Low-flow soft-tissue vascular malformations: Keys MRI and CE MRA findings

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Learning objectives

To evaluate the usefulness of MRI and CE MRA in patients with clinical suspicion of vascular malformations. To describe its characteristic findings, and to establish the keys to perform a suitable classification and a correct differential diagnosis.
Background

According with International Society for the Study of Vascular Anomalies Low Flow Vascular Malformations are classified in capillary, venous, lymphatic, capillary-venous and capillary- lymphatic. However, confusion with respect to terminology and imaging guidelines continues to result in improper diagnosis and treatment. Adequate imaging in association with clinical findings is crucial to establishing the correct diagnosis. Based on our experience with 47 patients from February 2008 to January 2009, we will describe the characteristic MR findings of these malformations.

Although the nomenclature of vascular lesions of soft tissue remains complicated, the classification of Muliken and Glowacki is most often used. This classification divides soft tissue vascular lesions into hemangiomas and vascular malformations. Hemangiom mas appear in early infancy, grow rapidly, and undergo involution. However vascular malformations, presumably, are present at birth, increase in proportion to the growth of the child, and do not regress spontaneously. Alternatively, malformations can be categorized as either high or low flow on the basis of hemodynamics flow characteristics. Vascular malformations without arterial components are considered to be low-flow lesions and include primarily venous, lymphatic, and mixed malformations. **Venous malformations** are dysplasias of small and large venous channels associated with a variable amount of hamartomatous stroma. Many venous malformations cause pain. Often patients will suffer from increasing symptoms in late childhood or early adulthood, and rarely regress. **Lymphatics malformations** consist of chylefilled cysts lined with endothelium. The most common locations include the neck and axilla where they are often cystic hygromas.
Imaging findings OR Procedure details

**Venous malformations (VM)** may be superficial (ie, intradermal or subcutaneous) that generally presents with a bluish compressible mass with no palpable thrill or audible bruit (Fig.1) or deep (ie, intramuscular or intraosseous) with no skin discoloration (Fig.2). The appearance of low-flow vascular malformations on MR imaging depends on the composition of lymphatic or venous components. The venous malformation will appear as a collection of serpentine structures separated by septations. This serpentine structures represents slow-flowing blood in the venous channels and appear as high signal intensity on T2 W images and intermediate signal intensity on T1 W images (Figs. 3,4,5,6,7). Phlebolits may be present and appear as round, low-signal intensity lesions on MR imaging (Figs.8,9,10). In cases of hemorrhage or thrombosis, heterogeneous signal intensity can be observed on T1 W images. Gadolinium-enhanced T1W images may show enhancement of the slow-flowing venous channels (Figs.11,12).

Venous vascular malformations have multifocal involvement, a tendency for orientation along long axis of affected extremities, absence of areas of signal voids, a tendency to follow neurovascular distributions, occasional extension into tendon sheats, osseous involvement and associated enlargement of adjacent subcutaneous fat (Figs. 13,14). Contrast-enhanced 3D MR angiography is also adequate to determine the morfoloy and the extent of venous malformation(Fig.15). Recognizing that the lesion is a low-flow vascular malformation is more important than determining whether the lesion is predominantly venous or lymphatic when making treatment decisions, because direct percutaneous sclerotherapy is describe as the treatment of choice for low-flow vascular malformations. Van Rijswijk et al performed dynamic contrast-enhanced MRI in attempt to better differentiate the various categories of vascular malformations.

Treatment depends on the location and extent of the venous malformations. When a lesion is localized (seudotumoral) and accessible, surgical excision results in excellent outcomes (Figs.16,17,18,19,20). More extensive lesions are often not amenable to resection or only partially resectable (Figs.13). To preserve function when vital structures are involved, a series of sclerotherapy procedures like bleomycin or other sclerosant agents is often performed.

**Lymphatic malformations** are benign vascular lesions and appear to arise from embryological disturbances in the development of the lymphatic system. They encompass a wide spectrum of abnormalities, including cystic lymphatic lesions, lymphangiectasis, and lymphedema. Cystic lymphatic malformations can be seen in any anatomic region but are more commonly seen in rich lymphatic areas, such as the head and neck, axilla, mediastinum, groin and retroperitoneum. In some patients, a venous malformation can be seen in association with a cystic lymphatic lesion.
Although like any other vascular malformation they grow slowly with the child, they may increase acutely in size and become symptomatic due to hemorrhage and infection (Fig.21). May be composed of very small cystic component a few millimeters in size (microcystic) or of larger cystic spaces (macrocystic). At MRI they appear as a multiseptated cystic mass that can infiltrate surrounding tissues.

The cysts are typically hypointense in T1 W images and hyperintense in T2 W images and may unilocular or multilocular (Figs.22,23,24). More heterogeneous signal intensity can be seen within cysts with proteinaceous content or prior hemorrhage (Figs.25,26). The cysts do not typically enhance, although the septa, which are vascularized, show contrast material uptake (Fig.24). There may also be enhancement of the venous component in mixed malformation (Figs.27,28). Whereas small lesions are generally amenable to excision with excellent results, large lesions that involve deep structures of the neck, tongue, and mediastinum entail the risk of multiple complications (Fig.21). Sclerotherapy is frequently used as an alternative to surgery for patients with macrocystic disease, and although it is not generally curative, approximately 40% of patients will have amelioration of their symptoms.
Images for this section:

**Fig. 1**

Superficial venous malformation. F 20 yo. Clinical photograph of the right arm shows extensive superficial venous malformation. Note the diffuse bluish discoloration of the skin that suggest a VM (red arrows). Note also the soft tissue lumps (yellow arrows).

