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Oral Vascular Malformations With Multiple Phleboliths Treated Using Sclerotherapy -A Report Of Two Cases

Dr. Gowri Shanmuganathan[1], Dr.G.V. Murali Gopika Manoharan[2]
[1](Post Graduate, Department of Oral Medicine and Radiology, Tamil Nadu Government Dental College and Hospital, Chennai, India)
[2](Professor, Department of Oral Medicine and Radiology, Tamil Nadu Government Dental College and Hospital, Chennai, India)

Abstract:
Vascular anomalies are developmental anomalies of the vascular plexus and should be differentiated from hemangiomas which are true neoplasm of the endothelial cells. It is often difficult to determine clinically and histopathologically whether the vascular lesion is a malformation or a neoplasm, with more than 50 % of the vascular anomalies being misdiagnosed. Imaging modalities such as MRI and USG play a crucial role in differentiating vascular malformations based on hemodynamic changes. The upsurge for simple and esthetic treatments for vascular malformation have increased more recently. This article gives a brief case report of sclerotherapy as an effective approach to treat vascular malformations in the oral cavity. In this report we present two cases of low flow venous malformation associated with multiple phleboliths treated using sclerosing agents. The patients were followed up regularly and there was good take at recipient site and also there was no recurrence.

Key Word*: Oral vascular malformations; Oral venous malformation; Multiple phleboliths; Sclerotherapy.*

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I. Introduction

Vascular malformations (VMs) are lesions of the vascular or lymphatic system that can affect any part of the body, but are common on the head and neck region. Vascular malformations (VM) are a collective term used to describe a set of lesions formed by an anomaly of the capillary, venous, lymphatic, and arterial system. These are structural anomalies of blood vessels without endothelial proliferation which most commonly manifest in childhood or adolescence and persist throughout life. They are classified according to the type of vessels involved (capillary, venous, arterial) and according to hemodynamic features (low flow or high flow).

Understanding these abnormalities in the head and neck is crucial for two main reasons. First off, the head and neck are where vascular abnormalities most frequently develop. Second, because of their anatomical position and the participation of important systems including the pulmonary, digestive, and visual, these lesions have particular implications. A thorough patient history is essential in making the right diagnosis because many of these vascular diseases have distinctive appearances. Additionally, the clinical history can aid in recommending the preferred imaging modalities and direct the doctor to the best course of action. Most frequently, ultrasonography is used for initial examination of the majority of vascular lesions. Magnetic resonance imaging (MRI) is then performed to further characterize the condition and determine its degree and structural involvement[1].The main reasons for treating vascular abnormalities are aesthetic issues, hemorrhaging events, or impairment of oral normal functioning. Laser therapy, steroid therapy, embolization, B-blocker therapy, sclerotherapy, surgery, and cryosurgery are few of the available therapeutic alternatives.

Our aim is to present two case reports of oral vascular anomalies showing the usefulness of sclerosing agents for the treatment of these lesions.

II. Case Description

Case 1:

A 48-year- old female patient reported to the Department of Oral Medicine and Radiology, Tamil Nadu Government Dental College and Hospital, Chennai with the chief complaint of swelling in left cheek region since birth. History revealed that patient has painless swelling in left cheek region since birth. The size of the swelling was gradually progressive in nature. Intraoral examination revealed multiple purplish well-defined swellings of size 3 x3.5 cm in left buccal mucosa and < 0.5 cm in left retromolar trigone and retrocommissural region. There

were no secondary changes, pulsations and discharge observed in the swelling. On palpation, the swelling was soft in consistency, compressible, reducible, fixed to the underlying tissue and non-tender (fig-1).

Based on the history and clinical findings, a provisional diagnosis of oral vascular malformation was arrived following which radiographical investigations were carried out. Based on the imaging features (fig 2 A-C) such as low flow vascularity and association with multiple phleboliths, a definitive diagnosis of venous malformation was arrived. The case was treated with intralesional injection of sclerosing agent, 1ml of 3 % sodium tetradecyl sulphate (fig-3). After 3 days on follow up, mucosal ulceration was seen which subsided gradually. During third week of follow up, the lesion had shrunken to a huge extent (fig-4A) and after six weeks, the lesion had completely resolved (fig-4 B). In both the cases, sclerotherapy was administered after test dose and after injection the patient was observed for 20 minutes, to avoid any untoward adverse events.

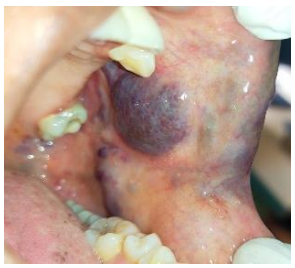

Figure 1: shows multiple well defined purplish swellings of size 3 x3.5 cm in left buccal mucosa and < 0.5 cm in left retromolar trigone and retro commissural area.

