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## RESEARCH ARTICLE

### VENOUS MALFORMATION IN THE OROFACIAL REGION:A RARE CASE REPORT

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#### ABSTRACT

Venous malformations are slow growing benign tumors of aberrant venous connections. They are clinically present in adults as bluish, soft, compressible lesions typically found on the face, limbs, or trunk. Rapid growth may occur during puberty, pregnancy or traumatic injury. Calcifications can be seen in these malformations. Here, we report a case of slow flow venous malformation in the floor of the mouth manifesting as a small bluish swelling which was subsequently treated with 3% sodium tetra decylsulfate. This paper provides an overall understanding regarding the presentation, investigations and management of small vascular lesions in the orofacial region.

## INTRODUCTION

Vascular malformations are slow growing, infiltrative, destructive, irregular vascular networks defined by their particular blood vessel type which are present at birth. A vascular malformation is a localized defect in vascular morphogenesis whereas a vascular tumor grows by cellular hyperplasia (Gresham, 2012). Based on their flow characteristics they are classified as slow-flow (capillary malformation, venous malformation, lymphatic malformation) and fast-flow (arteriovenous malformation) lesions (Joshua et al., 2014). Like other vascular malformations venous malformations (VMs) are present at birth. They affect 1% to 4% of individuals, and are the most common type of vascular malformations which appear as a bluish, soft, compressible lesion clinically and are found commonly on the face or trunk. Masses of veins and venules of different dimensions are present which are lined by a single endothelial layer (Colletti et al., 2014). Venous malformations expand and contract based on patient positioning as they are dependent lesions.

They often increase in size with puberty, hormonal changes, or infection and are proportional with the growth of the individual (Pappas, 1998). Phleboliths, are pathognomonic of VMs which occurs as a result of predisposition to thrombosis, and are diagnostic on Radiographs (Mulliken, 1982).

## CASE REPORT

A 33 years old female patient, reported to the department of Oral Medicine and Radiology with the chief complaint of painless swelling below the tongue of 6 months duration. She noticed the asymptomatic swelling 6 months back with no history of trauma which did not show any change in size. The past dental history revealed that she had undergone extraction of lower left molars due to caries 2 yrs back in a private clinic. Her past medical history was non contributory. On Intraoral examination a solitary diffuse swelling with bluish discoloration was present on right side of the floor of the mouth (Fig. 1&2), extending 0.5cm laterally from lingual frenum and antero posteriorly 0.5cm from base of tongue. The swelling was about 1.5x1 cm in size, with normal mucosa, smooth surface and ill defined borders. On palpation swelling was nontender, soft in consistency. On diascopic examination blanching was evident on compression & slow refill was present, no pulsation or bruit were evident.

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Fig. 1. and Fig. 2. Asolitary Bluish purple swelling on the floor of the mouth



Fig. 3. Occlusal radiograph revealing no calcifications & pleboliths



Fig. 4. Ultrasound of floor of the mouth showing Ill defined multiple thin walled cystic lesion noted in the inferior surface of tongue on the right side with vascularity on colour Doppler

Partial ankyloglossia was seen. Grade 1 gingival recession was present in 31,41, due to high labial frenal attachment, there was missing 18,28,36,37, & 48 and root stump in relation to 12. Crowding was seen in the lower anterior region with attrition of 31 and 41 and stains & calculus were evident. Salivary flow was adequate with normal duct orifices. Considering all the features a provisional diagnosis of vascular anomaly of the floor of mouth was considered with differential diagnosis of Venous malformation and hemangioma was given.

**Investigations and Diagnosis:** Routine blood investigations showed normal values, mandibular occlusal cross sectional radiograph (Fig. 3) showed no calcifications. Ultrasonography of floor of the mouth (Fig. 4) showed Ill defined hypoechoic areas with multiple thin walled cystic lesion in the inferior surface of tongue and floor of the mouth on the right side with vascularity on color Doppler suggestive of slow flow venous malformation on the inferior surface of the tongue. A final diagnosis of slow flow venous malformation on the floor of the mouth was made. Patient was treated with sclerotherapy by intralesional injection of 3.5ml of 3% sodium tetradecyl sulfate (Setrol) (an anionic surfactant used as a sclerosing agent IV injection which causes irritation to the intima of vein wall so that vein is permanently occluded by the development of fibrosis in the wall of the compressed vein).

Reduction in the size of the swelling was observed. Patient was asked to review after 15 days for 2<sup>nd</sup> dose of injection and is under follow up.

