A case series of intraosseous hemangioma of the jaws: Various presentations of a rare entity

Srinivasa R. Chandra 1, Eleanor Chen 2, Timothee Cousin 3, Dolphine Oda 4

1 MD, DDS, Clinical Assistant Professor, 1959 NE Pacific St, Department of Oral & Maxillofacial Surgery, University of Washington School of Dentistry, Seattle, Washington 98195-7133, USA
2 MD, PhD, Assistant Professor, Department of Pathology, University of Washington Medical Center, 1959 NE Pacific St Box 357705
3 DDS, Candidate, Class of 2017, 1959 NE Pacific St, Room D322, University of Washington School of Dentistry, Seattle, Washington 98195, USA
4 BDS MSc, Professor, 1959 NE Pacific St, Department of Oral & Maxillofacial Surgery, University of Washington School of Dentistry, Seattle, Washington 98195-7133, USA

Correspondence:
1959 NE Pacific St, Room D322
University of Washington School of Dentistry
Seattle, Washington 98195, USA
cousitim@uw.edu

Received: 14/08/2017
Accepted: 09/10/2017

Abstract

Background: Hemangiomas of the soft tissue are common in the head and neck area, especially in the tongue and in children under ten years of age. Intraosseous hemangiomas of the mandible and maxilla (IHM), on the other hand, are exceedingly rare and are not well characterized. This study presents six IHM cases focusing on the clinical, radiographic, and histologic characteristics.

Material and Methods: Six cases of IHM were retrieved from the archives of the Biopsy Services at the University of Washington. Clinical, radiologic, and histologic findings are described.

Results: A total of six cases of IHM were reviewed. The patient age range was 16 to 65; the group comprised three females and three males. All six cases presented as swellings, two caused tooth resorption, and one was associated with pain and numbness. Three of the six IHM were present in the body of the mandible, two in the area of the extracted right mandibular third molar, and one in the anterior maxilla between the right canine and lateral incisor. Radiographically, five were radiolucent and one was radiopaque. Of the five radiolucent, two were unilocular and three multilocular. The one radiopaque case was exophytic, simulating a large osteoma. Histologic features ranged from cavernous to a mix of venous and arterial types. Follow-up is available for all six cases ranging between one to seven years; only one case recurred within the first year post-surgery.

Conclusions: IHM are exceedingly rare; IHM do not present in a consistent manner both clinically and radiographically. It is therefore important to recognize the wide spectrum of IHM’s clinical, radiographic, and histological presentations.

**Key words:** Hemangioma, Vascular Malformation, mandible, maxilla.
Introduction
Hemangiomas are benign vascular tumors composed of endothelial cells and supporting cells that line blood vessels. Subsequent to the 1996 ISSVA classification of vascular lesions, it has been suggested that the term “hemangioma” was used erroneously in describing soft tissue vascular lesions and should be discarded in favour of two main types of vascular anomalies: Vascular tumors and vascular malformations (1). However, ISSVA classification has not been extended to osseous vascular lesions (2). Since the biological nature of these lesions is still controversial, we frequently refrain from defining them by any one category and refer to them as “benign vascular lesions” (3).

Soft tissue hemangiomas are common, especially in children under the age of ten (4-6). Intraosseous hemangiomas however, especially those of the mandible and maxilla (jaws) (IHM), are exceedingly rare (7). In other parts of the body, intraosseous hemangiomas comprise less than 1% of all hemangiomas (3,8) and occur more commonly in the vertebrae and craniofacial bone, including the skull and the jaws, followed by long bones (9-12). Women are slightly more susceptible to developing intraosseous hemangiomas (5,11,13,14). Regarding the jawbone, the mandible is affected three times as commonly as the maxilla (13). In the mandible, the body is the more common location, while the posterior mandible, the ramus, and the condyle are also sites affected (14). IHMs tend to occur in the first three decades of life,13 while that of other bones tends to occur in older patients with peak incidence in the fifth decade and an age range of two to 85 years (9,11,12). Clinically and radiographically, IHM can be a diagnostic challenge since a wide range of presentations has been reported. In this study, we present six cases of IHM, diagnosed between 2008 and 2015, with the objective of highlighting the variability in IHM’s clinical, radiographic, and histologic features.

