



## Management of Vascular Malformations of the Maxillofacial Region using Sclerotherapy and Sclerothrombotic Therapy along with Surgery

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### Abstract

**Introduction:** Haemangiomas and Vascular malformations of the maxillofacial region are formidable lesions causing considerable cosmetic deformity and grave functional debilitation. Technically, their surgical excision is difficult and challenging, being fraught with complications like hemorrhage, incomplete excision, facial disfigurement and recurrence. This retrospective longitudinal study has analyzed two techniques that are directed at treating these lesions.

**Aim:** To analyze the safety and long-term efficacy of single session sclerotherapy with Polidocanol for superficial cutaneous / mucosal low flow vascular malformations; and of sclerothrombotic therapy with hot water as a neoadjuvant modality followed by surgical excision, for deeper high flow vascular malformations of the maxillofacial region.

**Material and Methods:** A retrospective longitudinal analysis of a total of ten cases of Vascular Malformations of the maxillofacial region. Of them, five were superficial, low flow lesions which were managed using intralesional instillation of Polidocanol alone without any surgical intervention thereafter; and the remaining five deep, high flow lesions, which were managed first, by intralesional injection of hot water over two sessions, which was then followed by surgical excision of the lesion, within the next 72 hours.

**Results:** Complete clinical resolution of the lesions was achieved in all the cases.

**Conclusion:** Our results support the fact that single session sclerotherapy with Polidocanol may be considered as the primary modality in the treatment for superficial (cutaneous or mucosal) low flow vascular malformations. However, for deeper, high flow lesions, two sessions of intralesional injection of hot water followed by surgical excision within 72 hours of the first injection, may be the treatment of choice.

**Keywords:** Vascular anomalies; Vascular Malformations; Haemangioma; Sclerotherapy; Polidocanol; Hot water; Sclerothrombosis

### Introduction

Vascular lesions or anomalies are a heterogeneous group of blood vessel disorders which, based on their clinical behavior and endothelial cell characteristics, were classified by Mulliken and Glowacki in 1982 into two broad categories, Vascular Tumors, also referred to as Haemangiomas, and Vascular Malformations (VMs) [1,2]. This comprehensive classification was accepted and adopted by the International Society for the Study of Vascular Anomalies (ISSVA) in 1996. In 2013, the ISSVA Classification was reinforced with an additional review on syndrome-based classification [2].

Haemangiomas are true vascular neoplasms, usually seen in infancy and childhood, classically exhibiting increased number of normal or abnormal blood vessels lined with thickened subendothelial basement membrane laminae, and hypercellular / hyperplastic endothelial cells. They are characterized clinically by a rapid neonatal growth until the age of 6 - 8 months (proliferative phase) and then a slow regression by the age of 5 - 10 years (involuting phase), hall marked by diminishing cellularity, interstitial fibrosis and fibro fatty replacement [1].

Vascular malformations (VMs) on the other hand, are congenital lesions resulting from developmental anomalies due to errors in vascular and lymphatic morphogenesis, and comprise of abnormally formed channels that are lined by quiescent endothelium (normal endothelial cell turnover). 31% of VMs are found in the Head and Neck region [1]. These lesions are present at birth, though they often go unnoticed, and grow commensurate with the child with a normal rate of endothelial cell turnover. They increase in size during infancy through adolescence proportionally to the overall growth of the patient. Growth may also be triggered by trauma, infection and hormonal fluctuation as in puberty, pregnancy, use of oral contraceptives, etc. Histologically, there is no proliferative or involution phase and endothelial as well as mast cell activity is normal. Vascular channels are lined by mature endothelial cells which are surrounded by a normal reticulum and a single cell layer basement membrane [2]. These lesions grow as a result of alteration in the flow dynamics within and around the lesion resulting in the recruitment of collateral vessels and dilatation of the involved vessels.

Lymphatic, capillary, venous, and arteriovenous malformations make up the majority of vascular malformations. They may infiltrate skin, mucosa, muscles, joints, organ systems and sometimes even bones. Symptoms are dependent on the anatomic location of the lesion and can have a great impact on the patient's quality of life. Pain and swelling are common symptoms associated with VMs. With craniofacial lesions, cosmetic disfigurement may be more debilitating than functional limitations [3,4] often leading to psychological stress and feelings of embarrassment, anxiety, low self-esteem, and antisocial behavior. Moreover, these lesions are often associated with recurrent bleeding episodes, secondary infections and ulcerations. From these perspectives, the treatment of VMs is more than just a cosmetic concern. Based on flow characteristics which are readily characterized by thrills, bruits and increased warmth; and based on USG findings and CT angiography, Vascular Malformations may be classified into two groups. The first group consists of Low-flow lesions comprising of Capillary malformation, Venous malformation, Lymphatic malformation and Mixed lesions. The second group comprises of High-flow lesions consisting of Arterial malformation, Arteriovenous malformations (AVMs) and Arteriovenous fistulae (AVF).

