Arteriovenous Malformation of the Oral Cavity: A Case Report and Review of Literature

ABSTRACT

Arteriovenous malformations (AVMs) are developmental vascular anomalies that occur when the embryonic vascular network fails to differentiate. It is composed of abnormal communications between arteries and veins without a normal intervening capillary bed. Lymphatic, capillary, venous and AV malformations make up a majority of vascular malformations. Facial vascular malformations can cause dental emergencies that result in fatal or life threatening and disfiguring situations. Diagnosis of these lesions is essential and management of maxillofacial AVMs remains challenging. In this article, we report a case of intra-oral AV malformation of 16-year-old male patient that was treated with sclerotherapy using 1% sodium tetradecyl sulfate as sclerosant. The treatment was effective, within a short period of time after injection and no recurrence was observed one year postoperatively.

KEYWORDS arteryovenous malformation, vascular malformation, sclerotherapy, developmental anomaly.

INTRODUCTION

Vascular lesions are most common congenital and neonatal anomalies, that are present at birth and may not clinically manifest until late infancy or childhood. However, patient histories suggest that vascular malformations (VMs) also may be developmental. The prevalence of VMs is 1% at birth. 50% of lesions seen in head and neck region and bone involvement is seen in 35% of cases. Glowacki and Mulliken suggested that vascular lesions can be divided in two groups as hemangioma and vascular malformations, depending on histologic and clinical presentation. Many other terms such as arteriovenous aneurysm, arteriovenous shunt, and arteriovenous fistula have been used to describe AV malformations. They are composed of abnormal communications between arteries and veins without intervening capillary bed. AVMs can be asymptomatic or can cause functional disturbance. Their complications include severe hemorrhage secondary to tooth exfoliation, tooth extraction, blunt injury, trauma, biopsy and surgical intervention; and can result in death. In oral cavity AVMs are present on tongue, palate, buccal mucosa and lips.

AVMs are diagnosed by magnetic resonance imaging, ultrasonography, angiography, computed tomography scans. Depending upon anatomical location, extent and depth of lesion they are treated either by laser therapy, embolization, steroid injection, sclerotherapy, ligation of vessel, surgical removal or a combination of above treatment modalities.

In his paper, we present a case of vascular malformation-slow flow venous type involving the upper lip of 16-year-old male patient. The lesion was diagnosed by Color Doppler Ultrasonography, MRI and MSCT head and neck Angiography. The lesion was treated by injecting sclerosing agent i.e. 1% sodium tetradecyl sulfate (2 ml) into the lesion. A treatment was effective, a maximum of two injections of sclerosing agent resulted in significant reduction in size of the lesion compared to initial size within first 4 months after injection; with no evidence of recurrence postoperatively.

CASE REPORT

A 16-year-old male patient reported to the Department of Oral Medicine and Radiology with chief complaint of long standing swelling on the left side of
upper lip since 14 years. No history of trauma given by the patient. It was not associated with pain, bleeding or pus discharge.

Extra orally, a diffuse swelling was present on the left lateral aspect of upper lip measuring $3 \times 2 \times 2$ cm in size approximately; extending from 1 cm away from midline of upper lip till 2 cm distal to left corner of the mouth resulting in facial asymmetry. Overlying skin was normal (Fig. 1). On palpation it was soft in consistency and non-tender. Intra orally swelling was ill-defined, extending on mucosa in relation to canine and premolar region. Affected mucosa was bluish pink in color, smooth surface and thickened, on palpation lesion was soft, non-tender, compressible and non-reducible (Fig. 2).

**Hard tissue examination was not contributory**

Co-relating history, chronic nature of swelling being present around 12 years, its clinical presentation provisional diagnosis of AV malformation was given. In differential diagnosis lymphangioma, hemangioma was considered.

In order to confirm diagnosis and decide definitive treatment plan patient was subjected to further investigation such as Color Doppler USG of upper lip, magnetic resonance imaging and MSCT head and neck angiography of the maxillofacial region. In routine blood investigation, all values were normal.

USG revealed multiple cystic spaces within the lesion and hypo to anechoic serpigenous structures suggestive of multiple vascular channels within. On color flow imaging lesion showed intense vascularity and the presence of both arterial and venous flow. These features were suggestive of possibility of mixed AV malformation (Fig. 3).

An MRI was done for further evaluation and to assess true extent of lesion. The MRI confirmed well-defined, lobulated soft tissue lesion measuring $3.7 \times 1.8 \times 2.2$ cm in transverse, anteroposterior and craniocaudal dimensions. The mass was hypointense on the $T_1$ and hyperintense on the $T_2$ weighted images with multiple septae within on the $T_2$ weighted images (Fig. 4).

MSCT angiography of the maxillofacial region showed mild enhancement on delayed post-contrast images. No definite feeding vessel was noted. No obvious calcification within the lesion and underlying bone appeared normal (Fig. 5A and B). Thus, reports of above investigations were suggestive of a Vascular Malformation-Slow flow (venous) type.

Once the diagnosis of VM-slow flow (venous) was confirmed, sclerotherapy was opted as choice of treatment.

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**Fig. 1** A diffuse swelling present on the left lateral aspect of upper lip measuring $3 \times 2 \times 2$ cm in size, resulting in facial asymmetry.

**Fig. 2** A diffuse swelling extending on mucosa in relation to canine to premolar region. Affected mucosa bluish pink in color, smooth surface and thickened.

