

Hemangiomatous Ameloblastoma with Spindle Cell Proliferation: A Rare Case Report and Review of Literature

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Abstract

Ameloblastoma is a locally aggressive neoplasm with varied histological patterns. The histomorphologic variants of ameloblastoma bear no prognostic or biologic behavioral significance with possible exceptions of desmoplastic and hemangioameloblastoma. The present paper aims at reporting a case of 38-year-old male with a huge lesion present in the mandibular symphyseal region crossing the midline, histopathologically showing hemangiomatous component in follicular and plexiform ameloblastoma along with spindle cell differentiation in the ameloblastic follicles. Spindle cell differentiation in ameloblastoma is rarely reported and the prognostic significance is yet not clear. The present paper also reviews the literature since the diagnosis of this lesion is must as it may lead to surgical complications.

Keywords: Ameloblastoma, hemangioameloblastoma, odontogenic tumors, spindle cell tumors

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Submitted: 10-Nov-2021, **Revised:** 30-Dec-2021, **Accepted:** 21-Jan-2022, **Published:** 31-Mar-2022

INTRODUCTION

Ameloblastomas, according to the modified WHO classification, is a tumor arising from odontogenic epithelium without odontogenic ectomesenchyme. Hemangiomatous ameloblastoma is a solid ameloblastoma with a lot of extravasated red blood cells (RBCs) and large endothelial lined capillaries.^[1] Highly increased vascularity in a disease can itself affect the prognosis of the lesion, and in malignancies, it can induce tumor seeding and secondaries. Not much is known regarding the etiopathogenesis or biologic behavior of hemangioameloblastoma (HA), as there is a lack of report of many such cases. The paucity of reported cases in literature only adds to the distress of addressing this issue. On the contrary, Smith regards this entity to be histologically similar to one of the other recognized types of ameloblastoma and not as a distinct

histologic entity, as according to him, the blood supply to these tumors is variable and that circumstance other than the number and size of the vessels influences the blood supply.^[2] The biologic behavior of HA is thought to be similar to that of the conventional ameloblastoma. Yet, the variations seen in its presentation are intriguing even today. Whether such variations have any impact on treatment planning, and prognosis for the patient is not clear since not many cases with longterm followup of such lesions have been reported.

CASE REPORT

A 38-year-old male patient reported to the department of oral pathology with a chief complaint of swelling in lower anterior region of face for 8 years. The patient

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How to cite this article: Puri PD, Krishna A, Gosavi S, Nayyar V. Hemangiomatous Ameloblastoma with Spindle Cell Proliferation: A Rare Case Report and Review of Literature. J Oral Maxillofac Pathol 2022;26:132-3.

Access this article online

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10.4103/jomfp.jomfp_394_21

was apparently alright 8 years back. He gave a history of trauma, being hit by a bull in the submental region. He developed a small swelling in the same region which progresses to the present size. No treatment was taken at that time. Extraoral examination revealed well-defined swelling of approximate size 8 cm × 7 cm extending from right parasymphysis region to the body of mandible medio-laterally and supero-inferiorly from line joining the commissures to lower border of chin giving facial asymmetry [Figure 1]. Swelling was hard in consistency nontender, noncompressible and nonfluctuant with egg shell crackling at one place. Intraoral examination revealed well-defined swelling of size 5 cm × 4 cm seen on lower left alveolingival region obliterating both buccal and lingual vestibule extending from 43 to 36 region on buccal side whereas on lingual side extending from 42 to 35 region. Overlying mucosa was normal [Figure 2]. Orthopantomogram showed multilocular mixed radiolucent radiopaque lesion in the mandible extending from 37 to 43 crossing the midline

with thinning of lower border of anterior mandible giving soap bubble appearance [Figure 3]. Displacement of mandibular anterior teeth was also evident. Cone beam computerized tomography showed perforation at multiple sites with respect to inferior border of mandible and buccal and lingual cortical plate [Figure 4]. Magnetic resonance imaging revealed well-defined heterogeneous multiloculated enhancing solid cystic altered signal intensity lesion arising from the mandible with involvement of body of left hemimandible, symphysis and the left mandibular canal [Figure 5]. Angiography of head and neck revealed no feeding vessel to the tumor mass [Figure 6]. Differential diagnosis of ameloblastoma, aneurysmal bone cyst (ABC), and central giant cell tumor was made. On gross examination, resected segment of mandible was received of approximate size 8 cm × 9 cm extending from distal of 37 to distal of 45, grayish black in color with irregular and lobulated surface both buccal



