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Diagnostic Aids in Arteriovenous (AV) Malformations

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Abstract

The field of vascular anomalies and their clinical management is expanding and developments within the previous couple of years have improved the understanding of haemangioma's and vascular malformations, identification and treatment of vascular malformations may be a challenging endeavor for physicians, especially given the good concern and anxiety created for patients and their families. The goal of this study is to review of vascular malformations, organized by subtype, including capillary, venous, lymphatic and arteriovenous malformations by developing a transparent understanding of the clinical aspects, diagnostic tools and imaging modalities.

Keywords: Arteriovenous Malformation; diagnostic aids.

Introduction

Arteriovenous malformations (AVMs) are high-flow vascular malformations characterized by a complex vascular network that directly connects afferent arteries and efferent veins. The normal capillary network is not normally present, and arteriovenous shunts through these abnormal blood vessels tend to recruit new blood vessels over time. Therefore, AVM usually shows a history of natural progression, but spontaneous regression is purely anecdotal. They can stay calm throughout early childhood, and their bulky tendency often causes symptoms and signs that appear during the first 20 or 30 years of life. Hormonal changes (during puberty or pregnancy) and trauma can cause authoritative

progression of AVM. AVMs are highly aggressive entities that behave locally infiltrate like neoplasms. They do not metastasize, but can be fatal due to complications such as bleeding and heart failure. For these reasons, proper treatment of AVM is of paramount importance. Careful evaluation of clinical features and treatment options are essential for successful treatment. Complete "curative" surgical resection is most likely to recover. Early treatment is recommended to prevent the continuous and destructive growth that occurs in this type of lesion. Endovascular embolization is often performed prior to radical surgical removal of arteriovenous malformations due to the potential for intraoperative bleeding. On the other hand, the recurrence rate after embolization alone or after non-curative resection is said to be 80% or more [1]. There is considerable controversy over how to handle large AVMs that are not suitable for "fundamental" resection. Many experts advise against partial removal and / or recon touring because these procedures have an unacceptably high risk of inducing explosive growth of AVM debris. For these reasons, patients with large unrespectable lesions often go untreated, with devastating cosmetic and functional consequences. In the past, terms such as salmon patch, strawberry hemangiomas, port wine stains, and hemangiomas have been used to name vascular malformations and tumors, despite their different etiologies and etiologies, clinically, histopathologically, and therapeutically. It was often used obscurely without distinguishing the target aspect. It was not until 1982 that Mulliken and Glowacki developed the classification by combining the histological structure and appearance of the predominant endothelium with the biological behavior of vascular abnormalities. That classification was redefined by Mulliken and Young in 1996 and adopted by the International Society for the Study of Vascular Anomalies (ISSVA). In the new classification, vascular malformations were defined by their predominant component, separating the lesions into malformations and tumors. Malformations were subdivided into low and high flow. This system is today considered to be the reference classification. In the 1988 Hamburg classification, vascular malformations were defined according to the predominant vascular lesion (arterial, venous, arteriovenous shunt, or combined/mixed) and were divided into truncular or extratruncular depending on whether or not major axial vessels were involved. However, in a recent 2009 review, Hassane IN and Mulliken pointed out that the term hemangiomas were misused with a 71.3% chance, and nomenclature remains a serious problem and can cause confusion. Specify when prescribing certain treatments (such as propranolol). Genetic testing and diagnostic investigations based on the lesion's molecular biology, flow rate, and response to pharmacological treatment will be considered in the AVM classification in the near future.

Etiology: The etiology and most widely accepted theory of AVM hypothesizes that these lesions are due to an increase in the number of blood vessels caused by defects in angiogenesis, especially angiogenesis. The most common place for AVM is the brain, which is the most extensively studied place. It is unclear whether the results of examining brain AVM can be extrapolated to extra cranial AVM, but many angiogenic factors such as vascular endothelial growth factor and platelet-derived growth factor appear to play a role. This is the cause of the dynamic vascular dysfunction present in AVM and has been suggested to lead to early changes in

angiogenesis, as opposed to those caused by venous malformations. This is supported by the increased serum levels of metalloproteinase found in AVM. STAT proteins (transcriptional transducers and activators) are also involved in the pathophysiology of AVM angiogenesis. One group of these proteins, especially the STAT 3 protein, is the most active during the fetal period and may play an important role in angiogenesis.

