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Arteriovenous malformations

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ABSTRACT

Arteriovenous malformations (AVMs) are fast-flow vascular malformations composed of a complex vessel network directly connecting feeding arteries to draining veins. The intervening normal capillary network is absent. Proper diagnosis and treatment of AVMs is challenging and in need of an interdisciplinary team of experienced physicians. Careful analysis of the clinical features and evaluation of therapeutic options represent the basis for successful management of AVMs. This article will focus on the clinical and radiological findings and in particular on interdisciplinary management strategies of AVMs including minimally invasive endovascular and surgical treatment.

Introduction

Arteriovenous malformations (AVMs) have been particularly challenging to understand and manage. AVMs are fast-flow vascular malformations that comprise a complex network of primitive vessels directly connecting feeding arteries to draining veins. Arteriovenous shunting through these vessels has an inherent potential to increase and recruit new blood vessels. The intervening normal capillary network is lacking, either partially or completely. Peripheral AVMs occur virtually in any organ system with superficial, deep or combined distribution.

AVMs most commonly affect the head and neck (47.4%) followed by the extremities (28.5%).[1] Extracranial AVMs may be focal or diffuse.[2,3] This description best determines the outcome of AVMs following treatment. AVMs of the central venous system have different clinical presentation, natural history, and management approach, which is beyond the scope of this review.

The exact etiopathology of extracranial AVMs remains unclear. These uncommon lesions are usually sporadic, though genetic predisposition occurs in several syndromes such as capillary malformation–arteriovenous malformation (CM-AVM, mutation in RASA1 gene), CLOVES syndrome (somatic mutation in PIK3CA), PTEN hamartoma tumor syndrome (mutation in the PTEN gene), and hereditary hemorrhagic telangiectasia (HHT, mutations in ENG, ACVRL1, and SMAD4 genes).[4–7] The natural history of AVMs is typically progressive, and spontaneous regression is almost never seen. Recurrence after embolization or resection is reported to be greater than 80%, a rate more common than most cancers.[3,8]

Clinical findings and natural history

Arteriovenous malformations are usually congenital, though they may stay dormant during early childhood (Figure 1). Most of the superficial AVMs become evident during childhood. Only one-fifth of superficial AVMs are undetected before adulthood.[9] Young children often present with AVM in the dormant or quiescent states with any significant growth. A vascular stain may affect the overlying skin or mucosa. As they progress, increased pulsatility and tissue overgrowth become evident. Special physiologic conditions, like hormonal changes of puberty, pregnancy, or trauma, may trigger the expansion of AVM. The tendency to progress often leads to clinical manifestation in the second or third decade of life.[10–12] Chronic hemodynamic effects including local hypervascularity, steal phenomenon, and venous hypertension, predispose to many of the clinical features. The manifestations are variable in severity and include tissue overgrowth, hyperemia, pain, bleeding, ischemia, and in rare cases, high-output heart failure. When active, the AVM will grow, have large superficial telangiectasias, and will frequently bleed. Profuse, pulsatile bleeding that is prolonged and difficult to control is a hallmark of an AVM. The force and amount of bleeding is often out of proportion to the size of the bleeding site (millimeters).

Congestive heart failure is uncommon (occurs in less than 2%) and mainly affects children in the neonatal period with massive shunts or young adults.[9] Peripheral tissues can be affected with a wide range of symptoms from distal ischemia to gangrene, venous stasis dermatitis, and ulcer or gangrene caused by venous hypertension.[13] In some patients, symptoms may periodically worsen; the mechanisms are not very well understood. Endothelial progenitors and vascular factors are thought to be involved.[14]

The Schobinger clinical staging is a severity scoring system of AVMs and describes the natural progression of these lesions: stage I

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Table 1
Revised Richter–Suen classification system for extracranial arteriovenous malformations.[8]

Focal AVM	Multicentric (diffuse) AVM
Central nidus	Nidus difficult to detect or multiple
Single arterial feeder	Two or more arterial feeders
Firm	Compressible, rapid rebound
Single of skin involved if present	Diffuse skin involvement in affected region

is the quiescent stage. In stage II, the lesion becomes warmer, and larger with a thrill. Stage III comprises additional destruction like ulcers, hemorrhages, and bony lytic lesions, whereas the presence of heart failure constitutes stage IV. This classification scheme has been extrapolated by others to better define the outcomes of management.[8,11] In essence, focal AVMs with a single dominate feeder fair better from treatment than those crossing tissue planes with multicentric sites (Table 1).[3,8]

Radiological findings

Ultrasonography with color and spectral Doppler interrogation can reliably identify AV shunting and characterize the flow pattern in AVMs and arteriovenous fistulae (AVFs). Contrast-enhanced computed tomography (CT) scan is not typically required for soft tissue lesions, though it can readily detect bony involvement. Magnetic resonance imaging (MRI) allows assessment of the lesion and the adjacent soft tissue. MRI studies demonstrate dilated feeding arteries and draining veins without a discrete mass. On MRI, signal abnormalities may be present in the surrounding tissue representing fibrofatty changes, edema, and disorganized overgrowth. Osseous involvement manifests as overgrowth and lytic changes with diffuse abnormal bone signal.