Figure 1: Well-defined swelling in the lower anterior region of face giving facial asymmetry



Figure 2: Well-defined swelling on lower left alveolingival region obliterating both buccal and lingual vestibule



Figure 3: Orthopantomogram showed multilocular mixed radiolucent radiopaque lesion in the mandible crossing the midline with thinning of lower border of anterior mandible giving soap bubble appearance

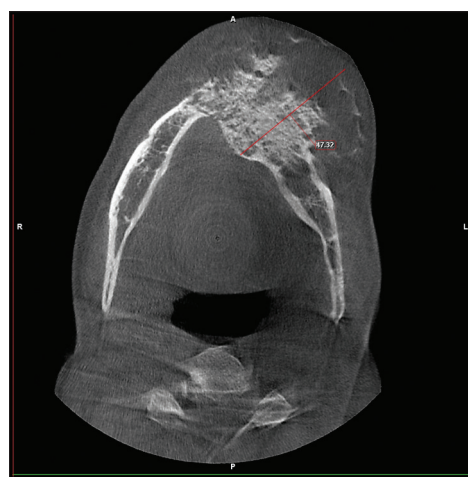


Figure 4: Cone beam computerized tomography showed mixed radiopaque radiolucent lesion causing expansion of buccal and lingual cortical plate

and lingual cortical plate expansion was evident [Figure 7]. Oozing of blood was present from the resected specimen. Histopathological examination showed cystic cavity lined by thin odontogenic epithelium comprising of basal cell layer which composed of tall columnar cells with reversal of polarity resembling preameloblast like cell. Epithelial cells were columnar to cuboidal with prominent stellate reticulum cells. This odontogenic epithelium also showed proliferation in the form of interconnecting anastomosing strands and follicles with peripheral tall columnar cell and reversal of polarity, central areas of the follicles showed presence of loosely arranged stellate reticulum like cell [Figure 8]. The stroma showed the presence of numerous vascular channels. The increase in vascularity was seen as small and large dilated engorged endothelial lined vessels and as areas of extravasation of RBCs in

the stroma and some of the follicles showed presence of spindle cell proliferation [Figure 9]. Vascularity was seen in the connective tissue as well as epithelium. Based on these features, a final diagnosis of hemangiomatous ameloblastoma with spindle cell proliferation was given.

DISCUSSION

The word “Hemangioameloblastoma” indicates the histologic variant of ameloblastoma in which the connective tissue is largely replaced by vascular tissue. It was first described by Kuhn in 1932 and is considered as a rare histomorphologic variant of ameloblastoma. However, this entity may mimic vascular lesions clinically and on radiologic tests and hemangiomas on histopathological examination,^[3] thus, creating a diagnostic impasse and surgical complications.

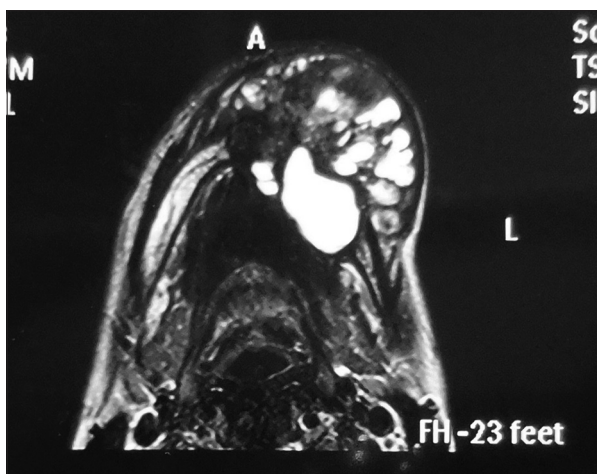


Figure 5: MRI revealed well-defined heterogeneous multiloculated enhancing solid cystic altered signal intensity lesion arising from the mandible with involvement of body of left hemimandible, symphysis, and the left mandibular canal

