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Hemangiomatous Ameloblastoma: A rare variant or secondary change in Ameloblastoma?

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Abstract

Hemangiomatous Ameloblastoma is a rare entity characterized by characteristics of conventional AM with the confluence of blood vessels and extravasated elements. It has not been recognized as a distinct entity or a variant of Ameloblastoma in the standard classification of head and neck tumors. Due to the rarity of lesions, not much is known about its clinicodemographic characterization and biological nature. The present article aims to report a case of the rare entity while emphasizing its pathogenesis.

Keywords: Odontogenic tumors; Vascular tumors; Histopathology; Pathogenesis

Introduction

At times, some neoplasms may comprise a prominent vascular component in association with the neoplastic cells of the tumor's derivative lineage. These exhibit numerous blood vessels and blood-filled spaces of various sizes with/without areas of necrosis. Some well-known examples of such neoplasms, that have been accepted as distinct histological entities, include angiomatoid malignant fibrous histiocytoma and the telangiectatic osteosarcoma **[1,2]**.

A similar vascular component may be observed, although rarely, in ameloblastoma (AM). The first such AM was described by Kuhn in 1932 as a combination of hemangioma and adamantinoma **[3]**. Over the years, such AMs have been reported as 'Combination of hemangioma and ameloblastoma', Adamantino-hemangioma, Ameloblastic Hemangioma, Hemangio-Ameloblastoma and Hemangiomatous Ameloblastoma (HA), and Vascular AM **[4]**. The World Health Organization (WHO) has grouped all the other histopathological variants such as follicular, plexiform, acanthomata's, basaloid, granular cell, and desmoplastic, under one category of AM. However, HA has not been recognized as a distinct entity or a variant of AM in the 4th edition of WHO Classification of Head and Neck Tumors **[5]**.

To date, only about 18 cases of HA have been reported in the scientific literature **[6]**. Therefore, not much is known about the characterization and clinical nature of this entity. The present report aims to elucidate various aspects of this rare entity while emphasizing its pathogenesis.

Case report

A 43-year-old male complained of pain and swelling in the maxillary right posterior region for two weeks. The patient had a diffuse swelling associated with pus discharge in the same region, 12 years ago. At that time, the maxillary right premolars were extracted, the lesion was enucleated and diagnosed as an infected odontogenic cyst. Except for hypertension for the past 13 years, the patient's medical history was unremarkable.

On intraoral examination, a diffuse swelling was noted on the hard palate extending from the maxillary right lateral incisor to the first premolar region **(Figure 1A)**. Obliteration of the buccal vestibule was noted, and the swelling extended up to the midline. On palpation, the swelling was found to be soft and fluctuant.

Orthopantomogram revealed an ill-defined multilocular radiolucent lesion extending from maxillary right lateral incisor to mesial aspect of the maxillary first molar. Spiked resorption of the roots of maxillary right lateral incisor and canine were noted, suggestive of a neoplastic lesion **(Figure 1B)**. The resorption associated with a canine was relatively more severe, giving rise to a 'floating tooth' appearance.

The provisional diagnosis was discerned as AM, while the differential diagnosis included central giant cell granuloma and odontogenic

myxoma. Because of extensive involvement along with spike-shaped resorption of the roots of maxillary teeth, malignant neoplasms such as clear cell odontogenic carcinoma and carcinoma of maxillary sinus were also considered in the differential diagnosis.

On aspiration, a small amount of blood-tinged fluid of watery consistency was elicited. Numerous RBCs with rouleaux formation were noted on wet mount examination. An incisional biopsy was undertaken. Macroscopically, the tissue was brownish to blackish in color, soft in consistency, having an irregular shape and surface texture.

Histopathologically, plexuses of odontogenic epithelium could be noted along with a highly vascular stromal component **(Figure 2)**. The odontogenic plexuses were lined by low to tall columnar ameloblast-like cells peripherally, while stellate reticulum-like cells were present centrally **(Figure 3)**. The stromal tissue is almost entirely comprised of endothelial-lined vascular channels of varying sizes. Extravasated blood elements were present abundantly in the stroma.