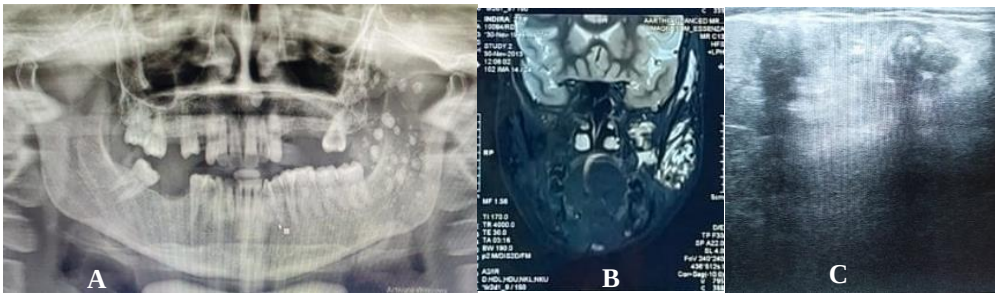

Figure 2: (A)Panoramic radiograph revealed multiple discrete radiopaque masses present in left retromolar trigone with lamellar pattern suggestive of multiple phleboliths. (B) MRI revealed Well defined space occupying lesion 8.3 x 4.8 x 3.4 cm extending completely replacing left masseter, extending into buccal space, left medial and lateral pterygoid and left temporalis muscle. (C) USG revealed relatively well defined heteroechoic lesion noted in the region of left masseter muscle with minimal internal vascularity and few phlebolith noted largest measuring 2.5 x 0.8 cm.

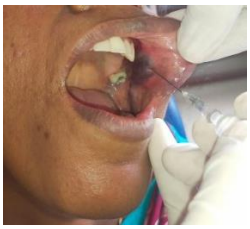

Figure 3 shows intralesional injection of 1ml of 3 % sodium tetradecyl sulphate.

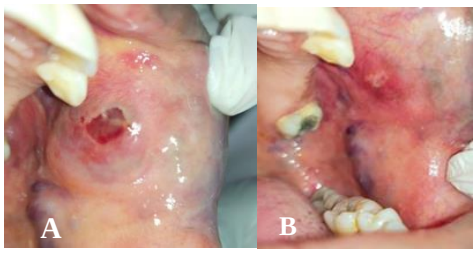

Figure 4: (A) shows change in color of the lesion with surface ulceration at third day after injection. (B) shows complete resolution of lesion noted at sixth week.

Case 2:

A 39-year- old male patient reported to the Department of Oral Medicine and Radiology, Tamil Nadu Government Dental College and Hospital, Chennai with the chief complaint of swelling in upper and lower lip and right and left side of tongue since childhood. History revealed that patient has swelling in left cheek region since childhood. Intraoral examination revealed multiple purplish well-defined swellings of size1.5 x1.5cm present in right and left side of the tongue and < 0.5 cm in upper and lower lip. There were no secondary changes, pulsations and discharge observed in the swelling. On palpation, the swelling was soft in consistency, compressible, reducible, fixed to the underlying tissue and non-tender (Fig-5).

Based on the history and clinical findings, a provisional diagnosis of oral vascular malformation was arrived following which radiographical investigations were carried out. Based on the imaging features (Fig 6 A-C) such as low flow vascularity and association with multiple phleboliths, a definitive diagnosis of venous malformation was arrived. The case was administered intralesional injection of 1ml of 3 % sodium tetradecyl sulphate (Fig-7). After 3 days, on review, mucosal ulceration was seen at the injection site. During third week of follow up, the lesion had shrunken to huge extent (Fig-8 A and B) and after six weeks, the lesion had completely resolved (Fig-8 C and D).

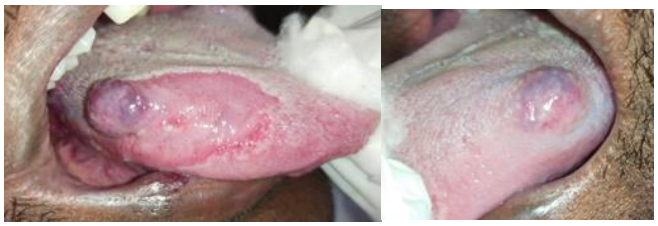

Figure 5: shows multiple well-defined swellings of size1.5 x1.5cm present in right and left side of the tongue and < 0.5 cm in upper and lower lip.

