Arterio Venous Malformation of Tongue Presenting with Bleeding in Pregnancy

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Arterio venous malformation (AVM) of the tongue is a rare craniofacial vascular anomaly. It occurs due to the failure of complete involution of fetal capillary bed leading to the development of abnormal connections between arteries and veins. It can present with different clinical presentations and radiographic findings. It may be overlooked at birth due to its innocent appearance. Progression of the AVMs is commonly induced by puberty, trauma, and pregnancy. Furthermore, some forms of treatment, including ligation of arterial feeders, partial excision, incomplete arterial embolization, and laser treatment can trigger progression of quiescent AVMs. Progression of AVMs in pregnancy can sometimes lead to torrential hemorrhage. This can endanger the life of the mother and fetus. We report a case of a 22-year-old primigravida referred to our institution at 35 weeks 3 days gestation with intrauterine fetal death and severe anemia, due to profuse bleeding from an AVM of the tongue.

Keywords: Pregnancy, Tongue, Vascular malformations

INTRODUCTION

Vascular malformations are seen in approximately 1% of the general population. However, many of them do not present for treatment.1 The first classification of vascular malformations was introduced by Glovacki and Mulliken in 1982. This classification was based on the structure and behavior of these malformations.2 According to this, vascular malformations were divided into arterial, venous, capillary, lymphatic and combined.3

Arteriovenous malformations (AVM) is a type of vascular anomaly where arterial and venous vasculature communicate with each other.4

AVM occurs as a result of failure of complete involution of fetal capillary bed. This results in the development of abnormal connection between arteries and veins leading to progressive vascular enlargement, venous hypertension, destruction of tissue, esthetic problems, and rarely cardiac decompensation due to high output state.

Trauma, puberty and pregnancy can induce proliferation of AVM. Those induced by trauma usually involve a single vessel. The congenital form of AVM usually involves multiple vessels. Most AVMs are present at birth, but become clinically significant later on in childhood.5,6

Pregnancy increases the risk of bleeding from an AVM. This can lead to increased maternal morbidity and sometimes mortality and can also endanger the life of the fetus.7

AVM may affect any part of the body, but most commonly seen in the intracranial cavity. Most common extracranial head and neck location of AVM is the auricle.

CASE REPORT

Mrs. R, a 22-year-old primigravida was referred to our institution at 35 weeks 3 days gestation with intrauterine fetal death, severe anemia and bleeding from AVM tongue. Her hemoglobin (Hb) at the time of reference was 3.6 g%. She gave a history of profuse bleeding from her tongue of 2 hours duration and oral cavity was packed with gauze. As she did not appreciate fetal movements, ultrasonography (USG) was done, which showed intrauterine death of the fetus and she was referred.

She gave a history of maxillofacial surgery 2 years back for cosmetic reasons. There was a history of surgery done for hemangioma of tongue 1 month back. A magnetic resonance imaging (MRI) taken at that time diagnosed AVM of the tongue.

On admission, she was conscious, well-oriented, facial dysmorphism present, had severe pallor, pulse rate 100/m,
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blood pressure 106/68 mm Hg. Examination of the oral cavity showed enlarged tongue with a reddish lesion 2 cm \times 2 cm on the left side, with no bleeding (Figure 1).

Obstetric examination revealed the uterus to be of 34 weeks size, not tense, not tender, Cephalic presentation, fetal heart was absent, uterus was acting.

P/v examination showed that the cervix was well effaced, 5 cm dilated, bag of membranes present, vertex at-1, pelvis normal.

Repeat investigations were sent, and 4 pints of blood products were transfused including 3 pints packed red blood cell, and 1 pint fresh frozen plasma.

Investigation reports showed Hb 3.3 g%, TC 21600, platelet count 1.58 lakhs/mm³. MRI tongue (Figures 2 and 3) 13/06/14 - high flow AVM involving tongue, tongue enlarged, multiple enlarged vessels within and around the tongue, more involvement of the left half of the tongue. Enlarged external carotid arteries, moderate narrowing of oropharynx and enlargement of ramus of left hemi mandible probably due to hyperemia.

USG - 26/07/14 single intrauterine fetal demise, anterior placenta, AFI 11 cm, no RP clots, estimated fetal weight 2.4 Kg, gestational age 34 weeks 2 days.

Labour was augmented with oxytocin. She delivered an IUD-FSB male baby of weight 2.32 kg, within 4 hours of admission, at 1 a.m. on 27 July, 2014. She was discharged on 01 August, 2014 after an oral and maxillofacial surgery consultation who advised her to review after 6 weeks.

