Review article

Venous malformation and haemangioma: differential diagnosis, diagnosis, natural history and consequences

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Abstract

Venous malformation (VM) is the most common form of congenital vascular malformation (CVM). VM presents at birth as an inborn vascular defect and never disappears/regresses spontaneously through the rest of life; it will continue to grow slowly at a rate that is proportional to the growth rate of the body. Haemangioma is *not* a vascular malformation but one of the vascular tumours originating from the endothelial cells; it develops after birth mostly in the infantile/neonatal period with a distinctive growth cycle: a proliferation phase of early rapid growth followed by an involutional phase of slow regression. Although the vascular malformation and vascular tumour belong to the 'vascular anomaly' together, both conditions are fundamentally different not only in their anatomical, histological and pathophysiological findings but also in their clinical courses. Therefore, an appropriate differential diagnosis of the VM is mandated not only from other kinds of CVMs but also from 'genuine' haemangioma. Appropriate diagnosis and assessment of VMs can be made based on clinical presentation and a proper combination of basic non-invasive studies in general but the presence of a mixed lesion involving other types of CVM lesions and the type of VM lesion, extratruncular and truncular, will dictate the need for further work-up with additional non- to less-invasive study or angiography. Otherwise, angiography is usually reserved for therapeutic planning and treatment.

Q1 **Keywords:**

Introduction

Venous malformation (VM) is one of the congenital vascular malformations (CVMs) known as an enigma of modern medicine through the century.[1–4] Such a notorious reputation was partly due to so many name-based eponyms (e.g. Klippel Trenaunay Syndrome, Parkes–Weber Syndrome, Sturge–Weber Syndrome) attempting to define such a complex vascular condition will only add to the confusion.

Name-based eponyms were used in the past to define various vascular malformations but due to the absence of a standardized classification system they often misled with regard to the underlying defect in these inborn errors.[5–8] For example, in the early 20th century many physicians including two French physicians, Klippel and Trenaunay, reported a unique condition of CVMs characterized by a syndrome of skin, soft tissue, bone and blood vessel lesions.[9]

With limited anatomical and pathophysiological knowledge due to the absence of appropriate diagnostic studies (e.g. angiography) then, these conditions were classified solely based on clinical findings and named after the describing physician (e.g. Servelle and Martorell syndrome).[10]

Although these name-based eponyms have been used extensively for many decades to classify various CVMs, these eponyms failed to provide essential information regarding the aetiology, anatomy and pathophysiology of these complex vascular conditions.[11,12]

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Nevertheless, each syndrome has important historical significance providing a clinical description of the primary vascular lesion along with its secondary non-vascular lesions and their clinical findings (soft tissue swelling, long bone growth discrepancy, etc.). These eponyms are still useful for certain combined forms of vascular malformations, although its role has been largely replaced by the newer classifications: ISSVA Classification[1,13,14] and (modified) Hamburg Classification.[6,7,12,15,16]

ISSVA Classification classified the CVMs as one of two 'Vascular Anomaly' components together with Vascular Tumour which is represented by the neonatal or infantile haemangioma,[17,18] and the CVMs were further classified based on the haemodynamic/flow status (Table 1). The Hamburg Classification classified all the CVMs based on its predominant type (e.g. venous, lymphatic and arterio-venous [AV] malformation) first and made a further subclassification of the CVMs based on the embryological characteristics of each malformation (Table 2).

These two abnormal vascular conditions: CVMs and vascular tumours/haemangiomas, however, are fundamentally different, not only in their anatomical, histological and pathophysiological findings, but also in their clinical courses, which emphasize the importance of a precise understanding of these two different conditions.

Therefore, proper understanding of the CVMs as a whole is mandated for VM diagnosis together with its differential diagnosis from haemangioma.

Definition and classification of CVM

CVM is an inborn vascular defect as the outcome of developmental arrest during embryogenesis[19–22] and presents at birth as various conditions of malformed vessels; it continues to grow at a rate that is proportional to the growth rate of the body regardless of its type.

Table 1 ISSVA* Classification of vascular anomalies, 1996, Rome, Italy

Tumours	Vascular malformation	
	Simple	Combined
Haemangioma	Capillary malformation (CM)	CVM
Other tumours	Lymphatic malformation(LM)	CLVM
	Venous malformation (VM)	LVM
	Arterio-venous malformation (AVM)	CAVM
		CLAVM

*ISSVA, International Society for the Study of Vascular Anomalies

Table 2 The modified Hamburg classification of congenital vascular malformations

Primary classification*
Arterial malformations
Venous malformations
Arteriovenous malformations
Lymphatic malformations
Capillary malformations
Combined vascular malformations
Anatomical/Embryological subclassification†
Extratruncular forms
Diffuse, infiltrating
Limited, localized
Truncular forms
Obstruction or narrowing
Aplasia; Hypoplasia; Hyperplasia
Obstruction due to atresia or membranous occlusion
Stenosis due to coarctation, spur or membrane
Dilation
Localized (aneurysm)
Diffuse (ectasias)

*Based on predominant vascular structure in the malformation
†Based on anatomy and developmental arrest at different stages of embryonal life: extratruncular form from earlier stages (no named vessels); truncular form from late stage (major named vessels)

VM is one of the most common CVMs, where defective development is limited to the venous system (Figures 1a and c).