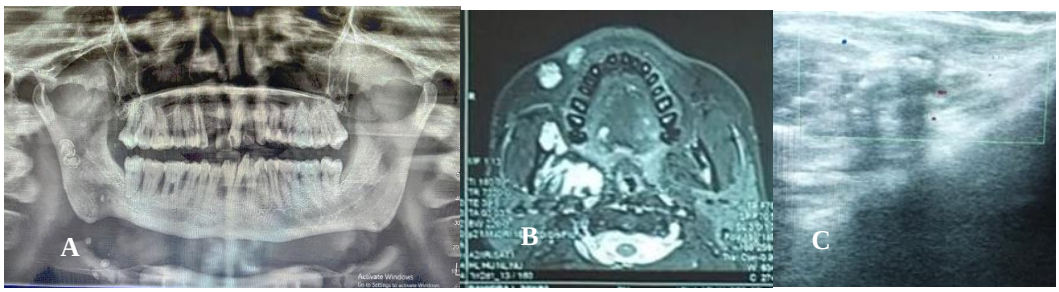

Figure 6: (A) Panoramic radiograph revealed multiple discrete radiopaque masses present in right submandibular region, right retromolar trigone region with laminated pattern. (B) MRI (T2W, T1W & T1fl 3d axial) revealed multiple (10-15) discrete STIR hyperintense lesions seen involving right upper lip, right buccal space (buccinator muscle), right sublingual space, right submandibular space, right infratemporal fossa (medial pterygoid muscle), right masticator space (masseter muscle), largest measuring 3.5 x 3.2 cms in right submandibular space. (C) USG revealed relatively well defined heteroechoic lesion with noted in right buccinator muscle shows minimal vascularity measuring 1.5 x 2.3 cm.

Figure 7: shows intralesional injection of 1ml of 3 % sodium tetradecyl sulphate.

Figure 8: A and B shows change in color of the lesion with surface ulceration at third day after injection. C and D shows complete resolution of lesion noted at sixth week.

III. Discussion

Vascular lesions in the head and neck include a wide range of pathologies. These constitute a variety of tumors and malformations ranging from simple capillary irregularities to complex structures involving arteries, veins and lymphatics[1]. Vascular anomalies usually affect the endothelium and related tissues that cause an aberrant and hamartomatous vessel growth. Based on the microscopic channel architecture, Virchow RLK classified vascular lesions in 1863[2]. Based on the embryological stage, the Hamburg classification system in 1988 categorized vascular malformation into truncular and extra-truncular lesions. The ISSVA classification for vascular lesions is widely recognized and regarded as the gold standard. Vascular lesions are divided into two major categories: tumours (true proliferative neoplasms) and malformations (defects in morphogenesis) according to the ISSVA (International Society for the Study of Vascular Anomalies) 2018 classification[3].

Vascular tumors

Benign
Infantile Hemangioma
Congenital Hemangioma
Tufted Angioma
Spindle Cell Hemangioma
Pyogenic Granuloma

Locally Aggressive/Borderline
Kaposiform Hemangioendothelioma
Retiform hemangioendothelioma

Malignant
Angiosarcoma
Epithelioid Hemangioma

Vascular malformations
Capillary
Macrocystic Lymphatic
Microcystic Lymphatic
Venous
Venolymphatic
Arterio-venous malformations
Arterio-venous fistula

Haemangiomas are more prevalent in infancy and occurs in 4–12% of all children[3].They are rarely seen at birth and manifest after a few weeks of life. They show a proliferative growth which is quicker than physical growth. Then the growth slows down and undergo involution before the first year of life. Vascular malformations are distinguished by the fact that they never exhibit signs of involution. They occur in 1 -1.5% of births. Vascular malformations can also be divided into low-flow lesions like capillary, venous, and lymphatic malformations and high-flow lesions like arterial, arteriovenous, and arteriovenous fistulas based on their hemodynamic characteristics. Some vascular abnormalities may be linked to disorders like Klippel-Trenaunay syndrome, Osler-Weber-Rendu syndrome, Sturge-Weber syndrome, or blue rubber bleb nevus syndrome[4].

Venous malformations (VMs) are part of the spectrum of vascular malformations characterized by histologically mature venous channels. Venous malformations morphologically and histologically resemble veins.

They are further classified as superficial or deep and as localized, multicentric or diffuse. Based on the depth and degree of ectasia of the lesion the skin or mucosa that covers such malformations varies in color[5]. They are continuous lesions involving several layers that are soft and compressible. They usually align with muscle groups or track along nerves or major vessels. When the lesion is in the head and neck, they can respond to changes in venous flow like Valsalva or occasionally compression of the ipsilateral jugular vein[6].

