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Plexiform Ameloblastoma: Report of 2 Cases and Review of Literature

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Abstract

Background: Ameloblastoma is an odontogenic tumor that originates from epithelial cells. Despite its benign classification, it exhibits local aggressiveness and the potential for unlimited growth. The 2022 World Health Organization (WHO) classification identifies five distinct subtypes of ameloblastoma, each displaying varying biological behaviors. Plexiform ameloblastoma is a subtype of conventional ameloblastoma that is characterized by a unique histopathological appearance. Unlike the typical follicular or acanthomatous patterns, plexiform ameloblastomas exhibit a network of interconnected strands or cords of odontogenic epithelium that resemble a "plexus" or network. This paper aims to present two clinical cases of conventional plexiform ameloblastoma, provide a review of the current literature, and detail the clinical, radiological, and histopathological features of this condition, as well as appropriate therapeutic approaches. **Case presentation:** This article presents two cases of plexiform ameloblastoma, a prevalent subtype of ameloblastoma, with one arising in the maxilla and the other in the mandible. The cases are analyzed in the context of existing literature to shed light on the unique features and clinical implications of this subtype. **Conclusion:** Plexiform ameloblastoma is a rare and benign odontogenic tumor that may reach gigantic proportions. Thus, ensuring an accurate diagnosis is crucial to minimizing the risk of local recurrences and obtaining an effective treatment.

Subject Areas

Pathology

Keywords

Ameloblastoma, Plexiform, Diagnosis, Differential Diagnosis

1. Introduction

Ameloblastoma is a slowly developing, histologically benign tumor that was first

identified by Cusack in 1827. The term "ameloblastoma" was adopted in 1929, combining "amel" for enamel and "blastos" for germ. This neoplasm accounts for approximately 1% of all tumors and cysts of the jaws and represents 13% - 78% of odontogenic tumors. It is most observed in regions such as Africa, India, and China [1]-[3].

While ameloblastoma does not exhibit a clear sex predilection, there have been occasional reports of higher frequencies in males. The average age of onset is typically between 30 and 60 years [4] [5].

The fifth edition of World Health Organization (WHO) classification of head and neck tumors (2022) categorizes five subtypes of ameloblastomas (extraosseous/peripheral ameloblastoma, metastasizing ameloblastoma, unicystic ameloblastoma, conventional ameloblastoma and adenoid ameloblastoma) based on their location, histological characteristics, and biological behavior [6] [7].

This paper presents two clinical cases of plexiform ameloblastoma, a prevalent variant of conventional ameloblastoma. It aims to provide a comprehensive review of the current literature, detailing the clinical, radiological, and histopathological features of this condition. Additionally, it will discuss suitable therapeutic approaches for managing plexiform ameloblastoma.

2. Cases Series

2.1. Case 1

A 54-year-old man presented at the oral surgery department complaining of a painless swelling in his right maxilla that had been appeared in the last 10 months. The patient reported no significant medical history.

Extraoral examination revealed tumefaction over the right maxillary region and obliteration of the corresponding nasolabial fold. No cervical lymphadenopathy was noted (See **Figure 1**).

Intraorally, a firm and non-fluctuant enlargement was observed, extending from the distal aspect of the canine to the maxillary tuberosity, affecting both the buccal and palatal cortical plates. The overlying mucosa appeared normal, exhibiting no secondary changes.

Noticeable mobility was observed in the right first upper molar, with altered vitality noted in the 14 and 16. While, the maxillary anterior teeth were found to be intact and non-mobile (See **Figure 2**).

Orthopantomography (OPG) and cone-beam computed tomography (CBCT) imaging revealed a large, well-circumscribed, unilocular radiolucency measuring approximately 5 cm in the right maxillary region. The lesion extended from the distal aspect of the canine to the tuberosity region completely resorbed with sinus invasion and perforation of the vestibular and palatal cortex from the16 to the retro-tuberosity region.

Notably, there was no evidence of root resorption in the teeth affected by the lesion, although root displacement was observed in the right maxillary first molar (See **Figure 3**).

