CASE REPORT

An unusual case of intraosseous vascular malformation of the maxilla mimicking fibrous dysplasia: a case report and literature review on imaging features of intraosseous vascular anomalies of the jaw

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Intraosseous vascular malformation (IVM) is a relatively rare pathological condition that may pose significant risks, such as excessive bleeding, during surgical procedures. We present a case of an 18-year-old female patient with firm swelling in the left maxilla. A bony expansion over the left half of the maxilla with preservation of the outer cortex and a ground glass appearance on CT images initially revealed a possibility of fibrous dysplasia. However, a tentative diagnosis of IVM was made based on the vascular nature of the lesion as well as the patient's surgical history and additional imaging findings. IVM should be included in the differential diagnosis of an expansile bony lesion with trabecular alteration. Through the literature review, it was found that imaging findings, such as a neurovascular canal widening on CT images and a hyperintense signal on T_1 weighted MR images, might be helpful in differentiating IVM from other pathologies.

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Introduction

Intraosseous vascular malformation (IVM) is a rare pathology that accounts for <1% of all bony tumours. It occurs most frequently in the vertebral column and calvarium and scarcely in the maxillofacial bones. Vascular lesions occurring in bone are generally referred to as central haemangiomas. In 1982, Mulliken and Glowacki¹ proposed a classification of vascular lesions, in general, into haemangiomas and vascular malformations. According to the literature review performed by Kaban and Mulliken,² the majority of the vascular lesions occurring in the maxillofacial bones were venous

malformations rather than haemangiomas. Thus, we defined vascular anomalies without endothelial cell proliferation in bone as IVMs rather than central haemangiomas in order to avoid ambiguity. The misdiagnosis of IVM before treatment could be disastrous because surgical procedures for the treatment of IVMs might cause excessive and unexpected bleeding. Therefore, the importance of a precise diagnosis in such cases can not be overemphasized.

Unfortunately, there have been no pathognomonic radiographic signs for IVM, and its radiographic appearance could be similar to many other bone lesions. Most cases demonstrated multilocular radiolucency (honeycomb or soap-bubble appearance), spoke-wheel

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appearance, presence of parallel- or tube-like arrangement, thin or coarse trabeculations and sunburst appearance.³⁻⁵ Moreover, IVM can present as a unilocular radiolucency.⁶ Overall, it might be difficult to distinguish IVM from other conditions such as ameloblastoma, osteosarcoma, aneurysmal bone cyst, central giant cell granuloma and fibrous dysplasia (FD).

We present a case of IVM mimicking FD on imagery, with particular emphasis on the differential diagnosis. We also reviewed the English literature with the goal of identifying imaging features common to intraosseous vascular anomalies of the jaw.

Case report

An 18-year-old female patient visited the Department of Oral and Maxillofacial Surgery (Seoul National University, Seoul, Republic of Korea) complaining of hard swelling in the left maxilla. In the patient's history, she had undergone a contouring surgery 8 years earlier owing to facial asymmetry; however, the operation had been unfortunately stopped because of excessive intra-operative bleeding.

A panoramic radiograph disclosed an expansile radiopaque lesion occupying her left maxilla (Figure 1). The lesion displaced the involved teeth inferiorly and elevated the floor of the nasal cavity and maxillary sinus. Trabecular alteration within the lesion was also noted. Contrast-enhanced CT images revealed an expansile lesion of the left maxilla with a ground glass appearance and coarse trabeculation occupying most of the left maxillary sinus and nasal cavity (Figure 2). A ground glass appearance and wide bony expansion with the original shape and outer cortex conservation were consistent with the imaging features of FD. In addition, infraorbital canal widening within the lesion was apparent, which was not consistent with the imaging features of FD (Figure 2b).

An MRI was then performed to gather additional information regarding the internal characteristics of the

expansile lesion (Figure 2c,d). The superomedial portion, seen as a ground glass appearance on CT images, showed a heterogeneously hyperintense T_2 signal with poor enhancement. In contrast, the inferolateral portion showed a markedly hyperintense T_2 signal and intense enhancement. T_1 weighted images showed evenly distributed hyperintense foci, indicating a residual fat component of the bone marrow. Intense enhancement and a high signal on T_1 weighted MR images supported the diagnosis of IVM rather than FD.

Based on the imaging features and her history of excessive intraoperative bleeding, angiography was performed to reveal the vascular nature and embolize the lesion. A large hypervascular tumour supplied by branches of the maxillary artery was demonstrated (Figure 3), and embolization was performed.

The following day, the patient underwent a contouring surgery of the left maxilla. Minimal hemorrhaging was encountered as a result of prior embolization, and the surgical procedure was uneventful.

