Intraosseous venous malformation in the anterior mandible: Report of a rare case

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Abstract

Intraosseous venous malformations are rare. It commonly affects persons in the second and fifth decades of life and shows female predilection. Among the facial bones mandible, maxilla, and nasal bones are commonly affected. The molar-bicuspid region is commonly involved in the lower jaw. The present case is in a female subject in her seventh decade who presented with an asymptomatic swelling in the mandibular anterior region. The history, clinical examination, and the computed tomography imaging of the lesion pointed toward a benign odontogenic tumor or an odontogenic cyst. Based on the provisional diagnosis segmental resection of the mandible was performed. However, the post-surgical histopathological investigation gave a diagnosis of intraosseous venous malformation of the mandible. Therefore, it is of prime importance to consider intraosseous vascular malformations in the differential diagnosis of anterior mandibular swellings to prevent any intra- or post-operative hemorrhagic complications.

Keywords: Intraosseous, mandible, venous malformation

Introduction

Vascular anomalies are among the most common congenital and neonatal dysmorphogenesis. In 1982, Mulliken and Glowacki classified vascular anomalies based on histology, biological behavior, and clinical presentation as tumors and malformations.[1] The International Society for the Study of Vascular Anomalies (ISSVA) classification system categorizes these lesions into vasoproliferative or vascular neoplasms and vascular malformations. Vasoproliferative neoplasms proliferate and undergo mitosis, whereas vascular malformations are structural abnormalities of the vasculature.[2]

Vascular malformations are benign vascular lesions that are present at birth. Previously intraosseous vascular malformation was termed as central hemangiomas. However, according to the present ISSVA classification, these lesions are termed as intraosseous venous malformation.[2] Intraosseous venous malformations are rare compared to soft tissue vascular malformations of the maxillofacial region.[5] They account for fewer than 1% of all intraosseous neoplasms and frequently involve the vertebral column and calvarium.[3] In the oral and maxillofacial region mandible and maxilla are frequently involved.[3] Intraosseous venous malformations are benign congenital lesions which becomes clinically visible by second or even fourth or fifth decade of life.[4] Females are commonly affected than males with a 2:1 ratio.[5]

Intraosseous venous malformations are found to affect the mandibular body more frequently when compared to other parts of the mandible and it is very rare for the lesion to affect the anterior mandible.[4]

Clinically, the patient may be completely symptom-free or may present with discomfort, pulsatile bleeding from the gingiva of the associated teeth, swelling with bluish discoloration of gingiva, mobility of teeth, derangement of the arch form or accelerated dental exfoliation.[6]

Radiographically, the lesion appears as multilocular or unilocular radiolucency with or without internal trabeculations. These trabeculations may produce honeycomb or soap bubble pattern.[7]

The radiographic appearance mimicks lesions such as ameloblastoma, residual cyst, odontogenic keratocyst, and osteogenic sarcoma.

The primary site and extent influence the management of the lesion. The various treatment options are sclerotherapy, embolization, resection of the involved bones, and combinations of these.[3]

Early diagnosis is essential for preventing uncontrollable hemorrhage which can be fatal. Therefore, dental practitioners should be aware of the clinical and radiologic features of this vascular bone lesion. This is a report of a case where the clinical features and imaging features of the lesion did not show characteristic features of intraosseous venous

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malformation. However, the histopathologic examination of the surgical specimen revealed the true identity of the lesion to be an intraosseous venous malformation. Therefore, the clinician should consider intraosseous venous malformation in the differential diagnosis of swellings involving the anterior mandible.

**Case Report**

A female subject in her seventh decade presented with the complaint of a swelling in the lower front teeth region since 4 months. History revealed sudden mobility and exfoliation of teeth in her lower anterior region 4 months back. Following this, she developed a swelling which was of primary incidence which started as a small swelling and gradually increased to the present size. The patient complained of slight interference of speech due to the swelling. No history of altered sensation or alteration in salivation, pain, bleeding or discharge from the swelling was elicited.

Extraoral examination revealed a solitary spherical swelling in the lower third of the face in the midline measuring about 3 cm in diameter extending superior-inferiorly from the lower border of lip to the inferior border of the mandible. The swelling extends 1.5 cm on either side of the midline. The skin over the swelling appears normal [Figure 1]. No ulceration or discharge was noted from the swelling. On palpation, the swelling was non-tender, hard and non-compressible. No local rise in temperature evident over the swelling.

