

## Intraosseous Hemangioma with unusual presentation

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### ABSTRACT

Intrabony vascular lesions are extremely rare conditions. The lesions include 0.5% to 1% of all intraosseous tumors. Females are more predilected than males with a female: male ratio of 2:1, usually affecting the second decade of life. Most common sites of occurrence of these lesions are in the vertebral column and skull, jaws are the rare location with the mandible being the quite rare location. The origin of hemangiomas is still doubtful. Many of the authors believe it as hamartoma but as per World Health Organization, it as a true benign neoplasm of vascular origin. The clinical presentation of hemangiomas is variable with atypical radiographic presentations. Due to the varied presentations of this lesion, the diagnosis becomes very difficult. But diagnosing central hemangiomas early is essential for preventing uncontrollable hemorrhage and even death during any surgical intervention. We present a case of intraosseous cavernous hemangioma which presented as periapical radiolucency with specks of calcification, quite a rare presentation. The case was managed by embolization followed by surgical resection of the body of the mandible.

**Keywords:** Cavernous, hemangioma, mandible, unilocular

### Introduction

According to the World Health Organization, Hemangioma is contemplated as a benign vascular neoplasm of endothelial origin.<sup>[1,2]</sup> Intraosseous hemangioma is an exceptionally infrequent condition that accounts for about 0.2% to 0.7% of all intraosseous tumors,<sup>[3]</sup> and is also a rare neoplasm discerned in the jaws.<sup>[4,5]</sup> These can affect any part of the body but in nearly 75% of cases, the skull and spine are the most familiar sites of occurrence.<sup>[1]</sup> The intraosseous variety of hemangioma affects the molar and premolar regions of maxilla frequently as compared with mandible, where it is an extremely rare site.<sup>[6]</sup>

The origin of cavernous hemangioma is elusive. Many authors have proposed it to be a hamartomatous lesion, whereas many

studies claim it to be a true benign neoplasm which is a result of endothelial proliferation and further differentiated into blood vessels.<sup>[5-8]</sup> While some studies have claimed it to be developmental or traumatic in origin,<sup>[9]</sup> Stanley in 1849 first described the intraosseous hemangioma<sup>[7]</sup> comprising the central and peripheral variety of intraosseous hemangiomas. The central hemangioma is immensely rare and comprises only 1% of all intraosseous tumors.<sup>[6,8]</sup> It originates in the medullary bone and extends toward the bony cortex,<sup>[9]</sup> whereas the peripheral variety originates in the vessels of the periosteum with a path of growth toward the medullary bone. These lesions generally are seen on the medullary cavity and barely affect the cortex, periosteum, and subperiosteal layers.<sup>[2,3,6]</sup>

Since these lesions can generate divergent radiographic images, they often are referred to as “great mimicker.”<sup>[4,10,11]</sup> Any minor surgical procedures such as biopsy or just extraction of single tooth without prior knowledge might lead to unwanted repercussions, which can cause catastrophic hemorrhages leading to death.<sup>[12]</sup> This case report describes the appearance of the

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**Figure 1:**OPG showing ill-defined mixed radiolucent area

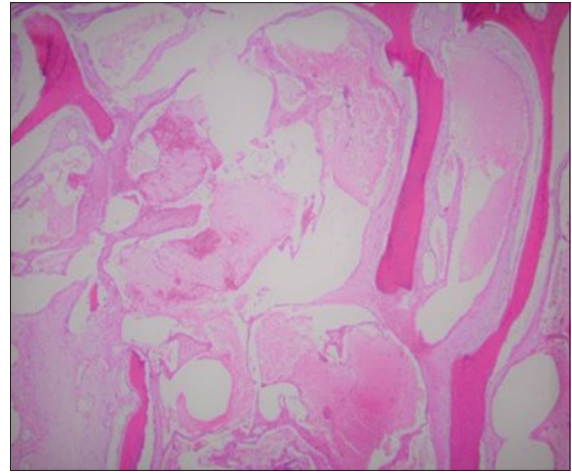
lesion which will be helpful for primary care physicians for early diagnosis of the lesion and its management.

