A-V malformation of nose: A rare challenging case

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ABSTRACT

Congenital A-V malformations of head-neck region are extremely uncommon lesions encountered in clinical practice. They may lead to a wide array of clinical effects from cosmetic disfigurement and recurrent life threatening haemorrhage to cardiac failure. Treatment of these lesions pose a challenge to reconstructive surgeons due to their extreme vascularity, high incidence of recurrence and involvement of adjacent vascular channels. The case presented here is a congenital A-V malformation treated successfully by thorough surgical excision and reconstruction by a well vascularised flap.

Key words: A-V malformation, MR angiography, Hemangioma, Nose, Scalping forehead flap

INTRODUCTION

Arteriovenous malformations (AVM) are a group of conditions related to errors of vascular morphogenesis, almost always present at birth and manifest later in life. AVMs are 20 times more common in intracerebral vasculature than in extracerebral sites. AVM is composed of a central nidus with anomalous arteriovenous shunts and a network of surrounding collateral vessels. Tissue from epicentre of an A-V malformation demonstrates close juxtaposition of medium sized arteries and veins. In time the veins become arterialized and exhibit intimal thickening, increased smooth muscle within the media and dilation of vasa vesorum. The arteries, primarily involved gradually dilate further and become tortuous. The veins also dilate and there is progressive fibrosis within the intima, media and adventitia.

Arteriovenous malformation of nose must be differentiated from haemangioma, commonest childhood tumour, affecting females more than males and may regress spontaneously. Histologically, haemangiomas show proliferation of endothelial cells in contrast to AVM which have none. AVM can increase rapidly secondary to infection, trauma, attempted ligation and hormonal influence such as puberty and pregnancy.

Herewith we present an unusual case of A-V malformation of nose involving the overlying skin and underlying structures. The patient presented with lesion in Schobinger stage III (Table 1).

After confirmation of diagnosis, the patient was treated with complete surgical excision and reconstruction by a well vascularised flap.

CASE REPORT

A 28 year old male presented with a diffuse swelling over the nose for last 7 to 8 years, gradually increasing in size and associated with episodes of bleeding. Examination of nose revealed a redish, irregular, diffuse swelling (10x6 cm in greatest dimension) involving the dorsum of the nose extending from root of the nose
overhanging the tip inferiorly and involvement of medial 2/3rd of both ala with a bleeding point over inferior most aspect of the swelling (Figures 1-4). Overlying skin was irregular and erythematous. On palpation, there was strong pulsation over the swelling with a marked rise of local temperature. The swelling was firm in consistency and non-tender. There was a palpable thrill over the swelling and auscultation revealed a continuous machinery bruit.

Anterior Rhinoscopy and Post nasal examination was normal. On examination of neck there was no palpable lymph node. Routine investigations were within normal limit.

Magnetic Resonance Angiography (MRA) showed a large soft tissue mass involving whole dorsum of nose and both ala with multiple curvilinear areas of flow voids within the lesion; no extension of lesion into the nasal cavity. Time of flight angiography of the lesion showed multiple small arterial feeds from bilateral facial and left lingual arteries. Tortuous draining veins were also noted.

After confirmation of diagnosis, the patient was prepared for excision of the lesion and reconstruction of the defect with scalping forehead flap under general anaesthesia.

Paramarginal incision was given surrounding the lesion→feeding vessels ligated→the swelling dissected

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Figure 1: Diffuse swelling involving dorsum of nose (frontal view)

Figure 2: Swelling involving whole dorsum of nose sparing part of ala (right lateral view)

Figure 3: Swelling involving whole dorsum of nose sparing a small part of ala (left lateral view)

Figure 4: Bleeding point over inferior most aspect of the swelling covered with a scab
off the underlying nasal bones and cartilages and excised with involved overlying skin and soft tissue→haemostasis secured→post excisional defect measured (Figure 5) and a scalping forehead flap planned accordingly with insetting part over the right lateral aspect forehead. Insetting part of the flap was elevated superficial to frontalis muscle and rest of the flap was harvested in the subgaleal plane (Figure 6).

Insetting part of the flap was sutured to the nasal defect and the flap donor site was covered with split thickness skin graft (Figure 7).

Specimen was sent for histopathological examination. Flap division and final inset was done after 3 weeks (Figure 8). Wound healed satisfactorily.

Histopathology revealed thick and thin walled tortuous vessels in a cellular stroma surrounded by fibromyxoid connective tissue. The lesion showed extensive spread across the skin and into the connective tissue.

Figure 5: Post excisional defect

Figure 6: Scalping forehead flap harvested

Figure 7: Flap insetting done with the margins of the defect

Figure 8: Flap division and final insetting was done after 3 weeks

Figure 9: Satisfactory aesthetic outcome
The postoperative evaluation performed at 1st, 3rd, 6th, and 12th month; showed satisfactory functional and aesthetic outcome (Figure 9).

DISCUSSION

Arteriovenous malformation may be congenital or acquired. While most congenital malformations are present at birth, they often come to clinical attention later in life. They frequently present in 2nd and 3rd decade of life and may manifest with pubertal growth spurt or pregnancy.

When a vascular mass presents with palpable thrill and a continuous bruit, it is usually associated with abnormal arteriovenous connection. An increase in local temperature over the lesion is a striking feature. There may be surface ulceration or episodes of bleeding from the swelling.

While in most instances, the diagnosis of A-V malformation can be made on the basis of clinical examination; the importance of MRA in delineating precise nature and extent of the lesion can’t be overemphasized. As the AVMs have a wide range of effects on the patients including cosmetic disfigurement and recurrent episodes of bleeding from the lesion, surgical removal followed by reconstruction is the treatment of choice.

The scalping forehead flap, described by Converse in 1942[^3] provides a large area of forehead skin of satisfactory thickness, colour and texture match for reconstruction of nasal defect.

The frontalis muscle, galea and their overlying skin are well vascularised by an extensive anastomotic network from supraorbital, supratrochlear and superficial temporal vessels. All these vessels except ipsilateral superficial temporal system ensure a rich blood supply to the flap.

CONCLUSION

Surgical treatments of A-V malformations pose a challenge to the reconstructive surgeons due to their location, proximity to vital structures and extreme vascularity. Preoperative accurate diagnosis, thorough surgical excision and reconstruction of the defect with a well vascularised flap offer the best mode of treatment of these lesions.

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Authors Contribution:

GRC : Concept and design of the case report, Manuscript preparation, Review of literature, Interpretation of case. RG: Assisted in Manuscript preparation, Submission of manuscript. RH : Collection of data, Review of study. SNB: Critical revision of manuscript; Overall review of literature.

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