Current Concepts in Classification, Diagnosis and Treatment of Vascular malformations

Essay

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List of Abbreviations

AVF	Arteriovenous fistula
AVFs	Arteriovenous fistulae
AVM	Arteriovenous malformation
AVM-LM	Arteriovenous malformation-Lymphatic malformation
AVMs	Arteriovenous malformations
B-FGF	Basic fibroblast growth factor
CLM	Capillary lymphatic malformation
CLVM	Capillary lymphatic venous malformation
CM	Capillary malformation
CMs	Capillary malformations
CM-AVM	Capillary malformation- Arteriovenous malformation
CSF	Cerebrospinal fluid
CT	Computed tomography
CTA	Computed tomographic angiography
CVM	Capillary venous malformation
CVM	Congenital vascular malformation
CVMs	Congenital vascular malformations
CW	Continuous wave
DMSO	Dimethyl sulfoxide
EC	Endothelial cell
EVOH	Ethylene Vinyl Alcohol
FLPDL	Flash Lamp Pulsed Dye Laser
FUGS	Fluoroscopic and ultrasound guided sclerotherapy
GI	Gastrointestinal
GLUT1	Glucose transporter protein 1
HIV	Human immunodeficiency virus

IF-α	Interferon- alfa
IH	Infantile hemangioma
IHs	Infantile hemangiomas
ISSVA	International Society for the Study of Vascular Anomalies
KHE	Kaposiform hemangioendothelioma
KHz	Kilo Hertz
KMP	Kassabach-Meritt phenomenon
KTP	Potassium-Titanyl-Phosphate
KTS	Klipple-Trenaunay syndrome
LM	Lymphatic malformation
LMs	Lymphatic malformations
LMWH	Low molecular weight heparin
LVM	Lymphatic venous malformation
MRA	Magnetic resonance angiography
MRI	Magnetic resonance imaging
NBCA	N-Butayl cyanoacrylate
Nd:YAG-Laser	Neodymium-Yittrium-Aluminium-Garnet laser
NICH	Non-involuting congenital hemangioma
OKT-3	Picibanil, a killed strain of group A Streptococcus pyogenes; also OK-432
PDL	Pulsed dye laser
PHACES	(P) Posrerior cranial fossa,(H) Hemangioma, (A) Aorta, (C) Cervical arteries, (E) Eye, (S) Sternum
POL	Poidocanol
PVA	Polyvinyl alcohol
PWS	Parkes-Weber syndrome.
RICH	Rapidly involuting congenital hemangioma
STS	Sodium Tetradecyl Sulfate

US	Ultrasound
VA	Vascular anomaly
VEGF	Vascular endothelial growth factor
VEGFR	Vascular endothelial growth factor receptor
VM	Venous malformation
VMs	Venous malformations
VMCM	Familial cutaneous and mucosal venous malformation

CHAPTER (1)

INTRODUCTION & AIM OF THE WORK

Introduction

The treatment of vascular anomalies is a relatively new and rapidly developing discipline that requires a broad interface between several surgical and medical specialities. ***(Kubiena et al, 2007)***

In 1982, *Glowacki and Mulliken* proposed a "biological" classification of vascular anomalies based on clinical behavior, histology, and histochemistry. Following this classification, which has been accepted by the *International Society for the Study of Vascular Anomalies (ISSVA)* in 1996, vascular anomalies are divided into *vascular tumors* (e.g., hemangiomas), in which the etiology is one of endothelial cell proliferation, and *vascular malformations*, in which a developmental error has caused abnormally formed vascular channels. ***(Kubiena et al, 2007)***

Vascular malformations are subdivided based on the channel type and on flow characteristics. This classification has helped to resolve the confusion regarding terminology in the field of vascular anomalies. ***(Kubiena et al, 2007)***

Congenital vascular malformations (CVMs), including arteriovenous malformations (AVMs), remain sophisticated despite efforts during the last century to improve their care. They have a wide range of clinical presentation and an unpredictable course. Complicated anatomical, pathological, physiological, embryonic and hemodynamic characteristics must be evaluated. High morbidity has been related to both surgical and nonsurgical treatments. There has been an associated high recurrence rate. ***(Lee et al, 2004)***

Among CVMs, AVMs have been particularly confusing because of their unpredictable nature. AVMs behave aggressively as a primitive type of CVMs because the majority belongs to the *extratruncular form* as the residual remnants of a developmental arrest in the early stage of embryonic life. They have a tendency to progress with a more destructive potency. ***(Lee et al, 2004)***

The primary effect of an AVM on the surrounding tissues is by the lesion itself, with compression and erosion. Secondary hemodynamic effects include a potential arterial steal phenomenon. The heart can be affected by high-output cardiac failure. Peripheral tissues can be affected in a wide range of changes from distal ischemia to gangrene, venous stasis dermatitis, and ulcer or gangrene caused by venous hypertension. ***(Lee et al, 2004)***

AVM, especially its infiltrating extratruncular form, has a high recurrence rate because of its origin from the mesenchymal cells at an early stage of embryogenesis. It retains the evolutional potential to grow, which is often represented clinically as a recurrence. Its behavior, therefore, is totally unpredictable, often responding to various stimulations such as injury or surgical intervention, as well as a systemic hormone effect. The result can be explosive growth. Improper treatment often stimulates dormant AVM to grow rapidly, making the condition worse. This recurrence and unbridled growth are the trademarks of AVM. ***(Lee et al, 2004)***

Complete eradication of the nidus of an AVM is the only potential cure. But this, however, is often difficult if not impossible. Radical resection to remove the lesion completely has been described as

"demolishing surgery". It is often accompanied by excessive blood loss in addition to serious complications. Thus, incomplete removal of the AVM is a frequent result of attempts to avoid the high morbidity associated with total excision. ***(Lee et al, 2004)***

New diagnostic technology, including less invasive imaging, has aided the differential diagnosis of CVM to provide a more precise diagnosis of AVM developed in different stages of embryogenesis. Contemporary diagnosis, based on the Hamburg classification, provides an opportunity for implementation of the new concept of a multidisciplinary team approach for managing AVM. This approach is based on a new classification scheme and diagnostic technology. ***(Lee et al, 2004)***

Embolo/sclerotherapy is a new therapeutic modality that is accepted as independent therapy, especially for surgically inaccessible lesions. It has also been implemented as preoperative or postoperative adjunct therapy. It has helped improve surgical results and to expand the role of surgical therapy. ***(Lee et al, 2004)***

Aim of the work

The principal goal of this review is to provide the up-to-date data on classification, diagnosis and management of congenital vascular malformations which are considered one of the most difficult to treat pathologies which needs multiple specialities to treat.

CHAPTER (2)

CLASSIFICATION