

# Late onset arteriovenous malformation of nose: a clinical report

Wajieha Saeed, Bushra Bashir, Faria Altaf

Department of Dermatology Unit I, KEMU/ Mayo Hospital, Lahore.

**Abstract** AVM's are errors of vascular morphogenesis, almost always present at birth, sometimes manifesting later in life. Head and neck is the commonest site, accounting for 60% of cases. AVM of head & neck is a rare but a life threatening benign condition. We are reporting a case of AVM on nose of a 60 years old patient, presenting during pregnancy at the age of 50 years.

**Key words**

Arterio-Venous Malformation, Vascular tumors, Embolization, CT angiography.

## Introduction

Vascular anomalies are a heterogeneous group of congenital blood vessel disorders, typically referred to as birth marks. They are subcategorized into vascular tumors and malformations.<sup>1</sup> Hemangiomas are the most common vascular tumor. AVM's are classified as fast flowing and slow flowing lesions. Fast flowing lesions are predominantly AV fistulas, whereas venous, capillary and lymphatic lesions produce slow flowing lesions.<sup>2</sup> AVM, representing a localized defect in vascular morphogenesis, is essentially an abnormal communication between an artery and vein, which bypasses the capillary bed.<sup>3</sup> AVM is composed of a central nidus with anomalous AV shunts and a network of surrounding collateral vessels.<sup>4</sup>

AVM of head and neck region is a rare benign condition but can induce a life-threatening

haemorrhage. AVM, usually present at birth, sometimes appear within weeks to years after birth, can increase rapidly secondary to trauma, pregnancy, puberty, infection, ligation or attempted excision.<sup>2,5</sup> They are often asymptomatic. Indication for treatment is disfigurement or bleeding.<sup>6</sup> Ultrasound, MRI and cerebral arteriogram are the choice of investigations.<sup>6,7</sup>

We present an unusual case of AV malformation, presenting at 50 years of age, expanding rapidly during pregnancy.

## Case report

60 years old married female presented to outpatient department with a 10 years history of swelling over the nose. There is history of painless erythematous patch on the left nasal ala and tip of nose since birth. 10 years back, when patient was in her last trimester of thirteenth pregnancy, she noticed a painless reddish small growth on the left side of her nose. It progressively increased in size for next 2-3 months with occasional history of nose bleed. 4 years later, the lesion expanded rapidly involving both alae and tip of nose leading an obvious cosmetic disfigurement.

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### Address for correspondence

Dr. Dr. Wajieha Saeed  
Department of Dermatology Unit-I,  
King Edward Medical University/ Mayo Hospital,  
Lahore.  
Ph: 03334210884  
Email: jiya114@hotmail.com



**Figure a**



**Figure b**

There is no history of trauma or surgery to the involved site. She took topical and oral treatment from local doctors and hakeem with no improvement. There is off & on history of headache and nasal bleed. Patient was diagnosed as being anti-HCV positive about 3 years, for which she didn't take any treatment. There is no similar history in family.

When the patient was seen, lesion was in second Schobinger stage (**Table 1**).

**Table 1** Schobinger Classification

Stage	Features
I	Cutaneous, bluish color, warmth
II	Expanding lesion, bruit, pulsatile
III	Pain, ulceration, bleeding, infection
IV	Cardiac failure

On examination, a purplish irregularly shaped, diffuse swelling 5x6 cm involving the dorsum of nose extending upto nasal alae, soft to firm in consistency, warm to touch and surface was smooth (Figure a, b). Pulsations were observed at the upper portion of plaque. There was no palpable thrill or bruit over the swelling. Skin colored bulge was seen in both vestibules, otherwise examination of nasal cavities was normal, as was the rest of head and neck examination. Cervical lymph nodes were not enlarged.

Routine investigations were insignificant. Patient was anti-HCV but LFTs and ultrasound abdomen within normal limits. Doppler ultrasound of the swelling showed a highly vascular soft tissue mass with multiple dilated tortuous vessels with high velocity and low resistance flow. MRI brain revealed numerous hypolucent arterial flow voids.

Diagnosis of low flow AVM was made. Patient was referred to plastic surgery department where an arteriogram has been advised

## Discussion

AVM may be congenital or acquired. Congenital AVM are extremely uncommon, though acquired are far more prevalent. Most of the congenital AVMs are present at birth, they often present later in life. Mean age at presentation is 19 years without sex predilection.<sup>4</sup> The natural course of AVM is early quiescence, late

expansion and ultimately infiltration and destruction of local soft tissue and bone. Hormonal changes are thought to influence the growth.<sup>1,4,5</sup>

While in most instances, the diagnosis of AV malformation can be made on the basis of a clinical examination; the imaging modalities are essential in identifying the extent of AVM.<sup>1,3</sup> MRI is helpful to differentiate AVM from other vascular lesions and to see intracranial extension. This will help in the treatment planning. An arteriogram or angiography is the gold standard modality to understand the angio-architecture of the lesion and to exclude any intracranial component.<sup>3,5,6</sup>

Treatment of AVM is complex because of its high flow, complex vascular anatomy and cosmetic issues, often requiring multiple disciplines and therapeutic options.<sup>4,5,7</sup> Intravascular embolization of AVM can be used alone or in combination with complex reconstructive techniques. Despite successful initial therapy, these lesions may recur many years later making vigilant management necessary.<sup>7,8</sup> Endovascular embolization; followed by surgical excision is the treatment of choice, to minimize the risk of recurrence. Post operation long term close follow up is required to detect early signs of recurrence.<sup>8,9</sup>

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