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Vascular Malformation Involving the Maxillofacial Region: A Case Report

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

Case Report

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ABSTRACT

Vascular anomalies constitute some of the most difficult diagnostic and therapeutic enigmas in the head and neck region and most of the congenital vascular malformations were often misdiagnosed and left untreated. These lesions are the result of an embryonic abnormality of the vascular system. Vascular lesions in the maxillofacial region are rare. These lesions can cause facial asymmetry, loose teeth, tinnitus, headache, and bruits and can be complicated by life threatening conditions such as haemorrhage and heart failure. This article presents a case of 25-year old female patient reported to the Department of Oral Medicine and Radiology with a chief complaint of swelling on the left side of the face. Based on the clinical findings and radiological investigations such as USG with Color Doppler, MR Angiography and CBCT the case was diagnosed as slow flow venous malformation involving the left maxillofacial region. So, the present case report depicts that apart from the clinical findings advanced radiological techniques like contrast enhanced Magnetic Resonance Imaging and CBCT will acts as a useful tool for the accurate diagnosis of Vascular Anamolies (VA).

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1. INTRODUCTION

"Vascular anomalies are a heterogeneous group of congenital lesions of abnormal vascular development and may occur anywhere on the body" [1]. Vascular malformation is a general term that includes congenital vascular anomalies of only veins, only lymph vessels, both veins and lymph vessels or both arteries and veins. Mullikan and Glowacki introduced "a simple classification in 1982 that was based on clinical, histochemical and cellular criteria to distinguish between the various vascular anomalies which include two distinct entities haemangiomas and vascular malformations" [2]. "Recently, a newer classification has been proposed by the International Society for the Study of Vascular Anomalies" (ISSVA, Revised 2018). "The ISSVA classification is a basic and standard classification of vascular anomalies with international acceptance, so its use infers the standardization of diagnosis and treatment" [3,4]. "Vascular Malformations are present since birth and never proliferate and involute in contrast to haemangioma. They expand continuously and relentlessly throughout life in pace with the growth of the patient. Venous malformations are the most commonly encountered vascular malformations and effect 1 to 4% of the individuals and clinically appear as bluish, soft, compressible lesions typically found on the face, limbs or trunk" [5].

2. CASE REPORT

A 25-year old female patient reported to the Department of Oral Medicine & Radiology, Lenora Institute of Dental Sciences with the chief complaint of swelling on the left side of the face since 15 to 16 years (Fig. 1). On eliciting the history of present illness, patient noticed the swelling irt left side of the face from 15 years. The swelling initially started as the size of a small peanut and gradually increased and attained the present size. There is a history of similar swelling in the same region at the age of 6 years for which she got operated and recurrence was seen 15 years ago, yet unable to elicit the details of the previous surgery with no history of paresthesia, no history of secondary changes but she gives the h/o pain especially while bending down and lying down on left side which was gradual in onset, mild, intermittent, piercing type of pain with increase in the size of the swelling. Patient gives h/o relief from the pain on itself and no history of functional inability.

On general physical examination, patient was conscious, coherent, co-operative & well oriented in the dental chair, moderately built and moderately nourished and extra oral examination revealed a diffuse swelling of size measuring approximately 3×4 cm seen involving the middle & lower one third of the face on left side. The swelling was soft to firm in consistency, non-tender on palpation and it was non compressible, non-reducible and non-pulsatile. On intra oral examination a diffuse swelling with bluish red discoloration measuring approximately 3×1.5 cm was seen involving the left buccal mucosa (Fig. 2) and another similar swelling was seen involving the lower lip (Fig. 3) on the left side measuring approximately 1×1cm and overlying mucosa was stretched with surface bluish red discoloration. The swelling was soft to firm in consistency, non-tender on palpation and it was pulsatile. Based on the clinical findings a provisional diagnosis of vascular malformation involving the left side of the face was given and a differential diagnosis of Hemangioma and Arteriovenous malformation was considered.

For confirmatory diagnosis, the patient was subjected to Ultrasound Color Doppler followed by Magnetic Resonance Angiography (MRA) and Cone Beam Computed Tomographic scan of the mandible to evaluate the bony abnormalities. Ultrasonography revealed well defined hypoechoic area measuring approximately 3×1.5cm in the subcutaneous region of the left side of the cheek (Fig. 4). On doppler, the vascularity shows slow flow and venous pattern while CBCT axial section showed loss of uniformity in the body of the mandible on the left side when compared to the right side and coronal section showed enlargement of the mental foramen on the left side due to the pressure of the neurovascular bundles emerging through the foramen (Fig. 5). 3D skull view revealed scalloping of the body of the mandible at the region of mental foramen on the left side due to the pressure of neurovascular bundles (Fig. 6).