Infantile or neonatal haemangioma, on the other hand, is a vascular tumour that originates from endothelial cells, and has no relationship with defective development. It usually appears in the early neonatal period. It has a distinctive growth cycle characterized by a proliferation phase of early rapid growth followed by an involutional phase of slow regression. Unlike CVMs, haemangiomas undergo self-limited growth followed by a subsequent involution that usually occurs before the age of 5–10 years in the majority of cases[13,14,17,18] (Figures 2a and d).

The terms 'capillary or cavernous haemangioma', which were originally used to define the location of infantile or neonatal haemangioma, has been erroneously used to describe VM lesions. However, the terms 'capillary or cavernous haemangioma' should no longer be used in order to avoid confusion between (a genuine) haemangioma and vascular malformation and prevent an incorrect diagnosis.[11,12]

'Angiodysplasia' is another popular older term used to describe CVMs but this term also has limited scope and utility, adding significant confusion to the classification so that it should be replaced with a correct term 'Congenital Vascular Malformation.'

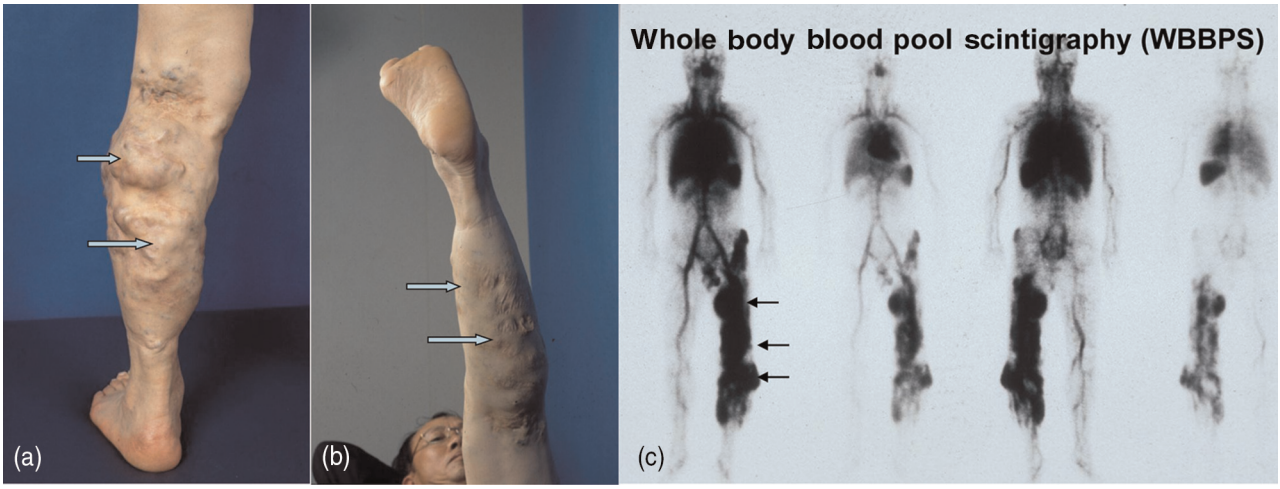

Figure 1 Panel a presents a venous malformation (VM) affecting the entire left lower extremity with significant symptoms; it is a diffuse infiltrating 'extratruncular' lesion possessing mesenchymal cell characteristics of steady growth. Panel b shows spontaneous collapse of the VM lesion when the leg is elevated (a), typical of a pure VM, is a significant clue to support its nature as predominantly VM in contrast to other kinds of congenital vascular malformations. Panel c illustrates positive evidence of abnormal blood pooling along the lesion by WBBPS (whole body blood scintigraphy) to confirm the lesion as VM (from Lee *et al.*[12])

The Hamburg Classification classified the CVMs into five different types based on its predominant vascular malformation component: arterial malformation (AM),[23,24] VM,[1–4] AV-shunting malformation (AVM),[25,26] lymphatic malformation (LM)[27,28] and combined vascular malformation. The most common combined (or mixed) malformation is haemolymphatic malformation (HLM)[29,30] (Table 2).