The removed bone fragment was sent for pathological evaluation, and a diagnosis of IVM was made based on the finding of numerous vascular channels filling marrow spaces among the bone trabeculae (Figure 4). 1 year after the contouring surgery, she had an orthognathic surgery to correct the facial asymmetry and malocclusion. She had a residual IVM on the left maxilla, but no expansion or regrowth has been detected during the 5-year follow-up (Figure 5). Further treatment will be considered if the lesion shows expansion causing facial deformity.

Discussion

Vascular anomalies often affect soft tissues of the maxillofacial regions, yet they are rarely found in hard tissues. The term "central haemangioma" has been used to describe various vascular lesions. However, haemangiomas and vascular malformations are classified as different disease entities with different histopathological features, clinical characteristics and natural course.¹

Figure 1 A panoramic radiograph shows an expansile radiopaque lesion occupying the left maxilla (black arrows). Note the downward displacement of the left maxillary occlusal plane and elevation of the nasal and maxillary sinus floor. R, right side of patient.

Figure 2 (a) An axial-enhanced CT image with bone window shows an expansile lesion of the left maxilla surrounded by an intact corticated rim. Note the ground glass appearance in the superomedial portion. High-attenuated foci of thickened trabeculae within the lesion are also noticeable. (b) Coronal CT image with bone window demonstrates widening of the infraorbital canal (arrow). (c) An axial T_1 weighted MR image shows high-signal foci throughout the lesion. (d) An axial fat-suppressed contrast-enhanced T_1 weighted MR image reveals intense enhancement of the lesion except in the medial portion of the lesion.

Haemangiomas are vascular tumours in infancy that have a rapid growth phase followed by gradual involution.^{1,7} Pathologically, haemangioma shows a well-defined, non-encapsulated mass of plump endothelial cells and pericytes demonstrating mitotic activity, especially during the proliferative state.⁸ Vascular malformations, in contrast to haemangiomas, are present at birth and grow insidiously with the patient throughout life. They show no active endothelial cell proliferation, and vascular channels in the lesion resemble the vessels of origin.⁸

In addition, the treatment strategies are different between haemangiomas and vascular malformations based on their characteristics. Aldrige *et al*⁹ asserted that primary intraosseous haemangiomas might not exist

and, if their existence would be proven, the general treatment concept of intraosseous haemangiomas might be “watchful waiting”. However, IVMs require surgical management. Therefore, the diagnosis of vascular lesions in bone, either haemangioma or vascular malformation, should be made with caution. The present case was classified as IVM rather than central haemangioma considering that it showed numerous vascular channels with no endothelial proliferation.

Although precise diagnosis of a vascular malformation is a critical factor for successful treatment, there are no radiographic findings of IVM that are pathognomonic. We reviewed the English literature to identify some radiographic features of IVM. The literature survey was conducted using PubMed and performed using the central keywords vascular malformation or haemangioma in combination with additional keywords such as jaw, maxilla, mandible, intraosseous or central. The initial search yielded 116 articles. The reports with radiographic images were included in the review, and a total of 82 cases were evaluated.

The most frequent imaging features were multilocular radiolucency, such as honeycomb or soap-bubble appearance, followed by a unilocular cyst-like radiolucency. Usually, delicate radiopaque striations and arrangements of these striations led to various multilocular patterns.^{10,11} A sunburst appearance or spoke-wheel appearance was also reported.^{12–14} In some cases, the lesion produced an alteration in the trabecular bone pattern that may be described as thin and sharp or thick and coarse.¹⁰ Some multilocular lesions showing mixed radiolucency with coarse trabeculation were similar to fibro-osseous lesions.^{11,15} Other mixed lesions located in the zygoma or orbital rim behaved in an unusual way and appeared as a benign tumour with focal growth.^{12,16,17}

Fan *et al*¹⁸ reported a case that was comparable to our case of coarse trabecular pattern on plain radiographs. However, this feature on plain imaging was due to the endosteal lysis or cortex erosion on CT images,

Figure 3 Pre-operative carotid arteriogram demonstrates hyper-vascularity throughout the lesion (white arrows).

Figure 4 (a) Routine histology shows marrow spaces filled with numerous vascular channels and bone trabeculae. (b) Thin-walled, enlarged vascular channels with a flat, single layer of quiescent endothelium are observed in between the bony trabeculae. The endothelium shows no signs of proliferation, mitotic figures, atypia, or tufting [haematoxylin and eosin stain, $\times 40$ (a) and $\times 100$ (b) magnification].

rather than actual inner trabecular alteration. Nagpal *et al*¹⁰ reported a case of IVM in the mandible with areas of increased radiopacity and a ground glass appearance on a panoramic radiograph. That case has been cited by many other researchers as a central haemangioma showing a ground glass appearance. However, only a small portion of the entire lesion showed a ground glass appearance and was not difficult to distinguish that lesion from FD.

