How much of this peripheral venous malformation is sclerosed? Our experience in discordance between imaging findings and clinical results in children

Poster No.: C-1420
Congress: ECR 2015
Type: Educational Exhibit
Keywords: Interventional vascular, Soft tissues / Skin, Paediatric, Ultrasound-Colour Doppler, MR, Ultrasound, Sclerosis, Education and training
DOI: 10.1594/ecr2015/C-1420

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Learning objectives

To review the clinical and imaging features of venous malformations in children.

To describe the changes found in ultrasound and magnetic resonance imaging (MRI) after percutaneous sclerosing treatment of peripheral venous malformations and to explain our experience in the follow up of these patients after sclerosis.
Background

Venous malformations (VMs) are the most common vascular malformations, with a prevalence of around 1% [1]. 40% of VM are located in the head and neck, many involving mucosal surfaces, and another 40% are located in the extremities [2].

VMs are present at birth although they may not be noticed until later in childhood. They are very variable in shape, size and tissue involvement, but they are all composed of dysplastic vessels with no cellular proliferation. They are classified as low-flow vascular malformations.

Depending on their location and size, they are frequently bluish in color in the overlying skin, soft and compressible and typically expand with Valsalva maneuver and dependent positioning.

They can manifest clinically as pain, swelling and functional limitations, mainly those involving deep cutaneous or intramuscular tissues. Other complaints may include cosmetic disfiguration.

Ultrasound should always be the first modality performed in a peripheral VM due to its advantages such as availability, low cost and lack of radiation. It can also differentiate between a high-flow and low-flow malformation, and if the latter, between a VM and lymphatic malformation. It should be performed with a high-frequency linear transducer (5-15 MHz).

Imaging features in ultrasound are very variable. VMs appear nearly always (98%) heterogeneous [3], usually (80%) hypoechoic [4] (Fig. 1 on page 5A), with anechoic channels visible in less than 50% cases (Fig. 1 on page 5B). Around 10% present as hyperechoic masses [4] (Fig. 1 on page 5C). Some VMs manifest only as a thickening of the subcutaneous tissues without a defined solid mass or anechoic channels visible. Doppler flow in VMs is low and monophasic (Fig. 1 on page 5D), and absence of demonstration of flow is frequent due to equipment limitations, and then flow is only discernible with compression and release of the lesion.

Phleboliths are pathognomonic of VMs but they are only seen in 16% of cases [4]. They are the result of thrombosis and calcification. In ultrasound they appear as hyperechogenic foci with acoustic shadowing, and they are easily depicted in conventional radiography or magnetic resonance imaging (MRI) (Fig. 2 on page 5).
Ultrasound may evaluate the full extension of superficial lesions, but MRI is the modality of choice to delineate the whole extension when affection of deeper tissues is suspected. MRI also allows flow assessment with dynamic contrast-enhanced sequences. The main disadvantage of MRI in children is the need of cooperation, which might imply the use of anesthesia.

The basic MRI protocol include T1-weighted and T2-weighted short tau inversion recovery (STIR) sequences, in at least two planes. Gadolinium enhanced sequences are also mandatory.

VMs are usually heterogeneous masses on all sequences, though lesions measuring under 2 cm tend to be homogeneous. They are predominantly hypo or isointense at T1W images and marked hyperintense in T2W. Areas of hypointensity in T2 can be the result of thrombosis, presence of phleboliths, or fibrous septa (Fig. 3 on page 6). MRI findings are sometimes not very specific and they must be correlated with ultrasound and clinical features. In cases of atypical findings, diagnostic phlebography with iodinated contrast medium can be performed. It should also be realized before interventional procedures as percutaneous sclerosis, in order to evaluate the volume of contrast needed to fill the malformation and therefore determine the amount of sclerosant to be injected (Fig. 4 on page 7).

Symptomatic VMs can be treated with percutaneous injection of sclerosing agents, in one or multiple therapeutic sessions. In our centre, we mainly use polidocanol 3% mixed with air in order to create a foam (Tessari’s method) and increase the time of contact of the product with the endothelium of the VMs, and reduce the washout. Under ultrasound-guided control, a 23G butterfly or 22G spinal needle is inserted in the VM and a phlebography is performed to evaluate extension and venous drainage (Fig. 5 on page 7). In VMs located in extremities, a pressure tourniquet can be used to occlude venous return and prevent rapid outflow of the sclerosing agent into the draining veins. Injection of the sclerosant is guided by ultrasound and fluoroscopy (Fig. 6 on page 8).
Fig. 1: Different features in ultrasound of VMs in different locations. 1A. Heterogeneous hypoechoic subcutaneous mass in vulva, the most typical appearance of VMs. 1B. Hypoechoic vascular channels are sometimes visible, as in this VM located in the plantar region. 1C. Sometimes, VMs appear as ill-defined hyperechoic masses like this subcutaneous lesion in the ankle. 1D. Monophasic low flow demonstrated in VM in thigh.

