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Venous malformations imaging: About 2 cases

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Abstract

Venous malformations include a spectrum of slow-flow vascular malformations that together are the most common forms of vascular anomalies. Their age of onset is variable. They are often asymptomatic, but they can become painful because of thrombophlebitis, articular or muscular involvement. Radiology plays a two-fold role: Making the diagnosis by a variety of imaging modalities and performing interventional radiology techniques to treat this condition.

We report 2 cases of venous malformations in a 9 year-old girl and a 4 year-old boy, both are superficial and localized in the left superior limb. Both of them appeared at birth, and pain was the main reason for consultation. Diagnosis was made with doppler US and CT angiography.

Keywords

Vascular malformations; Simple venous malformations; Pediatric; Thrombophlebitis.

Introduction

Congenital venous malformations (VMs) are the most common vascular malformation. They are characterized as slow-flow vascular malformations, and have an estimated incidence of 1–5 in 10,000 births and a prevalence of 1% [1,2]. More than 40% of VMs occur in the head and neck, approximately 40% occur in the extremities, and the remaining 20% occur in the trunk [3].

Doppler US is in most cases the first line imaging modality. However, MRI remains the gold standard for the exact diagnosis of VMs. CT is generally reserved for limited indications.

Therefore, the role of the radiologist in the management of these patients is two-fold: making the diagnosis with the use of ultrasound and magnetic resonance imaging, and performing sclerotherapy, which is the treatment of choice if conservative treatment fail to manage symptoms.

We report 2 cases of venous malformations in a 9 year-old girl and a 4 year-old boy, both are superficial and localized in the left superior limb.

Case Presentations

Case I

- A 9 year old girl with no previous medical history were referred to our department for a doppler US of the left superior limb after complaining of pain in her elbow pit.

- The interrogation revealed that the parents noticed a bluish, palpable compressible and slow growing mass in the elbow pit at the age of 18 months. It became painful 15 days before the consultation.

- The examination revealed a bluish elastic soft tissue swelling on the left elbow pit, non-pulsative, painful at compression.

- Doppler US of the superior left limb revealed a formation in the elbow pit containing multiple serpiginous structures taking color encoding, and presenting a venous spectrum on pulsed Doppler, some of which present intraluminal echogenic material; these structures are fed by a collateral of the cephalic vein. We also noticed the presence of shadowing phleboliths (Figure 1).

- In the absence of MRI, a CT Angiography of the superior left limb was performed and revealed a roughly rounded formation at the level of the external part of the elbow measuring 39.2 mm x 35.6 mm, surrounded by a pseudo capsule, and which contains serpiginous formations, not taking contrast at the arterial time, some channels are opacified at the portal time and seem to be fed by a collateral of the cephalic vein at the venous time, there is the presence of a calcification of 2 mm in diameter corresponding most probably to a phlebolith (Figures 2, 3, 4).

- Partially thrombosed venous malformation was taken as diagnosis

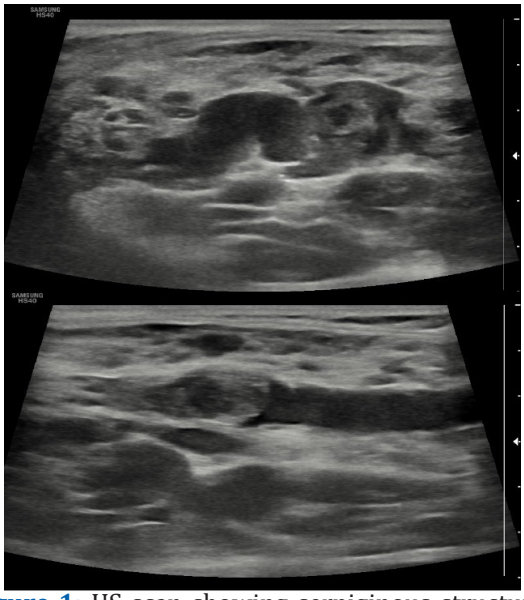

Figure 1: US scan showing serpiginous structures with intraluminal echogenic material.

