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Original Research Paper

Oral Medicine

Intraoral Venous Malformation-A Case report

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ABSTRACT The vascular anomalies consist of vascular tumours and vascular malformations. One among these are the venous malformations which constitute about 40% of all vascular malformations. Usually, the venous malformations present as localized lesions in the form of swellings/nodules with phleboliths, are diagnosed by ultrasonography, CT and MRI.

Summary: This is a report of a rare longstanding case of wide spread venous malformations of oral cavity and facial area with phleboliths.

KEYWORDS : Vascular malformation, Venous malformations, Phleboliths

INTRODUCTION

Vascular malformations (VM) are a group of benign lesions that may be present from birth affecting the vasculature. VM involving the head and neck region can affect capillaries, venules arteries, veins and lymphatics.[1]

ISSVA classification (2018) for vascular anomalies, categorize VM as simple capillary, lymphatic, venous, arteriovenous, combined and lesions which can be associated with other anomalies. These lesions are grouped as slow or fast flow VM based on blood flow rates demonstrated on ultrasonography and MRI scans.[2]

The incidence of VM's in head and neck region is around 1.5% of which the common is Venous malformations (VeM) (40%) followed by lymphatic malformations (12%), arterio-venous malformations (AVM) (8%), capillary malformations (4%).[3,4,5]

The VeM of head and neck, present as solitary swellings[6]/ nodules[7]/surface discolorations with phlebolith formation. Accurate diagnosis will help in preventing haemorrhagic complications.[8]

Literature search revels very few reports on extensive involvement of oral cavity with phlebolith formation. This is the report of a case of extensive VeM involving both extra and intraoral location.

Case report

A 63-year-old female patient reported with a complaint of swelling on right side of the face since childhood spread to the inside part of the mouth when she was around 15 years old. Initially the swelling was small and 1-2 in number on the skin of the right cheek and gradually increased in number and size. Patient gives a history of surgery for the same when she was 7-year-old. Since then, she noticed an increased in size, number and spread of the swelling to the oral cavity. Further, she has not taken any treatment for the same. No history of bleeding/discharge or any interfere with chewing or speaking. Patient is a known diabetic and hypertensive since 3 and 8 years respectively and is on medications.

On extraoral examination, multiple purplish fused nodules were observed on the skin of the right side of the face, involving area below the lower eyelid up to mandibular angle. The surface of the nodules appeared (Fig:1). On palpation they were non-tender, soft consistency, without any audible bruits on auscultation and no pus/bleeding on digital pressure.

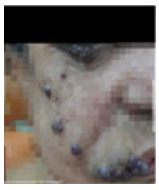

Fig: 1 Extra orally multiple purplish fused nodules

On Intraoral Examination revealed diffuse purplish swelling involving the right buccal mucosa extending up to the right soft palate region. The surface of the lesion appeared lobulated without any bleeding/discharge/ulcerative changes. Also, a purplish swelling was noted on the upper labial mucosa (Fig: 2). On palpation, the lesions were soft in consistency with pulsations with a pebbly texture and no bleeding/pus discharge on digital pressure and diascopy test was negative.

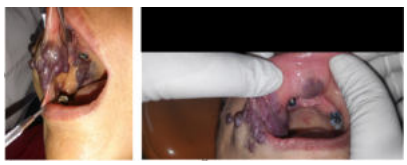

Fig: 2 Intraorally diffuse purplish swellings

A provisional diagnosis of VeM was considered.
AVM, capillary malformation, lymphatic malformation, hemangiomas and Kaposi sarcoma were considered in the differential diagnosis

Panoramic radiography reveled multiple phleboliths in the regions of right ramus and right masseter. PA skull, ruled out the calcifications in the skull region. No other bony abnormalities noted. Ultrasonography and MRI were performed which was suggestive of slow flow vascular malformation

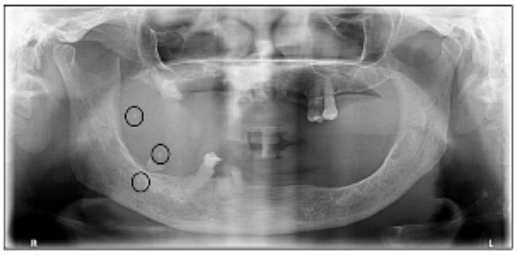

Figure 3: OPG reveals multiple phleboliths

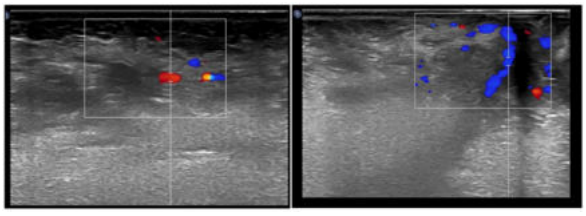

Figure 4: USG revealed vascular channels with both arterial and venous flow noted within soft tissue involving right cheek, buccal mucosa, lip suggestive of slow flow vascular malformation

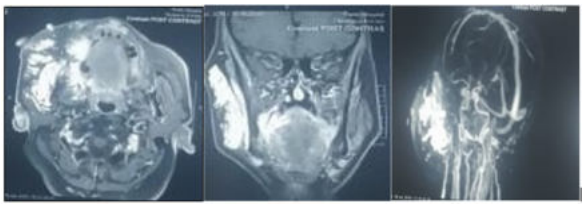

Figure 5: T2 weighted images show hyperintensity diffuse margins. T1-weighted sequences demonstrate hyperintensity in post-contrast T1-weighted sequences as a result of intralesional pooling of contrast agent.