Considering the ameloblastoma-like plexuses along with the highly vascular stroma, a final diagnosis of hemangiomatous ameloblastoma was imparted. A subtotal maxillectomy of the right maxilla was performed with a 1 cm margin and a palatal obturator was provided for the defect.

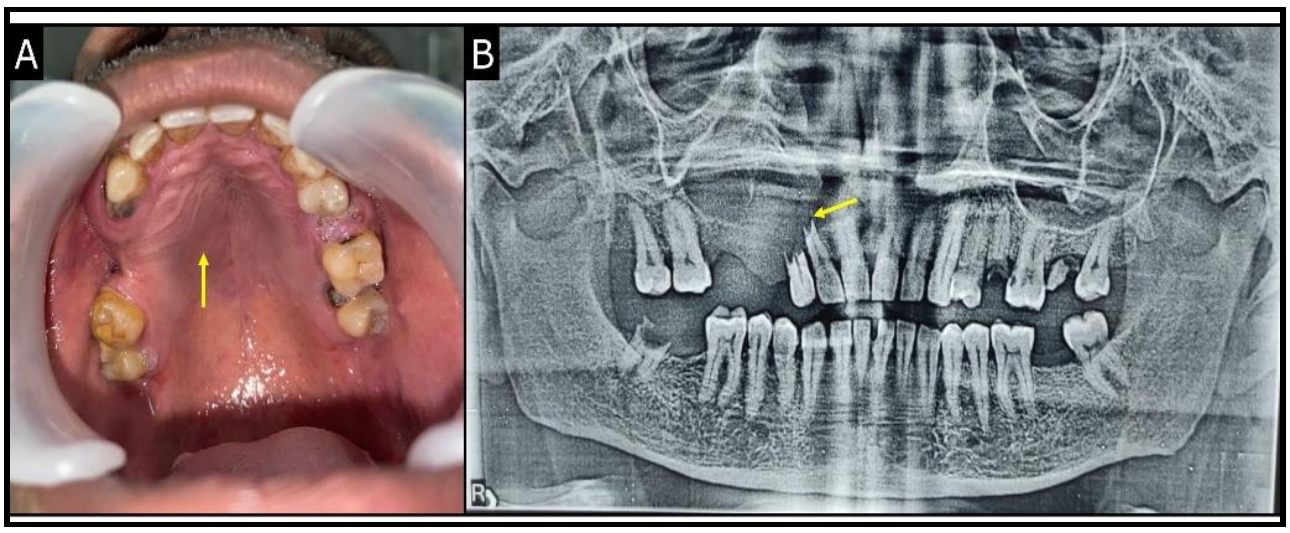

Figure 1: A) Intra-oral swelling in the anterior hard palate; B) Orthopantomogram exhibiting an ill-defined radiolucent lesion in the right maxilla with spiked resorption of roots of lateral incisor and canine (arrow)