An incisional biopsy was performed under local anesthesia at the site of the 17 and the spiceman was sent for histopathological analysis, which revealed: a plexiform ameloblastoma predominantly composed of epithelium arranged in long anastomosing strands and cords (See Figure 4 and Figure 5).

A partial maxillectomy was performed via the intraoral approach, with a safety margin of 1.5 mm of uninvolved bone. The patient's postoperative recovery was uneventful, and no signs of recurrence were observed during the 5-year follow-up examinations.

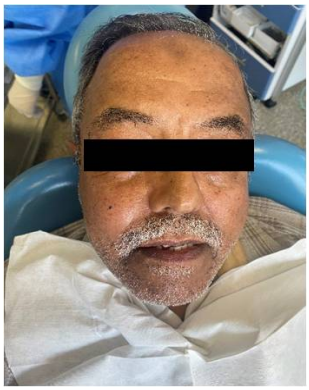


Figure 1. Extraoral presentation shows diffuse tumefaction involving the right maxillary region.

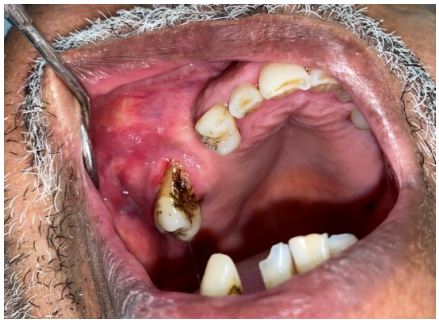
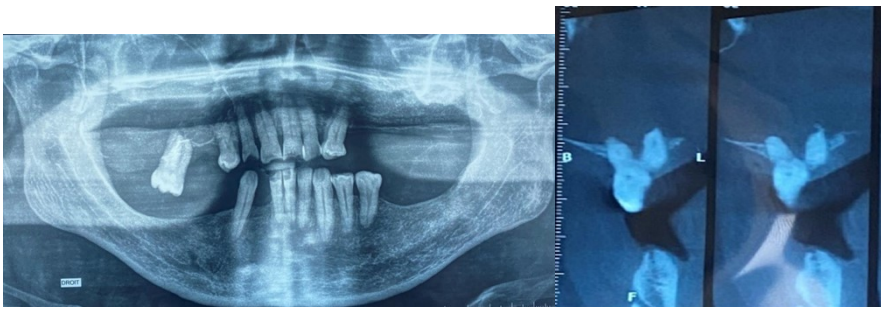
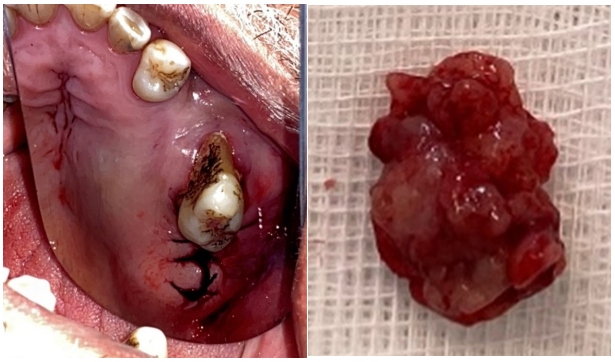


Figure 2. Preoperative intraoral image.



(a) (b)

Figure 3. Radiographic presentation: (a) Orthopantomography and (b) Cone-beam computed tomography show unilocular radiolucency in the maxillary posterior region causing expansion and perforation of the vestibular and palatal alveolar walls.



(a) (b)

Figure 4. (a) Post-operative view after biopsy; (b) Operative piece.

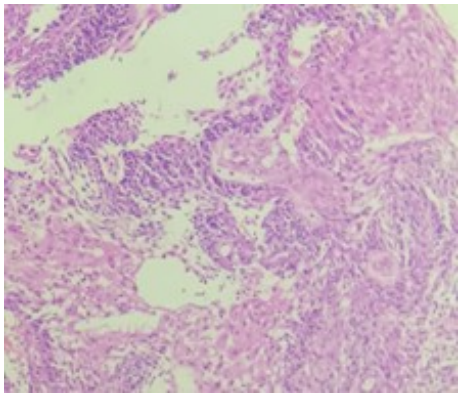


Figure 5. Histological slide confirmed the diagnosis of a plexiform ameloblastoma.

