Case Report

Hemangiomatous ameloblastoma: Case report with a brief review

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Abstract

Ameloblastoma is a benign epithelial odontogenic tumor with many histological variants. Hemangiomatous ameloblastoma (HA) is a very rare variant which shows unique histopathological features varying from conventional ameloblastoma. We present a case of a 35-year-old female patient with a swelling over right lower back region of jaw, showing mixed radiolucent-opacity. Incisional biopsy showed microscopic features of desmoplastic ameloblastoma showing extensive desmoplasia and compressed odontogenic epithelial islands. Excisional biopsy revealed ameloblastomatous areas with extensive vascular component microscopically. Based on these findings, a diagnosis of HA was made.

Keywords: Ameloblastoma, hemangiomatous, odontogenic, vascular component

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INTRODUCTION

Ameloblastoma is a locally destructive and invasive tumor that can recur despite adequate surgical removal. It occurs exclusively in the jaws, with a strong predilection for the posterior region of the mandible.[1] The clinical variants of ameloblastoma are solid or multicystic, unicystic, extraosseous/peripheral and desmoplastic type (the WHO classification).[2] A variable number of histopathological patterns of ameloblastoma exist and the common being follicular and plexiform patterns.[3] In the literature, a rare variant called hemangiomatous ameloblastoma (HA) was originally described as an ameloblastoma in which the tumor stroma contained spaces filled with blood or large endothelial-lined capillaries.[4] Only ten cases have been documented earlier in the literature. The HA in itself is a rare entity and due to the paucity of literature, the pathological knowledge about the tumor is superficial and remains elusive, and additional published cases will provide data for a complete clinical and prognostic profile of this lesion. This case report discusses the clinical, radiological and histopathological features of HA with the possible pathogenesis.

CASE REPORT

A 35-year-old female patient reported to the Department of Oral Medicine and Radiology with a chief complaint of swelling in the lower right back tooth region for the past 6 months. The medical and familial history was noncontributory. Dental history revealed tooth extraction of which the details were not disclosed by the patient.

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Extraoral examination revealed a diffuse solitary swelling of size 10 cm × 5 cm on the right side of the face. The swelling was extending anteroposteriorly from the corner of mouth to tragus and superoinferiorly from outer canthus of the right eye to the inferior border of the mandible. The color of the skin over the swelling is normal with the surrounding skin [Figures 1 and 2].

Intraoral examination revealed a single diffuse swelling measuring 1 cm × 2 cm in size extending from edentulous 43–47 region with obliteration of buccal vestibule [Figure 3].

A provisional diagnosis of ameloblastoma was given, and the patient was advised for radiological and hematological investigations. Hematological investigations were within the normal limits. Orthopantomograph revealed mixed radiolucent and radio-opaque appearance with ill-defined periphery extending from midline to 47 region to the inferior border of the mandible [Figure 4]. An incisional biopsy was done and sent for histopathological evaluation.

Microscopic examination of incisional biopsy showed odontogenic epithelial islands arranged as thin, long cords of different sizes and shapes in a desmoplastic connective tissue stroma [Figure 5]. The stromal desmoplasia with abundant collagen seems to compress or squeeze the odontogenic epithelial islands from the periphery giving the appearance of a kite tail. At higher magnification, peripheral layer of cuboidal cells present occasionally with hyperchromatic nuclei and central area showing cystic degeneration are shown in Figure 6. Based on these features, a diagnosis of desmoplastic ameloblastoma was made, and a wide surgical excision was advised.

Under general anesthesia, hemimandibulectomy was done and multiple sections were made from different locations of the gross specimen. The gross specimen was glistening, reddish brown in color indicating extensive vascularity, firm in consistency and measuring 7 cm × 5 cm [Figure 7]. Microscopic examination showed odontogenic epithelium arranged as anastomosing cords and sheets seen with a prominent vascular component containing numerous blood-filled areas [Figure 8]. At higher magnification, the peripheral cells of the cords showed columnar or cuboidal ameloblast-like cells with central stellate reticulum-like cells.

