Hemangiomatous Ameloblastoma: A Case Report of a Very Rare Type of Ameloblastoma

Mouli Chakraborty, Rajarshi Banerjee, Mayukh Misra, Ruma Saha, Mainak Ghosh, Neeraj Kumar

Department of Oral and Maxillofacial Surgery, Haldia Institute of Dental Sciences and Research, Haldia, West Bengal, India.

Abstract

A hemangiomatous ameloblastoma (HA) was present in the posterior region of the left mandible of a 23-year-old woman. The histological and radiographic features of this tumor differed from those of a conventional ameloblastoma. Its histopathologic features were consistent with those of a HA, and its standard radiologic features and computed tomography mimicked that of cystic lesions. The behavior and prognosis of the HA are uncertain due to the small number of documented cases and lack of long-term follow-up, but are thought to be similar to those of the conventional type. The relevant clinical, radiologic, and pathologic features of this case are presented with its typical treatment.

Keywords: Ameloblastoma, Hemangiomatous, Recurrence, Resection, Variants.

Introduction

Ameloblastomas are slow-growing solid or cystic benign, locally aggressive benign tumors of the jaws that consist of odontogenic epithelium in a connective tissue stroma. It expands severely to the cortical bones and may have a high recurrence rate.[1] Their polymorphic nature is reflected by the variety of recognized histologic patterns with which they may appear. The follicular and plexiform patterns are the main histologic types. Commonly encountered, histologic variants are acanthomatous and granular cell types. A mixture of these patterns is, however, usually found in a single tumor.[2] Other less commonly encountered histologic patterns include desmoplastic ameloblastoma, basal cell ameloblastoma, clear cell ameloblastoma, and unicystic ameloblastoma (UA).[3] Variations in histomorphologic patterns do not appear to have a significant bearing on the biologic behavior or prognosis of these tumors, with the possible exceptions of unicystic and desmoplastic types.[3,4]

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.[5] Lesions with similar histologic features that probably represented the same entity were documented in the early literature as adamantinohemangiomas,[6,7] ameloblastic hemangiomas,[8] and hemangio-ameloblastomas.[9] In 1966, Smith suggested that the HA should not be regarded as a separate histologic pattern because the blood supply to these tumors was variable and other circumstances may have affected the number and size of the blood vessels associated with them.[10]

The origin of the vascular component of the HA is not completely resolved, and the histologic and radiologic features differ from those of the accepted types of ameloblastomas. Additional published cases will provide data for a complete clinical and prognostic profile of this lesion. This article presents an ameloblastoma with clinical, radiologic, and histologic features, along with its treatment with those of an HA.

Case Report

A 23-year-old female reported in the Department of Oral and Maxillofacial Surgery, Haldia Institute of Dental Sciences and Research, with a gradually enlarging asymptomatic swelling in the posterior region of her left mandible (Figure 1a). She was aware of the lesion 1 year previously and reported that a foul-smelling yellow fluid had been draining from the lesion for the past few months. Her medical history was noncontributory.

Address for correspondence: Mouli Chakraborty, Dr. B C Roy Hospital, Balughata, Banbishnupur, Haldia, Purba Medinipur, West Bengal - 721645, India.
Email: mouli.chakraborty94@gmail.com

Received: Mar. 10, 2022; **Accepted:** Apr. 02, 2022; **Published:** Apr. 27, 2022

How to cite this article: Chakraborty M, Banerjee R, Misra M, Saha R, Ghosh M, Kumar N. Hemangiomatous Ameloblastoma: A Case Report of a Very Rare Type of Ameloblastoma. J Res Adv Dent 2022;13(3):3-6.

Access this article online

Website: www.jrad.co.in

DOI: https://doi.org/10.53064/jrad.2022.13.4.190

Extraoral examination revealed a diffuse swelling is seen in the lower third of the left side of face, ovale in shape, size approx. 7 × 5 cm, extending anteroposteriorly; from 1 cm behind the corner of the mouth up to angle of mandible, and superioinferiorly from ala-tragus line up to the lower border of mandible. Overlying skin was normal, no temperature rise, non-tender on palpation, firm consistency, and no pus discharge (Figure 1b). A single right submandibular lymph node was tender and palpable. Intraoral examination revealed vestibular obliteration irt 37, 38 region, and buccal cortical plate expansion with the feature of eggshell crackling, distoproximal pocket was present irt 38. No swelling was seen intraorally with no discharging of pus and overlying mucosa seemed normal. By aspiration 4 ml, straw colored clear fluid mixed with blood aspirated with 24G needle. Cystic fluid protein test – 6.92.

