Case Report

Acquired arteriovenous malformation of lip occurring as an occupational hazard: A case report with review of literature

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Abstract

"Vascular malformations" comprise a group of lesions, formed by an anomalous proliferation of angiovascular or lymphovascular structures. Arteriovenous malformations (AVMs) are high-flow malformation, characterized by direct communication between arteries and veins, lacking normal capillary network. AVMs are usually congenital. Acquired AVMs are reported to occur due to trauma or hormonal changes. Acquired AVMs, though rare in oral cavity, when present are persistent and progressive in nature and can represent a lethal benign disease. They are often associated with extensive blood loss, and an incomplete resection frequently leads to regrowth of the lesion often larger than its original size. We present a rare case of an acquired AVM diagnosed on contrast-enhancing computed tomography angiography and histopathology in a 65-year-old professional trumpet blower.

Keywords: Arteriovenous malformation, occupational hazard, upper lip

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Received: 09.03.2017, **Accepted:** 14.02.2018

INTRODUCTION

Arteriovenous malformations (AVMs) are the lesions with direct communications between endothelial-lined artery/arteries and vein/veins bypassing the capillary bed [Figure 1].[1-4] Head and neck AVMs are reported to occur in 0.1% of the population of which extracranial accounts for only 8.1%.[2]

AVMs may be congenital or acquired.[1,2,5,6] Acquired AVMs are usually posttraumatic with previous history of injury, trauma or surgery with higher incidence of hemorrhage.[1,2] Hemorrhage and disfigurement are common reasons requiring intervention in AVMs involving the head and neck region.[2]

Access this article online

Quick Response Code:

Website: www.jomfp.in

DOI: 10.4103/jomfp.JOMFP_4_16

The purpose of this case report is to describe the challenges in diagnosis of AVM and need for a prompt treatment of such cases.

CASE REPORT

A 65-year-old male reported with chief complaint of bleeding from a painless swelling of upper lip for 1 month. The swelling began as a peanut size, 5–6 years ago and gradually increased to the present size [Figure 2]. Dental, medical or family history was noncontributory. General physical examination showed all vital signs normal. The patient was a professional trumpet blower since 35 years.

On extraoral examination, upper lip appeared to be everted due to an oval swelling of size approximately 3 cm × 3 cm.

How to cite this article: Pandhare MN, Jyoti DB, Mandale MS, Suresh RB. Acquired arteriovenous malformation of lip occurring as an occupational hazard: A case report with review of literature. J Oral Maxillofac Pathol 2018;22:287.

Swelling was on the left side adjacent to midline, extending from vermillion border onto the labial mucosa. It showed central ulceration and bleeding points [Figure 2]. On palpation, swelling was soft to firm in consistency, with well-defined borders. It was immobile, nonreducible but slightly compressible. Swelling was pulsatile in nature with raised surface temperature. It bled on slight provocation.

Based on the history and clinical findings, a provisional diagnosis of vascular malformation (VM) was made, and in differential diagnosis, pyogenic granuloma and vascular tumors were considered.

The patient was investigated, and the following reports were sought:

- Complete hemogram revealed all the values within normal limits
- Electrocardiogram was normal
- Fine needle aspiration cytology revealed a hemorrhagic aspirate
- Doppler ultrasonography examination revealed a well-defined lobulated enlarged solid lesion of size 2 cm × 1.8 cm in upper lip with marked arterial flow within and dilation of feeding artery was noted
- Contrast-enhancing computed tomography (CT) angiography of neck showed:
    - Well-defined unencapsulated isodense lesion of approximate size 2.4 cm × 1.8 cm × 1.2 cm in the subcutaneous fibrofatty plane of upper lip in paramedian location slightly toward left side
    - The lesion showed brilliant enhancement on the arterial phase and early complete washout on the venous phase.

  Final impression:
    - Benign soft-tissue lesion or possibly AVM
    - The lesion derived its vascular supply through angular branch of left facial and nasopalatine branches of bilateral facial arteries [Figure 3].