Epidemiology: The prevalence of AVM is not well known, but hospital data range from 5 to 613 / 100,000 [6]. They are rare vascular malformations, accounting for only 1.5% of all vascular abnormalities]. Over 90% of AVM occurs intracranial. The exact incidence is unknown, but Visser et al. Extracranial AVM has been shown to occur in only 4.7% of 1131 patients with vascular abnormalities referred to the Center for Vascular Abnormalities for 14 years. Extracranial AVM most commonly affects the head and neck area (47.4%), followed by the limbs (28.5%). It has been reported that 50% of AVM in the head and neck is associated with the mouth, chin and face. In a retrospective review of 81 patients with head and neck AVM, Kim et al. He reported that the cheeks were the most common place (31%), followed by the ears (16%). Spreafico et al. However, no race or gender preferences have been shown have reported a male : female ratio of 1:1.5.

Angioarchitecture: It consists of: Arterial Feeder, Nidus, Draining Vein

Arterial Feeder: It can be single/multiple, It can be Pial/Perforating/Dural, Direct department deliver as teminal department, Indirect feeders deliver as passage.

NIDUS: The AVM Nidus is a compact tangle of dysplastic, skinny walled vessels of various duration connecting feeding arteries to draining veins

it could both be globular or conical in form and can be compact or diffuse.

Within the nidus, Arterial blood is shunted without delay into draining veins without passage via a normal, excessive resistance arteriolar capillary network.

Types of AVM

Type I: Unrelated dysplastic / incidental

TYPE II: Flow associated on proximal vessel

TYPE III: On distal small feeding vessel

TYPE IV: Intra Nidal Aneurysm

Sizes of AVMs: Cryptic, Occult, Micro AVM (much less than 1 cm), Small AVM (much less than 2.5cm), Moderate AVM (2 five-5cm), Large AVM (extra than five cm).

Hemodynamics: As a result of abnormal hemodynamic conditions, blood flow through the AVM is significantly higher than through the normal parenchyma, and the afferent and efferent veins gradually dilate and wind. AVM can be likened to a "sponge" of blood vessels that consume large amounts of blood and deprive the brain of normal circulation. This phenomenon is known as STEAL'S PHENOMENON.

Clinical Presentation of AVMs:

Brain AVM

- Hemorrhage
- Seizures
- Headache
- Neurological deficits
- Asymptomatic
- Pediatrics- hydrocephalus, heart failure

Orbital AVM

- Periocular mass, slowly increasing in size
- Pulsation positive
- Intermittent eyelid swelling
- Pain, proptosis, diplopia.

Maxillofacial AVMs

- Most patients with AVMs of maxillofacial soft tissue present with a facial deformity with an unclear boundary
- It originates from trauma and chronic involvement with formation of single fistulous tract between arteries and veins
- Telangiectasia
- Temperature of involved area is higher than the surrounding normal skin
- Objective signs are high blood flow
- In later stages the surfaces of AVMs turns into an ulcer and bleeding as blood steal, distension of jugular vein and an increase of pressure in superior vena cava cause widen heart boundary.

Intraosseous AVMs

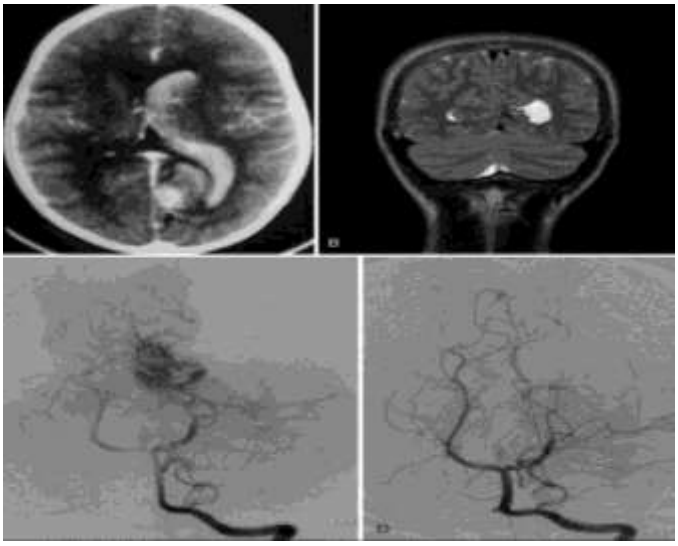
- Generally known as hemangioma of jaw.
- Acute bleeding occurs in the exchange of the primary and secondary teeth in children generally after tooth extraction.
- Intraosseous AVMs are located in molar areas with or without involvement of adjacent soft tissue.