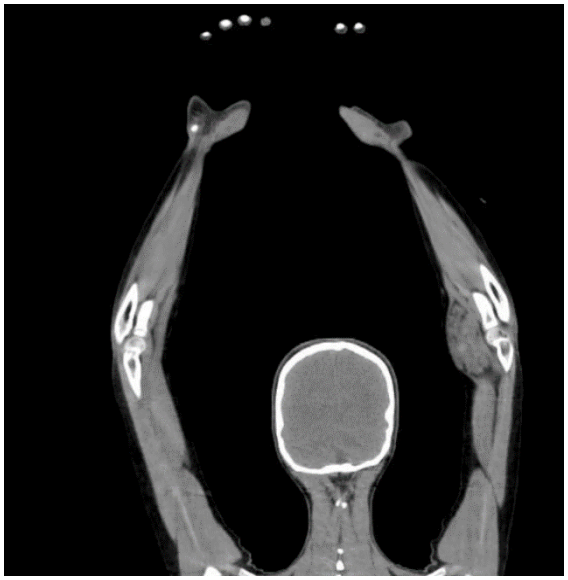

Figure 2: Coronal plane in a non-enhanced CT showing a well limited mass containing serpiginous structures in the left elbow.

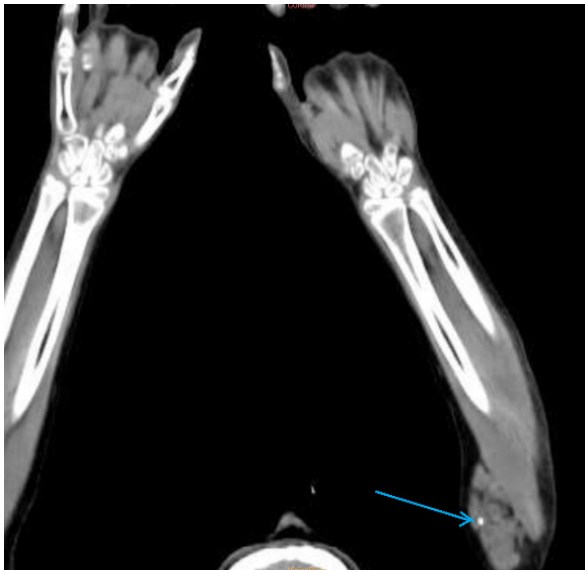

Figure 3: Coronal non enhanced CT show a calcification in the left elbow mass compatible with a phlebolith (blue arrow).

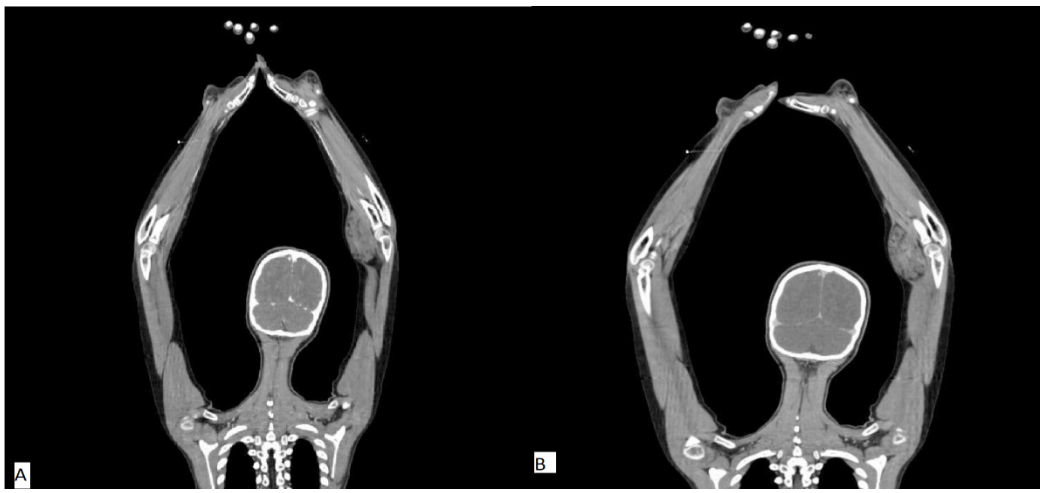

Figure 4: (A): Coronal plane in a contrast enhanced CT in the arterial phase showing no enhancement of the left elbow mass.
(B): Coronal plane showing a discrete enhancement of the mass in the venous phase.

Case II

- A 5 year old boy with no previous medical history were referred to our department for a doppler US of the left superior limb after complaining of pain in his elbow pit and in his on the lower 1/3 of his forearm.

- The interrogation revealed that the parents noticed at birth a bluish, palpable compressible and slow growing mass in the left elbow pit and in the lower 1/3 of the left forearm. It became painful 7 days before the consultation.

- The examination revealed a bluish soft tissue swelling on the left left elbow pit and in the left forearm, non-pulsative, painful at compression.

- Ultrasound exploration of the clinical swelling at the level of the lateral part of the left elbow revealed a formation of multiple serpiginous structures taking color encoding at the Doppler, and presenting a venous spectrum at the pulsed Doppler, a few of which present intraluminal echogenic material; these structures are fed by a collateral of the cephalic vein (Figure 5).

