

## Case Report

### Hemangiomatous ameloblastoma in maxilla: A report of a very rare case

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#### ABSTRACT

Hemangiomatous ameloblastoma (HA), which is a rare ameloblastic variant, is presented in a 15-year-old boy in the maxillary right molar region associated with unerupted canine and premolars. Radiologic and computed tomographic analysis was suggestive of cystic lesion. An histology picture confirmed the diagnosis of HA. There are less than eight cases documented in the literature and mostly are in the middle age with mandibular location. This is first ever reported case of HA in a 15-year-old boy with maxillary location. Due to less number of documented cases and no long-term follow-ups, clinical behavior and prognosis of this lesion are uncertain. In this case report, the clinical, histological, and radiographic features of HA are presented.

**Key Words:** Ameloblastoma variants, hemangiomas, unilocular radiolucency, vascular ameloblastoma

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#### INTRODUCTION

Ameloblastomas in young people (i.e., 19 years old and younger) are thought to be rare. They account for approximately 10–15% of all reported cases of ameloblastoma.<sup>[1]</sup> However, there is less accurate information available regarding its prevalence. It is a true neoplasm known for its destructive and recurrent behavior. On the basis of clinical and biological behavior, it is principally categorized into three types: solid or multicystic, unicystic, and peripheral. There are various numbers of histopathological patterns of ameloblastoma. The follicular and plexiform patterns are the main histologic types. Commonly encountered histologic variants are acanthomatous and granular cell types. Other less commonly encountered histologic

patterns include desmoplastic ameloblastoma, basal cell ameloblastoma, clear cell ameloblastoma, and unicystic ameloblastoma.<sup>[2]</sup> Although with the exception of desmoplastic and unicystic type, histopathological variation does not carry significance in terms of prognosis and biological behavior.<sup>[3]</sup> The hemangiomatous ameloblastoma (HA) was originally described as an ameloblastoma in which part of the tumor contained spaces filled with blood or large endothelial-lined capillaries.<sup>[4]</sup> Lesions with similar histologic features that probably represented the same entity were documented in the early literature as ameloblastic hemangiomas,<sup>[5]</sup> adamantinohemangiomas,<sup>[6,7]</sup> and hemangio ameloblastomas.<sup>[8]</sup>

To the best of our knowledge, there are less than eight cases documented earlier in the literature.<sup>[7-12]</sup> Other reported cases are mostly in the mandible and in the middle age. This is the first case of HA in a child with maxillary location. This article presents a rare form of ameloblastoma with clinical, radiological, and histological features consistent with those of an HA which will contribute to the literature in studying biological behavior and prognosis of the lesion.

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## CASE REPORT

A 15-year-old boy presented with a right maxillary swelling 3.5 cm in the width and 3 cm in length with a duration of 5 months with a history of gradual enlargement of the right jaw and the swelling had displaced the lateral wall of the nose on the right side up to the nasal septum. The patient noticed the swelling of face 5 months ago and pain on palpation. The swelling also encroached onto the upper alveolar process on the right side and adjoining hard palate up to the midline. The medical history was insignificant. Routine blood investigations such as hemogram, bleeding, and clotting time were normal. An intraoral examination revealed a firm, smooth, and nonfluctuating swelling that was bony hard in consistency and fixed to the right maxilla. The upper permanent canine, first and second premolar teeth were missing on the right side while deciduous

canine, first and second molar were present and they were mobile [Figure 1].