Factors increasing risk of bleeding

NIDUS: 82% small AVMs (less than 3cm), 29% medium sized AVMs (3-6cm), 12% Large AVMs (greater than 6cm)

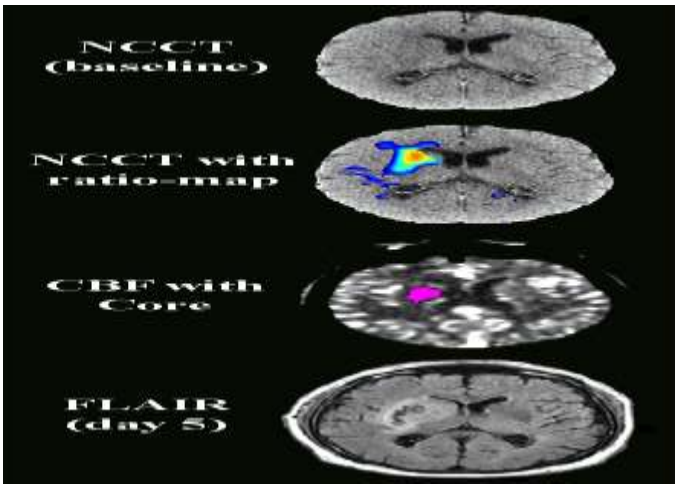
Diagnostic AIDS in AVMs

CT SCAN: CT is usually the first imaging modality used. AVM is suspected in young patients, if hematoma has lobar topography and if hyper dense serpigenous structures (calcifications) are seen, Parenchymatous Calcifications are observed in 20cases, related to intravascular thrombosis.



Non Contrast Computed Tomography

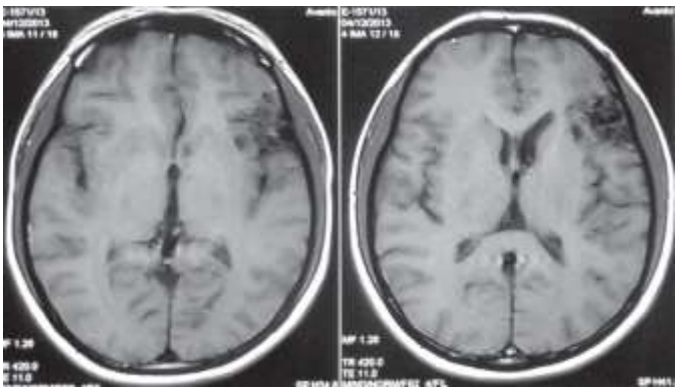
Head-to-face non-contrast computed tomography (NCCT) is an imaging test that uses rotating X-rays to create a cross-sectional image of the head and face and then uses it to create a three-dimensional image of the head and face. This image can be used to visually evaluate the anatomy of the skull and face and identify the underlying disorder**.**



MRI (Magnetic Resonance Imaging)

MRI is currently used to identify the underlying lesions in the case of unruptured AVMs or in the case of perilla hematomas days or weeks after bleeding. In T1 and T2-weighted images, there is no signal in the circulating vessels due to the flow void phenomenon. Blood vessels

are highlighted on T1-weighted images with gadolinium . The size and anatomical position of Nidas is accurately depicted by MRI.



MRA (Magnetic Resonance Angiography) :

MRA scan takes very clear, detailed pictures of the blood vessels-including arteries and veins.