Intraorally, hard tissue examination revealed missing 41, 42. Gingival recession noted in relation to 31, 32, and 33. Grade II mobility noted with 31, 32, and 43. On intraoral soft tissue inspection, a solitary dome-shaped swelling was evident in the anterior mandible [Figure 2]. The swelling involved the alveolar ridge, measuring about 4 cm × 3 cm. The swelling extended from the anterior alveolar ridge down to the labial vestibule causing vestibular obliteration. Mediolaterally the lesion extended from 43 to 32 regions. Lingually the lesion involved the lingual part of the alveolar ridge not involving the floor of the mouth. The surface appeared smooth. A mild bluish hue was evident on the swelling. No sinus opening was evident. The surrounding mucosa appeared normal. On palpation, the swelling was non-tender, hard in consistency. No bleeding or discharge on digital pressure was evident.

Aspiration of the swelling revealed frank blood of around 0.5 ml. Intra oral periapical radiograph revealed diffuse radiolucrency in relation to the alveolar bone of missing 41, 42. The internal structure of the lesion showed well-defined radiopaque septa in the superior part of the lesion and inferiorly the lesion revealed fine septae with reduced density of bone. The radiograph showed blunting of root apex in relation to 31 and 43, and there is also effacement of lamina dura around these roots. In addition, 32 showed resorption in the apical one-third of mesial part of root [Figure 3a].

The mandibular occlusal view showed a diffuse radiolucent lesion involving mandibular anterior region. The internal structure revealed thick bony septae in the middle of the lesion. Labially the lesion did not show any corticated border whereas lingually, it was bordered by a thick cortical bone. The
basal bone in relation to the missing anterior teeth appeared normal [Figure 3b].

The orthopantomogram revealed a diffuse radiolucent lesion involving the anterior segment of the mandible. The lesion extended from 43 to 32. Superiorly it extended from the alveolar ridge up to 1 cm above the inferior border of the mandible. Internal structure showed thin septae and there was root resorption in relation to 43. Extensive bone loss was seen in relation to 31 and 32 [Figure 4].

The computed tomography (CT) scan of the mandible with intravenous contrast showed a non-expansile osteolytic lesion of 2.6 cm × 2.9 cm causing focal erosion of anterior cortex of the alveolar process of the right anterior body of mandible, in the symphysis and right para-symphysis region [Figure 5].

Based on the history and imaging findings a differential diagnosis of ameloblastoma, central hemangioma, central giant cell granuloma, residual cyst, odontogenic keratocyst, and aneurysmal bone cyst were given.

Segmental resection of the mandible was performed under general anesthesia, followed by reconstruction with a free fibula graft. The surgical specimen macroscopically measured 4 cm × 2.5 cm whose cut section showed whitish and hemorrhagic areas and the cross section through bone showed a cavitary septal lesion measuring 4 cm.

Microscopic examination of the excised mass revealed capillaries of varying sizes lined by flat to high endothelial cells surrounded by skeletal muscle and thinned out scanty bone tissue. There was no atypia. Histopathologic diagnosis was suggestive of cavernous hemangioma of the mandible.

Discussion

Vascular malformations are congenital vascular anomalies due to anomalous development of vascular plexus. It may occur due to an arrest in the development of the mesenchyme primodia in the undifferentiated capillary network stage followed by the penetration of primitive vessels into the bone tissue which is replaced by mature vascular channels in the later stages. Various factors such as trauma, infection, and endocrine changes can lead to increase in size of the lesion with associated skeletal abnormalities such as changes in size, shape, or density of adjacent bone.

Primary intraosseous vascular malformation is rare, benign, slow-growing vascular neoplasms accounting for only 0.7% of all primary bone tumors. They occur more commonly in whites than in dark skinned people. The review of literature states that these lesions are usually solitary and occur more frequently in females. They are typically found in adults, and the peak incidence is reported between second and fifth decades of life.

The report is of a female subject in her seventh decade who is little older than reported in the literature.

Intraosseous venous malformations in the jaws are rare. According to Batsakis, only 60 cases reported with jaw involvement, with two-thirds occurring in the mandible. In the mandible, the most commonly affected region is the posterior body in the molar-premolar region and ramus region. The present case showed the involvement of anterior region of the mandible.