**DISCUSSION**

AVMs are fast-flow vascular lesions. They are composed of dysmorphic arterial and venous connections without an intervening capillary bed. They develop, due to the failure of regression of arteriovenous channels in the primitive retiform plexus. Thus, they are seen in early fetal development itself. The shunting between the high-pressure arterial and low-pressure venous channels account for the clinical picture and progression of the lesion.

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.

Transforming growth factor is involved in the induction of apoptotic endothelial cell death. A decrease in the apoptotic process may lead to the dysregulation of vascular
growth resulting in AVMs subsequently. They are more prevalent in the central nervous system, because neurons rarely undergo apoptosis.

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.

AVMs occur with equal frequency in males and females. About 40-60% of lesions are visible at birth. 30% of AVMs become clinically apparent during childhood. They may progress in severity through 4 different stages. The 1990 ISSVA-accepted Schobinger clinical staging is used to assess the severity of AVMs.

Stage I lesions are asymptomatic and have the appearance of a port-wine stain on involuting hemangioma. They usually last from birth until adolescence. The presence of a thrill, a bruit or increased warmth may suggest a high flow component in the AVM. Some AVMs may remain in this quiescent stage throughout the patient’s life.

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. On clinical examination, local temperature is increased and on palpation, a pulse or a thrill can be felt. On auscultation, a murmur may be heard. A close differential diagnosis at this stage is Kaposi’s sarcoma. Advancement to this stage is induced by puberty, trauma and pregnancy. Some form of treatments like ligation of arterial feeders, partial excision, incomplete arterial embolization, and laser treatment can also trigger progression.

Stage III - Grossly mimics Stage 2. Here deep destruction occurs with spontaneous necrosis, chronic ulceration, pain and hemorrhage. This stage is usually seen after years of progressive worsening.

Stage IV is characterized by cardiac decompensation. High output cardiac failure develops.

A number of cases of AVMs have been described by different authors. The largest series was reported by Kohout et al. They reported 81 cases of AVMs located in the head and neck area. The majority of the lesions were found over the cheek (31%) and the ear (16%). The rest of the malformations were located on the nose (10%), the forehead (10%), the upper lip (7%), the neck (5%), the mandible (5%), the maxilla (4%) and the scalp (4%).

Data regarding AVMs in pregnancy are limited. According to Robinson and Sabiston, bleeding from an AVM in pregnancy was most commonly seen in young women (20-25 years) and in primipara. In most cases, bleeding occurred between 15 and 20 weeks of gestation. However, it could occur at any stage of pregnancy including labour and puerperium. Cardiac failure can occur in cases of AVM in pregnancy, but the exact mechanism of this failure is not clear. In a normal pregnancy, the cardiac output increases by 30-40% by the 24th week, and this is caused by an increase in stroke volume and heart rate. When pregnancy and AVM coexist, it may result in nearly 150% increase in cardiac output. A number of cases have been reported with high output cardiac failure in pregnancies having AVM. So embarking on a pregnancy with preexisting AVM could be dangerous.

AVMs may show spontaneous regression in the postpartum period. Progesterone is the predominant hormone in pregnancy. Increased venous distensibility of pregnancy is due to progesterone. This could be the reason for progression of AVM in pregnancy.

Diagnosis of AVMs is by clinical findings and radiological features. Differential diagnosis includes other vascular malformations and vascular neoplasms. Plain radiography and CT Scans may not help as a diagnostic tool in these malformations. USG and colour Doppler evaluation helps in the initial assessment. Now-a-days, MRI is the investigation of choice. It shows the extent and lack 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 typical angiographic findings are marked hypertrophy and tortuosity of the feeding vessels. The Nidus of the lesion varies from large tortuous vessels to many small vessels. Collaterals have a corkscrew appearance. Parenchymal staining is absent when a contrast is used.

Treatment of AVMs may be difficult. Partial treatment results in recurrences. Hence, quiescent AVMs are followed up closely at 6 months or yearly interval and treated only in case of extreme pain, ulceration, bleeding and extensive enlargement. Ligation or proximal embolization of feeding vessels is avoided as far as possible as this may lead to the rapid flow from nearby arteries. Treatment of symptomatic AVMs is by palliative embolization whenever combination treatment cannot be performed. For AVMs not amenable to surgery, super selective arterial or retrograde venous embolization is used as the first choice of treatment. Different materials can be used for embolization. These
include coils, PVA foam, methyl methacrylate and silicone spheres and gel foam. Gelfoam particles are easy to handle with minimal risk of migration.  

CONCLUSION

AVM of tongue is a rare vascular anomaly. Pregnancy and partial treatment can cause progression of malformation. Profuse bleeding from the lesion in pregnancy can endanger the life of mother and fetus, which has happened in our case. Supportive measures and palliative embolization may help in such situations.

REFERENCES


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