## DISCUSSION

Vascular malformations are the result of errors of vascular development between the 4th and 6th weeks of gestation and are clearly distinct from hemangiomas, which are benign tumours with endothelial proliferation rather than the abnormal vascular morphogenesis seen in vascular malformations (Sinny Goel *et al.*, 2015; Takahashi *et al.*, 1994). Vascular malformations are always present at birth and enlarge in proportion to the growth of the child. They do not involute and remain present throughout the patient's life (Lane *et al.*, 2000). In 1982, Mulliken and Glowacki gave a classification of vascular birthmarks, and grouped them into two major categories: hemangiomas and malformations. The updated International Society for the Study of Vascular Anomalies/biologic classification classified vascular birthmarks into vascular tumors and vascular malformations (Garzon *et al.*, 2007; Manjunath *et al.*, 2014). A vascular malformation was divided as slow-flow (i.e., capillary, lymphatic, or venous) or fast-flow (i.e., arterial) lesions. When

combinations of these were seen, the malformation were called an arteriovenous malformation (AVM), lymphatico-venous malformation (LVM), or capillary-lymphatico-venous malformation (CLVM) (Manjunath *et al.*, 2014). One of the most common vascular malformations is Venous malformation. They are post-capillary lesions and have no arteriovenous (AV) shunts thus they exhibit a low flow rate. They demonstrate life long growth which is proportional with the body, and do not regress spontaneously. The incidence of venous malformation is approximately 1:5,000-10,000; among which approximately 40% of them occur in the head and neck regions (Buckmiller *et al.*, 2010; Jia Wei Zheng *et al.*, 2013). The venous malformation is speculated to be caused by developmental defects of the venous system. Its pathogenesis is not clear. In some patients with venous malformation syndrome (such as blue rubber bleb nevus syndrome) TIE2 receptor mutation was seen (Jia Wei Zheng *et al.*, 2013; Boon *et al.*, 2004). Autosomal dominant inheritance related to mutation of the 9P locus was seen in Familial venous malformation (Jia Wei Zheng *et al.*, 2013; Boon *et al.*, 1994). Somatic mutations in angiopoietin receptor gene TEK was considered in various single or multiple venous malformations which led to loss of TIE2 receptor function (Jia Wei Zheng *et al.*, 2013; Limaye *et al.*, 2009), and up regulated expression of other vascular endothelial growth factors such as  $\beta$ TGF and  $\beta$ FGF increased the severity of the lesion (Jia Wei Zheng *et al.*, 2013; Pavlov *et al.*, 2009). The expression of matrix metalloproteinase-9 was increased in intramuscular venous malformations, which suggested that venous malformations while expanding slowly have the capability for invasive growth and angiogenesis. The presence of Progesterone receptors in venous malformations, is considered to be one of the reasons for the rapid increase in the number of lesions when hormonal changes occur (Jia Wei Zheng *et al.*, 2013; Mavrikakis *et al.*, 2009). Histologically may be ectatic or micro-venular. As a result of stasis in these low-flow lesions calcification and formation of phleboliths occur through dystrophic calcification of thrombi. The thrombus when infected can cause pain and tenderness (Jia Wei Zheng *et al.*, 2013).

Venous malformations can be located superficial or deep, in single or multiple anatomical sites. The cheek, neck, eyelids, lips, tongue, soft palate, parapharyngeal space, and floor of the mouth are the commonly affected sites. The color of the skin or mucous membrane may be normal or appear blue or dark purple. It has illdefined boundary, and the lesion is soft, compressible and phlebolith can be palpated occasionally (Jia Wei Zheng *et al.*, 2013). When located superficially Venous malformations can be noted at birth. Deep intramuscular VMs are not seen at all. There are no symptoms when the lesion is small. Deformities of the face, lips, or tongue and functional disorders can occur when the lesion increases in size. In cases of trauma, secondary infection, abrupt hemorrhage of the lesions, or changes in hormonal levels can lead to pain, swelling, and even bleeding (Mavrikakis *et al.*, 2009). Venous malformations in parapharyngeal space, tongue, and soft palate may be accompanied with swallowing, speech, and airway problems. Venous malformations can also occur within muscles (such as the temporal muscle, masseter muscle and tongue muscle), which are known as intramuscular venous malformations. Some lesions in the pterygopalatine fossa and infratemporal fossa are difficult to detect when in the early stages of development (Jia Wei Zheng *et al.*, 2013). The best diagnostic scans include ultrasound (US), CT, MRI. CT scans are not good imaging modality for Vascular Malformations

(Jia Wei Zheng *et al.*, 2013). The two noninvasive imaging techniques that are most useful in the examination of vascular malformations are MR imaging and sonography. MRI using STIR and T2 weighted fat suppression without contrast is always completely diagnostic in defining the Venous Malformation (Jia Wei Zheng *et al.*, 2013). When the physical examination and clinical history are diagnostic or highly suggestive of a vascular malformation, the most important feature to be known is whether the lesion is a high- or low-flow vascular malformation. Most information needed to examine the lesion is available from a combination of T1-weighted, fat-saturated T2-weighted, and gradient-echo (flow-weighted) MR images. Gadolinium-enhanced images have been said to be helpful in differentiating between low-flow vascular malformations such as lymphangiomas and venous malformations (Lane F. Donnelly *et al.*, 2000). MRI demonstrates an intermediate T1-weighted signal, and markedly hyper-intense T2-weighted and STIR signals. A "bunch-of-grapes" configuration with septations is characteristic (Sinny Goel, 2015).