Case Reports
The radiographic and clinical findings of six cases of intraosseous hemangioma of the maxilla and mandible were reviewed by an oral surgeon (SC), and the hematoxylin and eosin (H&E) stained glass slides were independently reviewed by two pathologists (DO&EC). Statistical testing to determine any significant difference in our comparisons was not possible due to the small size of our series. IRB approval was obtained to perform this study.

-Clinical Findings
Of the 35,320 biopsies reviewed at the University of Washington Biopsy Services between the years 2008 and 2015, six cases were diagnosed with intraosseous hemangioma (an incidence of 0.017%). Table 1 summarizes the main clinical, radiographic, and histologic features of the six cases. There were three males and three females with an age range of 16 to 65 years (mean = 41). The age range of the three males was 28 to 46 years and of the three females it was 16 to 65 years. Five of the cases were in the mandible and one in the maxilla (case 3; Fig. 1C). In the mandible, three cases (cases 2, 4, and 5; Fig. 1B,D, E) were in the body of the mandible, and two were in the posterior mandible in area of the right mandibular third molar extending posteriorly into the ramus (cases 1 and 6; Fig. 1A,F). Five of the six cases were mildly expansile, while case five was significantly expansile with lingual extension into the floor of mouth (Fig. 1E). Five cases reported no clinical symptoms other than the expansion/swelling. Case four (Fig. 1D) reported pain followed by numbness and a mild expansion in an edentulous area. Cases one and six (Fig. 1A,F) were in the area of the right mandibular third molar extending to the ramus. However, neither of these cases reported symptoms of pain or numbness.

-Radiographic Findings
There is a wide range of radiographic presentations; five of the six cases were radiolucent (Fig. 1A-D,F), while case 5 was radiopaque (Fig. 1E). Three of the radiolucent cases were multilocular (cases 2, 4, and 6; Fig. 1B,D, F), and two were unilocular (cases 1 and 3; Fig. 1A,C). Cases 2 and 6 were associated with tooth resorption: the left mandibular first molar in case 2 and the right mandibular second molar in case 6 (Fig. 1B,E). The tumor in case 3 expanded and pushed the right maxillary lateral incisor and canine apart. Case 5 was completely radiopaque (Fig. 1E) and presented as an exophytic lesion protruding lingually into the floor of mouth. All six cases were isolated lesions.

-Pathologic Findings
The H&E-stained sections were reviewed by two independent pathologists (DO and EC) for a consensus of histologic features. Three were cavernous hemangiomas, two were venous hemangiomas, and one was an arteriovenous hemangioma. All six cases manifested large vascular spaces, four of the cases (cases 1, 3, 5, and 6; Fig. 2A,C,E,F) showed abundant erythrocytes within the vascular spaces. Case four (Fig. 2D) showed spaces with sparse erythrocytes, and case two (Fig. 2B) showed spaces with little to no erythrocytes, likely reflecting varying degrees of circulatory stasis. Three of the cases were cavernous hemangiomas (cases 3, 5, and 6). All cavernous hemangiomas (cases 3, 5, and 6, Fig. 2C,E, F) were made up of cystically dilated spaces, filled with erythrocytes and separated by dense connective tissue in some areas and by strands of bony trabeculae in other areas (Fig. 2C,E,F). Cases two and four (Fig. 2B,D) were made up of very large and tortuous vascular spaces lined by one layer of flat endothelial cells with sparse erythrocytes. Case one (Fig. 2A) had a mixture of arterial and venous channels lined by one layer of flat endothelial cells and
Intraosseous Hemangioma of the Jaws

Table 1: Summary of Clinical, Radiographic, and Histologic Features of Six Cases of Intraosseous Hemangioma of the Mandible and Maxilla.