Conventionally, VMs have been managed surgically with or without embolization. The procedure entails a surgical intervention, is time consuming and associated with technical difficulties and complications, such as inadvertent injury to important head and neck structures, intra operative bleeding and exsanguinations etc.

Though newer modalities of treatment of vascular lesions have been added to the options available, such as cryosurgery, compression, radiation, flash lamp pulsed laser, intralesional injection of fibrosing agents, interferon alpha-2b electrocoagulation and radiofrequency ablation [5-7], laser therapy [8-10] and so on, the older, time tested technique i.e. percutaneous / permucosal sclerotherapy [11] is still the mainstay in the treatment of these difficult lesions making them more amenable to easier surgical excision if warranted thereafter.

Sclerotherapy is a simple, popular, minimally invasive and cost-effective procedure, widely used in the management of hemorrhoids, superficial lower extremity telangiectasias and venous abnormalities such as varicose and reticulate veins [12]. It has also been found to be effective in craniofacial haemangiomas [13]. For smaller lesions, sclerotherapy by itself can lead to a complete cure, while for larger lesions; it may facilitate surgery by reducing lesion size and minimizing intraoperative blood loss [14-16]. It involves the direct injection of a small amount of an irritant solution / sclerosant into the abnormally dilated vessels and applying local compression to maintain the contact of the intravascular solution with the endothelial cells. Over the years, various sclerosants have been employed, such as sodium tetra decylsulphate, 5% phenol, sodium morrhuate, nitrogen mustard, boiling water, sodium psylliate, sodium citrate, invert sugar, absolute alcohol, hypertonic saline or hypertonic dextrose.

Our study analyzed a total of ten cases of Vascular Malformations (VMs). Of them, five were superficial, low flow VMs which were treated by a monotherapy technique, using intralesional Polidocanol sclerotherapy alone; and the remaining five were high flow, deep set VMs, which were treated by a bi-therapy technique, using neoadjuvant sclerothrombotic hot water instillation in two cycles, followed by surgical excision of the lesion

### Aim

To assess the efficacy of single session Polidocanol sclerotherapy for superficial, and hot water sclerothrombosis followed by surgical resection for deep vascular malformations of the maxillofacial region.

### Objectives

- To assess the efficacy of Polidocanol in the management of cutaneous and mucosal VMs in maxillofacial region, in terms of reduction in size and or complete resolution of the lesion, time taken for resolution and incidence of complications and of recurrence.
- To assess the efficacy of hot water sclerothrombosis, followed by surgical excision in the management of deep VMs in maxillofacial region, in terms of ease of injection of hot water, surgical excision, incidence of complications and recurrence.
- To evaluate the efficacy of the two methods with regards to utility of the procedures in different types of VMs.

### Patients and Methods

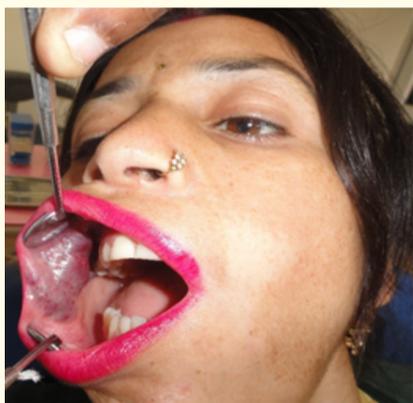
This retrospective longitudinal analytical study included cases treated between 2010 and 2016 and was based on our institutional protocol for the management of such lesions. Being a retrospective analysis of procedures carried out based on established institutional protocol, it was exempted from IRB permission.

The inclusion criteria were vascular malformations involving the soft tissues of the maxillofacial region. These included low flow as well as high flow lesions involving both superficial as well deeper soft tissues.

Other lesions i.e. intraosseous venous malformations and haemangiomas were excluded from this study. A total of ten patients (two male and eight females) were included in the study. The mean age range was from 28 to 52 years. The patient details are summarized as per as Figures.