2.2. Case 2

A 33-year-old woman presented with a right temporo-maxillary swelling that had been developing over a period of 2 years. Her medical history was unremarkable, and she was in good general health.

Upon extraoral clinical examination, facial asymmetry and a firm swelling were noted in the right mandibular ramus, extending into the temporal region. The patient exhibited limited mouth opening to one finger. No cervical adenopathy or sensitivity disturbances were observed during the examination (See **Figure 6**).

Panoramic radiography and CT radiography for visualizing soft-tissue extension were performed.

Radiographs revealed a well-defined, wide, multilocular radiolucent lesion involving the entire ramus and a part of the mandibular body. It extended from the distal aspect of tooth 47, with resorption of the root apices of the latter up to the ramus, encompassing the mandibular angle. There was complete destruction of the coronoid process, while the condyle remained preserved.

The lesion extended to involve the inferior alveolar canal, resulting in thinning of the basal border of the mandibular body and the posterior wall of the ascending ramus. Additionally, an extension of the lesion was noted in the temporal region,

reaching down to the floor of the orbit (See Figure 7).

A fine needle aspiration was ordered, and an incisional biopsy was performed under local anesthesia. The pathology report confirmed the diagnosis of plexiform ameloblastoma, and the patient underwent surgical resection (See Figure 8).

Surgical resection of the mandible was performed intraorally, transecting the entire ramus and part of the horizontal branch from the 47 with a safety margin of 1.5 mm of uninvolved bone (See Figure 9). Immediate reconstruction with an iliac graft was performed in the same session, followed by temporary maxillo-mandibular fixation for 2 months (See Figure 10).

The patient's postoperative recovery was uneventful, and no signs of recurrence were observed during the one-year follow-up examinations. However, 2 years later, a recurrence in the infratemporal fossa was diagnosed.

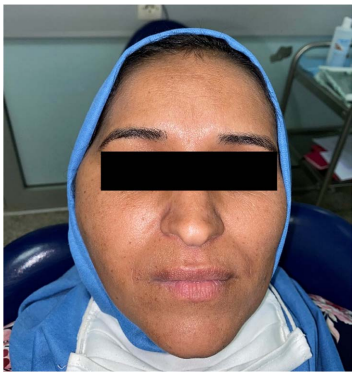
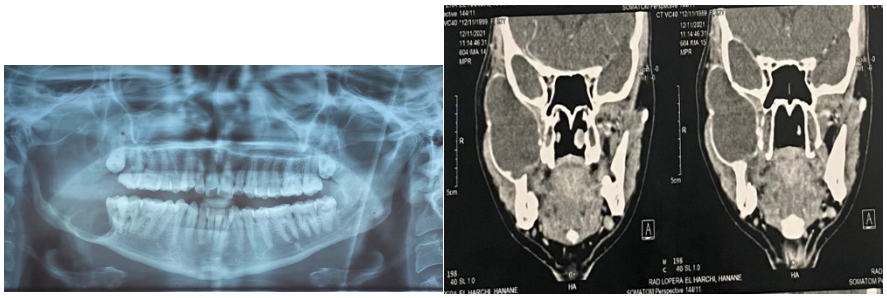


Figure 6. Extraoral presentation shows facial asymmetry and firm swelling in the region of the left ramus of the mandible, extending into the temporal region.



(a) (b)

Figure 7. Radiographic presentation (a) Orthopantomography and (b) Scanner show radiolucency extending from the right mandibular angle to the ramus, with destruction of the coronoid process and preservation of head of the condyle.

3. Discussion

Plexiform ameloblastoma is a subtype of conventional ameloblastoma, representing approximately 13% of all cases. It is the second most common type, with follicular ameloblastoma being the most prevalent at 64.9% and acanthomatous ameloblastoma at 3.9% [4] [7] [8].

