

A mixed neoplasm of intraosseous hemangioma with an ameloblastoma: a case of collision tumor or a rare variant?

Harshvardhan S. Jois,¹

Mohan Kumar K.P.,²

Mandali Satish Kumar,³

Shefali Waghrey⁴

¹Awadh Dental College & Hospital, Jamshedpur, Jharkhand; ²College of Dental Sciences, Davengere; ³MNR Dental College, Sangareddy, Andhra Pradesh;

⁴Panineeya Institute of Dental Sciences, Hyderabad, India

Abstract

Hemangiomas of the head and neck are considered to be benign tumors of infancy that are characterized by a rapid growth phase with endothelial cell proliferation, followed by gradual involution. Central hemangiomas are a rare occurrence and even rarer are the hybrid tumors of central hemangiomas with odontogenic tumors such as ameloblastomas. This paper reports a case of one such hybrid tumor in a middle aged adult clinical presenting as a mandibular swelling with indistinct mixed radiographic presentation and histopathologically comprising of intimately associated hemangiomatous vascular channels and typical ameloblastic areas. To the authors' knowledge this is the sixth case of such a hemangiomatous ameloblastoma which has been reported till date.

Introduction

Intraosseous hemangioma (IH) is a rare benign neoplasm amounting for 0.5% to 1% of all skeletal benign tumors^{1,2} and having an uncertain origin. Intraosseous hemangioma is considered as a hamartomatous malformation or a true endothelial tumor with production of blood vessels.¹ It mainly occurs in the vertebral column. The maxillofacial district is only occasionally involved, and studies illustrating IH of the mandible,^{3,4} upper jaw, nasal bones, or zygomatic bones are rarely reported.^{1,2} Central hemangiomas arising in the maxilla or mandible frequently present difficulties in differential diagnosis.⁵ More than one half of these cases occur in the first two decades of life. The tumor is typically a bone-destructive lesion which may be of varying size and

appearance, but is often suggestive of a cyst.⁶

The development intraosseous jaw hemangiomas is considered rare and even rarer is the existence of such lesions in combination with other pathologies such as ameloblastomas. Although ameloblastomas are relatively common tumors of maxilla or mandible, these tumors seldom appear with other co-incidental pathologies. Literature review shows rare occurrences of ameloblastoma along with central hemangioma of which only five cases have been reported till date with the most recent one being reported in the year 2001 by Van Rensburg and which has been termed as hemangiomatous ameloblastoma.⁷ This article discusses one such case of a hybrid tumor.

Case Report

A 42-year-old male patient reported to us with a complaint of a gradually enlarging asymptomatic swelling on the posterior region of his right mandible. The patient noticed the swelling about 6 months ago, which he said was slow in progression and grew inconspicuously to the present size over a period of 1 year. The swelling was not associated with paraesthesia. On extraoral examination, a single ovoid swelling was noticed on the right angle of mandible extending up to 1/3rd of the body of the mandible measuring about 3×2 cm in size. The borders of the swelling were ill defined and the skin over the swelling was normal. On palpation, the inspeitory findings were confirmed; the swelling was hard in consistency and was nontender. Intraoral examination revealed a firm, smooth, nonfluctuating swelling in the right retromolar region of mandible that extended into the buccal sulcus and anteriorly upto the mandibular first molar. On palpation, the inspeitory findings were confirmed; the swelling was hard in consistency, non compressible, non fluctuant and non tender in nature. It was not fixed to the overlying mucosa and electric pulp vitality test revealed that the teeth in the affected area were vital. The rest of his medical history was noncontributory. Based on the patient's chief complaint and clinical examination, a provisional diagnosis of ameloblastoma was made and differential diagnosis of odontogenic keratocyst was proposed.

An orthopantomograph revealed a mixed radiopaque-radiolucent lesion with diffuse borders and approximately extending from the roots of the right first premolar up to the postero-superior region of the ramus of the mandible.

Plain and IV contrast assisted computed tomography (6 mm axial slices, bone and soft tissue windows) with a dental protocol procured images in an axial plane which were

Correspondence: Harshvardhan S. Jois, Plot No. 670, Defence colony, P.O Sainikpuri, Secunderabad: 500094, Andhra Pradesh, India.
Tel. +91.996.614.2215.
E-mail: drhsjois@gmail.com

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later reconstructed in coronal plane. The mandible demonstrated an extensive expansile lytic lesion about 6.5 cm (antero-posterior) ×2.5 cm (transverse) involving the posterior part of the body and angle on the right side (Figures 1 and 2). The lesion was seen surrounding the roots of mandibular right second premolar, permanent mandibular right first molar and permanent mandibular right second molar with multiple thin and thick septae intervening the lesion (Figure 1). There was also an evidence of break in the lingual cortex along the postero-inferior border of the mandible (Figure 2). The soft tissue density (35-40 HU) component was noted to be enhancing moderately on IV contrast injection (90-100 HU).