## RADIOGRAPHIC FINDINGS

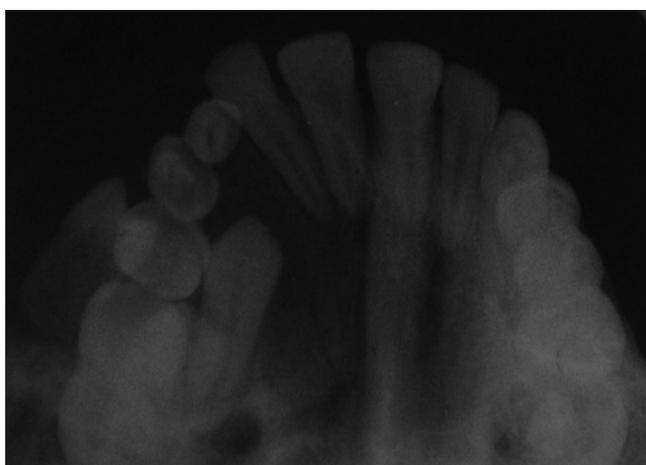
Radiographic examinations revealed well-defined radiolucency with sclerotic border in the maxillary canine region extending up to second molar surrounding the second premolar tooth bud. Permanent canine and premolars were unerupted and deciduous canine, first molars and second molar were retained [Figures 2 and 3]. Radiograph of the paranasal sinuses [Figure 4] showed a hazy right maxillary antrum with expansion and thinning of all its bony walls with the right upper canine tooth, pushed up to the floor of the orbit. Contrast enhanced computed tomography (CECT) of face suggestive of a well-defined expansile cystic lesion with 3.7 cm in width, 3 cm in length, and 3 cm in depth with a CT value in the range of 15–25 HU with corticated margin arising from



**Figure 1:** Facial aspect showing retained deciduous teeth and swelling



**Figure 2:** OPG showing well-defined radiolucency with sclerotic border in the maxillary canine region extending up to second molar surrounding the second premolar tooth bud



**Figure 3:** Occlusal radiograph showing permanent unerupted canine and retained deciduous teeth



**Figure 4:** Hazy right maxillary antrum with expansion and thinning of all its bony walls





**Figure 9 :** Post-op OPG

is part of the neoplastic process, represents a separate neoplasm, or is a hamartomatous malformation has not been satisfactorily resolved.

The stroma part of ameloblastoma consists of loose myxoid tissue which can undergo degeneration so blood vessels are left without any support. They become enlarged, and rupture leading to escape blood into empty stromal space.<sup>[5]</sup>

A traumatic incident such as a tooth extraction may provide a stimulus required for proliferation of epithelial cell rests in the periodontal ligament and subsequent tumor development.<sup>[6]</sup> Tissue damage is usually followed by repair, and this involves the formation of the granulation tissue in which proliferating endothelial cells and new capillaries are prominent. A disturbance in the repair of neoplastic odontogenic tissue may result in excessive granulation tissue formation or the development of an abnormal vascular component.

At the time of tooth development, numerous blood vessels furnish supply to outer enamel epithelium. It is probable that blood vessels are abnormally induced to become part of tumor or abnormal angiogenesis due to excess inductive influence can lead to overgrowth of vascular elements. It has been suggested that the HA represents a collision tumor. In this type, two separate tumors grow in the same area and collide with each other and the tumor elements intermingle.<sup>[8]</sup>

The HA observed here differs histologically and radiologically from a conventional ameloblastoma. Histologically, it consists of an ameloblastoma with a prominent vascular component while its conventional radiologic features are nonspecific.

A CT scan clearly demonstrates cystic features in this tumor, such as its expansile nature and soft tissue contents. According to Smith,<sup>[10]</sup> this HA is similar

histopathologically to conventional ameloblastoma and not a distinct entity. Vascular supply to this tumor is variable. Whether it is hamartomatous malformation or a separate neoplasm is not clear. Its vascular features are, however, well documented, and vascular variants of such neoplasm are accepted as separate histologic entities. Examples are the angiomatoid malignant fibrous histiocytoma<sup>[13]</sup> and the telangiectatic osteosarcoma.<sup>[14]</sup> The angiomatoid malignant fibrous histiocytoma combines the features of both a fibrohistiocytic tumor and a vascular tumor, while the telangiectatic osteosarcoma contains large blood-filled spaces, thrombus formation, organization, and massive areas of necrosis.

## CONCLUSION

Vascular lesions of the jaws are of much concern as fatality is documented following minor procedures. Cases have been reported in the literature as early in 1933 in which improper management led to rapid, fatal exsanguinations.<sup>[15]</sup> The biologic behavior of HA is thought to be similar to that of the conventional ameloblastoma, but because a few cases have been reported with no long-term follow-up so the pathogenesis and clinical features are not yet fully understood and biologic behavior cannot be predicted. Whether it should be given the status of distinct entity or just a variant of ameloblastoma, further research is needed.

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