### Case Report

A 47-year-old male patient came with a complaint of swelling in the right lower third of the face for the past 14 months. His history revealed that the swelling was sudden to start with and slowly increased to attain the present size. There was a history of mild paresthesia. The swelling was associated with mild on and off pain from the beginning but for the past 1 month, the mild persistent throbbing type of pain was present which was localized and aggravated on exertion.

Extra-oral examination revealed a solitary, roughly oval swelling present on the right lower third of the face extending from the angle of the mouth to the anterior border of the ramus anteroposteriorly, superoinferiorly from the line joining the angle of mouth to the tragus of ear to the lower border of the mandible, roughly measuring about 2 × 2 cm in size. The skin over the swelling was normal with no secondary changes. On palpation, the swelling was hard in consistency with mild pain and no lymph nodes were involved. Intra-oral examination revealed the swelling which was extending from 44 to 47 with complete vestibular obliteration, the color of the mucosa was normal, which was hard in consistency. There was a missing 45 with mobility of 46 and 47.

Orthopantomogram (OPG) revealed a single ill-defined, non-corticated mixed radiolucent, and radio-opaque lesion evident in the posterior part of the mandible measuring approx. 2 × 2 cm in the body of mandible extending from the apical area distal to 44 till the interdental area of 46 mesiodistally and from the alveolar crest of missing 45 till 2 mm above the lower border of the mandible superoinferiorly with unilocular appearance surrounded by coarse, dense, and well-defined trabeculae. The normal path of the inferior alveolar canal is altered into a serpiginous shape on the right side. The radiolucency is surrounded by a faint radioopaque line. Flecks of calcified areas can also be elicited in the distal aspect of the radiolucency just below the apical region of 46 [Figure 1].



**Figure 2:** Photomicrograph showing small vascular channels filled with red blood cells

Depending on the history and the clinical examination, we arrived at a diagnosis of residual cyst in relation to 45 with differential diagnosis of ameloblastoma, odontogenic myxoma, and aneurysmal bone cyst.

A biopsy was planned and the histopathology report showed that H- and E- stained slides revealed only blood and blood elements. No malignant cells or any other significant pathologic cells noted in the slides examined. The punch biopsy of the lesion revealed features of cavernous hemangioma without any giant cells or malignant cells.

Surgery was planned under general anesthesia. After the embolization of the lesion with ethiodol, segmental resection of the right body of the mandible was performed with L shaped osteotomy over the ramus and premolar region to get safe margins. Reconstruction was done with a 2.5-mm reconstruction titanium locking plate. The specimen was sent for histopathological examination, features were large and small vascular channels filled with red blood cells were seen and were separated by a network of dense bone [Figure 2]. The superficial epithelium showed features of mild hyperplasia. No other contributory pathology was seen. This concurred the previous diagnosis with additional features of intraosseous cavernous hemangioma. The case was under regular follow-up for the past 4 months.

### Discussion

Hemangiomas are vascular lesions more commonly seen in children if, at all affecting adults, it is mostly seen in the second decade of life, and with slightly more predilected toward females than males with a female to male ratio of 2:1.<sup>[2,13]</sup> Zlotogorski *et al.* and Smith *et al.* in their respective studies inferred that males and females are equally affected by hemangiomas with a ratio of 1.3:1.<sup>[2,7]</sup> Whereas studies conducted by Aldridge *et al.*, Kondylidou-Sidira *et al.*, Eliot and Castle, Fernandez *et al.*, 2003; Flis and Connor, Wu *et al.*, Bonini *et al.*, and Sakkas *et al.* Robson and many other studies showed female to be more predilected than men with a ratio of almost 2.<sup>[11-15]</sup> But in this case, the patient

was a 47-year-old male which was not in accordance with the study conducted by Smith *et al.* and Zlotogorski and did not coincide with the studies mentioned in the literature.<sup>[2,7]</sup>

Cavernous hemangiomas are mostly found accidentally in the skull or spine, but sometimes maxilla is seen to be affected. The mandible is an extremely uncommon site of occurrence. When detected in mandible, the body of the mandible is the most common site to be affected.<sup>[15,17]</sup> Anna *et al.* and Moore *et al.*<sup>[6,11]</sup> have mentioned maxilla to be more commonly affected than mandible but according to Langlais *et al.*, the mandibular site is common and accounts for about 2/3<sup>rd</sup> of intraosseous hemangiomas of jaws.<sup>[16,18]</sup> In our study, the presence of a lesion on the mandible itself was startling but the location in the angle of mandible was in agreement with the literature.