MR Angiography reported well defined lobulated heterogenous intensity lesions are noted largest measuring 6.2×6.1 cm diffusely involving the left masseter muscle, pterygoid space, infra - temporal fossa, pre auricular region and the temporalis muscle (Fig. 7a). Another similar intensity lesion 4.2×2.6 cm is noted in the subcutaneous tissues of the cheek near the angle of the mouth (Fig. 7b). Major neck vessels

are normal in caliber and no enlarged feeding vessels are noted and the lesion was diagnosed as slow flow venous malformation involving the left maxillofacial region (Fig. 8). Patient was advised surgical resection followed by cosmetic surgery as the lesion was extensive and involved deeper structures of maxillofacial complex.

Fig. 1. 25-year old female patient with swelling on the left side of the face

Fig. 2. left buccal mucosa

Fig. 3 Swelling seen in the lower lip

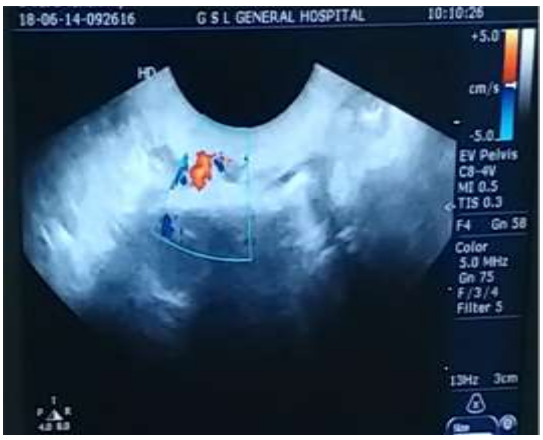


Fig. 4 Doppler test result

Fig. 5 Enlargement of the mental foramen on the left side due to the pressure of the neurovascular bundles emerging through the foramen

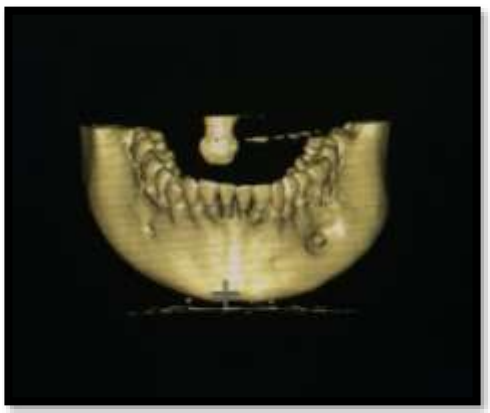
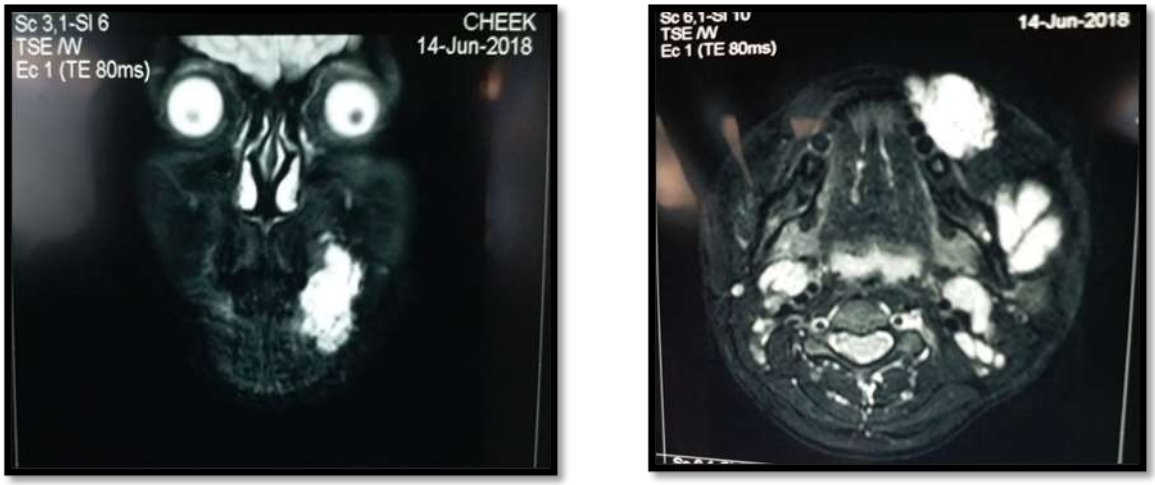


Fig. 6. 3D skull view



a b

Fig. 7 (a, b). Subcutaneous tissues of the cheek near the angle of the mouth

Fig. 8. Flow venous malformation involving the left maxillofacial region

3. DISCUSSION

"Vascular malformations (VMs) are a group of developmental anomalies of the vascular system and may involve any segment of the vascular tree, arteries, capillaries, veins or lymphatics. They frequently involve the head, neck and oral cavity. However, vascular anomalies are rarely seen in the maxilla and mandible" [6].