This is done using MRI Machine and MRA Scan does not use radiation but uses powerful magnets, invisible radio waves and a computer to scan the body and take pictures.

MRA–is a type of MRI that looks specifically at the body's blood vessels. Unlike a traditional angiogram, which requires inserting a catheter into the body, magnetic resonance angiography is a far less invasive and less painful test. During magnetic resonance angiography, you lie flat inside the magnetic resonance imaging scanner. This is a large, tunnel-like tube. In some cases, a special dye, known as contrast, may be added to your bloodstream to make your blood vessels easier to see. When needed, the contrast is given with an intravenous (IV) needle.

Angiography

Angiography is an imaging test that uses X-rays to view your body's blood vessels. The X-rays provided by an angiography are called angiograms. This test is used to study narrow, blocked, enlarged, or malformed arteries or veins in many parts of your body, including your brain, heart, abdomen, and legs. A coronary angiogram is an X-ray of the arteries in the heart. We will inject a liquid dye through a thin, flexible tube, called a catheter. The doctor threads the catheter into the desired artery from an access point. The access point is usually in your arm but it can also be in your groin. The dye makes the blood flowing inside the blood vessels visible on an X-ray and shows any narrowed or blocked area in the blood vessel. The dye is later eliminated from your body through your kidneys and your urine.

Types of Angiography

- Coronary Angiography – to check the heart and nearby blood vessels
- Cerebral Angiography – to check the blood vessels in and around the brain
- Pulmonary Angiography – to check the blood vessels supplying the lungs
- Renal Angiography – to check the blood vessels supplying the kidneys

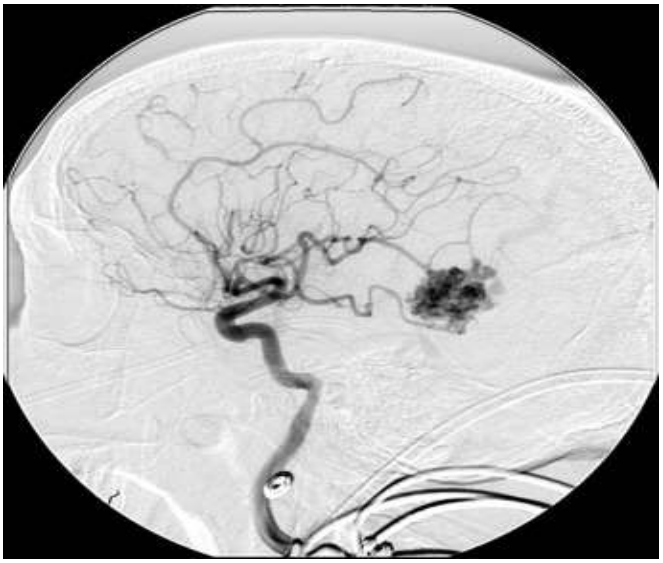
Multidetector Computed Tomography (MDCT)

MDCT angiography or MDCTA is now accepted widely as the standard technique for vascular imaging. MDCTA is a highly standardizable imaging technique. It provides three-dimensional information about the vasculature.

Digital Subtraction Angiography(DSA)

It is a fluoroscopy technique used to clearly visualize blood vessels in a bony or dense soft tissue environment. Images are produced using contrast medium by subtracting a "pre contrast image" or mask from subsequent images. Images are acquired by exposing an area of interest with time-controlled x-rays while injecting contrast medium into the blood vessels. The image obtained includes the blood vessels, together with all overlying and underlying structures. The images are

useful for determining anatomical position and variations, but unhelpful for visualizing blood vessels accurately. In order to remove the distracting structures to see the vessels better, first a mask image is acquired. The mask image is simply an image of the same area before the contrast is administered. The radiological equipment used to capture this is usually an X RAY intensifier, which then keeps producing images of the same area at a set rate (1 to 7.5 frames per second). Each subsequent image gets the original "mask" image subtracted out. (Mathematically, the incoming image is divided by the mask image.) The radiologist controls how much contrast media is injected and for how long. Smaller structures require less contrast to fill the vessel than others. Images produced appear with a very pale grey background, which produces a high contrast to the blood vessels, which appear a very dark grey. The images are all produced in real time by the computer or image processor, while the contrast is injected into the blood vessels.