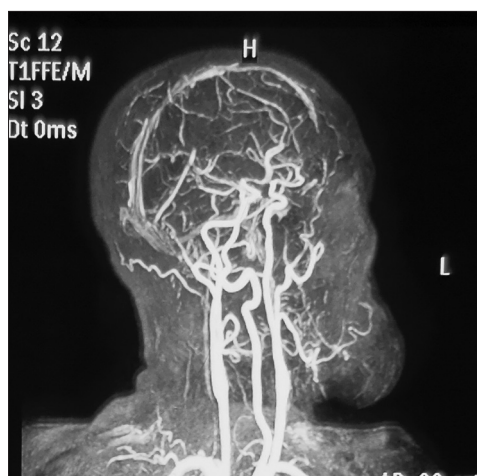


Figure 6: Angiography of head and neck revealed no feeding vessel to the tumor mass



Figure 7: Resected segment of mandible, grayish black in color with irregular and lobulated surface both buccal and lingual cortical plate expansion

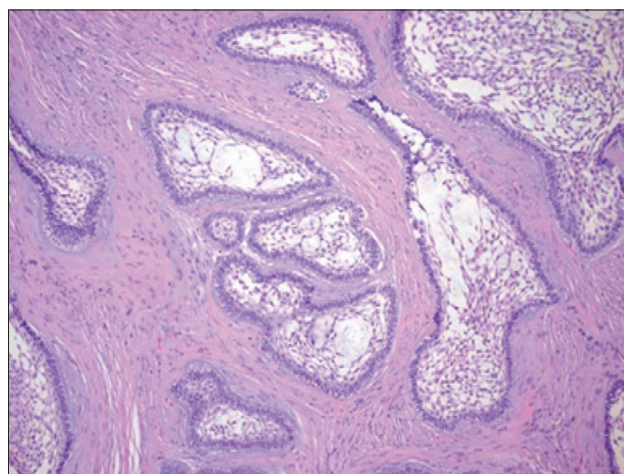


Figure 8: Odontogenic epithelium showed proliferation in the form of interconnecting anastomosing strands and follicles with peripheral tall columnar cell and reversal of polarity, central areas of the follicles showed the presence of loosely arranged stellate reticulum like cell

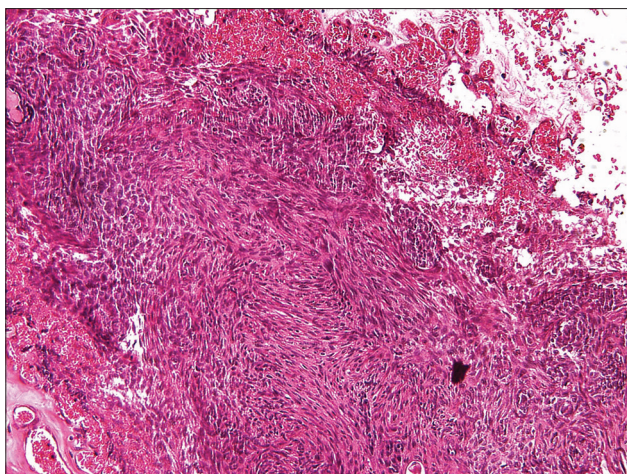


Figure 9: Increased vascularity seen as dilated endothelial lined vessels along with areas of red blood cells extravasation in the stroma and some of the follicles showed the presence of spindle cell proliferation

The origin of the vascular component is not yet clear but several theories have been thought of, including excessive stimulation of angiogenesis during tumor development. The outer enamel epithelium over preameloblasts is associated with high vascular structure providing much needed nutrition to the developing enamel organ. Somehow in HA, these vessels are abnormally induced to become part of the tumor. It has also been suggested to be a collision tumor, i.e., intermingling of two separate tumor entities. However, whether this component is a true neoplastic process or hamartomatous malformation or a separate tumor is not yet clear. Another theory is posttrauma, the epithelial rests of periodontal ligament may get stimulated leading to subsequent tumor development. Any such trauma is followed by repair leading to formation of granulation tissue in which neoangiogenesis is common.^[4] According to Lucas, the unusual vascularity is not due to neoplastic process, in the process of formation of stromal cysts in the ordinary type of plexiform ameloblastoma, the blood vessels often persist and dilate instead of disappearing; thus, it's likely to represent a purely secondary change.^[5]