Taking all precautions, excision of lesion was performed under general anesthesia. A single grayish white globular mass (2.8 cm × 2.5 cm × 1.5 cm) was received. Outer mucosal surface was smooth while inner surface was reddish with multiple tiny vessels and areas of hemorrhage.

Hematoxylin and eosin stained sections showed parakeratinized stratified squamous epithelium with varying degree of proliferation. Underlying connective tissue was loosely arranged with numerous thick- and thin-walled blood vessels arranged in a lobular pattern. Large vessels were surrounded by numerous proliferating small vessels. Proliferation of blood vessels was also seen in-between muscles. Mild degree of diffuse chronic inflammatory cell infiltrate was also present [Figure 4]. Based on clinicopathologic features, the lesion was diagnosed as AVM of the upper lip.

The patient when reviewed after a month, surgical wound had healed completely. He was warned not to blow trumpet

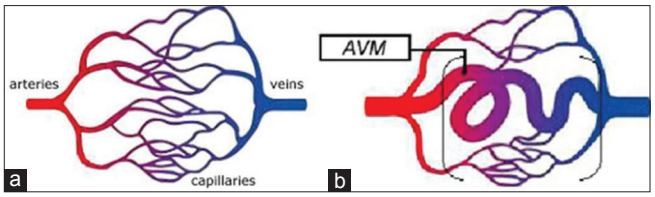


Figure 1: Image illustrating (a) normal connection between arteries and veins through capillary beds (b) while in arteriovenous malformation, direct communication between arteries and veins without capillary bed

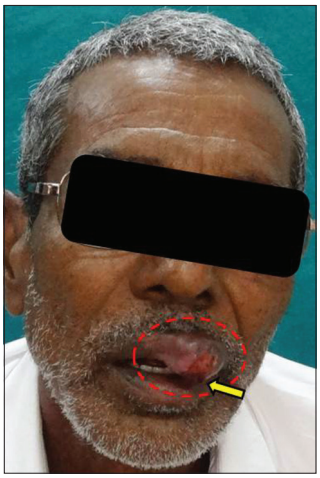


Figure 2: A clinical image showing everted upper lip with oval swelling of size approximately 3 cm × 3 cm with central ulceration and bleeding points

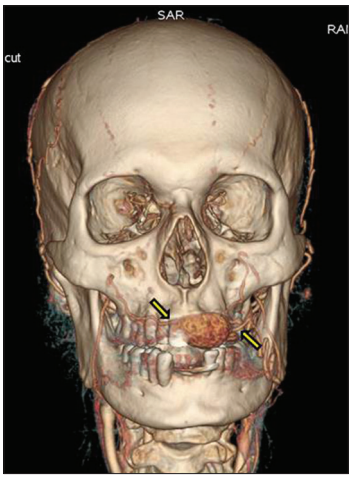


Figure 3: Three-dimensional reconstructive images: The lesion derives its vascular supply through branches derived from nasopalatine branches of bilateral facial arteries and angular branch of left facial artery

for at least 6 months. The patient was asymptomatic when reviewed after a year and had resumed his occupation of trumpet blowing.

DISCUSSION

Vascular anomalies are heterogeneous group of vessel disorders that may affect any segment of the vascular tree; arteries, capillaries, veins, or lymphatics or a combination of these.[1,2] Each anomaly is characterized by specific morphology, pathophysiology, clinical behavior and management approach.[1] They constitute some of the most difficult diagnostic and therapeutic enigmas in the head and neck region.[5] Hence, it is of utmost importance to establish a classification system[5] which will consider both diagnostic and therapeutic issues.

Early classification given by Virchow and Wagner was according to the pathologic appearance of vessel. Vascular growths were subclassified into angiomas and lymphangiomas. The biologic behavior and natural history of the vascular lesions were not considered.