The initial consensus, however, did not include capillary malformation (CM) in the classification since the lesion was believed to lack clinical significance and clinically was not equivalent to other CVMs. However, the CM was later included in the classification at the Denver consensus meeting in 1992[8] and at the Seoul consensus meeting in 1996.[31,32]

Each vascular malformation is further subclassified into one of two forms, based on its embryological development stage, as either an Extratruncular or a Truncular form (Table 2). The clinical behaviour of every vascular malformation is dependent on its unique embryological characteristics, specifically, the stage of embryogenesis at which developmental arrest occurs. This results in a wide range of clinical presentations, unpredictable clinical course and erratic response to treatment with the potential for high rates of recurrence among the extratruncular lesions.[33,34]

The 'extratruncular form' of CVM lesions arises when developmental arrest occurs in the 'earlier stage' of embryonic life while the vascular system is in the reticular stage. Extratruncular lesions are embryonic tissue remnants of mesodermal origin that retain the characteristics of the mesenchymal cells (angioblasts). It retains its potential to grow and proliferate when stimulated internally (e.g. menarche, pregnancy and hormone) or externally (e.g. trauma, surgery).[20–23,35–37] These lesions, therefore, carry a significant risk of recurrence, especially after suboptimal treatment.

Extratruncular lesions often present as either a (diffuse) infiltrating lesion or a limited lesion causing mechanical compression to surrounding tissues and organs in addition to their haemodynamic impact.

The 'truncular form' of CVM lesions arises when developmental arrest occurs during the vascular trunk formation period in the 'later stage' of embryonic development.[20–23,35–37] Truncular lesions have lost the embryonic characteristics of the mesenchymal cells along with the potential to grow and proliferate. Thus, these lesions carry minimal risk of recurrence.

Truncular lesions, however, are associated with more serious haemodynamic consequences related to the type of CVM (e.g. marginal and embryonic veins as truncular VM). These lesions often present as a persistent fetal remnant (truncal) vessel[38–40] that failed to involute normally (e.g. sciatic vein), or as a defective vessel trunk (e.g. vein web, venous aneurysm).[41–44]

The truncular lesion is further subgrouped as an obstructive or a dilated lesion. However, all truncular lesions are involved in a 'formed' vessel with various degrees of defective development (e.g. agenesis/rudimentary deep vein) ranging from incomplete or immature lesions (aplasia