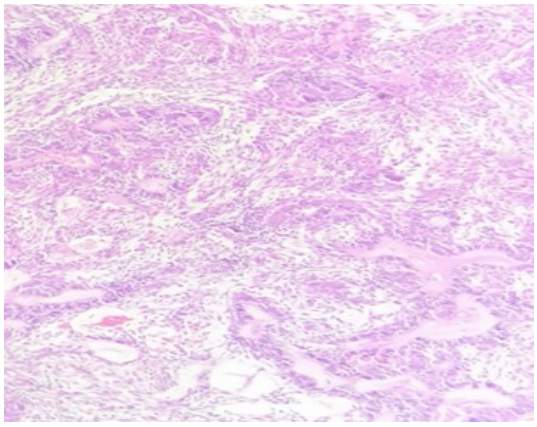
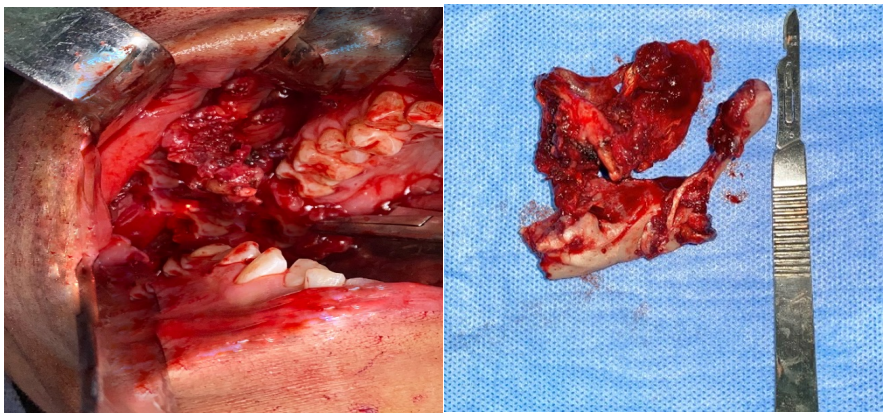


Figure 8. Histological slide confirmed the diagnosis of a plexiform ameloblastoma.



(a) (b)

Figure 9. (a) Post-operative view after excision of lesion; (b) Operative piece with sectioned mandible.

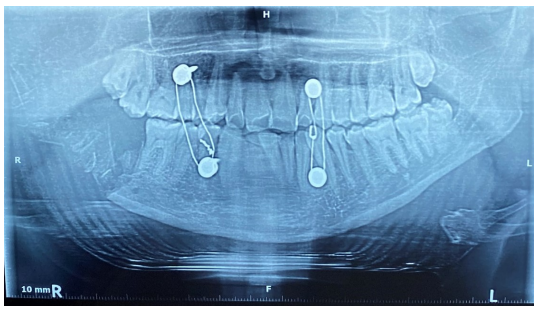


Figure 10. Postoperative radiograph to ensure graft stability after 2 months.

This subtype of ameloblastoma typically exhibits a preference for the ascending ramus of the mandible (70%) as noted in our second case, followed by the premolar region (20%). Around 10% - 15% of cases are associated with a non-erupted tooth. In the remaining 20% of cases, the tumors are in the maxilla, with the maxillary tuberosity being the most common site, as seen in our first case [3] [4] [5].

It often manifests clinically as a slow-growing, painless swelling accompanied by facial deformity, causing expansion of cortical bone, spreading of the lingual and/or buccal plates, and soft tissue involving. Sometimes it can be accompanied by malocclusion, tooth mobility and displacement, ulceration, periodontal disease and paresthesia of the affected area [9].

Radiographically, plexiform ameloblastoma may exhibit a well-defined unilocular or multilocular radiolucent image with a honeycomb or soap bubble appearance causing the thinning or expansion of cortical plates with scalloped margins and perforations with resorption of the involved teeth in advanced stages.