Panoramic and occlusal radiographs showed that a well-defined multilocular radiolucency is seen involving the left ramus and body of the mandible extending from distal aspect of 36 up to subcondylar region. Multiple thin, curved septa were seen giving a soap bubble appearance. Root resorption present irt distal root of 36, mesial and distal root of 37. Downward displacement of inferior alveolar canal was clearly visible (Figure 1c).

Computed tomography (5-mm axial slices, bone, and soft-tissue windows) showed a large space occupying lesion with bone destruction, in the left mandibular body, ramus region, and causing elevation of the ipsilateral masseter muscle. Large hypodense collection was seen at the level of 2nd, 3rd molar medullary expansion of left mandibular body with cortical thinning. Posterior cortex was paper thin and almost non-visualized extending superiorly along the ramus which showing thinning (Figure 1d). Primarily incisional biopsy report came as primordial cyst with fibro-osseous changes.

On the basis of the histopathological examination, left partial hemimandiblectomy was done with immediate reconstruction with a titanium recon. plate as patient could not afford vascularized free graft (Figure 2). Excisional biopsy report revealed a cystic cavity lined by odontogenic epithelium in a loosely arranged vascular connective tissue stroma which consists of small to medium sized endothelium lined congested capillaries. Focal epithelium was surrounded by cuboidal to flat ameloblast like cells with central stellate reticulum like cells. Based on these histopathologic findings, the lesion was diagnosed as HA (Figure 3).

The patient is under follow-up. The follow-up radiographs showed good healing of the bone in 6 months and there are no symptoms of recurrence still now.

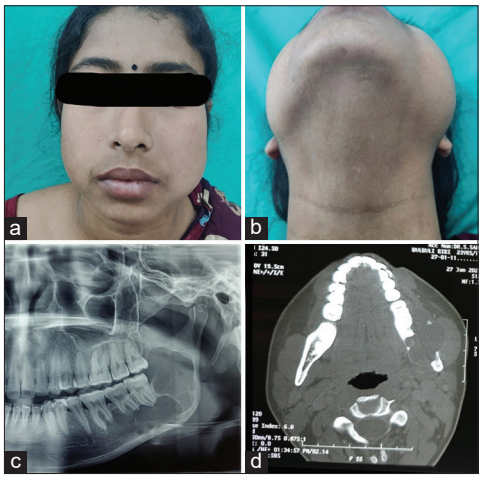

Figure 1: (a) Patient profile pre-op, (b) Extra-oral examination, showing swelling at the left side, (c) Pre-op OPG of the patient, (d) axial slice of the CT Scan, showing the lesion

Discussion

The origin of UA has remained a debate among researchers from several decades. It may arise *de novo* as a neoplasm or due to neoplastic transformation of non-neoplastic cystic epithelium.[1] According to Vickers and Gorlin, who conducted studies on such cases in the past, the diagnosis of ameloblastoma should satisfy the features such as (a) hyperchromatism of the basal cell nuclei of the epithelium lining the cystic cavity, (b) palisading arrangements of basal cells, (c) polarization of nuclei of the basal cells, and (d) cytoplasmic vacuolation of