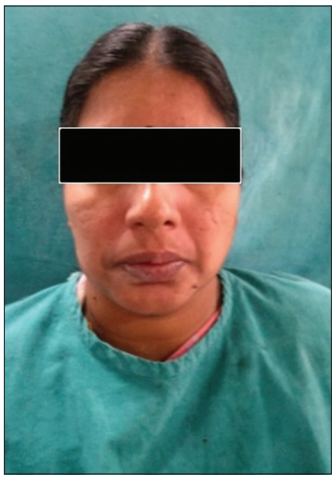

Figure 1: Clinical image showing extra oral view depicting swelling on the right side of the face with asymmetry

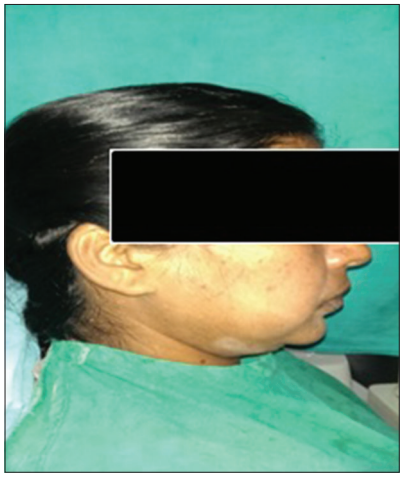

Figure 2: Lateral view showing a single diffuse swelling on right side of the face

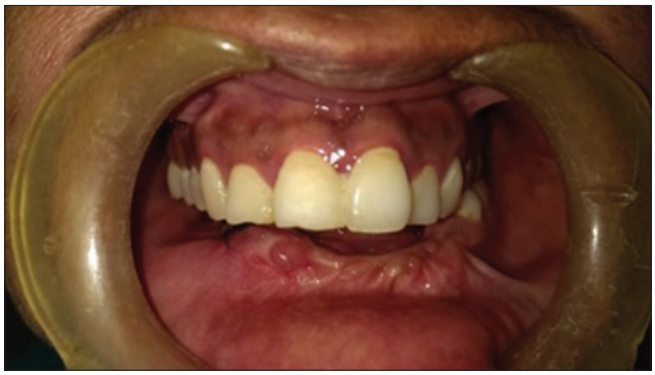

Figure 3: Clinical image shows intra oral view with obliteration of the buccal vestibule

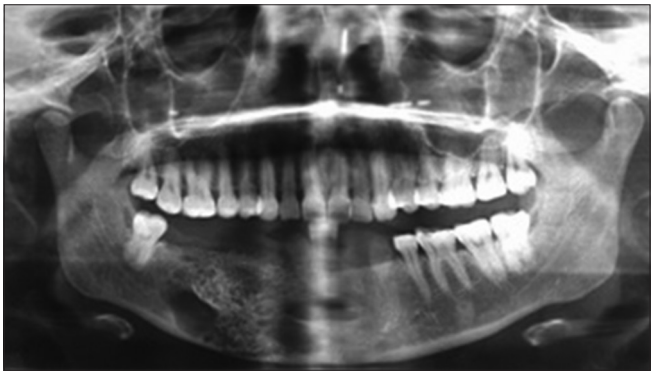

Figure 4: Orthopantamogram showing mixed radiolucent radio opaque lesion

The vascular component showed numerous endothelial lined channels and large blood-filled spaces with engorged red blood cells in the stromal component [Figures 9 and 10]. Taking all these features in count, a final diagnosis of plexiform HA was made.