"Vascular malformations are thought to result from developmental errors during embryogenesis, such as abnormal signaling processes that control apoptosis, maturation, and growth of vascular cells [4]. These errors lead to the persistence of vascular plexus cells with a certain degree of differentiation. There are four major categories of vascular malformations based on their flow characteristics: slow-flow (capillary malformation, venous malformation, lymphatic malformation) and fast-flow (arteriovenous malformation)" [7].

Clinical features: Both the genders are equally affected and always present at birth (though some may not be apparent until a later stage) never involute. Trauma, puberty & pregnancy may make the VMs to be active. VMs involving the soft tissue are usually soft, compressible and enlarge in size when venous pressure is increased. Phleboliths may be present in the lesion. Changes in the adjacent osseous structures are not uncommon & often cause pressure effect or distortion or both [2,8].

Venous malformations: "Venous malformations are part of a spectrum of vascular malformations

commonly found in adults. These are defined as malformations comprised of slow-flowing, abnormal dilated veins, and venous network. Clinically, these lesions appear as a soft, compressible, non -pulsating blue mass, typically present within the cutaneous tissues of the face, trunk, and limbs, although involvement of the viscera and bones has also been described, as seen in the present case. It is noted that two thirds of all vascular malformations are venous predominant" [8].

"Venous malformations are the most common type of vascular malformation, affecting 1% to 4% of individuals and can occur anywhere in the body, but are predominantly located in the head and neck (40% of cases), trunk (20%) and extremities (40%). VM's can occasionally be completely intraosseous and the mandible is the most common bone involved followed by maxilla as seen in the present case although nasal, and frontal lesions have also been reported" [9].

Imaging modalities include USG Color Doppler which is essential, non-invasive tool that is widely used to examine superficial vascular lesions. VMs are compressible with the typical multispacial, multicystic, and/ or partially solid heterogeneous echotexture (98%) and can be hypoechoic (82%), hyperechoic (10%), or isoechoic (8%) with respect to surrounding structures. Color Doppler imaging permits real-time analysis of arterial and venous flow and measurement of flow velocities and presents as monophasic waveforms with low flow in VM. MR Imaging is most valuable and it depicts the anatomic relation between the vascular lesion and adjacent organ nerves, tendons, and muscles. Slow-flow venous malformations have high signal intensity on T2-weighted images, whereas high-flow arteriovenous malformations and fistulas contain a signal void. "Angiography, particularly digital subtraction angiography (DSA), has a specific but limited role in the diagnosis of vascular lesions, but should not be used as a first line investigation. It is, however, useful for mapping out the blood supply of the lesion and in the assessment of the characteristics of flow of arteriovenous malformations" [10].

The choice of treatment depends on the depth, extent, and anatomical location of the lesion and the experience and preference of the clinician. The methods available are lasers, excision, sclerotherapy and combinations. Smaller lesions can be treated successfully by excision or sclerotherapy. Among all the sclerosing agents, absolute ethanol is the most effective one with the lowest recurrence rate but also with the most serious local and systemic side effects. Other agents used include polidocanol or sodium tetradecyl sulfate as foam sclerosants (0.5–3% concentration) and bleomycin. Resection must be complete to prevent recurrence and appropriate imaging is necessary to show the full extent of the lesion, particularly if the lesion is large. "Acquired adult venous malformations are commonly seen as small venous lakes in the lower lip and face and can be treated with laser photocoagulation or excision" [11]. "Soft tissue Er:YAG lasers present a significant advantage in management of vascular malformations since it causes negligible hemorrhage, disinfection of surgical wound and less post-operative complications which are advantageous to patients" [12].

4. CONCLUSION

Although vascular malformations are not so common, identification, diagnosis and reporting of such lesions is of dental significance as these lesions may have potential dental implications such as massive haemorrhage following extractions and minor surgeries as direct effect and indirect pressure effect. Thus, we as oral physicians should have adequate knowledge of the diagnosis and management of these lesions and referral should always be considered if required.

CONSENT

As per international standards or university standards, patient(s) written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

As per international standards or university standards written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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