the desmoplastic variant along with other variants like follicular, cystic, and acanthomatous ameloblastomas. This finding confirmed a unique hybrid variant, which we herewith report as the first of its kind to the best of our knowledge.

Desmoplastic ameloblastomas demonstrate more aggressive behavior than do the other variants of ameloblastomas. Various attributable facts could be

a) Their potential to grow to a large size
b) Likelihood of maxillary lesions to produce early invasion to the adjacent structures
c) The diffuse radiographical appearance and the histological findings of bone invasion

Finally, it is difficult to treat these ameloblastomas surgically because it is unfeasible to find the exact interface of the lesion with the normal bone.[12]

Smaller lesions can be managed with a conservative approach, whereas radical tumor ablation is preferred for larger lesions. For the unicystic type, this is usually enucleation, whereas for the solid and multicystic variants, it is marginal or segmental surgical excision. Because there are vital structures adjacent to the maxilla, a significant recurrence rate after local curettage has been reported.[17] Recurrences usually present even decades after surgery; therefore, following surgery, close follow-up is recommended during the first 5 years with an interval of once in a year and then once every 2 years with regular radiological investigations for the early detection of recurrence.[8,18] Concerning the present case, following the surgical management of the tumor, the patient is under regular follow-ups with no evidence of recurrence.

Conclusion

We herein presented a unique case of a hybrid variant of the ameloblastoma involving the maxilla with clinical, radiographical, and histopathological features that render it the first of its kind to have ever been reported. This report is to alert practitioners regarding the clinical behavior of such lesions, necessitating a prompt diagnosis and treatment for a better prognosis and prevention of recurrence.

Conflict of Interest: None declared.

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