DISCUSSION

Hemangiomatous ameloblastoma term refers to any ameloblastoma which has many spaces filled with blood or large endothelial-lined capillaries in its stroma.[3] The first case of HA was described in 1932 by Kuhn as a combination of hemangioma and adamantinoma.[5] Later in 1950, Aisenberg reported a similar lesion by the term admantinohemangioma.[6] Apart from these, they are also being reported under diverse nomenclature such as ameloblastic hemangiomas,[7] and hemangio-ameloblastomas.[8]

A search of literature revealed very few cases of HA and Table 1 shows review of all the cases until date. HA can occur at any age but is most commonly seen in the third and fourth decades of life with mandibular posterior region as the most common site which accords with the present case which had occurred in a 35-year-old patient in the right mandibular region. Previous reports showed male

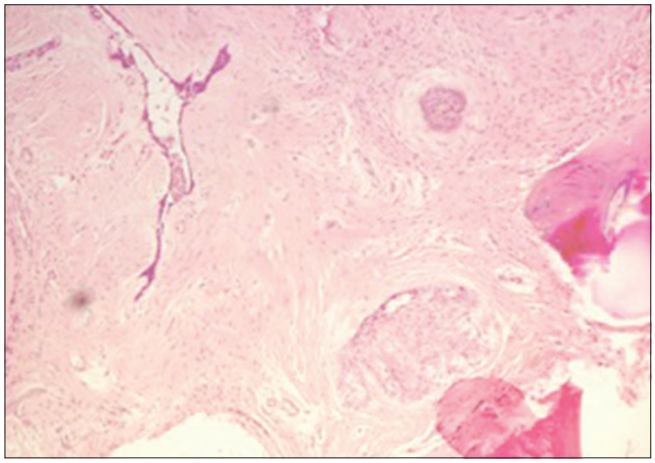

Figure 5: Histopathological image showing odontogenic epithelial islands arranged as thin cords (H&E, ×100)

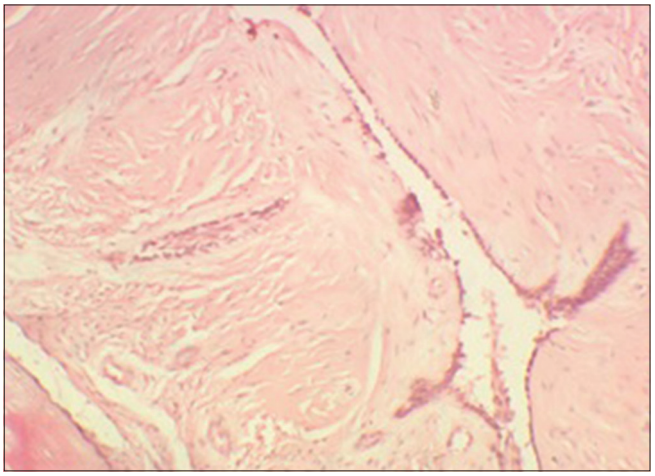

Figure 6: Histopathological image showing compression of odontogenic epithelial islands due to extensive stromal desmoplasia (H&E, ×200)

Table 1: Comparison of all cases of hemangiomatous ameloblastoma

Author	Age/sex	Location	Etiology	Radiography	Treatment done	Follow up	Prognosis
Aisenberg[6]	48/female	Right posterior mandible	-	-	Enucleation	-	Uneventful
Lucas, 1957[9]	43/female	Right mandible	-	-	Resection of affected portion	No complaint after follow-up	Uneventful
van Rensburg *et al*.[10]	26/female	Left mandible, 3rd molar region	-	-	Partial hemimandibulectomy Planned, but patient refused	No follow-up could be done	-
Ide *et al*.[11]	56/male	Anterior maxilla	-	-	Enucleation and curettage	6 months	Good healing
Avinash *et al*.[12]	31/male	Premolar- molar area of left mandible	Not mentioned	Well defined radiolucent lesion	Hemimandibulectomy	4 months follow-up with good bone healing	Not mentioned
Jois *et al*. (2012)[13]	42/male	Posterior region of right mandible	Not mentioned	Mixed radiolucent radioopaque lesion	Hemimandibulectomy	2 years follow-up	Not mentioned
Sharma *et al*.[14]	15/male	Right maxilla	Not mentioned	Well defined radiolucency with sclerotic border	Enucleation	Follow-up for 6 months	Not mentioned
Sarode (2013)[4]	18/male	Right mandible	Tooth extraction	Well circumscribed multilocular radiolucent lesion	Curettage	Patient lost for follow-up	Not mentioned
Rajmohan[15]	20/male	Right side of mandible	Tooth extraction	Soap bubble appearance	Hemimandibulectomy	Not mentioned	Not mentioned
Kasangari *et al*.[3]	35/male	Left posterior mandible	Tooth extraction	Well defined mixed radiolucent - radioopaque lesion	Patient refused for hemimandibulectomy. Enucleation done	Patient lost to follow-up	Not mentioned
Present case (2016)	35/female	Right posterior mandible	Trauma	Mixed radiolucent radioopaque lesion	Hemimandibulectomy	No recurrence	Under follow-up