In 1982, Mulliken and Glowacki introduced a biological classification based on their clinical appearance, histopathologic features and biologic behavior into two major categories: tumors and malformations [Figure 5].[1]

This classification was later redefined by Mulliken and Young and adopted by the International Society for the Study of Vascular Anomalies (ISSVA) in 1996.[7] Now, it has recently been revised at 20th ISSVA Workshop in Melbourne Australia, April 2014.[1,8]

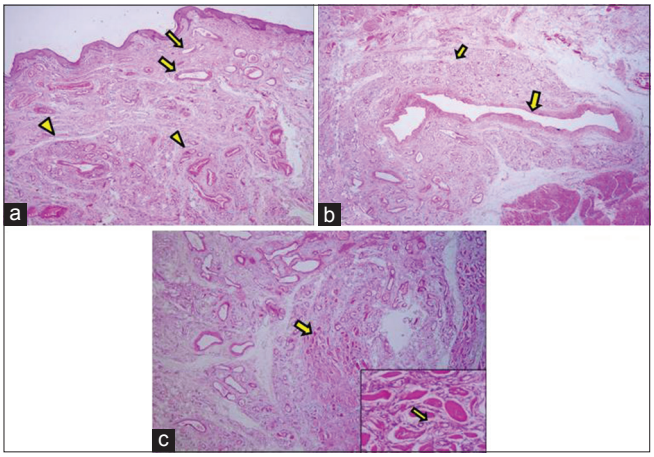


Figure 4: Hematoxylin and eosin stained sections. (a) Mixture of thick- and thin-walled blood vessels (arrow) arranged in lobular pattern (arrowhead) (H and E stain, ×40), (b) large blood vessels surrounded by proliferated small blood vessels; this large blood vessel (arrow) appeared to be a feeder artery (H and E stain, ×100), (c) intramuscular proliferation of blood vessels (arrow) (H and E stain, ×100). Inset showing the same in higher magnification (H and E stain, ×400)

Vascular tumors affecting the head and neck region are common, particularly involving the jaws. On the other hand, VMs are rare[2] but when present are persistent and progressive in nature and can represent a fatal benign disease.[1] Hemangiomas are the most common vascular tumors[1] and should be differentiated from VMs as therapy is different for each.[9]

Forbes *et al.* distinguished VMs as slow/low flow and high/fast flow lesions based on hemodynamic and contrast angiographic appearance.[1-3,5,9] Lymphatic and venous malformations belong to the first group, while AVMs to second group.[1,9]

AVMs are the most aggressive form of VMs, which can lead to significant deformity and functional impairment.[10]

Etiopathogenesis of arteriovenous malformation

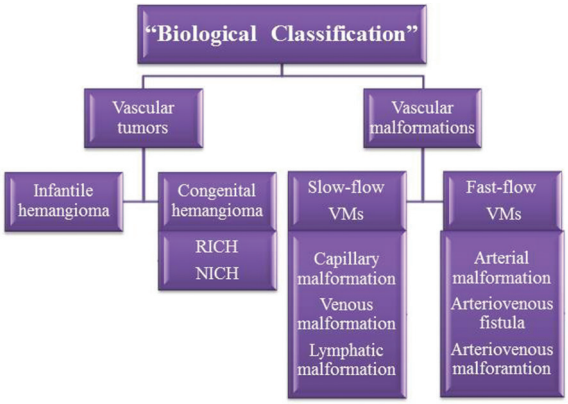
Little is known about the origin and pathogenesis of AVMs.