An incisional biopsy revealed typical odontogenic epithelium in a plexiform ameloblastomatous pattern with interspersed prominent endothelial-lined blood filled spaces (Figure 3). The ameloblastoma consisted of anastomosing cords and sheets of odontogenic epithelium in a loosely arranged vascular connective tissue stroma (Figure 4). The epithelium was surrounded by columnar ameloblast-like cells and contained stellate reticulum-like areas. There were numerous cavernous endothelial lined vascular channels in the connective tissue component of the ameloblastoma which were large in size and irregular in shape. Few typical ameloblastic islands were also seen in the adjacent areas. All the above mentioned features were sug-

gestive of a hybrid hemangiomatous ameloblastoma.

The patient underwent hemimandibulectomy along with the affected soft tissue resection onto the lingual side and immediate iliac graft reconstruction of the mandible. The post operative healing was uneventful and the patient has been on follow up for close to 2 years.

Discussion

After its first report by Kühn in the year 1932,⁸ several attempts have been made to explain the pathogenesis of the vascular component in ameloblastoma. The paucity of reported cases in literature only adds to the distress of addressing this issue. A theory suggests the probable induction and proliferation of blood vessels associated with the outer enamel epithelium during amelogenesis.⁹ 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.¹⁰ Alternatively, a traumatic incident such as a tooth extraction may also initiate the formation of an odontogenic tumor with simultaneous formation of

excessive granulation tissue or the development of an abnormal vascular component.⁹ Lucas believed that the unusual vascularity is due to the entire absence of vasoformative activity. According to him, 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.¹¹ It has also been suggested that the 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.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, 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.¹²

The mixed radiolucent-radiopaque appearance of this lesion radiologically attracts many differential diagnosis including odontogenic lesions such as desmoplastic ameloblastomas, odontogenic myxomas, ameloblastic fibromas or fibro-osseous lesions such as solitary

fibrous dysplasia, ossifying fibroma, central giant cell tumor or even a chronic sclerosing osteomyelitis.¹³ An important radiographic differentiation between fibro-osseous tumors, such as the ossifying fibroma and other lesions, is that these neoplasms are demarcated from their surrounding osseous bed by a thin line of lucency that represents the fibrous capsule.⁹ The radiograph in our case showed diffuse ill-defined borders merging into the mandible which radiologically looked more like an odontogenic myxoma or desmoplastic ameloblastoma. A previous study suggests the difficulty to distinguish such a hybrid variant from the desmoplastic ameloblastoma and other types of ameloblastomas or odontogenic tumors with conventional radiographic examination and computed tomography.¹⁴ Although another previously described case was well demarcated and corticated,⁷ without a surrounding radiolucent zone, our case in presentation showed an observable breach in the lingual cortex and extension into adjacent soft tissues.

Although there are several established histological entities which demonstrate a prominent vascularity, this unusual tumor with its close connection to proliferative odontogenic epithelium stands out distinctly from the others. The only close histological differential diagnosis could probably be a chronically infected plexiform ameloblastoma with abundant granulation tissue which necessitates the presence of large amount of chronic inflammatory cells.

The biologic behavior of this hybrid tumor is thought to be similar to that of conventional ameloblastomas, but whether the vascular component of the hemangiomatous ameloblastoma is part of the neoplastic process, represents a separate neoplasm, or is a hamartomatous malformation has not been satisfactorily resolved;⁷ nevertheless as very few cases have been reported in literature, there is still a lot to know about the clinical course and outcome of this unique hybrid tumor.

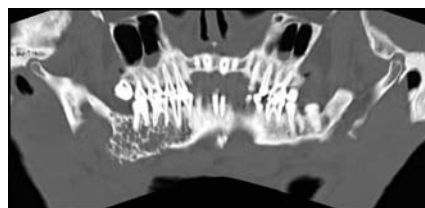


Figure 1. Panoramic reformatted computed tomography image of the lesion showing the extension of the lesion.



Figure 2. Axial computed tomography bone window showing the breach in the lingual cortical plate and extension of the lesion into the adjacent connective tissue.

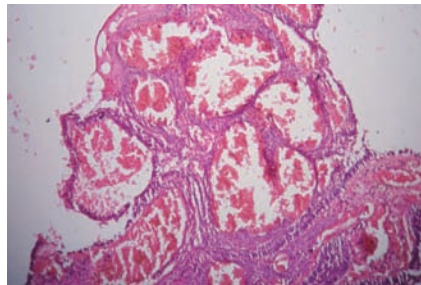


Figure 3. Photomicrograph of the tumor showing typical Plexiform ameloblastomatous areas interspersed with prominent endothelial lined vascular channels (hematoxylin-eosin, original magnification $\times 100$).

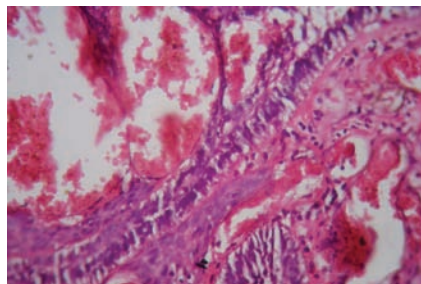


Figure 4. Photomicrograph showing tall columnar ameloblast-like cells in plexiform pattern and adjacent endothelial lined vascular channels (hematoxylin-eosin, original magnification $\times 400$).

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