Ultrasound exploration of the clinical swelling at the level of the ⅓ inf of the left forearm objectifies a formation containing multiple serpiginous structures taking color encoding on Doppler, and presenting a venous spectrum on pulsed Doppler; all compressible, a few of which contain phleboliths, these structures are fed by a collateral of the cephalic vein of the forearm (Figure 6).

- Partially thrombosed venous malformation was taken as diagnosis

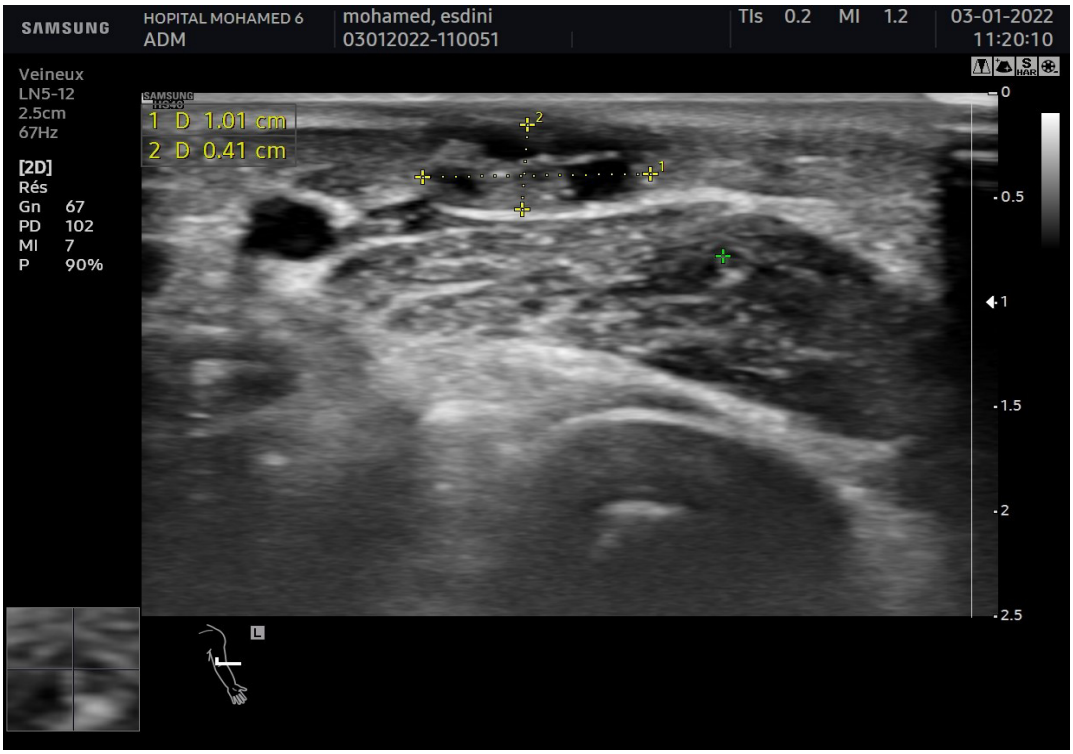

Figure 5: US image show a round mass containing serpiginous structures in the left elbow.

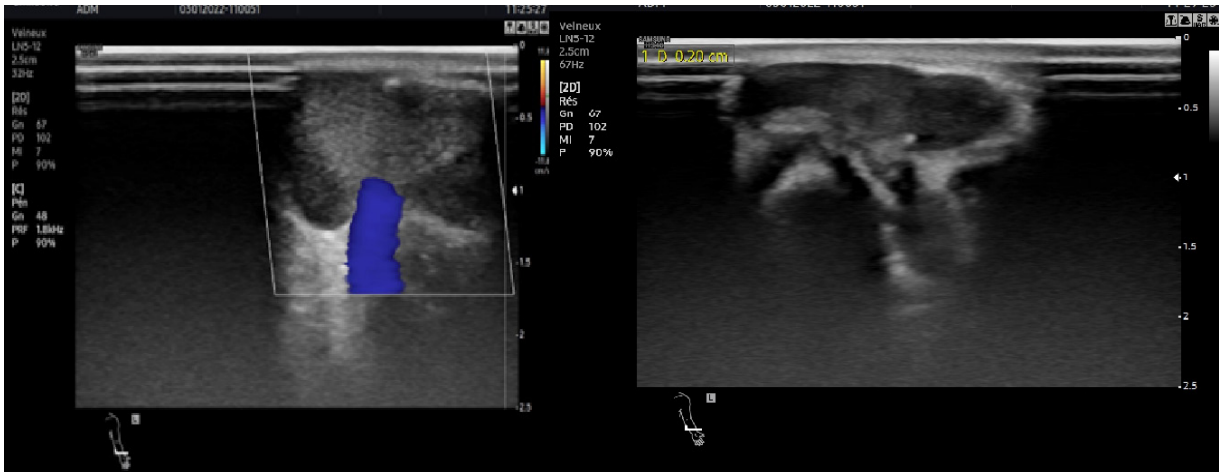

Figure 6: US scan showing large serpiginous venous structure in the left forearm, with echogenic intraluminal material.