VMs are due to an inborn error in vascular embryogenesis leading to the formation of abnormal endothelium and as a result, thin-walled, nonfunctional and ectatic veins. Over 90% of VMs are a result of a sporadic mutation, mostly of phenotypes arising from endothelial dysgenesis via the TIE2–PI3K (phosphoinositol-3-kinase)–AKT–mTOR (mammalian target of rapamycin) pathway. Mosaic mutations in this pathway causes VM-predominant overgrowth entities (known as the PIK3CA-related overgrowth spectrum or PROS), which include Klippel-Trenaunay syndrome (KTS) and CLOVES (congenital lipomatous overgrowth, vascular malformations, epidermal nevi, and spine/skeletal anomalies)[6].Venous malformations due to sporadic mutations are usually unifocal while hereditary forms (such as glomuvenous malformation and, rarely, blue rubber bleb nevus/bean syndrome) are multifocal anomalies[6].

Contrast imaging such as angiography and angioxerography demonstrate feeding vessels of the VMs. Peripheral vascularity as well as its velocity within the lesion for both arterial and venous nature of the flow can be appreciated with color-Doppler.

Phleboliths and VMs are rarely observed together. Rarely are they described as isolated calcifications or without a vascular lesion. Alterations in blood flow dynamics within the VM may result in a thrombus and phleboliths. Phleboliths appear as oval structures with concentric radiolucent or radiopaque laminations radiographically[7]. They can range from 1 to 5 mm in size, but can reach upto 1 cm. Incidence of phleboliths in the head and neck region are found to be between 5% and 20%. Phleboliths were present in both the cases reported herewith. Additionally, certain radiographic techniques may be required for the diagnosis and treatment of phleboliths, including computed tomography, magnetic resonance imaging (MRI), angiography, and Color-Doppler imaging[8].The association of hemangiomas with phleboliths has been reported in approximately 5% of all oral surgery cases. Among hemangiomas, spindle cell hemangioma often associated with phleboliths due to abnormal venous vasculature[1].

Sclerotherapy, electrochemical therapy, laser, liquid nitrogen cryotherapy, intralesional radiofrequency, compartmentalization, and surgical excision were among the management strategies used to treat low flow abnormalities.

Sclerotherapy is a first-line treatment option for low flow abnormalities that is efficient. Yamaki and Cabrera used foam sclerotherapy for the first time to treat symptomatic venous abnormalities. There have been reports of sclerosing agents such as bleomycin, pingyangmycin, ethanolamine oleate, 5% sodium morrhuate, picibanil or OK-432, 100% ethanol, doxycycline, Lauromacrogolaethoxysklerol or polidocano, and sodium tetradecyl[9].

Most common approach for sclerotherapy is intralesional route. The benefit of this approach is that it has an inherent predisposition to remove the complete vascular network feeding the lesion, expressing the possibility for regenerative somatic development to replace the vascular abnormality. The average volume for sclerotherapy is 0.5–1.5 ml at a therapeutic interval of 3 weeks. For the management of venous malformations, foam is recommended over fluid sclerotherapy because it has better and longer-lasting contact, resulting in a successful treatment with lower concentrations[10]. Both of our cases resolved completely following single intralesional injection of 3% sodium tetradecyl sulfate.

According to European Guidelines, known sclerosant allergy, pulmonary embolism, DVT, localized or generalised infection, and persistent immobilisation or bed rest were the absolute contraindications to sclerosant therapy. Pregnancy, breast-feeding, high thromboembolic risk, peripheral occlusive disease of the artery, and high allergy susceptibility were relative contraindications[11]. Pain with injection, post-sclerotherapic pigmentation, urtication, superficial thrombophlebitis, cutaneous necrosis, temporary visual disturbances, nerve damage, pulmonary embolism, anaphylaxis, deep vein thrombosis, cardiac arrest, and systemic hypotension were the reported side effects of sclerotherapy.

For many decades, Sodium tetradecyl sulfate has been used as a sclerosing agent with a high level of safety and efficacy. Chemically, this drug is distinct from other commonly used sulfur-containing pharmacologics that normally have a strong allergic potential. It works by inducing a localized inflammatory reaction which leads to obliterative thrombosis of hemangiomatous space that result in subsequent fibrosis of the endothelial spaces and finally regression of the lesion without affecting the bone[12].

IV. Conclusion

Management of vascular lesions are challenging. The ideal way to make clinical decisions is in a multidisciplinary setting and treatment options should include conservative, medicinal, interventional, and surgical methods. The majority of patients are treated with interventional techniques. Despite the fact that there

are many different treatment options, sclerotherapy has been shown to be a successful method of treating them with a small toolkit, cost-effectiveness, and minimal complications. It is also capable of producing excellent aesthetic results in a limited number of sessions and is well-accepted by patients.

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