In T2-weighted images, venous malformations can appear as "venous lakes" (Jia Wei Zheng *et al.*, 2013). Occasional flow voids corresponding to phleboliths are the additional features determined (Sinny Goel *et al.*, 2015). According to the features of the draining veins, venous malformations are divided into four types (Puig, 2005) Type I, isolated malformation without venous drainage; Type II, malformation with drainage into normal veins; Type III, malformation with drainage into dilated veins; and Type IV, dysplastic venous ectasia (Jia Wei Zheng, 2013; Puig *et al.*, 2005). Sonography is useful in examining soft-tissue masses that are suggestive of hemangiomas or vascular malformations (Lane F. Donnelly *et al.*, 2000). Vessel density on Doppler sonography has been used in differentiating other types of masses from vascular malformations (Lane F. Donnelly *et al.*, 2000). The Doppler characteristics of vascular malformations are helpful in differentiating low- from high-flow vascular malformations by evaluating the blood flow, velocity, and vessel resistance together with surrounding morphology. Hemangioma Color Doppler ultrasound reveals hypochoic lesion with intermittent color picking and show a high peak arterial doppler shift whereas venous malformations show the features of slow flow lesions and needle placement during percutaneous sclerosis can also be guided by sonography (Lane F. Donnelly *et al.*, 2000; Donnelly *et al.*, 1999). Other potential imaging sequences that have been advocated as useful in the evaluation of vascular malformations include MR angiography, venography, and lymphangiography (Lane F. Donnelly *et al.*, 2000). In the above case we considered the differential diagnosis of hemangioma which are usually present at birth and can be diagnosed by 1 year, they show rapid growth until 6-8 months and in volute by 5-9 years of age. The important signs for the diagnosis of hemangiomas include detection of disappearance of the blood on finger pressure and appearance of blood after removal of finger pressure. A deep hemangioma presents with an overlying bluish tint. Venous malformations are present at birth but often not diagnosed until the second decade of life. Venous malformations show slow growth throughout life with increase in response to infection, trauma, or hormonal fluctuation and they do not involute. The overlying skin may appear normal or possess a bluish discoloration. Venous Malformations are compressible whereas an Arteriovenous Malformation will be warm on palpation, pulse or thrill will be palpable due to its high

vascular flow. Color Doppler Ultrasonography can differentiate low flow lesions like venous malformation from high flow lesions like Arteriovenous Malformation and hemangiomas which show high peak arterial doppler shift. The management of Venous malformations includes observation, irradiation, electro coagulation, cryotherapy, low-dose aspirin, sclerotherapy, surgical excision, or combinations. Sclerotherapy is commonly the preferred treatment method for venous malformations. It can be used alone or combined with surgery and/or laser therapy. OK-432, ethanol, bleomycin, doxycycline, sodium tetradecyl sulfate, 5% sodium morrhuate, pingyangmycin (PYM), anhydrous ethanol, lauromacrogol and hypertonic saline, alone or in combination are agents used for sclerotherapy. Hypertonic saline sodium chloride 23.4% is one of the best-known sclerosant, it causes dehydration to endothelial cells and red blood cells. The drug is readily available, lacks allergenicity, and is inexpensive. Boiling water and warm hypertonic saline solutions have been variously used in treating venous malformations. They act by releasing heat energy that destroys the anomalous vascular tissues. More than boiling water, boiling saline retains heat energy which can adversely affect the vascular endothelial tissues which lead to necrosis and obliteration of the vessel's lumina, which regularly promotes coagulation of blood and death of the vessels. Other sclerosing agents work by destroying the endothelial cells of blood vessels, accelerating protein coagulation in the blood of the lesions, promoting platelet adhesion to the vascular wall during thrombosis formation and causing vascular occlusion through thrombotic mechanisms. The potential complications of sclerotherapy include mucosal ulcerations, swelling, infection, transient nerve palsy, blood loss, and anaphylaxis (Jia Wei Zheng *et al.*, 2013; Yousif *et al.*, 2016). For type I and type II lesions, mild sclerosants such as pingyangmycin can be considered; for type III and type IV lesions, strong and aggressive sclerosants such as ethanol is more suitable due to the fast vein drainage (Jia Wei Zheng *et al.*, 2013). For large lesions, multiple treatments are necessary. After treatment with sclerosing agents recurrence is common which might be due to insufficient doses and the presence of residual lesions. For patients with venous malformations in the floor of the mouth as in our case, base of the tongue, oropharynx, and larynx, appropriate methods include sclerotherapy, Nd: YAG laser treatment, surgery and combined treatments should be selected based on the extent of the lesion. For large tongue lesions, sclerotherapy can be first conducted followed by surgery, laser and further sclerotherapy is subsequently done (Jia Wei Zheng *et al.*, 2013).

## Conclusion

Small Intraoral Venous malformations are difficult to diagnose and manage. The reported case small in size similar to deep hemangioma was diagnosed as slow flow venous malformation on the floor of the mouth and treated with 3% sodium tetradecylsulfate successfully, this highlights the importance of proper investigations, diagnosis and treatment of small vascular lesions intraorally.

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