These findings confirmed the diagnosis of VeM.
The patient refused treatement and was advied for follow up after 3 months.

DISCUSSION

VeM affects both males and females equally and generally present since birth. They increase in size due to hormonal changes, infection or trauma.[9] Post trauma increase in the size of the lesion can be due to the presence of any residual lesions or vascular nidus which stimulate neovascular recruitment by inducing ischemia.[10] The history in the present case was on similar lines.

VeMs can be localized to certain areas of the skin and mucosa. However, in the present case the lesions were multiple and widespread involving the right side of the face and buccal mucosa and labial mucosa.

VeM can present as small isolated lesions or complex growths involving different parts of the body. They are characterized as slow flow lesions with abnormal venous network. The incidence of venous malformations is rare is roughly about 2 in 10,000 births.[11]

Venous anomalies mostly occur sporadically, but families with dominant inheritance have been identified in the pathogenesis. The mutation in chromosome 9p21 has been noted causing ligand-independent activation of an endothelial cell-specific receptor tyrosine kinase altering angiogenesis in the venous anomalies.[12]

Clinically VeM manifest as a bluish, soft, compressible lesions involving different tissues and organs which was true in the present case. The lesions swell with an increase in the venous pressure.[1]

AVM's are congenital lesions of arteries and veins communicating by a central nidus presenting as an asymptomatic birthmark or warm pink-bluish lesion which are latent in the childhood and pulsatile in adolescence. Lesions are firmer in comparison to VeM's when on compression they do not empty readily.[3]

Capillary malformation usually presents at birth and persists throughout lifetime and appear as a macular, pink or purple stain with a cobblestone appearance and distortion of facial features. Port-wine stain with history of seizures and leptomeningeal involvement called Sturge-weber syndrome is a feature of capillary malformation however these were ruled out.[13]

Lymphatic malformation is a congenital disorder which never regress in size, small, dome-shaped usually pink/yellow/bluish. This may be caused by either a venous component or an intralesional haemorrhage which enlarges significantly leading to anatomic distortion. It expands and contract based on the amount of lymphatic fluid present. [14] However in the present none of these features were present.

Hemangiomas present since birth, they remain asymptomatic with a female predilection and appear as solitary, deep red or blue-purple lesions. Common site is anterior tongue. The osseous involvement rare[15]unlike the present case.

Kaposi sarcoma present as multiple bluish-purple lesions growing slowly over years and develop into painless tumor nodules. Common site is palate. It is a disease of late adult life with male predilection and common in individuals infected by HIV. Cutaneous lesions are rare [16]. None of the features were part of our patient.

Phleboliths are noted in VeM's as a result of inflammation and venous stasis due to the intralesional calcifications.[17]

VeMs can be diagnosed by Doppler US, CT, MRI, and direct phlebography.

The Doppler US helps in differentiating VeMs from other vascular anomalies. Usually, they present as hypoechoic or heterogeneous lesions in most of the cases. Anechoic channels are visualized in less than 50% of cases, whereas isoechoic thickening of the subcutaneous tissues without a solid mass are sometimes the only feature. Doppler US classically demonstrates monophasic, low-velocity flow lesions and in 20% of lesions, no flow is demonstrated, this absence of flow is attributed to thrombosis or may be caused by equipment limitations.[18]

The MRI is excellent for defining the extension of venous malformations and relationship to adjacent structures. VeM's are usually hypointense or isointense, cases of haemorrhage or thrombosis, evaluating the circulatory portion of the malformation are also observed at T1-weighted images. In T2-weighted MR imaging, the extension of the malformation into adjacent structures is usually clearly delineated. Hypointensity is related to thrombosis, septation inside the malformation, and phleboliths.[18]

Direct Phlebography is a procedure in which radiographs of the veins, is taken after a special dye is injected into the veins. It is the gold standard for diagnosing deep venous thrombosis and atypical VeM's. Opacification of abnormal venous cavities allows confirmation of the diagnosis of venous malformation and exclusion of other soft-tissue tumours. Three different phlebographic patterns can be observed with VeM opacification, the most common of these is a cavitary pattern with late venous drainage without evidence of abnormal veins, the second is a spongy pattern with small "honeycomb" cavities and late venous drainage, and the third is rapid opacification of dysmorphic veins.[18]

Various methods have been used to treat venous malformations, including surgery, sclerotherapy, laser therapy, cryotherapy, electrocoagulation treatment, treatment with copper needles and conservative treatment such as head position elevation, local compression, anti-infection therapy, pain control.

These lesions can undergo secondary thrombosis formation and have a tendency to bleed. This makes it important to diagnose and manage these lesions.

CONCLUSION

VeM's are esthetically unpleasing and most patients desire it to be treated but the surgical complications of significant hemorrhage are a matter of concern. These lesions, when seen in the oral cavity may be an additional source of bleeding due to high chances of trauma, either iatrogenically or self-inflicted. Therefore, the dental surgeon should consider the differential diagnosis of bluish-black mass in the maxillofacial region.

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