Arteriovenous Malformation of Tongue: A Case Report and Review of Literature

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ABSTRACT

The intracranial arterial or AVMs most commonly present in the head and neck region are usually overlooked when present at birth owing to their innocent appearance. Associated important clinical signs are warmth, a palpable thrill and a bruit. They may also be associated with complications, like ischemic changes, indolent ulceration, pain and bleeding. Here, we present a case of AVM of tongue in a 27-year-old male patient which was diagnosed based upon his oral lesions.

Keywords: Arteriovenous malformation, Vascular anomalies.

INTRODUCTION

Arteriovenous malformation (AVM) occurs due to the failure of complete involution of the fetal capillary bed resulting in the development of abnormal connections between arteries and veins (arteriovenous shunting). This results in progressive vascular engorgement, venous hypertension, expansion and destruction of tissue, producing obvious esthetic problems and, rarely, cardiac decompensation due to high output state.1,2 The lesions may be located anywhere in the body, but most commonly affect intracranial cavity. AVM outside cranium is rare and the auricle is most common extracranial head and neck location.3 We hereby report a case of AVM of the tongue in a young male.

CASE REPORT

A 27-year-old male patient (Figs 1 and 2) visited Department of Oral Medicine and Radiology, Syamala Reddy Dental College, Bengaluru, India, with a chief complaint of swelling on the tongue. The patient's history revealed that the swelling was of pinhead sized when it was first noticed around 10 to 12 years back and it has gradually increased to attain the present size. It was associated with the stiffening of the tongue on awakening and subsided during the rest of the day. There was no history of pain, difficulty in phonation, deglutition and altered taste sensation, and there was no history of trauma, bleeding or ulceration of the tongue (Fig. 3).
compressible, non-fluctuant, no fluid thrill was present and it was not fixed to the underlying structures (Fig. 4). Based on the history and clinical findings, a clinical diagnosis of vascular malformation was made and a differential diagnosis of lymphangioma, lingual varix and mucocele were considered. Patient was subjected to complete blood investigations which were revealed to be normal and Doppler ultrasonography, the findings of which revealed a well-defined multicystic lesion of size 1.2 x 1.0 cm on the anterior aspect of the tongue with both the venous and arterial flow noted within; predominantly of arterial type, and a dilated feeding artery noted posterior to the lesion (Figs 5 and 6). By considering clinical and investigation findings, a final diagnosis of arteriovenous malformation of the tongue (AVM) was made. Since the lesion was asymptomatic, the patient was followed up and asked to report if any changes are noted.

**DISCUSSION**

AVMs are fast-flow vascular lesions composed of dysmorphic arterial and venous vessels connected directly to one another without an intervening capillary bed.

AVMs may arise during early fetal development, due to the failure of regression of arteriovenous channels in the primitive retiform plexus. The embryologic theory explains the predominance of AVMs in the head and neck region, since the
early embryo is composed mainly of cephcal structures. In addition, the cheeks and ears, the most common facial sites of AVMs, have higher surface area to volume ratio than other facial structures during early embryonic development. Although most cases of AVMs are sporadic, there are few inherited syndromes whose molecular genetics have 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. There are also other hereditary cases of AVMs that are autosomal dominantly transmitted, such as hereditary hemorrhagic telangiectasia. It is evident that transforming growth factor-β is involved in the induction of apoptotic endothelial cell death. There is also research to support the idea that a reduced apoptotic process may cause the dysregulation of vascular growth in AVMs. It is interesting that AVMs are much more prevalent in the central nervous system than elsewhere because neurons, having lost the ability to multiply, rarely undergo apoptosis. The milieu in which neurons reside may similarly affect the vascular endothelium to explain the high incidence of intracranial AVMs.

In addition, it is evident that even in early embryogenesis, arterial and venous endothelial cells (ECs) are distinguished by distinct ligands and receptors; namely, arterial ECs with transmembrane ligand ephrin B2 and venous ECs with the receptor for ephrin B2, EphB4. The reciprocal signaling between these two types of vessels is critical in the formation of capillary beds. Thus, it is proposed that a defect in ephrins or their receptors may be a causative factor in the formation of AVMs. Although the cause of AVM expansion is unknown, it has been proposed that expansion may result from dormant primal arteriovenous communications, dilation of normal latent arteriovenous shunts by increased pressure and flow, and local ischemia caused by trauma. AVMs occur with equal frequency in males and females. About 40 to 60% of lesions are visible at birth, and 30% become clinically apparent during childhood. They are more common in the head and neck area than in other locations. AVMs may progress through four different stages and can be scored by severity using the 1990 ISSVA-accepted Schobinger clinical staging. Stage I lesions are in the quiescent phase, are asymptomatic, and usually last from birth until adolescence. During this stage, the AVM may not be apparent or have the appearance of a port-wine stain or involving hemangioma. The presence of increased warmth, a bruit, or thrill suggests a high-flow component. Some AVMs remain quiescent throughout a patient’s lifetime. Stage II, the progressive phase, most commonly begins during adolescence. This stage represents expansion, when the vascular lesions enlarge and darken, deforming the integument and invading deep structures. Histologically, both arteries and veins undergo progressive dilatation, thinning and fibrosis. On examination, local temperature is increased, a pulse or thrill can be palpated, and a murmur is heard on auscultation. Tortuous draining vessels appear and elongate. Skin changes mimicking Kaposi sarcoma, known as Stewart-Bluefarb syndrome, may develop in lower extremity AVMs. Conversely, Kaposi sarcoma may be misdiagnosed as an AVM. Advancement to this stage is commonly induced by puberty, trauma and pregnancy. In addition, some forms of treatment, including ligation of arterial feeders, partial excision, incomplete arterial embolization and laser treatment, can trigger progression of quiescent AVMs. During stage III, which grossly mimics stage II, deep destruction occurs with spontaneous necrosis, chronic ulceration, pain and hemorrhage. Lytic bone lesions may occur. This phase usually develops after years of progressive worsening. Finally, stage IV is defined by cardiac decompensation. High-output cardiac failure may result from increased blood flow in large AVMs.

AVMs are diagnosed by clinical findings and radiologic features. The differential diagnosis of AVMs includes other vascular malformations, vascular neoplasms and, in rare cases, other neoplasms. Auscultation of the lesion and bedside Doppler scanners are helpful screening tools. Ultrasonography and color Doppler evaluation are often performed initially to assess the flow characteristics. In addition, the presence of flow voids on MRI will help to confirm the presence of “fast-flow” vessels. MRI/magnetic resonance angiography and angio computed tomographic scans are noninvasive methods of visualizing the vascular components and defining the extent of the AVM. An arteriogram is done in patients with arterial malformation not for diagnosis, which is easily done clinically but to know the major feeder artery prior to embolization. Treatment of AVMs may be difficult. Partial treatment usually results in recurrences that may be more difficult to manage than the initial malformation. Therefore, quiescent AVMs that are not disfiguring or impairing function should be followed closely, avoiding premature partial treatment whenever possible. Extreme pain, ulceration, bleeding and extensive enlargement of the malformation are indications for treatment. Palliative embolization may be used to treat symptomatic AVMs when combination treatment cannot be performed.

REFERENCES