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Fig. 2: Phleboliths. 2A. Hyperechoic focus in the VM (arrow), with evident acoustic shadowing consistent with phlebolith. 2B. X-ray shows a rounded and well defined calcification (arrow). 2C. Gradient echo T2W sequence of the VM depicts two markedly hypointense foci of phleboliths (arrows).

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Fig. 3: 4-years-old boy with subcutaneous and intramuscular VM in the right calf. 3A,C. A lobulated and well defined mass of intermediate signal in T1w is shown. 3B. The mass demonstrates hyperintensity in STIR, with areas of focal hypointensity due to
phlebolith (arrow). 3D. After gadolinium administration, the VM shows heterogeneous enhancement.

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Fig. 4: 5-years-old girl with a superficial VM affecting the thumb pad. 4A. Digital subtraction phlebography performed by direct puncture of the VM, depicts its venous drainage. 4B,C. T1 W and contrast enhanced MRI of the lesion shows absence of osseous or tendon infiltration, and presence of intense enhancement.

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Fig. 5: Material for sclerotherapy. Puncture of the VM is performed with a 23G butterfly or 22G spinal needles of different lengths for deeper locations. After administration of contrast for phlebography, introduction of the sclerosant foam is performed. The foam is created by mixing a variable proportion of polidocanol and air (1:2-4) through a three way stopcock.

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Fig. 6: Digital substraction phlebography and ultrasound guided sclerotherapy in an intramuscular VM in the thigh. 6A. First, contrast enhanced phlebography is performed in part of the VM. 6B. Sclerosant foam is administrated, shown as a negative contrast because it contains air. The previously administered contrast is pushed and delineates some small draining veins (arrow). 6C,D. Phlebography and sclerosis of an independent part of the VM is performed. The previous needle and sclerosed part is depicted and substracted (asterisks). 6E. Heterogeneous hypoechoic VM is shown (open arrows). 6F. During introduction of the foam, we can see how the air fills part of the VM (open arrows), leaving deeper parts inaccessible to ultrasound.

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Findings and procedure details

After sclerotherapy, a delay of several weeks is necessary to evaluate therapeutic response, allowing time for the inflammatory changes to resolve. It is recommended that a control with MRI should not be performed before 6 months after treatment [2].

In case of good response, a progressive shrinkage of the VM should be observed.

In ultrasound, besides the decrease in size (which in big VMs is sometimes difficult to assess), diffuse heterogeneous hyperechogenicity is observed in almost all cases. We should find less hypoechoic channels visible, if present, and lower compressibility of the VM treated (Fig. 7 on page 11).

In MRI, sclerosed VMs demonstrate heterogeneous signal intensity in T1 and STIR, sometimes with more hyperintensity in T1 and hypointensity in STIR. Thick septa and more enhancement of the lesion compared with the non treated parts are frequently seen (Fig. 8 on page 11, Fig. 9 on page 12).

Most VMs need few or many sclerotherapy procedures for a satisfactory treatment. Imaging controls between procedures can be performed with ultrasound or MRI. Considering our patients are children and the need for anesthesia in many of them, we usually perform ultrasound for control, with the disadvantage of the operator dependency. We evaluate by MRI for control mainly when we are not having good results in partial or total relief of the clinical symptoms. This statement produces a considerable warp in comparing previous and post-procedure MRI in our patients, as we get more MRI studies in those "difficult" cases, often with disappointing results.

On the other hand, some VM like facial superficial lesions or small cavitary VMs, are probably those in which we obtain better response. As a result of that, these patients are not usually controlled by imaging but clinically (complaints are often only aesthetic), and we frequently do not get feedback in imaging of the treatment success if only one session is needed.

As far as we can confidently evaluate the size and changes in ultrasound, we try to avoid the use of MRI for smaller children who need general anesthesia. We reserve MRI for the diagnosis, and control of MVs with little results.
As it has already been published [5, 6, 7], spongy patterns of VMs are more difficult to treat, especially when they are intramuscular. In these cases, we have found greater changes in ultrasound than in MRI, regardless there might be no clinical relief (Fig. 10 on page 13).