<table>
<thead>
<tr>
<th>Case</th>
<th>Age</th>
<th>Sex</th>
<th>Site</th>
<th>Clinical</th>
<th>Radiograph</th>
<th>Histology</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>65</td>
<td>F</td>
<td>RM-Posterior and ramus</td>
<td>Area of extracted right mandibular third molar</td>
<td>Unilocular-RL</td>
<td>Arteriovenous hemangioma</td>
</tr>
<tr>
<td>2</td>
<td>44</td>
<td>M</td>
<td>LM-Body</td>
<td>Tooth resorption</td>
<td>Multilocular-RL</td>
<td>Venous hemangioma</td>
</tr>
<tr>
<td>3</td>
<td>46</td>
<td>M</td>
<td>Between the maxillary right canine and lateral incisor</td>
<td>Teeth pushed apart</td>
<td>Unilocular-RL</td>
<td>Cavernous hemangioma</td>
</tr>
<tr>
<td>4</td>
<td>49</td>
<td>F</td>
<td>RM-Body</td>
<td>Pain and numbness</td>
<td>Multilocular-RL</td>
<td>Venous hemangioma</td>
</tr>
<tr>
<td>5</td>
<td>28</td>
<td>M</td>
<td>LM-Body</td>
<td>Exophytic lingual plate (recurred)</td>
<td>Radiodense osteoma-like</td>
<td>Cavernous hemangioma</td>
</tr>
<tr>
<td>6</td>
<td>16</td>
<td>F</td>
<td>RM-Posterior and ramus</td>
<td>Area of extracted right mandibular third molar &amp; tooth resorption</td>
<td>Multilocular-RL</td>
<td>Cavernous hemangioma</td>
</tr>
</tbody>
</table>

Abbreviations: RM, right mandible; LM, left mandible; RL, radiolucent.

Fig. 1: Radiological features of Intraosseous Venous Malformation of the Mandible and Maxilla. A: This CT scan image demonstrates a unilocular expansile radioluency at the right posterior mandible in area of extracted tooth #32. B: This panoramic image demonstrates a multilocular expansile radioluency in the left body of mandible between teeth #18 and #20. Note apical resorption of tooth #19. C: This is a portion of a panoramic radiograph demonstrating a unilocular expansile radioluency in the right maxilla between teeth #6 and #7, which is pushing them apart. D: This CT scan image demonstrates a multilocular and mildly expansile radioluency in the right body of the mandible in an edentulous area. E: This panoramic image demonstrates a large radiopaque expansile nodule in the lingual left body of the mandible protruding into the floor of mouth simulating a large osteoma. F: This panoramic image demonstrates a large and multilocular expansile radioluency in the right posterior mandible in area of extracted tooth #32 extending posteriorly into the ramus. Note distal root resorption of tooth #31.
Fig. 2: Histological features of Intraosseous Venous Malformation of the Mandible and Maxilla. A: Vascular spaces filled with erythrocytes and lined by one layer of flat endothelial cell. The vascular spaces represent veins and arteries (inset: mainly arteries) (H&E stain, x100). B: Vascular spaces intertwining and lacking erythrocytes. They are lined by one layer of flat endothelial cells. The vascular spaces represent veins (H&E stain, x100). C: Large vascular spaces filled with erythrocytes and lined by one layer of flat endothelial cells. They are separated by strands of fibrous connective tissue and bony trabeculae (H&E stain, x100). D: Venous-type vessels coalescing with each other. They are lined by one layer of flat endothelial cells and have sparse amount of erythrocytes. The vascular spaces represent veins (H&E stain, x100). E: Large vascular spaces of variable sizes filled with erythrocytes and lined by one layer of flat endothelial cells. The spaces are separated by fibrous connective tissue and bony trabeculae (H&E stain, x100). F: Large vascular spaces of variable shapes partially filled with erythrocytes and lined by one layer of flat endothelial cells. They are separated by bony trabeculae (H&E stain, x100).

Discussion
In general, intraosseous hemangiomas are rare, constituting less than 1% of all hemangiomas and most commonly occur in the vertebrae followed by the skull bones. IHM, on the other hand, are exceedingly rare. They are also clinically and radiographically challenging to diagnose. This study presents six cases of IHM exhibiting variability in clinical and radiographic findings and several distinct histologic patterns, providing more insight into the behaviour of this rare condition.

Clinically, intraosseous hemangiomas of the bones in general and that of the jaws tend to occur slightly more commonly in females with a ratio of 3:2 female to male.
In conclusion, we present six cases of IHM with an overall benign clinical outcome, except for one case which recurred within one year of treatment. This recurrence suggests that although hemangiomas of the jaw are benign, they have a small potential for recurrence. Importantly, IHM do not present in a consistent manner both clinically and radiographically. It is therefore important to recognize the wide spectrum of IHM’s clinical, radiographic, and histological presentations.

References

Acknowledgements
We would like to thank Dr. Thomas Morton for kindly providing some of his cases to this case series.

Conflicts of Interest
The authors have declared that no conflict of interest exist.