Vascular Malformations of the Orofacial Region

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Abstract

Vascular malformations are the benign lesion of the blood vessels or vascular elements and are considered errors of vascular morphogenesis, displaying abnormal dilatations and channels but no increased cell turnover. Vascular malformations are always present at birth and enlarge in proportion to the growth of the child. They do not involute and remain throughout the patient’s life. These lesions occur with equal sex predilection and do not favor any races. Vascular lesions of the face are not very common. Most of the oral and oropharyngeal lesions tend to occur in the tongue and the floor of the mouth. In most cases, the diagnosis of vascular malformations is based on clinical evaluation. Here a case report is presented of 16-year-old male patient with vascular malformation of the orofacial region. The clinical presentation, radiological finding are discussed with emphasis on recent advances for diagnosis of the same.

Keywords: Vascular malformation, tongue, CT scan.

INTRODUCTION

Vascular malformations are common lesions accounting for approximately 7% of all benign tumors, the majority of which develop in the head and neck region. In general, vascular lesions were separated into two major groups, infantile hemangiomas and vascular malformations. Hemangiomas are characterized by endothelial cellular hyperplasia, proliferation and rapid growth which is seen for 6 to 8 months after birth and then regress to a variable extent. On contrary, vascular malformations have a normal endothelial cell cycle and do not involute. Generally, vascular malformations such as lymphangiomata, hemangiomas and arteriovenous communications in the head and the neck region represent only an esthetic problem. Nevertheless, in the majority of cases when localized in the tongue, these lesions can produce clinical problems consisting of spontaneous hemorrhage from the mouth. Although uncommon, progressive asymmetric growth of the tongue (macroglossia) can also be seen.

CASE REPORT

A 16-year-old male patient reported to the department of Oral Medicine and Radiology, MA Rangoonwala college of Dental Sciences and Research Center, Pune, complaining of swelling in the lower facial region and recurrent episodes of bleeding from the mouth since childhood. It was reported that the swelling was initially smaller in size and gradually increased to present size, with growth being very slow and continuous. Patient gives history of melena and hematemesis during childhood. There was no history of anaphylaxis, no neonatal or prenatal problems, bleeding disorder, petechiae, ecchymosis, jaundice, or any drug injection or overdose. There is history of spontaneous bleeding from tongue and gingiva. On general examination, patient was poorly built and moderately nourished.

On extraoral examination a diffused swelling was seen on the right and left middle 1/3rd facial region, lateral nasal region bilaterally with upper and lower lips and chin region causing swelling of the upper and lower lips. There is also erythematous area on the lower lip and chin region which is irregular in shape extending from the vermilion border of lower lip to the entire chin region and involving submental region. The erythematous area is intermixed with purplish spots (Fig. 1).

On palpation the swelling was soft in consistency, non-tender with no evidence of pulsation or bruit. On intraoral examination tongue appears enlarged reddish purple in color with rough nodular surface having deep fissures and enlargement of all the papilla’s giving a strawberry appearance (Fig. 3) with bleeding on touching, and causing difficulty in speech and mastication. Other intraoral findings being a small reddish purple erythematous growth seen on the left side of the soft palate about 3 × 3 cm in size with well-defined borders and tumor like gingival enlargement in the lower anterior teeth region (Fig. 2) involving 31, 32, 33, 41, 42, 43 respectively was firm in consistency covering almost entire teeth surface and bleeding on touch and probing.

OPG radiograph reveals dense radiopacity in mandibular anterior region which could be attributed to the soft tissue mass, enlargement of inferior alveolar canal in lower left premolar region and alveolar bone loss (Fig. 4).

CT scan (Fig. 5) reveals hypertrophied soft tissues noted in face and neck region with multiple dilated, tortuous vessels noted in hyoid, infrahyoid, bilaterally parapharyngeal, submandibular, submental levels extending up to thoracic inlet level. There are two large vessels noted supplying these dilated tortuous vessels at infrahyoid level. Tongue is also hypertrophied and shows increased vascularity on contrast administration. On the basis of clinical examination and CT scan a diagnosis of vascular malformation of the soft tissue of Face, Tongue and Neck region was given. However, a doppler ultrasound and arteiography was advised but the patient refused for further diagnostic procedure and treatment.

Vascular malformations are extremely common lesions accounting for approximately 7% of all benign tumors. Although the head and neck constitute less than 14% of the total surface area of the body, approximately 50% of all vascular malformations occur in these regions. In the majority of cases vascular malformations are present at birth, and tend to increase in size during early childhood. Intracranial locations are relatively common and often associated with potential complications of great concern that may occur in case of rupture or bleeding. Generally, extracranial vascular malformations represent only an esthetic problem and normally do not require treatment.