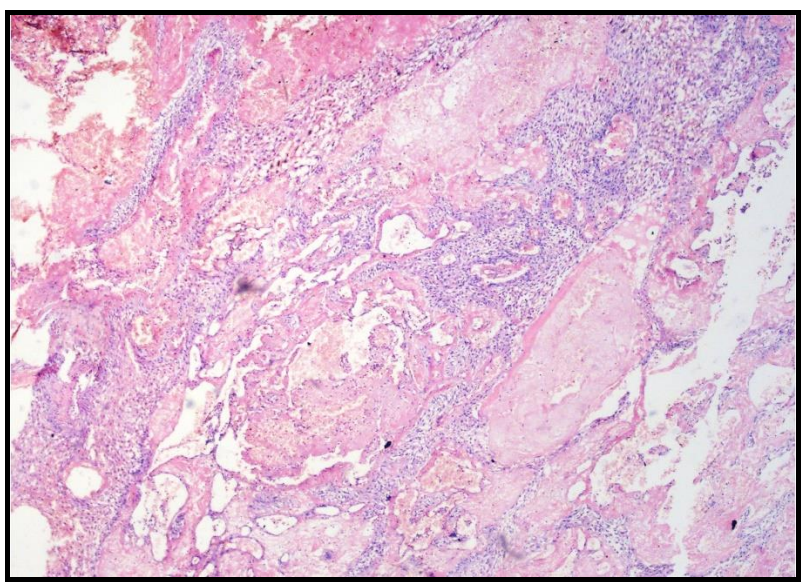

Figure 2: Odontogenic plexuses confluent with highly vascular stroma. (H and E, 4x)

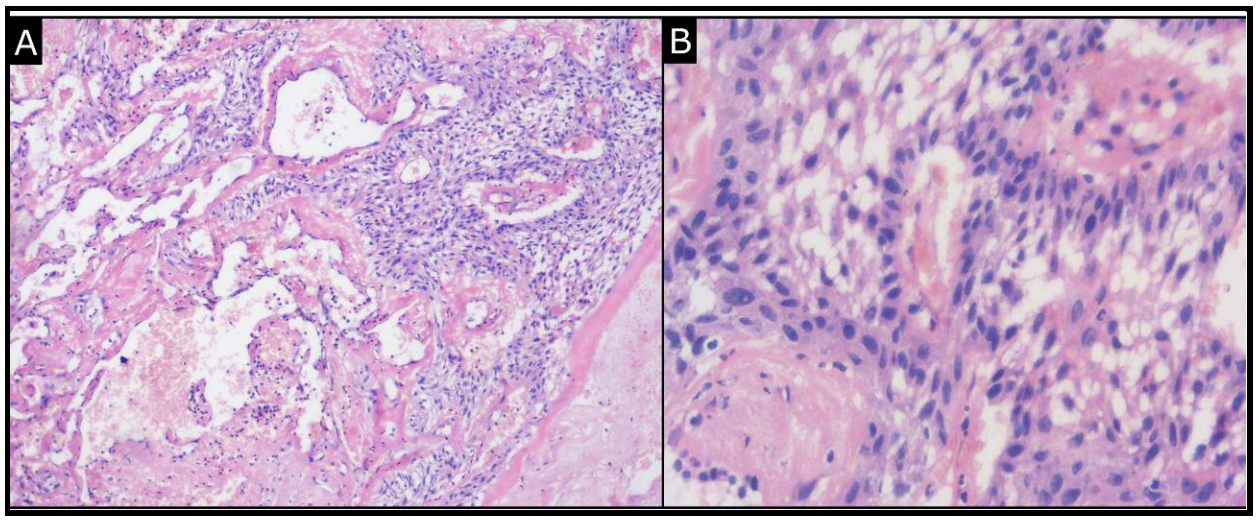

Figure 3: A) Odontogenic plexuses with endothelial lined vascular channels (H and E, 10x); **B)** Peripheral ameloblast-like cells with central stellate reticulum-like cells (H and E. 40x)

Discussion

Only a few reported cases of HA exist in literature which could be attributed to various reasons- (i) The entity is actually rare with a very low prevalence rate, (ii) There is a lack of awareness amongst pathologists owing to the modicum of reported cases of HA, and therefore, they are diagnosed as one of the conventional variants of AM, (iii) Some pathologists do not consider HA as a distinct variant of AM **[7]**. Either way, as the number of reported cases of HA increases, more information would shed light on its demographic and clinical characteristics.

HA can occur in patients of any age. However, unlike other variants of AM which commonly occur in the second or third decade, HA tends to occur in slightly older individuals in the third or fourth decade **[4]**. It exhibits a slight male predisposition, with a male: female ratio being 7:4. These demographic observations from previously reported cases of HA accord with our case, which occurred in a 43-year-old male. Most of the cases reported to date have occurred in the mandibular posterior region while only two cases have been reported to occur in the maxilla **[4,6,8]**. The present case consisted of HA occurring in the maxilla, which is an unusual site of occurrence for the rare entity.

