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 Mandal Satish Kumar,2 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 hemangiomaticous vascular channels and typical ameloblastic areas. To the authors’ knowledge this is the sixth case of such a hemangiomaticous 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 tumors1 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.2 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-incident 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 hemangiomaticous 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 consequentially 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 inspector 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 inspector 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 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 inspector 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 later reconstructed in coronal plane. The mandible demonstrated an extensive exsudation of the right first premolar (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 posterior-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 incisal 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 amelo-
blastoma.

The patient underwent hemimandibulecto-
my along with the affected soft tissue resec-
tion 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 com-
ponent in ameloblastoma. The paucity of
reported cases in literature only adds to the
distress of addressing this issue. A theory sug-
gests the probable induction and proliferation
of blood vessels associated with the outer
coronal epithelium during amelogenesis.9 Few
believe that this neoplasm represents a colli-
sion 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 odonto-
egenic tumor with simultaneous formation of

excessive granulation tissue or the develop-
ment 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 for-
mation of stromal cysts in the ordinary type of
plexiform ameloblastoma, the blood vessels
often persist and dilate instead of disappear-
ning; thus, it’s likely to represent a purely sec-
ondary change.11 It has also been suggested
that the excessive stimulation of angiogenesis
during tumor development, by inductive influ-
ces such as those that occur during odonto-
genesis or by other factors, may result in the
overgrowth of vascular elements in the odonto-
egenic ectomesenchyme or in adjacent connec-
tive 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 influ-
ences the blood supply.12

The mixed radiolucent-radiopaque appear-
ance 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 radiographicdif-ferentiation between fibro-osseous tumors,
such as the ossifying fibroma and other
lesions, is that these neoplasms are demarcat-
ed from their surrounding osseous bed by a
thin line of lucency that represents the fibrous
capsule.3 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 exami-
nation and computed tomography.14 Although
another previously described case was well
demarcated and corticated,7 without a sur-
rounding radiolucent zone, our case in presen-
tation showed an observable breach in the lin-
gual cortex and extension into adjacent soft
tissues.

Although there are several established his-
tological entities which demonstrate a promi-
nent vascularity, this unusual tumor with its
close connection to proliferative odontogenic
epithelium stands out distinctly from the oth-
ers. The only close histological differential
diagnosis could probably be a chronically
infected plexiform ameloblastoma with abun-
dant granulation tissue which necessitates the
presence of large amount of chronic inflamma-
tory 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 ameloblas-
toma is part of the neoplastic process, repre-
sents a separate neoplasm, or is a hamartoma-
tous 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.

References

1. Cuesta Gil M, Navarro-Vila C. Intraosseous
   hemangioma of the zygomatic bone. A
2. Fechner RE, Mills SE. Tumors of the Bones
   and Joints. Washington DC: Armed Forces
3. Alves S, Junqueira JL, De Oliveira EM, et
   al. Condylar hemangioma: report of a case
   and review of the literature. Oral Surg Oral
   Med Oral Pathol Oral Radiol Endod