A rare case of lingual artery anastomosis: A case report and literature review

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Abstract
Arteriovenous malformation (AVM) is defined as a convolute of abnormally connected arteries and veins, where the capillary bed is missing. Intracranial cerebral AVMs are 20 times more common than extracerebral. Focal lingual artery AVMs, especially at carotid bifurcation are rarely reported in the literature. Magnetic resonance imaging, angiography, digital subtraction angiography are the current modalities of diagnosis. Surgical resection can be an effective treatment for vascular malformation; however, the potential for large volume blood loss poses a challenge. In addition, the inability to identify the surgical margins of the nidus can result in recurrence. Present case was diagnosed based on radiological imaging as lingual AVM at the bifurcation of carotid artery. The paper discusses the case of a AVM of the tongue in a 23-year-old female patient at the level of carotid bifurcation. The paper further discusses the various imaging modalities used to diagnose the condition and review the latest literature on the current management concepts. Utmost care needs to be exercised when AVMs are suspected. Radiological investigations are must to know the location and extent of the lesion before attempting surgical intervention like biopsy or excision, which poses life-threatening complications like massive hemorrhage and damage to vital structures.

Keywords
Arteriovenous malformation, carotid bifurcation arteriovenous malformations, tongue

Introduction
Arteriovenous malformation (AVM) of the tongue is an extremely rare lesion of the craniofacial vascular anomaly. Despite the high prevalence of head and neck AVMs, the lingual artery AVMs is very rare and meagerly reported in the literature.[1] Radiological imaging and surgical management of lingual AVMs, especially giant lesions are extremely challenging because topographically these may be very concealing and may lead to impairment of lingual functions.[1,2] Early clinical diagnosis, assessment of anatomical extension of AVM is of paramount importance in the management of this condition and further preventing the complication.

Case Report
A 23-year-old female patient reported with a chief complaint of bleeding from tongue since 1-week. History revealed that the lesion was present on the tongue since 3 years which was small in size and gradually increased in size to attain present size with recurrent bleeding on provocation. Medical and family history was noncontributory.

On intraoral examination, a diffuse purplish red swelling involving the anterior 1/3rd of tongue bilaterally measuring around 5 cm x 4 cm in size superoinferiorly extending from dorsum of tongue to the ventral surface [Figure 1], anteroposteriorly from 1 cm posterior to the tip of tongue to the anterior border of posterior 1/3rd of tongue, which was irregular in shape with fading border. Tongue was coated with food debris. On palpation, the swelling was non-tender; soft to firm in consistency. Diascopy test was positive.

Based on history and clinical examination a provisional diagnosis of hemangioma was made and other differential diagnosis like hematoma or AVM was considered. The possibility of Kaposi’s sarcoma was considered, but since the patient did not show any signs or symptoms of immunodeficiency, this was kept as the last differential diagnosis.

The patient was advised routine blood examination like bleeding time, clotting time, and random blood sugar, which were within the normal limit. The patient was further tested for...
HIV, hepatitis B, C, D, which all proved negative. The patient was then sent for further specialized investigations such as magnetic resonance image (MRI) and angiography.

Axial and sagittal T2-weighted MRI images at the level of tongue showed multiple serpiginous voids most likely vessels [Figure 2]. Axial fat sag images showed hypointense serpiginous lesions. T1 axial images of the tongue showed hypointense lesions [Figures 3 and 4]. Axial post contrast enhanced T1-weighted image demonstrating large vascular channels within the substance of the tongue [Figure 5].

Arteriography revealed tangles of tiny vessels on bilateral sides of tongue suggestive of nidus with dilated arterial feeders from bilateral external carotid artery amounting to high flow lesion. Venography showed draining veins to bilateral internal jugular veins [Figure 6]. The rest of the soft tissue and bones were normal.

On the basis of history, clinical examination, and radiological investigations the patient was diagnosed to have lingual artery AVM at the level of carotid artery.

**Management**

The patient was referred to an interventional radiologist for embolization. The procedure was carried out successfully and the patient was recalled for review. Patient was lost in follow-up.