Being a retrospective analysis, the patients were selected and placed into two Groups, depending upon the diagnosis and subsequent treatment i.e. the flow pattern, extent and depth of the lesions. Patients with superficial mucosal/cutaneous, easily accessible lesions were placed in Group A, while deep set, high flow lesions were placed in Group B. The procedures carried out on these patients were as per the institutional protocol with permission of the institutional review board.

Group A patients were treated by single session sclerotherapy using intra lesional injection of Polidocanol in an outpatient setting (Figure 1A - 1F, Figure 2A - 2D, Figure 3A - 3B). Whenever possible, the lesion was first isolated using black braided silk sutures to prevent sclerosant spill, reducing its diffusion into adjacent unaffected tissues, and also to prolong its contact with the endothelial cells of the abnormal vascular channels (Figure 1B,2B). 2 - 3cc of 3% undiluted Polidocanol depending upon the size of the lesion, was slowly injected into the sides as well as the center of the lesion, using 1 cc syringes, after infiltrating the area with a local anesthetic. Immediately following the injection, the lesion swelled and became tense (Figure 1C). A mild ooze of blood was noted from the perimucosal / percutaneous puncture sites, which soon stopped after application of local pressure over the area (Figure 1D). The silk sutures were left in situ. The patients were put on anti-inflammatory and analgesic medications for five days and cold compress was advised externally over the region.



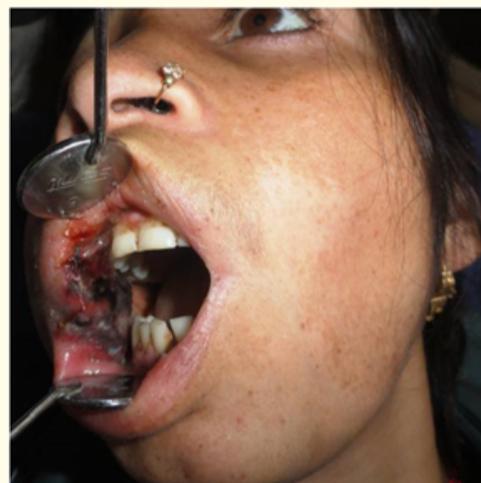
**Figure 1A:** A well demarcated, deep bluish - purple, raised, irregularly shaped, friable and pebbly surfaced patch on the right buccal mucosa, which was continuous with the buccal vestibule and its reflection into the alveolar and gingival mucosa in the right upper posterior quadrant.



**Figure 1B:** A Lesion following its isolation using black braided silk sutures, to prevent sclerosant spill into the adjacent unaffected mucosa.



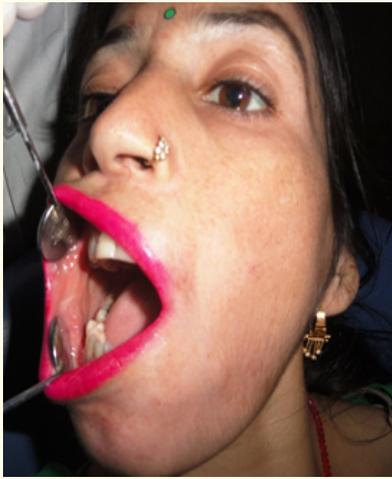
**Figure 1C:** After injection of 3cc of Polidocanol into the sides and center of the lesion.



**Figure 1D:** Necrosis and sloughing of mucosal tissue at the site of sclerosant injection, and considerable local edema and swelling of the cheek seen after 5 days of the single session sclerotherapy.



**Figure 1E:** By the third week, the area of whitish grey slough was restricted to the center of the area and the swelling of the cheek and lips had regressed.



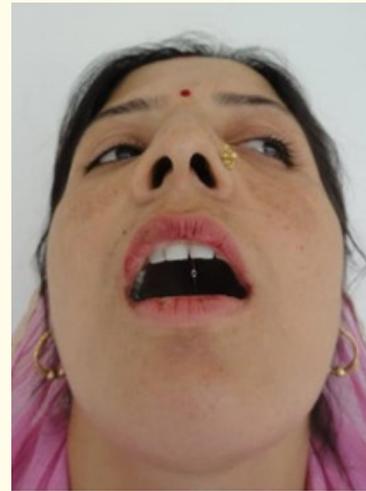
**Figure 1F:** Complete elimination and total resolution of the vascular lesion by the fourth week following the single session sclerotherapy.unaffected mucosa.