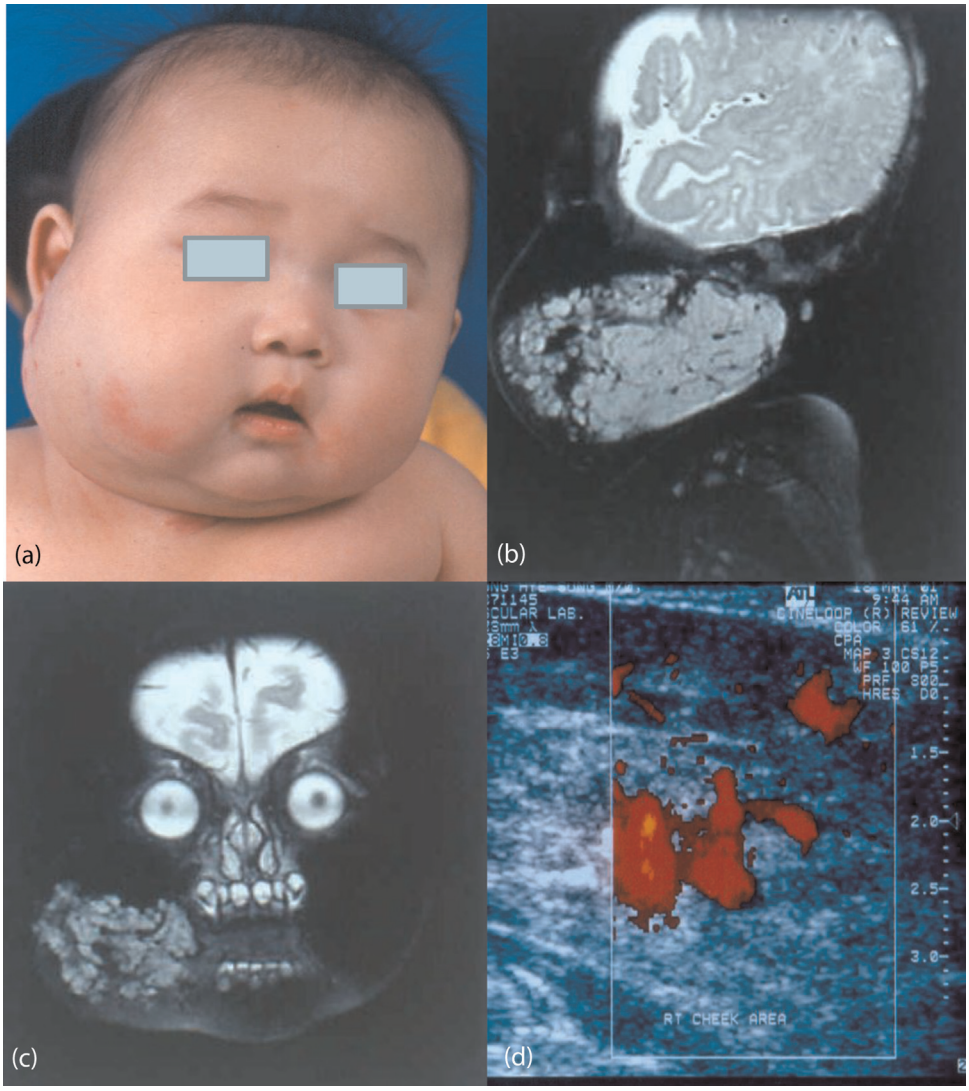

Figure 2 Panel depicts clinical condition of (infantile) haemangioma as rapidly expanding lesion along the right cheek, first noted a few months after birth, which may be mistaken for a venous malformation (VM). Panels b and c show typical magnetic resonance imaging findings of (infantile) haemangioma, affecting right cheek with prominent vascularity that helps differentiate it from common VM. Panel d presents duplex ultrasonographic findings of active/high blood flow through the lesion in contrast to what would be found in a VM lesion (from Lee *et al.*[12])

or hypoplasia) to overdeveloped lesions (hyperplasia).

The Hamburg Classification provides clinicians with the most critical information regarding this embryological status of various vascular malformations. Embryological subclassification into one of two different subtypes, truncular or extratruncular, allows clinicians to predict the clinical course, response to treatment and likelihood of recurrence.[1,12]

The ISSVA classification system based on the haemodynamic status of the CVM was introduced by Mulliken *et al.*[13,14] Using the traditional name based eponyms, CVMs are further classified as 'fast flow' and 'slow flow' lesions. This flow-based classification is useful from a clinical management point of view but remains confusing and cumbersome due to its combination with various traditional name-based eponyms; the continued use of name-based nosology and syndrome-based classification allows further confusion with current classification systems. Besides, the AVMs are classified to 'AV malformation and AV fistula' despite malformation in 'fistulous' condition to allow AV shunting. Currently, many specialists across the Atlantic have adopted parts of this classification system and have combined it with the Hamburg Classification.

Diagnosis of CVMs

A new concept on the CVM mandated a precise diagnosis to provide accurate information on its histo-pathological, haemodynamic and embryological characteristics; this new information in turn allowed a new prospect to its management with a new view.

Now we know there are many different vascular malformations with different clinical significances and we no longer consider them as an enigma since we have enough knowledge to verify many different aspects of each CVM lesion either existing alone or as combined with other CVMs.[1,12]

Many newly developed tests, mostly non-invasive, can now provide precise diagnosis of each CVM to confirm the clinical impression.[45,46]

The diagnosis *per se* is now feasible only with limited combinations of a few non- to less-invasive tests in the majority of CVMs: duplex ultrasonography,[47,48] magnetic resonance imaging (MRI),[49,50] whole body blood pool scintigraphy (WBBPS),[51,52] transarterial lung perfusion scintigraphy (TLPS)[53,54] and radionuclide lymphoscintigraphy,[55–58] etc. (Table 3).

The invasive tests (e.g. arteriography, phlebography) in general are seldom needed for the diagnosis *per se*, and could be saved till needed as a road map for the treatment except differential diagnosis with haemangioma or AVM.[1,46]

Precise defining of the type (e.g. VM, LM, HLM and AVM) and nature (e.g. truncular or extratruncular lesion) of each CVM involved should follow a detailed assessment of its extent, severity and its secondary impact on the related systems/organs, which is also now feasible with non- to less-invasive tests alone in its majority.[12,46,59]

Table 3 Diagnostic tests for venous malformation

I. Non- to less-invasive studies
Magnetic resonance–T2-weighted image (MRI) study
Duplex ultrasonography
Whole body blood pool scintigraphy (WBBPS): transvenous angioscan utilizing radioisotope-tagged red blood cells
Transarterial lung perfusion scintigraphy (TLPS)*: transarterial angioscan utilizing radioisotope-tagged microsphere albumin
Air plethysmography
MR venography (MRV)and/or MR arteriography (MRA)*
Computerized tomographic (CT) study with contrast and 3D reconstruction
Radioisotope (RI) lymphoscintigraphy*
Ultrasound lymphangiography*
MR lymphangiography*
II. Selective invasive tudies
Ascending, descending and/or segmental venography
Percutaneous direct puncture phlebography
Standard and/or selective arteriography*
Percutaneous direct puncture lymphangiography*

*Optional for differential diagnosis of AVM and LM

Differential diagnosis with haemangioma

True haemangioma is a vascular tumour belonging to the vascular anomaly together with vascular malformation; the female to male ratio is 3 to 5:1. Haemangioma is usually not present at birth and generally appears suddenly during the early neonatal period as a rapidly growing tumour. However, haemangioma has a distinctive pattern of self-limited growth as previously mentioned, with initial rapid growth during the early proliferate phase followed by slow regression through the long involution phase. It is generally resolved spontaneously with minimum morbidity before reaching the age of 7–9 years.[13,14,17,18,60,61]

In contrast, a CVM lesion is always present at birth (even though initially it may not be apparent) and generally distinctive on birth as an inborn error; they all steadily grow commensurably in proportion to general/systemic body growth unless complicated (e.g. bleeding, infection); it never disappears nor regresses. It carries an equal gender distribution.