Vascular Tumors			Vascular Malformations		
Benign Tumors	Borderline Tumors	Malignant Tumors	Simple	Combined	Associated with Other Anomalies
- Infantile Hemangioma - Endothelial Cell Proliferation - GLUT-1 Marker positive	Hemangio-endothelioma	Angio-sarcoma	Venous Malformation (VM) Blue rubber bleb nevus Syndrome Glomovenous malformation	CM + VM CM + LM CM + AVM LM + VM	Klippel-Trénaunay-Syndrome
Congenital Hemangioma Excessive Angiogenesis with Capillary Lobules GLUT-1 Marker negative Fully developed at birth	Others	Others	Lymphatic Malformation (LM) Macrocystic Microcystic Mixed Cystic	CM + LM + VM CM + LM + AVM CM + VM + AVM	CLOVES Syndrome
Tufted Angioma			Capillary Malformation (CM) Teleangiectasia Nevus Simplex Others	CM + LM + VM + AVM	Sturge-Weber-Syndrome
Spindle Cell Hemangioma			- Arterio-Venous Malformation (AVM) -		Parkes-Weber-Syndrome
Epitheloid Cell Hemangioma Others			Arterio-Venous Fistula (AVF) Hereditary Hemorrhagic Teleangiectasia (HHT)		Others

Abbreviations: CM, capillary malformation; VM, venous malformation; LM, lymphatic malformation; AVM, arterio-venous malformation.

Figure 7: ISSVA classification of vascular anomalies [5].

Discussion

- The International Society for the Study of Vascular Anomalies (ISVA) has developed a widely accepted categorization system for vascular anomalies (Figure 7), which are divided into two basic categories: vascular tumors and vascular malformations. Simple malformations (composed primarily of one type of vessel, such as arterial, venous, capillary, or lymphatic), combined malformations (comprising more than one type of vessel), malformations of major named vessels, and vascular malformations associated with other anomalies are all types of vascular malformations [1].

In both our cases, the vascular malformations were classified as simple venous malformations.

– Simple venous malformations (VMs) are vascular abnormalities that are made up of aberrant veins that can be localized or diffuse. Slow-flow vascular malformations are the most common type, with an estimated incidence of 1–5 per 10,000 births and a prevalence of 1% [1,2]. VMs can be superficial, infiltrating the dermis and subcutaneous tissues, or they can be deeper, infiltrating muscle and bone. The head and neck localisations account for more than 40% of VMs, with the extremities accounting for another 40% and the trunk accounting for the remaining 20% [3].

– In our cases, the VMs were both superficial and occurred in the superior left limb.

– Superficial VMs are bluish in colour, non-pulsative, show no local increase in temperature, are compressible and typically increase in size during Valsalva manoeuvre , and grows in proportion to the child [2,3]. Venous malformations are often asymptomatic, but they may become painful in case of : 1) compression of local structures including intraosseous, intraarticular, and nervous involvement, 2) superimposed infection, and 3) venous stasis resulting in localized intravascular coagulopathy (LIC) [2,4].

Localized intra vascular coagulopathy (LIC) is due to stasis and abnormal venous endothelium. Coagulation abnormalities are widespread, and they are often related to the size of the VM. Many patients

with extensive venous malformations have been found to have elevated D-dimer levels. D-dimer is a sensitive marker for thrombus development and fibrinolysis, and it can be utilized as a negative predictive marker to rule out a VM [5]. Risk factors for the presence of LIC include lesions with large surface area or volume and the presence of phleboliths.

In both our cases, both VMs were bluish and became painful because thrombophlebitis.

– Histologically, venous malformations are composed of thin walled, dilated, sponge-like abnormal channels of variable size and thickness.

– No biopsy were made in our cases.

– Imaging plays [5] an important front-line role in the evaluation of vascular malformations not only for confirmation of diagnosis but for evaluation of the extent of disease.