**Fig. 3** Color Doppler USG showing intense vascularity and presence of both arterial and venous flow.
SOTRADECOL 1% [Sodium tetradecyl sulfate 10 mg/ml] BIONICHE Pharma U.S.A. was sclerosant used to induce vascular sclerosis. 2 ml of 1% sodium tetradecyl sulfate was directly injected into the middle of the lesion; slow infiltration using insulin needle. A week later, another injection was given in the same dose into the lesion. A patient complained of mild pain at the site which subsided two weeks later. At 2-month follow up visit, the lesion showed involution in size with localized fibrosis. In the next follow up 2 months later, regression of tissue fibrosis was observed. The lesion had become in size. At 1-year follow up, the patient had not developed any complication and lesion had significantly reduced in size, the results were esthetically satisfying to the patient. The patient was under regular follow up and no recurrence of lesions was observed 1 year postoperatively.

In this case, sclerotherapy was effective definitive treatment, achieving total regression of lesions.

**DISCUSSION**

Vascular malformations comprise of more than 35% vascular lesions. Males and females are equally affected. Angelo et al. stated that most affected age group is adolescents aged 13-18 years. AVM may find on any site of the body, but one third of VM are recorded in head and neck region, both intracranial (i.e. occipital, dural, cerebral) and extracranial (i.e. maxillofacial region)\(^5\). Kohout et al. reported 81 AVMs located in head and neck as cheek (31%), ear (16%), nose (10%), upper lip (7%), mandible (5%), neck (5%), scalp (5%) and maxilla (4%)\(^6\). In oral cavity tongue is the most common site followed by the palate, gingiva and buccal mucosa. This can interfere with mastication, speech, deglutition and often get traumatized resulting in ulceration and secondary infection.

Clinically malformation presents as bluish, compressible pulsatile mass with thrill or bruit. An erythematous blush or port wine stain may be present on overlying skin. A bruit may be discernable but some intraosseous lesions do not produce audible bruit\(^7\). Sometimes numbness of the affected area is present due to neurosensory changes\(^7\). Intra oral lesions may be asymptomatic in many cases. Intraosseous VM near alveolar bone results in mobile teeth, widened periodontal ligament spaces, facial asymmetry\(^8\).

The most severe complication is profuse and uncontrollable bleeding with teeth exfoliation, dental extraction, biopsy, trauma. Spontaneous bleeding is also a hazard.

Mulliken and Glowacki in 1982 classified vascular lesions in two types; as hemangioma and vascular malformation\(^9\). Hemangioma exhibits endothelial proliferation with rapid growth, followed by involution and 90% of cases resolve themselves before 9 years of age. The second type VM is present at birth and remains throughout life affecting 1% of newborn infants. They become more visible over time, even if not seen in early stages and may expand at puberty. They show no signs of involution with age. This type does not exhibit proliferation of endothelium.
VMs are subdivided based on vessel involved; as either Slow flow type (capillary, venous, lymphatic) or High flow type (arterial).

VMs on radiographs seen as lytic lesions with soap bubble or sun-ray appearance. Doppler ultrasound is recommended for examination and can be best supplemented with MRI and Computed tomography. Angiography is the gold standard used to identify VMs, its contributing vessels and flow characteristics.

Management of these lesions is challenging and requires a multidisciplinary approach. Treatment varies depending on the location and the flow characteristic of the lesion. Intraoral lesions can be treated by laser therapy, embolization, cryotherapy, sclerotherapy, steroid injection, surgical removal and combination of above modalities.

Sclerosants, i.e. sodium morrhuate, sodium pylsate, ethanolamine oleate, sodium tetradecyl sulfate have been used to manage low-flow lesions and have a lowest recurrence rate. They induce severe intimal inflammatory reaction leading to thrombosis and shrinkage of vascular anomaly. These agents should be injected with care as they may trigger allergic and anaphylactic reactions; also affect normal structure if it flows in neighboring normal tissue, resulting in tissue necrosis. They are ineffective in high flow lesions as sclerosants are rapidly removed from the malformation and lesion recur due to collateralization.

Embolization followed by surgical excision by surgical excision is the most conventional approach. Embolization is a method of occluding contributing vessels and has been used since 1930 to treat VMs. Several materials such as polyvinyl alcohol, gelfoam, cyanoacrylate, collagen, muscle have been used. These agents are directly placed into contributing vessel through a catheter inserted for angiography. Blockage may result in subsequent enlargement of collateral vessels to the area. Complications reported are hyposthesia of nerves, reflux of embolizing material into the carotid artery.

Surgical ligation of parent vessel is preferred in an emergency situation but it prevents subsequent angiograms and embolization procedure.

Surgical resection of AVM can result in profuse bleeding and incomplete resection may lead to tumor regrowth.

In this case administration of 1% sodium tetradecyl sulfate into the lesion, was an effective treatment. It provided rapid and safe involution of lesion, using non-surgical method and esthetic recovery of patients. No lesion recurrence was observed postoperatively.

CONCLUSION
Vascular malformations in head and neck are complex pathologies that are unlikely to resolve on their own, and appropriate treatment need to be chosen. The clinicians should be aware of their potential life threatening complications and management. Sclerotherapy is a feasible treatment method and can effectively resolve vascular malformation. Early detection of lesions and intervention, with adequate follow up is required for potential recurrence of vascular malformation.

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CONFLICT OF INTEREST
The authors declare that they have no conflicts of interest.

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REFERENCES