The haemanigoma occurs in the range of 2–3% among newborns and then steadily increases to about 10% by the end of the first year of life. However, the incidence of CVM is reported to be 1.2% by Tasnadi *et al.*[62] This is higher than other congenital malformations (e.g. congenital hip dislocation and dysplasia).

Therefore, when a vascular anomaly is first identified during the neonatal period, the differential diagnosis should include both CVM and haemangioma, which is often specified as either neonatal or infantile haemangioma (Figure 2).

In most cases, a careful history and physical examination provide enough data to clinically distinguish a vascular malformation and haemangioma. Distinction between the two diagnoses is often made using additional non-invasive laboratory studies (e.g. duplex ultrasonography). These studies are useful especially for a deeply seated haemangioma mimicking VM. Tissue biopsy is useful in situations where the D.D. includes sarcoma.[1]

Differential diagnosis with other CVMs

Once haemangioma is excluded and a diagnosis of CVM is confirmed, further characterization of the

type of CVM is the next step.[1,12] The VM remains the most common type of CVM. VM lesions occur as a pure VM or as a mixed lesion combined with other types of CVMs which include CM, LM and AVM as mentioned above.[12,46,63]

The majority of VM lesions are pure/predominant VM lesions with no other circulation systems involved. However, approximately 15–20% of VMs are mixed lesions, and most frequently combined with LM and CM, often known as Klippel–Trenaunay Syndrome (KTS)[64,65] (Figure 3). When the VM lesion is further combined with AVM besides LM and CM, this mixed lesion is often called Parkes-Weber Syndrome (PWS).[66,67]

The presence of an occult AVM, therefore, must be determined when the VM presents as a mixed type in view of the virulent nature of AVMs. AVM lesions compared with VM lesions are more destructive, exhibit unpredictable behaviour and carry high morbidity. TLPS is a study useful for diagnosing and screening for the presence of an AVM. This study also assesses the extent of shunting of cardiac output through the AVM lesion when located in an extremity.[53,54]

Diagnosing the presence of an LM[68,69] is not as critical as it is with an AVM for the HLM cases. However, the presence of an LM lesion cannot be always determined by MRI alone; LM lesions are sometimes indistinguishable from VM lesions