**Figure 2C:** Reduction in the swelling at the end of two weeks.



**Figure 2A:** Isolation of the VM of the upper lip followed by injection of 1cc of Polidocanol in the center of the lesion.



**Figure 2D:** Complete resolution and disappearance of the VM by the end of the third week.



**Figure 2B:** One week following sclerotherapy, showing necrosis and sloughing in the region of the lesion and edema and swelling in the surrounding areas.



**Figure 3A:** VM of the nose in a middle aged male patient.



**Figure 3B:** After 4 weeks of single session sclerotherapy. Complete resolution and elimination of the lesion.



**Figure 4A:** Middle aged female patient presented with a diffuse swelling in the right lower border of mandible region.

On the fifth day following sclerosant injection, there was observed considerable local edema and swelling in the region of the lesion (Figure 1D), with necrosis and sloughing of mucosal tissue from it. Pain was not severe and was effectively managed with analgesics. The patients were also prescribed a course of antibiotics. The sutures were removed on the fifth day. By the second week, the swelling, inflammation and sloughing of tissues began to subside and by the third week, the swelling had completely regressed with an area of whitish slough restricted to the center of the area (Figure 1E). In some cases, just a healing crust remained at the end of two weeks (Figure 2C). There was a complete resolution of the vascular lesion by the fourth week following the single session sclerotherapy, with practically no evidence of the lesion (Figure 1F,2D,3B). Successful results with this modality of treatment were obtained even with cutaneous vascular malformations over the face (Figure 3B), with complete resolution of the lesions and no accompanying complications such as infection at the injection site or necrosis or dehiscence of the skin in this region. All patients were followed up for a period of two to five years and none of these cases reported recurrence of the vascular lesions.

Group B patients with more extensive, deeper, high flow lesions, were managed first by sclerothrombosis with hot water (2 cycles) followed by its surgical excision under general anesthesia, within 72 hours of the injection (Figure 4A-4F, Figure 5A-5D). In all our cases, clinically satisfactory results were obtained post operatively. All patients were followed up from one to five years post treatment. One case showed a mild, slow growing, much smaller recurrence, which was confirmed with a CT angiogram, however no intervention was undertaken.



**Figure 4B:** CT angiogram showing a vascular lesion involving Facial and Lingual arteries.



**Figure 4C:** Post hot water injection.



**Figure 4D:** Submandibular approach for excision of the lesion and ligation of the feeder vessels.



**Figure 5A:** Painless, bulbous, soft swelling, present since childhood, showed a recent increase in size over the past six months, following the use of oral contraceptives.



**Figure 4E:** Two years post-op appearance.



**Figure 5B:** 72 hours following intralésional instillation of Hot water, showing a decrease in size of the swelling.



**Figure 4F:** Two years post-op CT Angiogram.

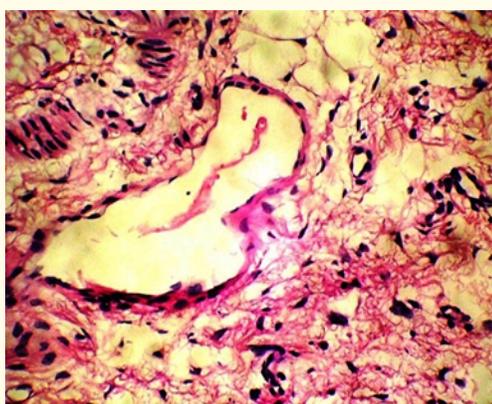


**Figure 5C:** Excision of the mass.



**Figure 5D:** 2 yrs postoperative appearance, showing no recurrence of the lesion.

Histopathological examination of excised specimen revealed numerous irregularly dilated vascular channels lined by a single layer of flattened endothelial cells, dispersed within fibro-collagenous connective tissue stroma, consistent with the diagnosis of Vascular Malformations (Figure 6).



**Figure 6:** Histopathological examination of excised specimen revealed numerous irregularly dilated vascular channels.

## Results

There was complete resolution of the vascular lesions in all patients in Group A as well as Group B except for the minor recurrence in one case in Group B after a follow up period of two to five years.

## Discussion

Management of VMs depends on several factors, such as its flow characteristics, depth, location and anatomical site, extent of involvement and proximity of other structures in the region [18]. These characteristics dictate the choice of the treatment modality to be employed i.e. sclerotherapy, sclerothrombotic therapy, micro embolism, surgery etc.