Congenital arteriovenous malformation

This occurs as a result of lack of differentiation of arteries, veins and capillaries during vascular development.[1,2,5] Most are present at birth but come to clinical attention during second or third decade of life.[9] Defects in transforming growth factor-β signaling and a genetic two-hit hypothesis are the prevailing theories to the pathogenesis.[1,10]

Familial arteriovenous malformation

Although rare,[4,5,9] most of the cases are sporadic. There are few inherited syndromes whose molecular genetics has been recently elucidated. A mutation in gene RASA1, expressing p120-Ras GAP, on chromosome 5q, has been identified in families with congenital malformations associated with AVMs.[11]



(RICH-Rapidly involuting congenital hemangioma; NICH-Non-rapidly involuting congenital hemangioma)

Figure 5: A biological classification by Mulliken and Glowacki

Acquired arteriovenous malformation

Trauma, ischemic event secondary to thrombosis, ectasia and hormonal changes (puberty and pregnancy[5]) can induce aberrant proliferation of the arteries-veins and trigger the manifestation of its troublesome symptoms.[1] They may also occur following surgery, invasive diagnostic procedures or arterial aneurysm.[4] When secondary to trauma, the lesion is usually supplied by a single vessel, as compared to the multiple vessels often seen with congenital etiology.[2]

AVM has been reported to develop in areas (buttocks) subjected to prolonged pressure in bedridden patients. Ischemic tissue damage due to prolonged pressure may lead to necrosis in the deep subcutis. Subsequently, in the process of tissue repair and angiogenesis, arteriovenous anastomoses may form, leading to bizarre lesion.[6]

In present case, ischemic tissue damage due to prolonged pressure by trumpet probably leads to development of arteriovenous anastomoses.

Clinical features

AVMs are the most common type of clinically significant VM with predisposition in males twice frequently than females. The lesion is often recognized clinically between the ages of 10–30 years.[12] They commonly involve the limbs but can occur in any organ of body.[2] In the oral cavity, these can present at any site, but most commonly reported on anterior two-third of the tongue, palate, gingiva and buccal mucosa.[1]

Patients usually present with a localized swelling and bleeding tendency. These lesions present as a pulsatile mass, some demonstrating a palpable thrill and occasionally some of these will be able to be auscultated for a bruit. The patient is often able to detect a "whirring sound" or will claim to hear their heartbeat within the lesion. Occasionally, local hyperthermia and functional impairment due to arterial steal and ischemia may be observed. Shunting of blood diminishes nutritive flow, which may result in skin necrosis, ulceration and bleeding.[1-3,11,13]

Review of the available reported cases of acquired AVM in maxillofacial region summarized in Table 1 showed predominance in the tongue (27.27%), and involvement of upper lip (9.09%) has been documented. The other sites included face, lower lip, premaxilla, maxilla and nasolabial region. The age distribution was ranging from 13 to 43 years (mean 29.62 years). Both genders were equally affected (M: F = 1:1). Typically acquired AVM presented as asymptomatic, soft-firm pulsatile swelling.

Intraosseous VMs near the alveolar bone presented with pericoronal bleeding, mobile teeth and sometimes occlusal anomalies.[1]

AVMs may progress through four different stages and can be scored by severity using the ISSVA accepted clinical staging system (Schobinger 1990). Lesions progress from Stage I (quiescence) to Stage II (expansion) with increasing pulse and thrill. Advancement to next stage is commonly induced by trauma, puberty and pregnancy. In addition, some forms of treatment including ligation of arterial feeders, incomplete excision and laser treatment can trigger progression of quiescent AVMs. Stage III is characterized by local destruction associated with pain, ischemia and necrosis. If left untreated, AVMs can progress to Stage IV (decompensation) with progressive vascular engorgement, venous hypertension and rarely cardiac decompensation due to high output state.[2,10,11]

Histopathological features

Histologically, AVM is a nonencapsulated aggregation of blood vessels in the submucosal tissues. This submucosal vascular complex contains mixture of abnormal thick- and thin-walled arteries and veins which has been seen in the present case. Mixture of blood vessels in connective tissue is suggestive of shunt between arteries/arterioles and veins/venules. The vessels are lined by flat endothelial cells lining. Internal elastic lamina of arteries may be reduplicated, interrupted and distorted. The muscularis mucosa has a significant variation in thickness. In close proximity, feeder arteries and veins may be seen.[4,13,14]