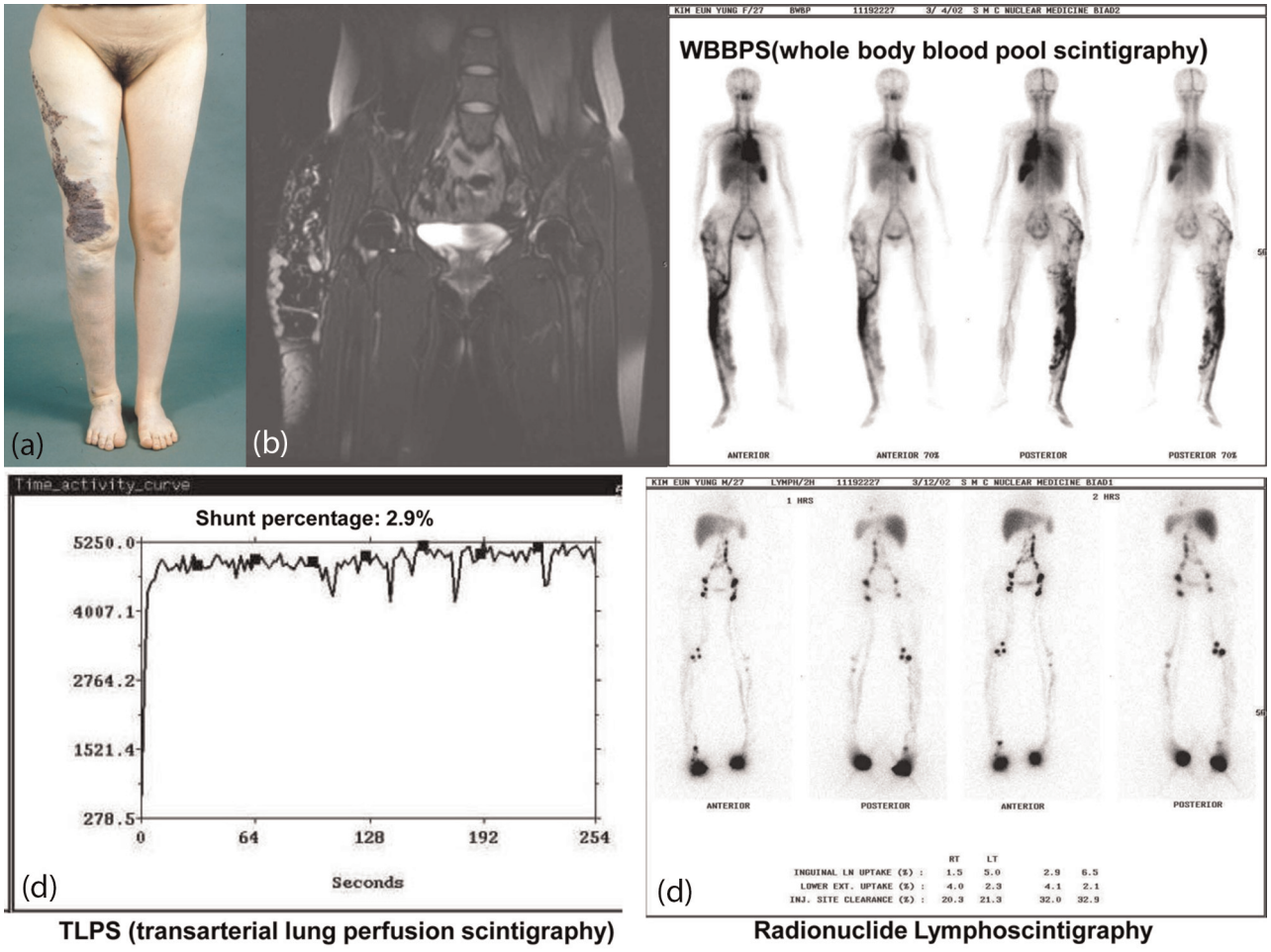

Figure 3 Panel a presents clinical appearance of VM (venous malformation) lesion affecting the right lower extremity as a haemolymphatic malformation, mixed with lymphatic malformation and capillary malformation, often known as Klippel–Trenaunay syndrome. Panel b depicts magnetic resonance imaging finding of extratruncular VM lesion diffusely infiltrating in the soft tissue and muscles of right lower extremity. Panel c shows WBBPS (whole body blood pool scintigraphy) findings of massive abnormal blood pool throughout entire right lower extremity; this WBBPS effectively ruled out any additional lesions throughout the body. Panel D illustrates transarterial lung perfusion scintigraphy study that is negative for abnormal AV shunting – 2.9% is within normal range. Panel e delineates radionuclide lymphoscintigraphy findings of anatomically normal, but functionally abnormal double (deep and superficial) lymphatic transporting vessel, visualized along right lower extremity. This finding is consistent with a clinical finding of chronic lymphoedema secondary to hypoplasia of the superficial lymphatic system which is well compensated by the deep system (from Lee *et al.*[46])

even on MRI. However, T2-weighted MRI imaging remains the gold standard in diagnosis of VM lesions (5). VM lesions are readily detected on WBBPS (Figures 2 and 3)

WBBPS utilizes radioisotope-tagged red blood cells to detect abnormal blood pooling in VM lesions (with blood volumes as little as 1.0 mL) throughout the body when present.[51,52] LM lesions are not detected with WBBPS. Thus, the combination of T2-weighted MRI imaging and WBBPS will allow accurate diagnosis of a VM lesion combined with an LM lesion.

Percutaneous aspiration (using ultrasound guidance or direct puncture) and fluid analysis of a mixed VM and LM lesion is also useful in determining the nature of the lesion.

Diagnosis of VM

After the diagnosis of a VM lesion is made, further identification of its subtype as extratruncular or truncular is required in order to identify its embrylogical and haemodynamic characteristics. Both extratruncular and truncular VM lesions have distinctively different characteristics regarding its natural history and clinical behaviour.[20–23, 35,36,37]

Extratruncular VM lesions

These lesions naturally carry a high risk of recurrence if not treated radically on the contrary to truncular lesions due to their unique evolutional power to grow when the condition should meet as mentioned previously.

Extratruncular lesions are further subdivided into diffuse, infiltrating type and localized, limited type. Diffuse, infiltrating extratruncular lesions may cause symptoms due to compression of the surrounding structures (bones, muscles and nerves). They have a notorious reputation by namely pain due to embryological proximity of nerves and veins. They may also produce significant haemodynamic impact on the involved venous system that is dependent on lesion size and location.

Truncular VM lesions

These lesions (e.g. popliteal vein aneurysm or ectasia, femoral vein hypoplasia and iliac vein agenesis) no longer exhibit their embryonic characteristics and lack the potential for proliferation on the contrary to extratruncular lesions.

However, truncular lesions have haemodynamic consequences due to congenital valvular incompetence, obstruction (atresia, hypoplasia) or dilation/aneurysm formation with associated risk of thromboembolism (e.g. marginal vein [MV]).