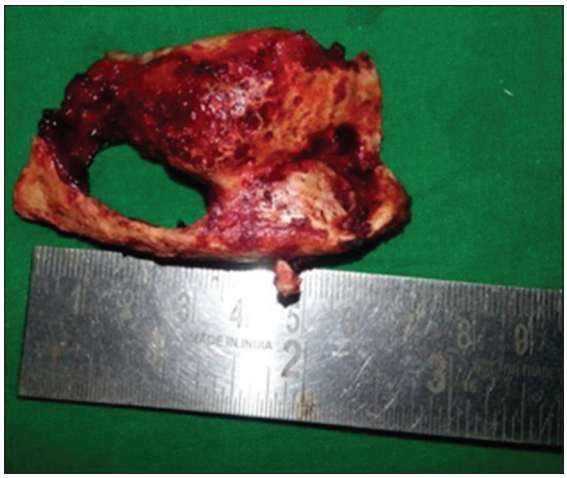

Figure 7: Gross specimen

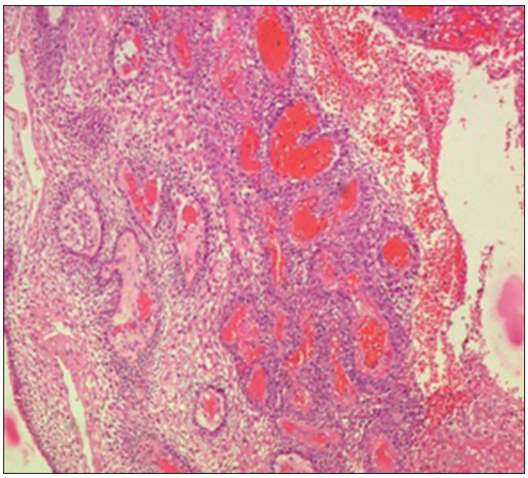

Figure 8: Histopathological image showing odontogenic epithelial islands with a prominent vascular component (H&E, ×100)

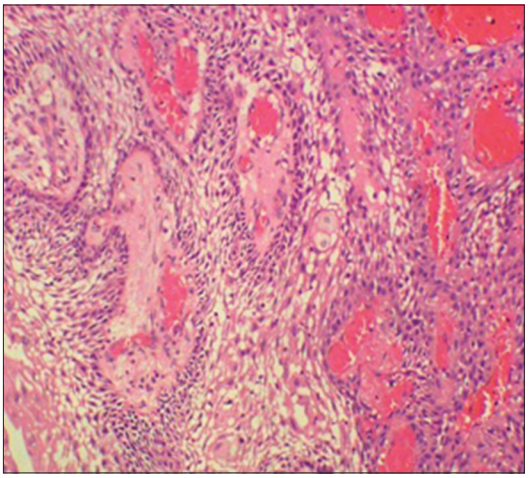

Figure 9: Histopathological image showing typical plexiform ameloblastomatous component interspersed with large blood filled spaces (H&E, ×200)