IVM has the potential to be confused with several other lesions owing to its diverse radiographic features. Particularly in the present case, it was very difficult to figure out IVM as final diagnosis because several image findings were comparable with those of FD. The expansile bony lesion of the left maxilla showed a ground glass appearance in the superomedial portion on the CT images, which was a representative feature of FD. A heterogeneous enhancement with high-attenuated foci of thickened trabeculae might be misdiagnosed as an immature stage of FD. Moreover, the lesion showed bony expansion with preservation of the original outer maxilla cortex and did not cross the intermaxillary suture. These findings were typical features of FD. Some authors propose that a sunburst appearance of an IVM might help distinguish vascular lesions from FD.^{10,14} However,

IVMs rarely show a sunburst appearance, and working through the differential diagnosis remains a challenging process.

Nevertheless, there have been several useful imaging findings that helped to distinguish IVM from FD in the present case. First, infraorbital canal widening was detected on the CT images. When the IVM involves the canal of the nerve and vessel, the canal can be enlarged. Enlargement of the mandibular foramen, mental foramen and mandibular canal has been reported in previous reports of IVM involving the mandible.^{19,20} On the contrary, craniofacial FD often displaces or narrows the neurovascular canal.^{21,22} Hence, searching for neurovascular canal widening with caution is worthwhile when IVM is in the differential diagnosis. Second, the present lesion showed high-attenuated foci with low-attenuated background on the CT images, corresponding to thickened trabeculae mixed with fatty bone marrow. This “polka-dot” pattern was described as a feature of IVM in the vertebra,^{23,24} which would resemble an immature stage of FD. Third, IVM usually exhibits a high signal intensity on T_1 weighted MR images because of the presence of fat within the bone marrow,^{23,25} whereas FD shows a low signal on T_1 weighted MR images.²⁶ Although the signal intensity of IVM on T_1

Figure 5 Panoramic radiograph obtained 5 years after the orthognathic surgery shows no evidence of regrowth of the residual lesion located in the left maxilla. R, right side of patient.

weighted MR images might vary depending on the amount of residual fatty bone marrow,^{12,27} it should also be included in the differential diagnosis of expansile lesions with hyperintense signal on T_1 weighted MR images. Finally, the intense enhancement of the present case was noted on MR images, although this finding was not evident on CT images. IVMs may show intense enhancement on MR images, which reflects the vascular nature,²⁸ whereas FD lesions generally do not have this feature. MRI is currently regarded as the most effective imaging modality to demonstrate IVMs, and it also helps to differentiate between low-flow and high-flow lesions on the basis of MRI findings, such as flow voids and enhancement pattern.²⁹ Angiography is an effective method to differentiate vascular lesions from other diseases. However, it is not the primary choice of imaging modality owing to its invasiveness.

Management of vascular anomalies depends on the size of the lesion, location, flow characteristics, functional compromise and patient age.^{7,9} Medical therapies include steroid or anti-angiogenic medications, and surgical procedures include laser therapy, injection of sclerosing agents, cryosurgery, radiation therapy, embolization, curettage and surgical resection.^{7,30} The effectiveness of these treatments and their indications, in contrast to soft tissue vascular anomalies, are not yet clearly defined. Surgical procedures for IVMs have a risk of significant bleeding owing to the rich collateral

circulation in the lesion. To prevent unwanted outcomes, an exact diagnosis is crucial. After the diagnosis of a vascular malformation is made, the following preparations should be considered prior to the surgery: embolization of its feeding main vessel, preparations of packed red blood cells for transfusion and the usage of various antihemorrhagic agents. Relatively simple dental procedures, such as an extraction of a tooth or biopsy from the lesion, may also cause uncontrollable bleeding. The patient in the present case had a history of contouring surgery that was terminated unexpectedly early in another institute due to the excessive bleeding during the operation.

The lesion in the present report might have been misdiagnosed as FD owing to its radiographic findings, especially the wide bony expansion over the left half of the maxilla with preservation of the outer cortex and the ground glass appearance on the CT images. It is important to be familiar with the various imaging aspects of IVM to include it in the differential diagnosis of suspicious lesions.

In conclusion, the correct diagnosis of IVM is very important to assure safe surgical procedures. Imaging findings such as neurovascular canal widening, polka-dot pattern on CT images and hyperintense signal on T_1 weighted MR images are useful to differentiate IVM from other diseases.

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