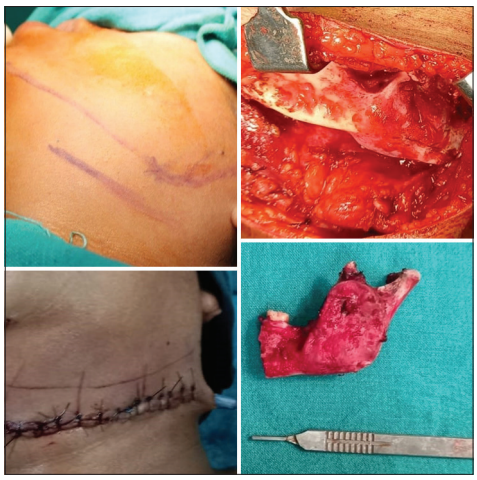

Figure 2: Intra-op photographs of the patient

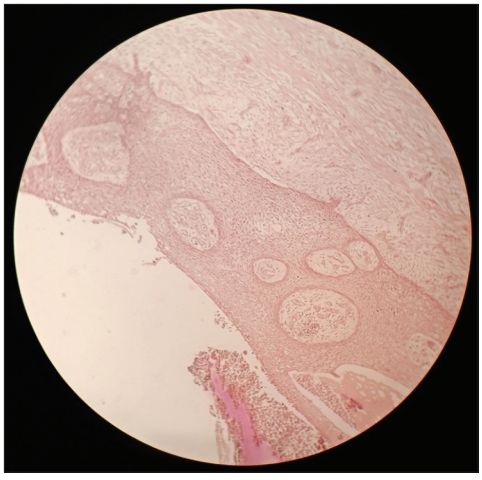

Figure 3: Histopathological examination of the excised specimen

the basal cells.[11] Our case satisfies all the criteria of Vickers and Gorlin.

The radiographic appearance of UA is mostly unilocular radiolucency; however, multilocular radiolucencies may also appear especially in some recurrence cases. Differential diagnosis of dentigerous cyst and odontogenic keratocyst is considered to be most common.[12] On histopathological examination, it was finally diagnosed to be a case of hemangiomatous proliferation in UA.

Various theories have been put forth to explain the pathogenesis of the vascular component in ameloblastomas. During amelogenesis, many capillaries are associated with the outer enamel epithelium. They furnish the profuse blood supply necessary for enamel completion. It is probable that in the HA these blood vessels are abnormally induced to become part of the tumor.[7] It is also said that excessive stimulation of angiogenesis during tumor development, by inductive influences such those that occur during odontogenesis or by other factors, may result in the overgrowth of the vascular elements in the odontogenic ectomesenchyme or in the adjacent connective tissue.

Alternatively, 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.[7] 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.

It had been suggested by Oliver *et al*. that the HA represents a collision tumor. In this type, two separate tumors grow in the same area and collide, and the tumor elements intermingle.[9]

Smith regarded the HA as histologically similar to one of the other recognized types of ameloblastoma and not as a distinct histologic entity. He thought that the blood supply to these tumors was variable and that circumstances other than the number and size of the vessels influenced the blood supply.[10] Adisa *et al*., in 2010, reported a case of HA in mandible and suggested repeated surgical intervention as a possible etiology for its development.[13] Whether the vascular component of the HA is part of the neoplastic process, represents a separate neoplasm, or is a hamartomatous malformation which has not been satisfactorily resolved. A very few cases of HA have been reported in the literature. Van Rensburg *et al*., in 2001, reported a case with histologic features consistent with those of a UA, but radiologic features and computed tomography mimicking a fibro-osseous lesion and magnetic resonance imaging suggesting a vascular lesion.[14] Sharma *et al*. reported a case of HA in maxilla in 2012 in a 15 years old.[15] Sarode *et al*. reported a case of intraluminal plexiform HA in a UA.[16] Hegde *et al*. reported hemangiomatous proliferation in a case of mural and luminal plexiform UA and plexiform ameloblastoma of mandible.[17] Kasangari *et al*. reported a case of HA in mandible of a 35-year-old female patient.[18] In our case, 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 radiologic features are nonspecific but suggestive of a fibro-osseous lesion.

Compute tomography (CT) scan clearly demonstrates cystic features in this tumor, such as its expansile nature and soft-tissue contents and proliferative endosteal reaction, that cause a fibro-osseous–like appearance on conventional radiographs. The panoramic radiograph finding in the HA of a lesion resembling a fibro-osseous lesion is nonspecific. Fibro-osseous lesions can appear radiolucent, opaque, or of mixed opacity, depending on the degree of calcification within the lesion. An important point of radiographic differentiation between larger fibro-osseous tumors, such as the ossifying fibroma and other lesions, is that these neoplasms may be demarcated from their surrounding osseous bed by a thin line of lucency that represents the fibrous capsule.[7] The HA is moderately well demarcated and corticated, without a surrounding radiolucent zone.

Tumors that demonstrate vascular features are, however, well documented, and vascular variants of these neoplasms are accepted as histologic entities. Examples are the angiomatoid malignant fibrous histiocytoma[19] and the telangiectatic osteosarcoma.[20] 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.