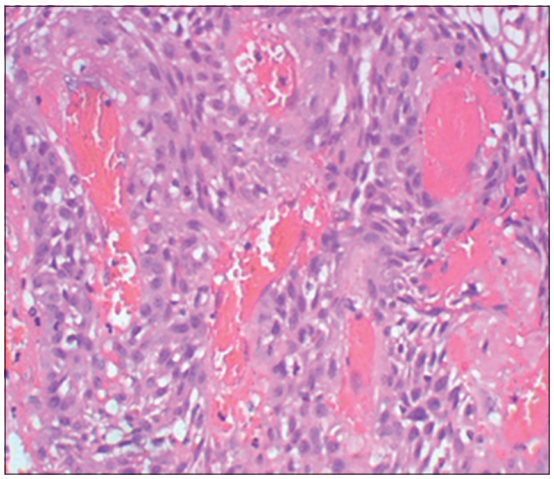

Figure 10: Histopathological image showing ameloblastomatous component with endothelial lined channels and engorged red blood cells (H&E, ×400)

preponderance,[3,4,11-15] but in this case was a female. In the present case, the radiographic appearance showed mixed radio-opaque– radiolucent lesion which is consistent with other cases reported by Kasangari *et al*.,[3] Harshavardhan *et al*.,[13] and Rajmohan *et al*.[15]

Contrary to the cases reported in the literature previously, the present case was primarily diagnosed as to be desmoplastic ameloblastoma on incisional biopsy, but on excisional biopsy, a final diagnosis of HA was made. The discrepancies between diagnosis of incisional and excisional biopsies and the extensive vascular component in excisional biopsy might be due to the following reasons: (1) Incisional biopsy might not give the representation of the entire lesion. (2) Any trauma during incisional biopsy and subsequent disturbance in the repair may result in excessive granulation tissue or abnormal vascular component. (3) The time elapsed between incisional and excisional biopsy was 2 months during which there could be stimulation of angiogenesis by inductive influence of various factors. Due to the paucity of reported cases in the literature, the origin of its vascular component is not clearly understood and is still debatable. There are various theories of the pathogenesis of HA, but none clearly concluded on the vascular component, whether it represents a part of the neoplastic process, a separate neoplasm or a hamartomatous malformation.[14]

One of the theories states that during amelogenesis, capillaries associated with the outer enamel epithelium providing necessary nutrition for enamel completion are abnormally induced and result in their abnormal proliferation. Such proliferated vessels possibly turn into a tumor component.[6] Another theory suggests that any traumatic incident such as a tooth extraction may provide the stimulus for the proliferation of epithelial cell rests in the periodontal ligament and subsequent tumor development.[6] In normal conditions, tissue damage is repaired with the

formation of granulation tissue in which proliferating endothelial cells and new capillaries are prominent, and any disturbance in the repair of neoplastic odontogenic tissue may result in excessive granulation tissue formation or the development of an abnormal vascular component.[12]

It is also suggested that excessive stimulation of angiogenesis during tumor development by inductive influences such as those that occur during odontogenesis or by other factors may result in the overgrowth of vascular elements in the odontogenic ectomesenchyme or in adjacent connective tissue.[12] Kasangari *et al*.,[3] Gargi *et al*.[4] and Rajmohan *et al*.[15] have opined that history of tooth extraction might be the etiological factor for the development of the vascular component. The other cases reported in the literature did not make a mention of the probable etiological factor.

A few believe that this neoplasm represents a collision type of tumor where two separate tumors grow in the same area and collide, and the tumor elements intermingle.[8] According to Lucas, 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.[9] 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, and according to him, the blood supply to these tumors is variable.[16]

Lesions such as hemangiomas, telangiectatic osteosarcoma, angiomatoid malignant fibrous histiocytoma should be included in the differential diagnosis. In hemangioma, there are large dilated blood vessels without ameloblastomatous component. In telangiectatic osteosarcoma, there are large spaces filled with blood and huge areas of necrosis, whereas, angiomatoid malignant fibrous histiocytoma contain characteristics of a fibrohistiocytic tumor and a vascular tumor.