Truncular lesions can be subdivided into aplasia or hypoplasia,[41,70,71] as well as obstruction, or dilation/aneurysms.[44,72,73] Immature/incomplete/abnormal development of the main axial veins result in aplasia, hypoplasia or hyperplasia of the vessel (e.g. agenesis/rudimentary femoral vein) or as a defective vessel: obstruction (e.g. vein web, spur, annulus or septum) or dilation (e.g. popliteal or iliac vein ectasia/aneurysm).

One of the unique truncular VM lesions is persistent embryonic veins such as the MV or the sciatic vein when a fetal (truncal) vessel fails to undergo normal involution.[38–40, 74]

Truncular lesions of obstructive nature (webs, hypoplasia) may have different haemodynamic impacts on their relevant venous systems depending upon their location, extent/severity and natural compensation through collaterals. Chronic venous insufficiency develops in the territory drained by the truncular vein. Stenosing truncular lesions produce venous obstruction leading to a reduction in venous drainage. Membranous obstruction of the (suprahepatic) inferior vena cava in primary Budd–Chiari syndrome is an example of a primary obstructive VM affecting a major vein.[42,43]

Truncular VM lesions may also occur in veins with the same embryological origin or draining the same territory (e.g. stenosing lesions of the extracranial jugular veins, superior vena cava, and azygos vein system along the main outflow pathways of the cerebro-spinal venous system).[75,76]

Avalvulia, or absence of valves, is another form of hypoplasia that produces venous reflux. Together with atresia of the venous trunks and venous aneurysms, they are relatively common. The incidence of aneurysm has been reported to be 4% in nearly 490 cases of congenital anomalies of the venous system.[41,77,78]

Diagnostic evaluation of VM

Clinical evaluation

The diagnosis of a VM, both extratruncular and truncular subtypes, can often be made with a careful history and physical examination alone. Therefore, proper clinical evaluation of patients with VMs is essential. A thorough history, including a detailed birth and family history, must be taken.

VM lesions occur throughout all areas of the body, and therefore complete examination of all skin surfaces including the genitalia is essential. Patients should also be examined in the standing position and posture and gait evaluated to assess the dynamic impact of the VM lesion. Limb length discrepancy (with and without pelvic tilt) and compensatory scoliosis will probably not be missed on examination with this approach.[79–81]

The physical examination should include careful assessment (inspection, palpation and auscultation) of both the arterial and venous systems including a detailed pulse exam, making a note of any oedema, skin changes, varicosities, pigmentation or ulcerations. An enlarged or longer extremity, digital anomalies and asymmetric growths of any part of the body must be recorded.

The 'limb elevation test' should also be performed for all extremity VM lesions. In this test, the VM lesion bearing limb demonstrates a reduction in both limb and lesion swelling with leg elevation (Figure 2).

Furthermore, initial baseline evaluation should include an assessment of VM lesion associated acute complications (e.g. superficial and deep vein thrombosis [DVT], pulmonary embolism [PE]) and chronic complications and sequelae (e.g. limping and scoliosis with pelvic tilt). An accurate record of these findings is useful for regular periodic follow-up after treatment.[1,34,46]

Assessment of direct and/or indirect secondary effects of the primary VM lesion on various organ systems (e.g. bone, soft tissue and muscles) should be performed using comprehensive clinical evaluation and appropriate laboratory studies especially in situations where the VM lesion exists as an HLM lesion. Appropriate assessment of the involved organ systems such as the gastrointestinal system (e.g. GI bleeding and malabsorption syndrome), cardiopulmonary system (e.g. pleural effusion), musculoskeletal system (e.g. long bone length discrepancy, scoliosis and pelvis tilt) and genitourinary system (e.g. lymph leak) will require involvement of additional surgical specialties.

Abnormal long bone growth as a consequence of intraosseous and extraosseous VM lesions involvement should be evaluated routinely with appropriate radiological studies.[79–81]

Confirmation of the presence of a VM lesion is usually made with a combination of various non-invasive to minimally invasive studies which include Duplex ultrasonography, MRI and WBBPS as described above. The minimally invasive studies are generally adequate to diagnose and assess the extent and severity of the VM lesion prior to treatment (Table 3).

Non-invasive tests

Duplex scanning is the first diagnostic test of choice for non-invasive evaluation of all patients with VMs, involving the limbs, to assess the deep and superficial veins, to identify any aberrant vein, obstruction, dilation or valvular incompetence and define the feeding artery or draining veins of the VM (Table 3).[47,48,82,83] Q6

Other non-invasive studies, such as plethysmography, segmental pressure measurement and pulse volume recordings should be used selectively and clinical correlations with abnormal findings (e.g. outflow obstruction) need to be established.[1]

Standard plain X-ray is still useful to identify abnormal findings in the soft tissue, and other malformation-related abnormalities along the skeletal system. It is also useful to detect the phlebolith to confirm the diagnosis of VM.