The clinical features can vary depending on the site of the lesion, flow rate and type of blood vessel involved. Most of the patients present with a firm, painless, slowly progressing swelling. Hypermobility or exfoliation of the associated teeth with asymmetric expansion of the jaws, pulsatile bleeding form the gingival sulcus of the tooth in the region of lesion, bluish discoloration of the gingiva may also be seen during clinical examination. Bruit or pulsation can be detected if the cortical plates are very thin. In the present case, the patient reported with a painless swelling involving the anterior mandible with mobility and exfoliation of the associated teeth. No bruit was appreciated on auscultation.
Investigations include conventional radiography followed by CT and magnetic resonance imaging (MRI). These imaging modalities help the clinician to characterize the lesion and to delineate the anatomic extent. Conventional radiographic examination reveals the periphery of the lesion as either well defined and corticated or ill defined. The center of the lesion may appear as a multilocular or unicocular radiolucency with or without internal trabeculations. These individual loculations can be either small (honeycomb) or large (soap bubble). Rarely the radiograph shows radiating spicules at the expanded periphery giving a sunburst appearance. Vascular malformations appear isodense in a non-contrast enhanced CT image. In contrast enhanced CT it appears as a lobulated heterogeneously enhancing well-circumscribed solid mass. Prominent dystrophic veins may also be identified.[9]

In the present case, the radiographic features were suggestive of a radiolucent lesion with wide loculations and resorption of roots of associated teeth. Contrast CT image showed a non expansile osteolytic lesion with focal erosion of anterior cortex of the alveolar process of the mandible. The variable appearance of the lesion on the radiograph may lead to a differential diagnosis of ameloblastoma, central hemangioma, residual cyst, odontogenic keratocyst, aneurysmal bone cyst and central giant cell granuloma.

The site of occurrence and radiolucent appearance of the lesion favored central giant cell granuloma. Central giant cell granuloma usually affects younger age group. It appears radiographically as a well-defined unicocular or multilocular radiolucency lesion with fine septa. In the present case the patient belongs to an older age group and the radiographic appearance of the lesion also was not in favor of central giant cell granuloma.[7]

Ameloblastoma radiographically presents as a decreased radiodense lesion with a cortical border and internal bony septa. It causes cyst like expansion and thinning of adjacent cortical plate. CT images reveal regions of perforation of expanded cortical plate. It commonly affects age group between 20 and 50 years.[1] The absence of cortical border and well defined internal septa and anterior mandible with the lesion crossing the midline were not in favor of ameloblastoma.

Aneurysmal bone cyst affects younger individuals and more commonly affects mandibular molar and ramus area. Radiographically it manifests as well defined circular radiolucent lesion with ill-defined septa.[7] The site of the lesion and radiographic findings were not in favor of aneurysmal bone cyst.

The absence of well-defined cortical border in the radiographic finding of the lesion and presence of resorbed roots of the associated teeth excluded cystic lesions from the list.

Microscopically this condition can be classified based on the size of the vessel involved into cavernous which contains dilated thin-walled vascular spaces, capillary containing small tortuous vessels and mixed variety. Most of the intraosseous vascular malformations reported in the mandible are either of the capillary or mixed type. Cavernous type is mostly seen in the skull. Based on the literature, only 25 cases of cavernous type have been reported in mandible since 1975.[4] In this case, the post-surgical histopathologic report suggested cavernous type of hemangioma which is now termed as venous malformation.[3]

The management depends on topography of the pathology and patient demographics. Sclerotherapy and scalpel excision are the recommended management strategies. Besides these two techniques, a wide variety of methods like irradiation, electrocoagulation, cryotherapy, intravascular magnesium or copper needles, and compression have also been described in literature.[9] In the present case, segmental resection of the mandible was performed followed by reconstruction with free fibula graft.

**Conclusion**

The anterior mandible is a site for odontogenic and non-odontogenic tumors. Due to the varied pathogenicity and clinical features, management of the lesion becomes challenging. Advanced imaging with CT and MRI scans is very helpful in differentiating these bony lesions. This case report highlights the need to consider vascular lesions in the differential diagnosis of anterior mandibular swellings.

**References**

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