The diagnosis of ameloblastoma is often delayed, probably because of its slow-growing character. It can also mimic other tumors of the mandible, making accurate diagnosis challenging [10] [11].

In the first case, the negative pulp sensitivity test prompted the consideration of a periapical cyst. Additionally, due to the localization a differential diagnosis was established with keratocyst.

In the second case, the intact mucosa, and no paresthesia led to a differential diagnosis including odontogenic keratocyst, aneurysmal bone cyst, or a giant cell tumor. The definitive diagnosis can be confirmed only through histopathology.

Histopathologically, plexiform ameloblastoma is presented as a well-differentiated palisaded cells found around the periphery and networks of epithelium. Nuclei of the palisaded cells are typically polarized away from the basement membrane and the epithelium displayed a stellate, arranged as a tangled network of anastomosing strands, enclosing cysts of various sizes [12] [13].

The term plexiform refers to the appearance of anastomosing cords of epithelial elements within the stroma [9] [14] [15], which is consistent with the anatomopathological description in our cases showed anastomosing sheets and cords of odontogenic epithelium.

Management of plexiform ameloblastoma is still controversial. Various treatment methods of ameloblastoma have been suggested in relation to the tumor type and clinical presentation [5] [16].

Radical surgery like segmental resection is thought to be associated with lower recurrence rates but often requires extensive reconstructive surgery immediate or delayed of the defect with tissue grafts and/or prosthetic rehabilitation [17] [18].

Several grafts can be used for reconstruction purposes. Autogenous grafts are considered the gold standard. However, donor site morbidity, restricted availability, and higher surgical time are all disadvantages [5] [19].

Generally, conservative treatment (includes enucleation, curettage, and marsupialization) associated or not with adjuvant treatments (chemical cauterization with Carnoy's solution, cryotherapy, and curettage of the bone after enucleation) is reserved for the unicystic form [5] [20]-[22].

Targeted therapies, particularly those targeting BRAF V600E mutations, represent a promising new approach for treating ameloblastoma, either as primary treatment or as adjuvant therapy to reduce tumor progression and limit the

morbidity associated with surgical interventions. This type of treatment aims to specifically target the molecular pathways involved in tumor growth. However, the systemic side effects linked to BRAF and/or MEK inhibitors can hinder their effectiveness, especially in the context of benign tumors like ameloblastoma, where adverse effects such as fatigue and skin rashes may compromise the patients' quality of life. This highlights the importance of thorough research to optimize the use of these therapies, exploring dosing strategies, combination therapies, or identifying biomarkers to predict treatment responses, with the goal of tailoring therapeutic options to the specific characteristics of both the tumor and the patient, thus promoting personalized medicine [22]-[24].

In both cases, resectative surgery is recommended, followed by reconstructive surgery to provide reasonable cosmetic and functional outcome to the patient.

The recurrence of ameloblastoma seems to depend on several factors, such as type of ameloblastoma, method of treatment of the primary lesion, the extent of the lesion and the site of origin [25].

Multicystic ameloblastoma with its histological variants has a much higher rate of recurrence than unicystic ameloblastoma. The reason is thought to be due to the numerous micro extensions the tumor has projecting into the bone.

The identification of certain immunohistochemical markers such as CD10 and Ki67 could be used to establish prognosis. CD10-positive tumors with a high Ki67 index are usually associated with a high recurrence rate [26] [27].

Studies conclude that tumors larger than 6 cm, the histological follicular tumors, the presence of radiological multilocular lesions or basal cortical bone rupture, invasion of soft tissue or adjacent anatomical structures as observed in the second case where extension into the soft tissues was a significant indicator of recurrence [18] [25] [28].

4. Conclusion

Plexiform ameloblastoma is a rare and benign odontogenic tumor that may reach gigantic proportions. Thus, ensuring an accurate diagnosis is crucial to minimizing the risk of local recurrences and obtaining an effective treatment.

Ethics

Informed consent was obtained from all individual participants included in the study.

Conflicts of Interest

The authors declare that they have no conflict of interest.

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