Most of the cases reported opted for a wide surgical excision (hemimandibulectomy) as the treatment of choice which is similar in the present case. Surgical complications due to extensive vascularity in such vascular lesions should always be kept in mind during treatment planning. The present case has been followed up over a period of 15 months and has shown no recurrences until date. Previous reports also showed no recurrences or complications for varied periods of follow-up.

CONCLUSION

Due to the paucity of cases reported in the literature, with no long-term follow-up, the biological behavior cannot be predicted. HA with the extensive vascular component may be fatal during surgical procedures. The present case demonstrates unique histopathological pattern, and other vascular lesions should be included in the differential diagnosis. Further research is needed to exactly to know the origin, biological behavior and nature of HA.

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.

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Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Gomes CC, Duarte AP, Diniz MG, Gomez RS. Review article: Current concepts of ameloblastoma pathogenesis. J Oral Pathol Med 2010;39:585-91.
2. Barnes L, Everson JW, Reichart P, Sindransky D, editors. WHO classification of tumours series. In: Pathology and Genetics of Head and Neck Tumous. Lyon: IARC Press; 2005.
3. Kasangari MD, Gundamaraju K, Jyothsna M, Subash AV, Aravind K. Hemangiomatous ameloblastoma- A case report of a very rare variant of ameloblastoma. J Clin Diagn Res 2015;9:ZD08-10.
4. Sarode GS, Sarode SC, Vaidya K. Intraluminal plexiform hemangioameloblastomatous proliferation in unicystic ameloblastoma: An unusual case report. Indian J Dent Res 2013;24:390-2.
5. Kuhn A. A combination of adamantinoma with hemangioma as a central jaw tumor. Dtsch Mschr Z 1932;50:49-56.
6. Aisenberg MS. Adamantinohemangioma. Oral Surg Oral Med Oral Pathol 1950;3:798-801.
7. Shafer WY, Hine MK, Levy BM. A Textbook of Oral Pathology. 4th ed. Philadelphia: W.B. Saunders; 1983.
8. Oliver RT, McKenna WF, Shafer WG. Hemangio-ameloblastoma: Report of a case. J Oral Surg Anesth Hosp Dent Serv 1961;19:245-8.
9. Lucas RB. A vascular ameloblastoma. Oral Surg Oral Med Oral Pathol 1957;10:863-8.
10. van Rensburg LJ, Thompson IO, Kruger HE, Norval EJ. Hemangiomatous ameloblastoma: Clinical, radiologic, and pathologic features. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2001;91:374-80.
11. Ide F, Horie N, Shimoyama T, Sakashita H. So-called hybrid odontogenic tumours: Do they really exists. Oral Med Patol 2001;6:13-21.
12. Avinash PT, Dhirendra GS, Sudhir SB, Pariera T, Sandhya AT. Hemangiomatous ameloblastoma: A case report of a rare variant of ameloblastoma. Int J Contemp Dent 2010;1:16-9.
13. Jois HS, Kumar K *P* M, Kumar MS, Waghrey S. A mixed neoplasm of intraosseous hemangioma with an ameloblastoma: A case of collision tumor or a rare variant? Clin Pract 2012;2:e5.
14. Sharma VK, Verma SK, Goyal L, Chaudhary PK. Hemangiomatous ameloblastoma in maxilla: A report of a very rare case. Dent Res J (Isfahan) 2012;9:345-9.
15. Rajmohan M, Prasad H, Shanmugasundaram N, Tamil Thangam P, Ilayaraja V, Anuthama K. Hemangiomatous ameloblastoma: A rare variant. J Orofac Res 2014;4:63-6.
16. Smith JF. The controversial ameloblastoma. Oral Surg Oral Med Oral Pathol 1968;26:45-75.