AVM Associated Syndomes

1. Sturge-Weber Syndome
2. Rendu-Osler-Weber Syndome
3. Wyburn-Mason Syndome

Diagnosis

Grading

Classification of AVM Based On Diagnostic Grading Scales

A. Luessenhop-gennarelli Anatomical grading (1977)
B. Spetzler- Martin (1986)
C. Garretson
D. Nataf
E. Vienna Classification

Luessenhop-gennarelli Anatomical grading: This is the anatomical grading of supratentorial cAVM described in 1977, according to the degrees of surgical difficulty for total obliteration, graded into I–IV. Luessenhop-Gennarelli grading of cAVM is based upon the number of directly participating arteries for which there is a standardized nomenclature.

Spetzler-Martin (SM) Grade

A common method of grading cerebral AVMs is the Spetzler-Martin (SM) grade. This system was designed to assess the patient's risk of neurological deficit after open surgical resection (surgical morbidity), based on characteristics of the AVM itself. Based on this system, AVMs may be classified as grades 1 - 5. This system was not intended to characterize risk of hemorrhage.

AVM size	Adjacent eloquent cortex	Draining veins
< 3 cm = 1	Non-eloquent = 0	Superficial only = 0
3 – 6 cm = 2	Eloquent* = 1	Deep veins = 1
> 6 cm = 3		

"Eloquent" is defined as areas within the brain that, if removed will result in loss of sensory processing or

linguistic ability, minor paralysis, or paralysis. These include the sensorimotor cortices, deep cerebellar nuclei, cerebral peduncles, thalamus, hypothalamus, internal capsule, brainstem, and the visual cortex.

The risk of post-surgical neurological deficit (difficulty with language, motor weakness, vision loss) increases with increasing Spetzler-Martin grade.

Supplemented Spetzler-Martin (SM-supp, Lawton-Young) Grade

A limitation of the Spetzler-Martin Grading system is that it does not include the following factors: Patient age, hemorrhage, diffuseness of nidus, and arterial supply. In 2010 a new supplemented Spetzler-Martin system (SM-supp, Lawton-Young) was devised adding these variables to the SM system. Under this new system AVMs are classified from grades 1 - 10. It has since been determined to have greater predictive accuracy that Spetzler-Martin grades alone.

Variable	Spetzler-Martin Grading Scale		Supplemental Grading Scale	
	Definition	Points	Definition	Points
AVM size	< 3 cm	1		
	3 – 6 cm	2		
	> 6 cm	3		
Deep venous drainage	No	0		
	Yes	1		
Eloquence	No	0		
	Yes	1		
SM Grade Subtotal		(1 - 5)		
Age			< 20 years	1
			20 – 40 years	2
			> 40 years	3
Unruptured presentation			No	0
			Yes	1
Diffuse			No	0
			Yes	1
SM-Supp Grade Subtotal				(1 - 5)
SM-Supp Total				(1 - 10)

Nataf grading

In this 4-point grading, angiographic parameters are used for the determinants of the bleeding risk.

- GRADE Ia- 13% hemorrhage
- GRADE Ib- 38% hemorrhage
- GRADE II- 48% hemorrhage
- GRADE III- 90% hemorrhage

Vienna Classification

	Grade 0	Grade I	Grade II	Grade III
Feeders		Pial	Pial+perforator	Perforator
No. Of feeders	1-2	Greater than 2		
Nidus (cm)	Less than 2	2-4	Greater than 4	

The algorithm is similar to that of spetzler and martin for use in arteriovenous malformation surgery, allowing the comparison of surgical and endovascular feasibility.

Conclusion

Although advancements in the last few years have improved our understanding of vascular malformations and haemangiomas, the field of vascular anomalies and their clinical management is still growing. Despite this, doctors may find it difficult to diagnose and treat vascular malformations, particularly given the anxiety and concern they cause for patients and their families. By gaining a clear grasp of the clinical features, diagnostic instruments, and imaging modalities helps us to overcome this.and we recommend several more studies to understand diagnostic and treatment modalities in arteriovenous malformations.

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