An intralesional sclerosant injection, causes marked tissue irritation and necrosis, localized inflammatory reaction with swelling, disruption and destruction of the endothelium of the vessel wall, causing platelets and cellular debris to attach to the lining of the vessels, followed by formation of an intravascular coagulum leading to obliterative thrombosis of the blood vessel [19]. Subsequently, fibrosis of the vascular endothelial spaces takes place, eventually leading to disappearance of the treated vessels by histiocytic digestion, with consequent regression of the lesion and tissue contraction [19].

Advantages of sclerotherapy are that it is a simple and inexpensive procedure, entailing no loss of blood, and that it can be carried out on an outpatient basis with no need for hospitalization and with a minimal armamentarium. Disadvantages are occasional anaphylactic reactions, post-operative pain and burning sensation, local swelling, inflammation and tissue necrosis, peripheral nerve injury, hemoglobinuria, thromboembolism, infection and delayed muscle fibrosis [20,21]. Its application should always be carried out in the center and deeper portion of the lesion in order to avoid extensive tissue necrosis of adjacent normal areas [22]. In our case series, necessary precautions were taken during the application of the sclerosing agent in the form of silk sutures on the periphery in order to contain the sclerosant within the area of interest.

In our study, five patients with superficial mucosal/cutaneous low flow vascular malformations, were managed using intralesional Polidocanol monotherapy injections. This sclerosant is a mixture of ethers, macrogols, and fatty alcohols that induces endothelial damage through multiple mechanisms. Its clinical efficacy is equivalent to Sodium Tetradecyl Sulfate (STS) but with less severe adverse effects [23]. Furthermore, due to its anesthetic effect, it does not cause much pain [24].

Boiling water sclerothrombosis acts by releasing heat energy that destroys the anomalous vascular tissues. The heat energy adversely affects the vascular endothelial tissues leading to disruption, necrosis and obliteration of the vessel's lumina, which promotes coagulation of blood eventually leading to collapse and blockage of the channels in the anomalous angiomatous tissues with subsequent destruction of these vessels [25]. Since, water in itself has no sclerosing properties per se, intralesional injection acts primarily due to the transfer of heat injury and therefore produces a much lesser inflammatory response in the surrounding tissues.

The major issue in the management of a high flow VM is the difficulty in restraining the intralesional sclerosant / thrombotic / embolization agent within the lesion. The high blood flow through the lesion tends to flush out the sclerosant into the surrounding areas [26], thus reducing its efficacy and also risking collateral damage to the adjacent structures e.g. skin, eyes etc. In order to avoid these deleterious effects of agents like Sodium tetradecyl-sulphate and Polidocanol in the management of high flow VMs, hot water proves to be more efficacious in terms of (a) a rapid onset of action, which is heat dependent endothelial damage, thus promoting instant intralesional coagulation; (b) minimum collateral damage as the hot water quickly loses heat on injection into the lesion and the damage due to the outflow from the lesion is minimal [27]. However, due to negligible sclerosing property of hot water, its use as a standalone therapy in management of such lesions may be inefficacious, coupled with the fact that the high flow VMs tend to quickly develop collateral circulation on disruption of flow through the lesion.

In our study, the five patients in whom the VMs were larger and deep-seated, with high flow characteristics as demonstrated by Doppler examinations, were managed using hot water injection into the lesion, and followed by surgical excision later. In these cases, sclerotherapy alone would not have sufficed, as there was a need for a more encompassing technique that would not only collapse and obliterate the abnormal blood vessels to avoid undue hemorrhage, but also extirpate the residual fibrosed tissues. Two hot water injections were given over a period of 24 hours and the lesion was surgically removed within 72 hours of initiation of therapy. All these five cases were those of high flow VMs involving the facial and / or lingual artery. There was complete resolution of all the five lesions, except for one which showed a mild recurrence after two years.

## Conclusion

Based on our experience we conclude that sclerotherapy, which is a relatively simple, conservative and inexpensive treatment modality, can be safely and effectively used in superficial low flow lesions of the maxillofacial region. For patients with larger, deep seated and high flow vascular malformations, intralesional sclerotherapy using hot water in two cycles followed by surgical excision within 72 hours of the intralesional therapy can be taken up as a definitive treatment, having proved to be effective in management of such lesions, without the need of more advanced procedures/instrumentation. However, considering the challenging and ominous nature of these lesions, larger studies may be undertaken to corroborate these findings.

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