The biologic behavior of HA is thought to be similar to that of the conventional ameloblastoma, but because few cases have been reported, the pathogenesis and clinical features are not yet fully understood and biologic behavior cannot be predicted with certainty. The treatment of such cases is vital due to its high recurrence, for which enucleation, marginal and aggressive resection, is needed. CT helps in detecting the extension of the lesion.[21] UA has better prognosis than other variants, but sometimes the prognosis is bad as it can breach the periphery of the capsule and can infiltrate the surrounding bone behaving more aggressively at any stage.[22]

Conclusion

There have been very few cases reported in the literature of HA. Very few cases have been reported in the literature, in which improper management led to rapid, fatal exsanguination. It is likely that surgical accidents involving vascular lesions go unreported. This case clearly demonstrates the distinctive histologic, conventional, and special imaging features of an HA. We believe the HA should be regarded as a distinct histologic entity.

Declaration of Patient Consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal.

Financial Support and Sponsorship

Nil.

Conflicts of Interest

There are no conflicts of interest.

References

1. Neville BW, Damn DD, Allen CM, Bouquot JE. Oral and Maxillofacial Pathology. 2nd ed. Philadelphia: WB Saunders Co.; 2002. p. 611-6.
2. Krammer IR, Pindborg JJ, Shear M. Histological Typing of Odontogenic Tumors. Berlin: Springer-Verlag; 1992. p. 11-4.
3. Robinson L, Martinez MG. Unicystic ameloblastoma: A prognostically distinct entity. Cancer 1977;40:2278-85.
4. Philipsen HP, Ormiston IW, Reichart PA. The desmo- and osteoplastic ameloblastoma. Histologic variant or clinicopathologic entity? Case reports. Int J Oral Maxillofac Surg 1992;21:352-7.
5. Stones HH. Oral and Dental Diseases. 3rd ed. Edinburg and London: E & S Livingstone; 1957. p. 836.
6. Thoma KH. Oral Pathology. St. Louis: CV Mosby; 1944.
7. Aisenberg MS. Adamantinohemangioma. Oral Surg Oral Med Oral Pathol 1950;3:798-801.
8. Shafer WY, Hine MK, Levy BM. A Text Book of Oral Pathology. 4th ed. Philadelphia: WB Saunders; 1983.
9. Oliver RT, McKenna WF, Shafer WY. Hemangioameloblastoma; report of a case. J Oral Surg Anesth Hosp D Serve 1961;19:245-8.
10. Smith JF. The controversial ameloblastoma. Oral Surg Oral Med Oral Pathol 1968;26:45-75.
11. Rajendran R, Sivapathasundaram B. Shafer's Textbook of Oral Pathology. 6th ed. Netherlands: Elsevier; 2009. p. 276.
12. Kasat VO, Karjodkar FR, Ladda R. Plexiform unicystic ameloblastoma – A case report and review of literature. J Health Res 2014;1:103-6.
13. Adisa AO, Lawal AO, Olusanya AA, Arotiba JT. Hemangiomatous ameloblastoma: Report of a case. Niger Dent J 2010;18:73-5.
14. 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.
15. 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.
16. 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.
17. Hegde U, Sheshanna SH, Chandan SN, Mahima VG. Enriched vascularity in ameloblastomas, an indeterminate entity: Report of two cases. J Cancer Res Ther 2015;11:946-9.
18. 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:D08-10.
19. Enzinger FM. Angiomatoid malignant fibrous histiocytoma: A distinct fibrohistiocytic tumor of children and young adults simulating a vascular neoplasm. Cancer 1979;44:2147-57.
20. Matsuno T, Unni KK, McLeod RA, Dahlin DC. Telangiectatic osteogenic sarcoma. Cancer 1976;38:2538-47.
21. Unlu G, Tari V, Alan H. Unicystic ameloblastoma in 8 years old child: A case report review of unicystic ameloblastoma. Int Dent Med Disord 2008;1:29-33.
22. Kiran Kumar KR, George GB, Padiyath S, Rupak S, Kumar KR, George G, *et al.* Mural unicystic ameloblastoma crossing the midline: A rare case report. Int J Odontostomat 2012;6:97-103.