The most important fact to consider, over imaging changes of decrease in size, heterogeneity in signal or echogenicity, or greater enhancement, is the clinical results. On the other hand from the unfortunate cases like the above mentioned, we have found surprising imaging results in patients who affirm feeling less discomfort, with little change in MRI but more evident changes in ultrasound (Fig. 11 on page 14).

It is difficult to evaluate treatment effect in clinical parameters, because patient satisfaction is subjective and can be easily under or overestimated by their parents or even their doctor. We have found that objective decrease in size on MR imaging or ultrasound is less common than subjective clinical improvement in symptoms. We believe that in many cases a minimal reduction or changes in echogenicity might indicate a change in the physiologic characteristics of the VM that implies a clinical improvement.

We think that MRI is useful for a first diagnostic evaluation and to determinate the extension of a VM, but it is not required to show treatment benefit.

Our experience and opinion need replication in a study to investigate long term clinical outcome and imaging changes in these patients treated with sclerotherapy.

It is important to consider that a multidisciplinary approach is needed for an appropriate management of these patients, including pediatric radiologist, pediatricians, plastic surgeons and dermatologists.
Fig. 7: Changes in ultrasound after treatment. Superficial VM in the chest wall of a 3-years-old boy. 7A,B. VM previous to sclerotherapy. A hypoechoic round nodular mass is visualized affecting subcutaneous tissue and muscle. Filling of Doppler color with decompression is noted. 7C. After one treatment with polidocanol, the mass is smaller and more heterogeneous, with areas of hyperechoic tissue and areas of hypoechoic channels of the VM showing permeable parts of the VM. 7D. After a second treatment, the VM is yet smaller, non-compressible and predominantly hyperechoic.

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Fig. 8: 16-years-old girl with an small subcutaneous VM in the right shoulder. 8A-C. MRI prior to sclerotherapy. A small subcutaneous mass above the clavicle is noted (arrows), of intermediate signal in T1, markedly hyperintense in STIR and with heterogeneous nodular enhancement after gadolinium administration. 8D-F. Six months after treatment, the VM has decreased in size, with diffuse enhancement.

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Fig. 9: 9-years-old girl with an extensive VM affecting vastus medialis and lateralis muscles. 9A-C Pre-treatment images. An isointense in T1 (9A) and hyperintense in STIR (9B) lobulated intramuscular mass is noted. After gadolinium administration (9C), only subtle enhancement is demonstrated. 9D-F. MRI post-esclerotherapy. Minimal shrinkage of the malformation is noted, with higher intensity in T1 (9D) and lower in STIR (9E), with thicker septa (arrows). Greater enhancement (9F) is evident.

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Fig. 10: 10-years-old girl with an extensive VM affecting the left gluteus region. 10A-C. Pre-treatment images. A well-defined lobulated subcutaneous and intramuscular mass affecting gluteus muscles, isointense in T1 (10A) and hyperintense in STIR (10B). 10C. Ultrasound shows an ill-defined heterogeneous hypoechoic mass, with extension difficult to assess. 10D-G. Images after various sclerotherapy procedures. 10D,E. After 4 procedures, the mass has slightly decreased in size, is more heterogeneous and thicker septa are depicted. 10F. After 8 procedures, no change from the previous MRI is noticed. 10G. Greater echogenicity of the VM is seen in ultrasound. Unfortunately, our patient did not have clinical symptoms relief.

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Fig. 11: 11-years-old boy with a VM in quadriceps muscle. 11A,B. MRI previous to sclerotherapy shows a big hyperintense in STIR mass with heterogeneous enhancement. 11C. In ultrasound the VM appears hypoechoic with some hyperechoic foci due to presence of phleboliths (arrow). 11D-E. After some procedures of sclerosis, the mass has a minimal shrinkage, with more intense enhancement. 11F. Sclerosed parts of the VM show hyperechogenicity, while some others are still compressible and hypoechoic (11G).

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Conclusion

Shrinkage of the VM is the main change in imaging when there is a good response to sclerotherapy. Apart from the decrease in size, we found more evident changes in ultrasound to differentiate treated parts of the VM from those left untreated/recanalized.

MRI is in any case superior to determine the global extension of the VM, but we think it is not required to show treatment benefit.

We found clinical relief in many patients after sclerotherapy despite discrete changes in imaging.