Investigations

The diagnosis of AVMs can usually be made correctly on the basis of clinical history and examination alone.[2] The appropriate combination of noninvasive to minimally invasive tests should follow in order to confirm or exclude the clinical impression.[1] A diagnostic aid for the evaluation of an AVM is a positive fine needle aspiration although this may not distinguish between high- and low-flow lesions. This technique is safe, efficient and reduces the possibility of fatal or profuse bleeding reported with biopsies.[5]

There have been recent advances in the evaluation of AVMs with imaging modalities.[2] The first investigation usually performed is ultrasound and Doppler examination which allows immediate differentiation between a low- and high-flow lesion.[2] Although CT scan can provide excellent pictures of the malformations and delineate the extent of involvement, it may not always differentiate AVM from other VMs.[2] Magnetic resonance imaging has become the investigation of choice since it depicts the extent of these

Table 1: Reported cases of acquired arteriovenous malformation in maxillofacial region

First author, year	Age/sex	Site and duration	History (cause)	Clinical features	Pathogenesis	Investigations - USG, CT scan, CECT scan, MRI, angiography, color Doppler	Treatment and recurrence
Martines, 2009	32/female	Right side and base of tongue Few weeks	23 weeks 2 days of gestation	Swelling, elevation of the lateral floor Mucosal telangiectasia No pulsations	Due to the effect of progesterone causing smooth muscle relaxation, increased venous distensibility, AVM dilatation and rupture	An AVM with its feeding arteries (lingual artery, internal maxillary artery)	Spontaneous postpartum regression No recurrence
Atmaja Nair, 2014	22/female	Left side of the tongue	35 weeks 3 days of gestation Maxillofacial surgery 2 years back for cosmetic reasons Surgery done for hemangioma of tongue 1 month back	Enlarged tongue with a reddish lesion 2×2 cm on the left side, with no bleeding		Enlarged tongue and multiple enlarged vessels (external carotid arteries) within and around the left side of tongue	Advised a review after 6 weeks
Manjunath, 2014	25/male	Left side of face and base of tongue	Father having similar asymptomatic swelling on back	Multiple nontender soft swellings Lacked prominent pulsation	Defects in TGF-β signaling and a genetic two-hit hypothesis	Hyperintense images with no flow voids	Combined approach of embolization and surgery
Smith, 2008	23/female	Left side of maxilla 3 months after surgery	Treatment of cleft lip and palate with an advancement Le Fort I maxillary osteotomy and sagittal split mandibular set back osteotomy 3 months back	Pulsatile tinnitus	Arise as a result simultaneous laceration of an artery and vein. The resulting hematoma is then organized by endothelial proliferation and endothelial lined channels between the artery and vein. Blood is then shunted directly from the high-flow arterial system to the low-pressure venous system	A large, convoluted niche of abnormal vessels within the left infratemporal fossa involving the left pterygoid muscle	Several small platinum coils placed to occlude the vessel No recurrence
Hassard, 1985	2 cases	Upper lip	Trauma	Swelling	After trauma/injury subsequently, in the process of tissue repair and angiogenesis, arteriovenous anastomoses may form, leading to bizarre lesions	-	Complete surgical excision, following embolization
Darlow, 1988	43/male	Paranasal sinus	Blunt trauma	-		-	-
Vadisha Bhat, 2013	31/female	Right premaxillary soft tissue Since 6 months	Dental treatment on the right upper premolar 1 month before	Soft, compressible, nontender, pulsatile swelling 3×2 cm		Well-defined rounded lesion in right premaxillary and nasolabial region. Multiple enlarged flow voids seen within it	Selective embolization with gel foam, followed by complete surgical excision with ligation of the vessels No recurrence
Kao ST, 2014	-	Face	Facial laceration	-			Selective embolization or surgical exploration
Sandhya Ramachandra, 2014	23/male	Left naso-orbital region Since 1 year	Road traffic accident, 3 years ago	Painless swelling, Increased temperature pulsatile Bruit		Multiple vascular channels in the swelling with high flow and turbulence with enlarged draining vein (superior ophthalmic)	Lost to follow-up
Han HH, 2015	13/male	Lower lip Since 5 years	Lower lip laceration 5 years ago	Swelling		Expanded mass fed by the mandibular branch of both facial arteries and drained to the posterior facial vein	Complete surgical excision, following embolization No recurrence

