Hemangiomatous ameloblastoma: Case report with a brief review

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

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. The clinical variants of ameloblastoma are solid or multicystic, unicystic, extraosseous/peripheral and desmoplastic type (the WHO classification). A variable number of histopathological patterns of ameloblastoma exist and the common being follicular and plexiform patterns. 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. 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.

Ameloblastoma is a benign epithelial odontogenic tumor with many histological variants. Hemangiomatous ameloblastoma 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.

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CASE REPORT

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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.
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. The first case of HA was described in 1932 by Kuhn as a combination of hemangioma and adamantinoma. Later in 1950, Aisenberg reported a similar lesion by the term admantinohemangioma. Apart from these, they are also being reported under diverse nomenclature such as ameloblastic hemangiomas, and hemangio-ameloblastomas.

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

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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[7]  | 43/female| Right mandible          | -                            | -                             | Resection of affected portion   | No complaint after follow-up| Uneventful              |
| van Rensburg et al.[8] | 26/female| Left mandible, 3rd molar region | -                            | -                             | Partial hemimandibulectomy      | No follow-up could be done  | Uneventful              |
| Ide et al.[9]   | 56/male | Anterior maxilla        | -                            | -                             | Hemimandibulectomy              | 6 months                   | Good healing            |
| Avinash et al.[10] | 31/male | Premolar- molar area of left mandible | Not mentioned | Not mentioned | Enucleation and curettage       | 4 months follow-up with good bone healing  | Not mentioned            |
| Josi et al. [2012][11] | 42/male | Posterior region of right mandible | Not mentioned | Not mentioned | Hemimandibulectomy              | 2 years follow-up          | Not mentioned            |
| Sharma et al. [12] | 15/male | Right maxilla           | Not mentioned                | Not mentioned                | Enucleation                     | Follow-up for 6 months     | Not mentioned            |
| Sarode (2013)[13] | 18/male | Right mandible          | Tooth extraction             | Tooth extraction             | Curettage                       | Patient lost for follow-up | Not mentioned            |
| Rajmohan[14]    | 20/male | Right side of mandible  | Tooth extraction             | Soap bubble appearance      | Hemimandibulectomy              | Not mentioned              | Not mentioned            |
| Kasangari et al.[15] | 35/male | Left posterior mandible | Tooth extraction             | Well defined mixed radiolucent - radiopaque lesion | Patient refused for hemimandibulectomy | Patient lost to follow-up | Not mentioned            |
| Present case (2016) | 35/female | Right posterior mandible | Trauma                       | Mixed radiolucent radiopaque lesion | Hemimandibulectomy              | No recurrence              | Under follow-up          |
preponderance,\textsuperscript{3,4,11-13} 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 \textit{et al.},\textsuperscript{3} Harshavardhan \textit{et al.},\textsuperscript{13} and Rajmohan \textit{et al.}\textsuperscript{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.\textsuperscript{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.\textsuperscript{4} 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.\textsuperscript{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.\[13\] Kasangari et al.,[13] Gargi et al.[14] 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.\[16\] 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.\[16\] 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.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

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