Regarding the pathogenesis of vascular malformations, the abnormal development of the vessels of the head and neck region seems to be responsible for anomalous vessels formation. The normal vasculature development of the head and neck results from three different stages. The first (endothelial stage) is characterized by the formation of endothelial lakes; the factors influencing this phase of development may be responsible for hemangiomas, whilst arteriovenous malformations may be caused by a sudden arrest of the second step, also called “network-forming” stage. In this stage, intercommunicating capillary channels develop in a vascular network connecting the endothelial lakes and mural muscular elements appear in the wall of rudimentary channels. Mature arteries and veins complete the development in the last stage, forming the
definitive vascular system, coursing in the predefined territories.\(^6\)

On the basis of histological and embryological aspects, vascular abnormalities can be divided into two categories—hemangiomas and vascular malformations. Vascular malformations can be further divided into low-flow and high-flow lesions. Lymphatic and venous malformations belong to the first group, while arteriovenous malformations and anomalous branches of the arterial system are considered high-flow lesions.\(^7\) Patients with vascular anomalies of the tongue may present different nonspecific symptoms, related to the size and extent of the lesions. Frequently, the patient reports recurrent mouth bleeding due to microtraumatic events, such as biting of the tongue, teeth brushing, chewing, etc. Although the diagnosis of vascular malformations may be suspected on the basis of subjective symptoms and clinical features, diagnostic imaging is required in order to define the origin and extension of the anomalies, to differentiate between low- and high-flow lesions, and to define whether the lesions are isolated or part of syndromes such as the Sturge-Weber disease, or ataxia-van-tangiectasia syndrome.

Many imaging techniques have been proposed for the diagnosis and assessment of vascular malformations of the head and neck. Scintigraphy with Tc-99 labelled red blood cells can easily identify hemangiomas, but arteriovenous and/or abnormal development of arteries and veins can be misdiagnosed.\(^8\) Computed tomography before and after intravenous administration of contrast medium has been the main imaging diagnostic procedure for a long time, being able to identify both hemangiomas and other vascular anomalies. CT scans with the use of spiral and multirow system show a high-sensitivity in the depiction of vascular abnormalities and are also able to differentiate, with multi-phasic acquisition, the high-flow from the low-flow vascular malformation, as seen in this case. It is important to remember, however, that the ionizing radiation used with CT, particularly in studies with multiphasic protocols, may produce a great exposure to the subjects that in the majority of cases are young. Although angiography constitutes a better diagnostic technique, providing direct identification of the origin and distribution of vascular abnormalities and permitting the therapeutic management of some lesions (i.e. embolisation), nevertheless, it is a very invasive technique. Moreover, therapeutic actions during the diagnostic procedure are rarely possible, especially in cases of large vascular malformations, when extensive collateral vascularization is present, as in our case, providing little effects of treatment. Furthermore, the threat of necrosis of the entire tongue may be significant.\(^9,10\)

MRI can be used as an excellent technique in the diagnosis of vascular malformations, either by itself or before angiography with embolization. It offers a good depiction of vascular structures, permitting the differentiation between high-flow and low-flow vascular abnormalities. High-flow lesions show typical signal flow voids both in T1 and T2 sequences, with the appearance of serpentine images. Low flow lesions, on the other hand, also appear as meandering structures characterized by low signal in spin echo T1W sequences, but with moderate and homogeneous increased signal in T2W images.\(^7,11,12\) Multi-planar images using Angio-MR techniques provides the whole course of abnormal vessels, the origin of the anomalous branches and an adequate pretherapeutic planning, when the possibilities of embolization is considered.

Therapy is differentiated according to the kind of malformation: capillary malformations are best treated by laser therapy,\(^13\) while the main therapy for venous malformations localized in the extremities consists of compression. Bulk resection by means of sclerotherapy (alcohol, ethibloc) or laser photo-coagulation is often utilized in the head and neck region.\(^14-16\) Smaller, localized venous malformations can be excised. Cystic lymphatic malformations are treated surgically, although sclerosing therapies (Ethibloc, OK432) are also recommended by some authors.\(^17,18\) Arteriovenous malformations (High-flow lesion) are treated by embolization combined with surgical excision. Surgical therapy alone leads to a high-frequency of recurrence by means of collateral vessels. Ligation of the main vascular pedicle prohibits embolization in case of recurrence, thus not recommended. Combination therapy becomes an essential modality.\(^19\) In large resections it may be necessary to reconstruct the defect with a free, revascularized tissue transfer.\(^16,20\)

CONCLUSION

Vascular lesions of the face are not very common. Most oral and oropharyngeal lesions tend to occur in the tongue and the floor of the mouth. In most cases, the diagnosis of vascular malformations is based on clinical evaluation. Computed tomography with contrast medium has been the main imaging diagnostic procedure to identify both hemangiomas and other vascular anomalies. The use of spiral and multirow system shows a high sensitivity in the depiction of vascular abnormalities and are also able to differentiate between high-flows from the low-flow vascular malformation. Although angiography constitutes a better diagnostic imaging but it is a very invasive technique. MRI can be used as an excellent technique in the diagnosis of vascular malformations; it offers a good depiction of vascular structures, permitting the differentiation between high-flow and low-flow abnormalities. A vascular malformation occurs less frequently and in many instances no treatment or conservative management is preferred.

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