Minimally invasive tests

Computed tomography with intravenous contrast[1,84,85]

Computerized tomographic venography is recommended for evaluation of obstructed veins and other truncular anomalies of large veins in the chest, abdomen or pelvis (Table 3). Computed tomography accurately identifies the underlying pathology, confirms venous obstruction or extrinsic compression and delineates anatomical variations and extent of venous thrombosis.[86]

MRI and MR angiography[49,50,87,88]

MR imaging and MR venography are recommended for evaluation of VMs. The test is reliable; it confirms the extent and type of the VM, delineates feeding and draining vessels, distinguishes between different soft tissues (muscle, fat) and the vascular structures. The imaging modality is highly accurate in diagnosis of DVT.[86]

WBBPS: transvenous angioscan utilizing radioisotope-tagged red blood cells[51,52,89,90]

WBBPS is an optional test to screen for multiple VM lesions scattered throughout the body. It allows qualitative and quantitative evaluation of the VM lesion especially during the course of multisession sclerotherapy as a cost-effective measure. It is an excellent tool for routine follow-up and to assess the progress of treatment and the natural course of the VM lesion. It can exclude a combined

VM–LM where the absence of an abnormal blood pool over the lymphatic lesion is the typical finding.

TLPS: transarterial angioscan utilizing radioisotope-tagged microsphere albumin[53,54]

TLPS is not indicated for evaluation of the VM lesion. Its major function is to rule out the presence of a combined AVM lesion. TLPS can detect micro-shunting of an AVM lesion which can often be missed on conventional arteriography.

Invasive tests

Selective invasive study (e.g. arteriography and phlebography) is often not needed for routine diagnosis of the VM lesions as mentioned previously and can be reserved for therapeutic planning and treatment (Table 3). Arteriography and venography may be required in the setting of a mixed VM lesion to determine and assess the nature of the CVM components. This is especially true for a mixed VM lesion combined with an AVM.[8]

However, invasive tests may be required for diagnosis when non- to minimally invasive tests (e.g. computerized tomographic and/or MRI) fail to confirm the diagnosis or to delineate important diagnostic details.

For example, an obstructive truncular VM lesion along the iliac vein often needs more precise anatomical information. Ascending phlebography combined with IVUS studies is essential for proper management. Descending phlebography is an indispensable tool to assess deep venous reflux along the pelvic veins and/or sciatic veins.[91–93]

Direct puncture phlebography is also very useful to identify a large efferent vein of extratruncular lesions.

In the paediatric age group less than two years old, only minimum necessary diagnostic procedures (usually non-invasive studies) are indicated in general to confirm the lesions as the VMs and rule out the AVM lesion. Complete assessment with angiography is often not required and if needed, can often be delayed until the child is older or fully grown. Angiography is required for urgent and immediate treatment in the rare occasion when a VM lesion is located in a life-threatening area which compromises normal functions (e.g. seeing, breathing, hearing, and eating) or is limb threatening.

Diagnosis of VM-coagulopathy

A potential risk of coagulopathy beyond pain has been largely neglected despite new evidence which shows this pain/ache along the VM lesion is closely linked to 'intravascular coagulation phenomenon' together with the phleboliths. The pain is a further warning sign for increased risk of more serious complication (e.g. venous thromboembolism) and subsequent morbidity to cause extensive damage throughout its natural course.[94–97]

Among the many types of truncular VM lesions, the lateral embryonic or MV in particular accompanies quite high risk of DVT and PE, which is often fatal.[38–40,74] Together with its counterpart, extratruncular VM lesions, MV causes various degrees of pain and thrombosis within a lesion resulting in PE besides severe bleeding especially during surgical procedures.

A 'localized intravascular coagulopathy'[98,99] is the most common form of coagulopathy involved in the VM so that proper understanding on this coagulation issue remains essential for diagnosis and management of VM.

Coagulation profile and D-dimer levels are particularly indicated for extensive VM lesions.[100–103]The following laboratory studies form an essential part of the patient's work-up:

- D-dimer-quantitative assay;
- Fibrinogen;
- Platelet count;
- PT, APTT;
- Thrombophilia screening for high-risk malformations (e.g. lesions involving the orbit) (in the presence of thrombophilia and depending on the risk of the specific procedure, adequate anticoagulation should be provided).

Coagulation studies are not routinely included in the evaluation of extratruncular VM lesions. However, scattered phleboliths combined with extensive VM lesions are generally indicated for a full coagulation study especially when the treatment is planned. The finding of consumptive thrombocytopaenia and leucopaenia would need further coagulation study to decide anticoagulation therapy.

Conclusion

VM lesions often occur as mixed lesions involving other types of CVM lesions. The presence of a mixed lesion and the type of lesion present will dictate the need for further work-up and non-invasive study or angiography.

Appropriate diagnosis and assessment of VMs can be made based on clinical presentation and

non-invasive investigations. Angiography is usually reserved for therapeutic planning and treatment.

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