USG: Ultrasonography, CT: Computed tomography, CECT: Contrast-enhancing computed tomography, MRI: Magnetic resonance imaging, AVM: Arteriovenous malformations, TGF: Transforming growth factor

lesions and also allows differentiation between various VMs. Angiography is useful in poorly defined lesions and for embolization before surgery. It demonstrates the flow characteristics, feeding vessels and dangerous anastomoses.[1]

Radiographically, intraosseous AVMs appear as varying-sized osteolytic lesions with a "soap bubble" or "sun ray" appearance, often with indistinct borders.[3]

In the present case, the patient was diagnosed clinically as having a VM and further investigations including histopathology and imaging helped us to characterize the type of malformation as a high-flow AVM.

Differential diagnosis

The differential diagnosis of AVM includes vascular neoplasms, other VMs and in rare cases, other neoplasms.[11] AVM should not be confused with hemangiomas which are characterized by an initial rapid growth of endothelial cells followed by spontaneous involution. Spontaneous involution of VM has never been reported.[9] Hemangiomas are present at the time of birth and involute themselves and if persist in adulthood are known as residual hemangioma.

Pyogenic granuloma and hematoma can be considered in differential diagnosis of AVM. They are nonpulsatile and histopathologically do not show mixture of arterioles and venules.

Therefore, when evaluating a lesion with a clinical manifestation of pulsatile swelling and spontaneous bleeding, the first step would be to rule out a vascular anomaly before any intervention. Secondly, it is important to determine whether the lesion is a tumor or malformation. In most cases, differentiation between the two can be achieved by obtaining a proper history and physically examining the patient,[3] while histopathology further provides confirmation.

Thus, it is of great therapeutic importance to correctly classify and differentiate AVM from other vascular lesions.[9]

Treatment

AVMs present a therapeutic challenge because of their hemodynamic characteristics and their growth modality.[1] They have to be treated according to their histopathology, location and hemodynamic features as shown radiographically with angiography.[9]

AVMs when asymptomatic, they do not require treatment, but if it is associated with pain, ulceration, bleeding or heart problem, therapy is necessary. Multimodal treatment including preoperative sclerosing agents or embolization followed by complete surgical resection is still the most conventional modern approach to treat these lesions.[1,2]

Surgical resection is often associated with extensive blood loss, and an incomplete resection frequently leads to regrowth of the tumor to sizes that are often larger than its original size. Proximal ligation of the parent vessel is avoided as it is ineffective and may aggravate the problem making future endovascular therapy difficult.[1]

Based on relative indications, our patient was planned for preoperative embolization followed by complete surgical excision. Since then, the patient is on follow-up with no subsequent consequences.

CONCLUSION

Acquired AVMs are quite rare in the head and neck region. They may present as a dental emergency due to their potential for uncontrollable bleeding when performing dental procedures such as tooth extraction, biopsy or during the natural exfoliation of a primary tooth. They may result in alarming hemodynamic manifestations such as venous engorgement, distal ischemia and high-output cardiac failure**.** Hence, early diagnosis and prompt treatment of such cases is must. A good clinical history with imaging usually delineates the lesion well and gives a chance to make a careful decision of intervention. The surgical management is complex and requires elaborate planning and multidisciplinary approach. This case report provides an overview of posttraumatic/acquired AVMs in the head and neck.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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