Various plausible explanations have been put forth in order to explain the pathogenesis of HA. One school of thought believes that HA represents a true collision tumor, comprising of two distinct neoplasms growing in the same area **[9,10]**. These neoplasms comprise hemangioma and ameloblastoma that intermingle and compose the collision tumor, HA. However, whether to consider the hemangiomatous component as a true neoplasm or a hamartoma is still a matter of debate.

During normal odontogenesis, the outer enamel epithelial cells of the enamel organ are abutted by numerous blood vessels of the dental follicle. It is believed that an inductive stimulus leads to abnormal angiogenesis, ultimately causing the overgrowth of blood vessels. This inductive stimulus may be provided in the form of trauma which leads to an exaggerated granulation tissue response in an attempt to repair the odontogenic tissue. Traumatic incidents such as dental extraction or other minor surgical procedures have been suggested as possible sources of the stimulus by Sarode et al., Rajmohan et al., and Kasangari et al. **[11-13]**. A history of extraction of maxillary right premolars as well as enucleation of a cystic lesion in the same region, twelve years ago, was also elicited in the present case.

Like conventional AM, our case of HA also exhibited an ill-defined multilocular radiolucent lesion. HA can be differentiated from benign fibro-osseous lesions which are well demarcated and consist of radiopaque foci. However, the dense vascular tissue and surrounding periosteal reaction may give the impression of a mixed radiographic appearance in some cases **[14]**. In such cases, the bulk of lesions should be considered from a comparison of the radio dense areas to avoid misinterpretation.

Histopathologically, the conventional plexiform AM comprises interconnecting plexuses of odontogenic epithelium with small stromal spaces in between **[5]**. At times, the stroma may get degenerated owing to hypoxia and other related factors. Lucas et al. opined that the blood vessels often persist while the stroma degenerates in a conventional plexiform AM **[15]**. As a result, the blood vessels are left without any resilient structures such as collagen fibers to support them. They tend to dilate in the available space resulting in enlargement. Subsequent rupture of these dilated and engorged blood vessels can lead to the accumulation of blood elements in the stromal spaces. Overall, these events could account for the histopathological picture noted in HA. Lack of supporting stroma would also account for the soft consistency of the lesion noted clinically as well as during grossing.

The exact biological behavior and necessary treatment required for HA have not yet been clearly established. In most of the reported cases, hemi mandibulectomy was performed with satisfactory results **[4,6]**. While the two cases of HA in the maxilla were treated with enucleation and curettage, long-term follow-up data about the outcome was not provided **[4,8]**. We believed that HA should be treated as conventional AM, and thus, subtotal maxillectomy was determined to be a more appropriate treatment for the present case. Complications that may arise due to the highly vascular nature of the lesion must be kept in mind while obtaining an incisional biopsy as well as treating HA. Aspiration of lesions should precede the biopsy procedure, as it could caution the surgeon beforehand.

Reconstruction with fibula graft was planned for the present case, however, because the patient did not consent to the procedure, a palatal obturator was provided as an alternative to close the defect. Long-term follow-up of the present case as well as in other reported cases would provide more insight into the appropriateness of treatment procedures in HA.

Conclusion

HA may be considered as a rare variant of AM in which inductive stimulus such as trauma leads to extensive proliferation of vascular components. It exhibits a predilection to occur in the mandibular posterior region in the third to fourth decade of life. The present case would serve to add to the limited literature available with respect to HA, especially in the maxilla, for which only two cases have been reported so far. Although the histopathological features of HA are pathognomonic, whether to consider it as a distinct entity or a variant of ameloblastoma is yet unclear. Clinicopathological data with long-term follow-up of more cases of HA would provide more insight into this matter.

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