The pathogenesis of intraosseous hemangioma is elusive. Some believe it to be true neoplasms and some hypothesize it to be caused as a result of trauma.<sup>[19,20]</sup> Many studies have been reported in which the patients have given previous history of trauma before developing an intraosseous hemangioma. However, in our case, there was no history or predisposition for development of the intraosseous hemangioma.

Patients with intraosseous hemangiomas are usually asymptomatic. But sometimes patients might report with discomfort and facial asymmetry because of the inflammation of the soft tissue.<sup>[15]</sup> Apart from this, other signs and symptoms like pain, paresthesia, vestibular obliteration, expansion of the buccal and lingual cortex, bluish discoloration along with oozing or pulsatile bleeding from the gingiva, deranged occlusion, mobile teeth, and hastened exfoliation of the primary teeth is also elicited.<sup>[12,21,22]</sup>

A sensation of pulsation is felt frequently in patients having lesions with high vascular pressure. Large expansile lesions extending to the abutting soft tissues might have audible bruits and blanching on pressure.<sup>[21,22]</sup> Despite being a benign lesion, if a precise clinicoradiographic diagnosis is not given, minor surgical intervention might cause life menacing hemorrhage and even death.<sup>[6,23]</sup> Also in this case, there was facial asymmetry and discomfort along with vestibular obliteration and the mobility of tooth in the region affected.

The radiographic appearance of hemangiomas is not pathognomonic and can be very confusing for the clinician and a radiologist in reaching a definitive diagnosis.<sup>[20]</sup> These lesions are called “great mimicker” owing to its tendency to mimic other infrabony lesions.<sup>[21]</sup> Lesions may show variable radiographic appearances namely multilocular radiolucency with loculations resembling honeycomb or soap-bubble pattern, unilocular radiolucency, exhibits internal radiopaque striae, which radiate from the center of the lesion resembling spokes of a wheel, and the presence of parallel or tube-like arrangement of radiopaque striae is an important feature of cavernous hemangioma.<sup>[12,16]</sup> Many lesions show multiple, irregular, spotlike radiolucencies.<sup>[18]</sup> The coarse trabecular pattern perpendicular to the surrounding

bone gives the sunburst appearance. The buccal or lingual cortical expansion might be elicited along with resorption, displacement, and untimely exfoliation of teeth. The inferior alveolar canal may appear to be widened.<sup>[14]</sup>

Nagpal *et al.*<sup>[8]</sup> in his case report described the variable appearance of the lesion in a different projection.<sup>[22]</sup> Zlotogorski *et al.*<sup>[2]</sup> in their study reported that 96% of lesions were mostly radiolucent and 66% of them showed a multilocular pattern, and the radiopaque striations usually constituted several or many locules.<sup>[23,16]</sup> The classical feature is the “polka-dot” appearance with cortical expansion.<sup>[2]</sup> Langland *et al.* and Langlais *et al.*<sup>[6,11]</sup> reported unilocular radiolucency in central hemangiomas of the jaws.<sup>[16,18,24]</sup> Friedrich *et al.*<sup>[13]</sup> reported a case of intraosseous vascular malformation of infraorbital rim which appeared as a radiopaque mass on cone beam computed tomography. Abdullah *et al.*<sup>[14]</sup> reported a case of expansile destructive trabeculated intraosseous hemangioma of nasal bone. In our case, we saw a mixed radiolucent-radiopaque appearance consisting of a well-defined radiolucency with flecks of calcified areas in the distal aspect of the radiolucency just below the apical region of 46 which was a rare radiographic finding and usually not elicited in cavernous hemangioma of jaws.