For many patients with venous malformations, ultrasound (US) may be the first-line imaging modality. The appearance might range from a focally dilated vein (comparable to adult varicosities) to a fluid-filled cavitary lesion with septations, which is more prevalent [6]. The presence of echogenic debris or shadowing phleboliths is extremely specific for venous malformation diagnosis. On real-time US imaging, the cystic areas are compressible, and Doppler interrogation reveals little to no flow. Doppler sonography can help distinguish low-flow lesions like venous malformations from higher-flow lesions like arteriovenous malformations or other soft tissue malignancies by providing additional information. Doppler flow may be increased by Valsalva motions and compression. Arterial flow from neighbouring arteries can be visible within and around the VM. Ultrasound is an excellent screening exam because of its inexpensive cost, absence of ionizing radiation exposure, and lack of procedural anesthesia. Ultrasound, on the other hand, has several limitations, such as difficulty visualizing large, invasive, and/or deeper venous abnormalities and determining connections to surrounding tissues.

In both cases, diagnosis were made by doppler US after revealing multiple serpiginous structures taking color encoding, and presenting a venous spectrum on pulsed Doppler, some of which present intraluminal echogenic material; these structures are fed by a collateral of the cephalic vein. We also noticed the presence of shadowing phleboliths.

– Computed tomography (CT) is generally reserved for patients with suspected osseous involvement with their venous malformation. On CT, venous abnormalities usually have low attenuation and delayed diffuse enhancement. Phleboliths and b one alterations such as cortical thinning and focal bone deterioration associated to the venous malformation are better seen with CT [1].

CT angiography had been performed for our 9 year-old patient and revealed a delayed enhancement of the malformation with the presence of phleboliths with no osseous involvement.

– Magnetic resonance imaging (MRI) [5] remains the diagnostic imaging gold-standard for venous malformations [7]. Studies have shown the sensitivity and specificity to be 98.9% and 90%, respectively.

MRI allows for a more accurate assessment of the disease's size and spatial resolution, as well as the link between the venous malformation and surrounding anatomic tissues [12]. Venous malformation typically show a high T2 signal with intra-lesional fluid-fluid levels. Decreased T2 signal can be seen with area of hemorrhage, thrombosis, and/or phleboliths. With contrast enhancement, there is characteristic delayed heterogeneous enhancement. Flow-sensitive sequences will assist detect nearby veins and venous outflow, as well as rule out an arterial component to the malformation. Unlike US, MRI may require coordination with procedural sedation, especially for extensive scans or for young children [13]. MRI is also a useful tool to evaluate the joint for patients who have venous malformations involving the extremities. It is important to establish if there is an intra-articular component to the venous malformation [12]. Intra-articular involvement can carry a risk of hemarthrosis, thrombosis, and thinning of the cartilage.

– Treatment of patients with VMs rely firstly on a medical management, which include compression garments, anti-inflammatory medication, and low molecular weight heparin [9].

If medical management fails, sclerotherapy is often considered first-line treatment.

Sclerotherapy aims to induce the vessel's fibrosis due to obliteration of the lumen [14]. The endothelial layer of a vessel is encountered first by the sclerosant. Sclerosing solutions vary widely in their chemical nature and in their ability to inflict damage to the endothelial and the deeper muscular and connective tissue layers [14].

Common sclerosants for venous malformations include dehydrated ethanol and detergent sclerosant such as sodium tetradecyl sulfate (STS), polidocanol, sodium morrhuate, and ethanolamine [9].

Sclerotherapy involves a radiological guided cannulation of the venous malformation, typically using a 20- or 22-guage angiocatheter. After the venous malformation has been cannulated, contrast is injected to opacify the venous malformation and assess for any venous outflow or contrast leakage into surrounding structures such as the skin, soft tissues, and muscle. Large outflow veins are particularly concerning because they can result in not only venous thromboembolism, pulmonary embolism, and systemic sclerosant distribution, but also difficulty achieving the desired sclerosis of the malformation due to escaped sclerosant [9,15,16]. These risks can be mitigated by judicious application of a tourniquet during the procedure [9,15]. Contrast leakage into surrounding tissues and skin increases the likelihood of skin breakdown and ulceration [9,15].

The use of compression garment following sclerotherapy is mandatory for long-term success of the sclerotherapy procedure.

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Conclusion

The radiologist has an important role in the diagnosis and therapy of VMs. Doppler US is generally the first imaging modality to be performed if VMs are suspected. However, in order to make a precise diagnosis and to have a complete mapping of the lesions, MRI remains the Gold standard tool. Interventional radiology techniques are now considered first line alternative tools to manage VMs if conservative management fails.

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