Angiography reveals dilatation of the feeding arteries with tortuosity, AV shunting, and even greater dilatation of draining veins (Figure 2). No defined soft tissue enhancement is detected in AVMs. In a diffuse or early-stage AVM, a blush with early venous opacification may be noted rather than a discrete nidus. Cho et al.[15] classified the peripheral AVMs into 4 types according to their angiographic morphologies with implications for therapeutic approach and outcomes.

Interdisciplinary treatment

Treatment of AVM is very challenging and requires extensive experience and an interdisciplinary approach. These trans-spatial lesions are prone to recurrence, and interventions can be associated with serious side effects. Necrosis of locally involved and adjacent normal tissue may occur. Wound healing is compromised by the inherent hypoxic environment of AVMs as intervening capillaries (carriers of oxygen rich nutrition) are absent.

Treatment planning relies on proper evaluation of the shunt type, clinical and hemodynamic burden, and detailed understanding of local anatomy. The main goal of AVM therapy is often maximal control of shunting and palliation of the clinical manifestations. Cure is possible in some of the favorable angiographic types and arteriovenous fistulas. In diffuse AVMs, the objective is to prevent the treatment from being worse than the disease. Thus, chronologically and site-specific staged procedures are common in this population to prevent local tissue damage and to capture recurrent disease early.

Embolization can be combined with resection, particularly for a localized nidus. Ligation of feeding arteries or proximal embolization only provides temporarily benefit and is eventually counterproductive. Incomplete therapy is not recommended as it typically leads to expansion of the AVM with recruitment of more vessels, collateralization, and expansion.

Minimally invasive endovascular approach

Percutaneous embolization is considered the first line of management of AVMs. This minimally invasive technique can be planned as the primary treatment or as an adjunct to surgical resection. For extensive AVMs, the treatment is staged over multiple procedures. For successful occlusion of an AVM via the transarterial route, the AVM arterial supply must be selectively targeted closing “the nidus” and early draining veins while maintaining the patency of normal branches, as AVMs share blood supply with the surrounding tissue.[16] The ideal embolic agent should be safe and allow for controlled deployment, penetration of the nidus into draining veins, and permanent occlusion. Unfortunately, such an ideal agent does not exist. Liquid (e.g., ethanol) and semiliquid (e.g., N-butyl cyanoacrylate glue and Onyx) agents are generally considered as permanent agents. The lack of visibility and risk of serious complications are the disadvantages of ethanol application. The use of glue demands extensive experience, high polymerization time, extended period of injection, high radiation doses, and the need to immediately retract the microcatheter to prevent it from “gluing” to the vessels themselves. Onyx, an ethylene vinyl alcohol copolymer dissolved in various concentrations of dimethyl sulfoxide (DMSO) and opacified with micronized tantalum powder, has been primarily used for brain-AVMs. Onyx provides enhanced nidal penetration, forming a cast that plugs the nidus permanently. The use of Onyx in peripheral AVMs is still limited and time-consuming.[17,18]

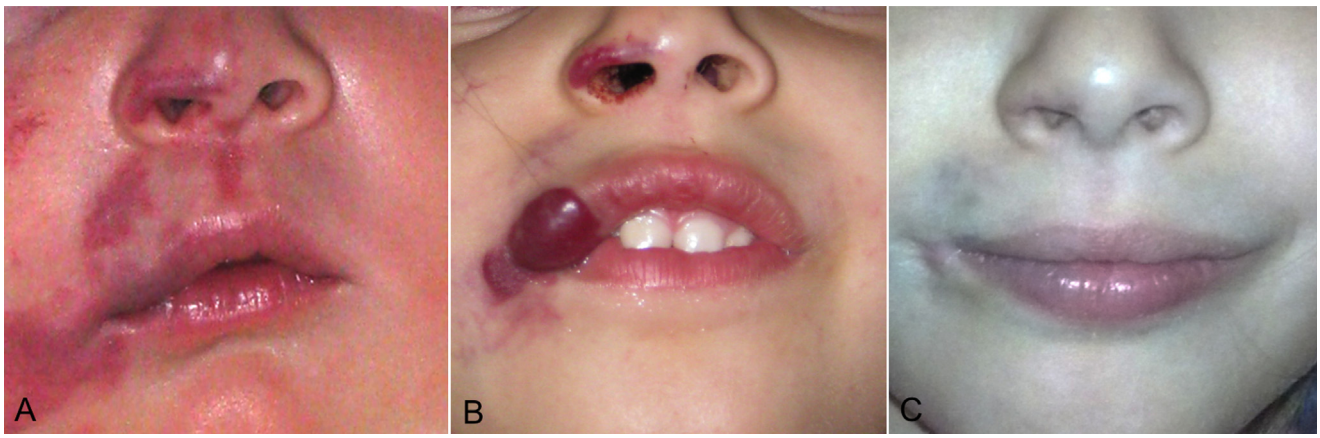

Fig. 1. Photographs of a young female with continuous and destructive growth, an arteriovenous malformtion of the midface. A red blush can be seen at infancy (A) that progresses to highly vascular masses of the nasal tip and lip (B). Improvement can be seen following multimodal treatment with surgery with preoperative embolizatio and gentle yag laser therapy (C). (Color version of figure is available online.)