Microscopically, congenital hemangiomas, depending on whether the microscopic size of the capillaries is small or large have been classified as capillary hemangiomas or cavernous hemangiomas, respectively. Cavernous type is composed of larger vessels and sinuses lined by thin endothelium and sparse stroma and the vascular spaces are lined by endothelium without muscular support.<sup>[18]</sup> Histopathologically, hemangioma has been classified into (1) capillary, (2) cavernous, and (3) mixed variant (15). Yao K *et al.*<sup>[16]</sup> described the development of hemangioma in three stages (1) early: Highly vascular, (2) intermediate: Exhibits blood clotting, and (3) terminal: Various stages of ossification.<sup>[26-28]</sup> Like in the studies conducted, in this case, the histopathologic features constitute large and small vessels which were filled with red blood cells and were separated by a network of dense bone which was following intermediate the stage explained by Yao K *et al.* The superficial epithelium showed features of mild hyperplasia.

Angiography has been used as a standard diagnostic tool for a definite diagnosis of a vascular lesion like hemangiomas. It also differentiates between the Arteriovenous malformations and central hemangioma.<sup>[25]</sup> Computed tomography (CT) angiography helps in revealing feeder vessels and vascular abnormality, which has got an important role to play in reaching a definitive diagnosis.<sup>[12]</sup> Dentici *et al.* reported that hemangiomas in angiography, appeared as well-circumscribed lesion and exhibited intense tissue staining, which had a lobular pattern. The angiogram in this case also exhibited a similar pattern explained by Dentici *et al.*<sup>[17,18]</sup>

Treatment in case of cavernous hemangiomas is usually indicated if the patient has got an esthetic problem, growing mass, pain, and continuous bleeding. Clinical observation is indicated in cases

with a minimal facial deformity and asymptomatic patients.<sup>[12,29]</sup> According to literature, there are numerous treatment modalities based on (1) hemorrhage control, (2) complete eradication of the lesion, and (3) prevent recurrence.<sup>[12]</sup>

Therapeutic alternatives include Surgery, curettage, radiotherapy, and embolization.<sup>[12,30]</sup>

Intralesional injection of sclerosing agents like boiling water, sodium morrhuate, and sodium tetradecyl sulfate can be given for extensive lesions. But studies have proved that intralesional would not be much effective because of the bony nature of the lesion. Systemic or intralesional corticosteroids can also be used along with angiogenesis inhibitors, such as interferon in selected cases.<sup>[31]</sup> The patient's age, systemic health, medical history, and clinical findings should be taken into consideration before the surgery is planned. Radiation is given for inaccessible lesions. According to Jaffe, the growth of lesion may be controlled by radiotherapy, but osseous deformity can only be corrected by surgery.<sup>[12,32]</sup> Radiation and cryotherapy also have been tried but have high-risk side effects including possible malignant transformation and loss of innervation, respectively.<sup>[9]</sup>

The most favored treatment modality in the case of hemangiomas is surgical excision with reconstruction. Prognosis after complete excision is excellent and recurrence is usually rare.<sup>[7,29]</sup> Presurgical embolization before surgery of big lesions is necessary to minimize the surgical bleeding.<sup>[3,32,33]</sup>

Hence, in our case also, embolization was done with ethiodol and then surgery was planned under general anesthesia followed by segmental resection of the right body of the mandible with L shaped osteotomy over ramus and premolar region to get safe margins. Reconstruction was done with a 2.5-mm reconstruction titanium locking plate.

## Conclusion

Cavernous hemangiomas have a bewildering presentation and have varying clinicoradiographical appearances. Consequently, any minor surgical intervention might lead to complications like hemorrhage and even death of the patient. Hence, the knowledge regarding the clinical aspects and radiographic appearance is important for the dental professional and the treatment should be carefully planned to avoid complications.

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## Conflicts of interest

There are no conflicts of interest.

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