Pathogenesis, treatment modality and prognosis of hemangiomatous ameloblastoma are not completely understood till to-date as only very few cases have been reported.^[2] However, it is thought to be of benign nature similar to conventional ameloblastoma. The lesion has to be differentiated from ABC, vasoformative lesions, telangiectatic osteosarcoma before any treatment options are planned.^[6]

Ameloblastomas are known to display varied histologic patterns. Along with hemangiomatous component, the

present case also displayed fascicles and eddies of spindle cells in the center of the follicles. Spindle cell differentiation has been previously described in ameloblastic carcinoma which was first described as a separate entity in 1999 by Slater under the heading “low-grade spindle-cell ameloblastic carcinoma.”^[7] However, others described it as a rare variant of spindle-cell ameloblastic carcinoma resulting in extensive metastasis and unfavorable outcomes.^[8] The present case, however, had no malignant features. No such reports of spindle cell differentiation in benign ameloblastoma were found on an extensive research using the various search engines. Thus, it could be the first case of spindle cell differentiation in ameloblastoma.

Zarbo 2003 documented a case of malignant ameloblastoma, spindle cell variant which is treated only by curettage and excision.^[9] The present case was, however, treated by complete surgical resection. Epithelial spindle cell variants of ameloblastoma, benign or malignant, are not recognized in any classification scheme, nor have they been documented. The spindle cells were immunohistochemically analyzed in the reported cases of ameloblastic carcinomas and out of the 12 reported cases, 7 were positive for cytokeratins thus suggesting the epithelial origin of the cells rather than mesenchymal component.^[10-14]

REVIEW OF LITERATURE

Till date, 11 reported cases are found in the English literature to the best of our cognition [Table 1].

Among the reported cases till date, HA has been found commonly in males in the age range of 15–56 years. Clinically, it manifests in the posterior lower jaw most commonly in the form of swelling causing expansion of the cortices and sometimes perforations. The present case, however, is the first to be reported in the mandibular anterior region. Radiographically, it is a multilocular lesions similar to conventional ameloblastomas, however, it may show large cystic areas as in the present case mimicking an ABC or a vasoformative lesion.

We suggest trauma to be an important factor in the origin of hemangiomatous component since the reviewed cases had history of extraction few years back from the same site in 6 cases out of 11 and one case had not mentioned about any past dental history. Further, in the present case, there was history of bull's horn hitting the chin region, i.e., the location of the neoplasm. Thus, it could be concluded that the vascular component occurs as a result of trauma resulting into neoangiogenesis.

Table 1: Comparative review of Hemangiomas of Ameloblastoma cases

| Year | Author | Age of patient | Sex | Site | Treatment |
|------|-------------------------------------|----------------|--------|--------------------------|--|
| 1950 | Aisenberg ^[15] | 48 | Female | Right posterior mandible | Enucleation |
| 1957 | Lucas ^[5] | 43 | Female | Right mandible | Resection of affected portion |
| 2001 | Van Rensburg et al. ^[3] | 26 | Female | Left posterior mandible | Partial hemimandibulectomy planned but patient refused |
| 2010 | Tamgadge et al. ^[16] | 31 | Male | Left posterior mandible | Enucleation and curettage |
| 2011 | Karagir and Ranpise ^[6] | 32 | Male | Left posterior mandible | Surgical resection |
| 2012 | Sharma et al. ^[17] | 15 | Male | Right posterior maxilla | Enucleated |
| 2012 | Harshvardhan et al. ^[18] | 42 | Male | Right posterior mandible | Hemimandibulectomy |
| 2013 | Sarode et al. ^[19] | 18 | Male | Right posterior mandible | Curettage |
| 2014 | Rajmohan et al. ^[20] | 20 | Male | Right posterior mandible | Hemimandibulectomy |
| 2015 | Kasangari et al. ^[4] | 35 | Female | Left posterior mandible | Enucleation |
| 2015 | Hegde et al. ^[21] | 18 | Female | Right posterior mandible | Partial resection of the mandible |
| 2017 | Our case | 38 | Male | Anterior mandible | Resection of the mandible |

CONCLUSION

Lesions with extensive vascular component may lead to mortality even following minor surgical procedures. Our case clearly demonstrates the characteristically unique histologic pattern of HA with spindle cell differentiation. We suggest trauma as an origin of hemangiomas of Ameloblastoma since most of the reported cases in literature had history of extraction few years back in the same site and our case also had a history of trauma.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient (s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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