**Discussion**

Vascular malformations are common lesions accounting for approximately 7% of all benign tumors. Although the head and neck constitute <14% of the total surface area of the body, approximately 50% of all vascular malformations occur in these regions.\(^3,4\)

AVMs are fast-flow vascular lesions. They are composed of dysmorphic arterial and venous connections without an intervening capillary bed.\(^5,6\) They develop early in fetal life due to defective retiform plexus formation. Thus, they are seen in early fetal development itself. Clinical condition progress as a
AV malformation

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Figure 5: Axial post-contrast enhanced T1-weighted image demonstrating large vascular channels within the substance of the tongue

result of shunting of blood between high pressure arteries low pressure veins.[5-7]

A number of cases of AVMs have been described by different authors. 81 cases of head and neck AVMs were reported by Kohout et al. The most of AVMs are found in cheek (31%), ear (16%), and nose (10%). The forehead (10%), the upper lip (7%), the neck (5%), the mandible (5%), the maxilla (4%), and the scalp (4%) account for the remaining AVMs.[4,6]

According to the Hamburg classification, congenital vascular malformations are classified into arterial, venous, arteriovenous, lymphatic malformations, and complex combined vascular malformations.[4,8] Subdivision of vascular malformations is based on the vessels involved, (i.e. capillary, venous, lymphatic, arterial). This classification simplifies older and confusing terminology. Thus, a port-wine nevus is now considered a capillary malformation. The term “lymphangioma” is actually a lymphatic malformation and a “cavernous hemangioma” that fails to involute is considered a venous malformation.[4,8] A more proper term for “arteriovenous fistula” is an arterial malformation with fistula. Although the anomaly can consist of a single type of vessel, combinations also occur.

Most cases of AVMs are sporadic. However, there are a few inherited syndromes seen along with AVMs. A mutation in gene RASA 1, on chromosome 5q, expressing p120-Ras GAP, has been identified in families showing AVMs, and some associated congenital malformations. In hereditary hemorrhagic telangiectasia, AVMs are transmitted in an autosomal dominant fashion.[4,4]

Transforming growth factor plays a vital role in the induction of apoptotic endothelial cell death. A decrease in the apoptotic process may lead to the deregulation of vascular growth resulting in AVMs subsequently. They are more prevalent in the central nervous system, because neurons rarely undergo apoptosis.[1,4,9]

In early embryogenesis, distinct ligands and receptors are present on arterial and venous endothelial cells. The reciprocal signaling between these arteries and veins is very important in the formation of capillary beds. Hence, a defect in ligands or receptors can lead to the formation of AVMs.[4]


Frequency of occurrence of AVMs is similar in both males and females. About 40-60% of lesions are visible at birth. 30% of AVMs become clinically apparent during childhood. The 1990 ISSVA-accepted Schobinger clinical staging is used to assess the severity of AVMs.[4,4]

Stage I: Lesion is asymptomatic with a port wine stain on involuting hemangioma.

Stage II: The progressive phase begins during adolescence. This stage represents expansion and invasion of deep structures. Progressive dilatation, thinning, and fibrosis of arteries and veins is seen histologically.

Stage III: Mimics Stage II with spontaneous necrosis, chronic ulceration, hemorrhage, and pain with deep destruction of tissue. Our case with hemorrhagic manifestation fits into this stage.

Stage IV: Is characterized by cardiac decompensation. High output cardiac failure develops. The flow characteristics (low-flow vs. high-flow) vascular malformations indicate their vascular nature with high-flow usually characteristic of arterial vessels. High-flow lesions tend to cause more destructive skeletal changes in the head and neck area than low-flow lesions, perhaps as a result of the hemodynamic characteristics of the high-flow malformation. The flow rate can be used as an indicator of the proper therapy.[6,10]

Diagnosis of AVMs is by clinical findings and radiological features. Plain radiography and computed tomography scans may not help as a diagnostic tool in these malformations. Ultrasonography and color Doppler evaluation helps in the initial assessment.[6,10] Nowadays, MRI is the investigation of choice. It shows the extent of invasion of these lesions. It also provides
multiplanar images and differentiates between high and low flow lesions. Presence of flow voids on MRI helps to confirm fast flow vessels. An arteriogram may be done prior to embolization to know the major feeder vessel. It also shows the flow characteristics and dangerous anastomoses. The present case was diagnosed as a high flow AVM based on MRI and angiogram.

Embolization is one of the treatment options and is aimed at the nidus or central feeding vessel of the malformation; this method as a sole treatment for the AVM can result in recruitment of collateral vessels, making further embolization or surgical resection impossible. Embolization represents a promising alternative treatment in cases of surgical inaccessibility or when surgery would cause an unacceptable deformity. Surgery with embolization can avert large-volume blood loss; however, this method is not always successful. Stereotactic radiosurgery (SRS) is a common treatment modality for brain AVMs. For giant AVM of the tongue can be treated by cyber knife image-based SRS.

Conclusion

Early clinical diagnosis and radiological investigations pose a real challenge especially when less intimidating symptoms. Convincing a patient for intervention or surgical management would be a herculean task. However, early management of giant lesions is vital to prevent complications and reduce morbidity and mortality. Hereby we urge the professionals to invent more safer and effective modalities of management.

References