**Fig. 0**

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Fig. 2

Deep venous malformation of the right tight. F 7 yo. Asymmetry and pain of the superior and middle part of the right calf. Note no skin discoloration (arrows).

Fig. 0

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Venous malformation. Same patient of fig1. Extensive venous malformation of the right superior limb. SPAIR coronal MRI shows a hyperintense mass (red arrows) with a mixed multi-septated and honeycomb appearance, along the distribution of the muscles.
Venous malformation. Same patient of fig1. Axial T1 W MRI. Red arrows shows the intermediate signal intensity of VM, some of them with seudotumoral aspect (yellow arrows) that correspond to soft tissue lumps of the clinical photograph of fig1.
Venous malformation of the left plantar region. T2 W coronal MRI clearly demonstrates the malformation within the plantar muscles and subcutaneous fat (asterisk). Note the phlebolith (red arrow).

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Venous malformation of the left plantar region. STIR W coronal MRI clearly demonstrates the malformation within the plantar muscles and subcutaneous fat (asterisk). Note the phlebolith (red arrow).

Fig. 6

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Venous malformation of the left plantar region. Contrast enhanced MRA shows the uptake contrast of the VM (arrow)
M 13 yo. Left submaxilar tumor (asterisk). Note also the involvement of the floor of the tongue (black arrow)
Venous malformation of the left side of the neck (asterisks) CT of the neck clearly demonstrate the phleboliths (yellow arrows) but does not clearly show the extension and the limits of the VM.

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Venous malformation of the left side of the neck. Axial T2 W MRI better demonstrates the extension of the VM (asterisks). Yellow arrows show the phleboliths. MRI is superior to CT in demonstrating the extension and limits of the vascular malformations.

Fig. 0

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Fig. 0

Venous malformation of the posterior right thigh. Gadolinium axial Fat Sat T1 W MRI demonstrates partial perfusion of the VM (red arrows).
Venous malformation of the right calf. Gadolinium axial Fat Sat T1 W MRI demonstrates partial perfusion of the VM (red arrows). Asterisks show the non contrast enhanced areas due to non vascularized tissue.
Venous malformation of the right thigh. Same patient of fig.2. T2 W coronal MRI demonstrates a multi-septate high signal intensity lesion (asterisks). Note the orientation of VM along the axis of right lower extremity and within the muscles and the subcutaneous fat.

Fig. 0

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M 13yo with an intramuscular venous malformation in the left arm. STIR axial MRI shows hyperintense lobulated mass (asterisk) and osteolytic bony lesion of the ulna (red arrows). Sometimes we can see this “aggressive” behavior vascular malformations.
Fig. 0

M 13yo with an intramuscular venous malformation in the left arm. Contrast enhanced MRA shows the VM (arrow)
Fig. 0

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Fig. 17

F 4 mo. Localized venous malformation of the right thigh. Axial (left image) and sagittal (right image) T2 W MRI demonstrate an hyperintense localized mass within muscles (asterisks). Yellow arrows shows septan within the VM. Phleboliths (blue arrows).

Fig. 0

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Fig. 18

F 4 mo. Same patient of fig. 12. Soft, painless lump in back of right leg at birth with progressive increase in size (asterisk).

Fig. 0

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Fig. 0

F 4 mo. Same patient of fig.12. Well demarcated hyperintense muscle lesion (asterisks). Phleboliths (yellow arrow) and septa (blue arrow)
Fig. 20

F 4 mo. Same patient of fig. 12. Postsurgical localized venous malformation.

Fig. 0

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Fig. 21

F 4 mo. Diffuse macrocystic lymphatic malformation with increase in size from birth due to hemorrhage into cysts (arrows).

Fig. 0

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Fig. 22

Left posterior mass in the neck without skin discoloration (arrow). Axial T2 W MRI shows homogeneous hyperintense unicocular lymphatic malformation of the left posterior cervical space (asterisk)

Fig. 0

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Right hemifacial mass (white arrows) with retroauricular extension (yellow arrow). Same patient at 2 yo (left image) and at 8 yo (right image). Note increase size of the lesion.
Fig. 24

Same patient of fig. 23. Left image: axial T2 W MRI shows homogeneous hyperintense multilocular lymphatic malformation of the right side of the neck (yellow arrows). Right image: coronal T1 W Fat Sat with gadolinium shows absence of contrast enhancement (red arrow)

Fig. 0

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Fig. 0

Same patient of fig. 21. Axial T1 W MRI of the thoracic region. Large lymphatic malformation with hypeintense lesions that probably represent methemoglobin (asterisks)
Fig. 26

Same patient of fig.21. Axial T2 W MRI of the thoracic region. Large lymphatic malformation. Note fluid-fluid levels. Upper high hypeintense fluid levels probably represent methemoglobin (yellow arrows). Lower fluid areas are darke and probably represent clot or hemosiderin (red arrows)

Fig. 0

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Right retroocular venouslymphatic malformation causing protrusion of the eyeball, orbit deformation and redbluish discoloration of the skin (asterik).
Same patient of fig. 27. Left image: Axial T1 W MRI of the orbit shows an retroocular intra and extraocular mass with isointense tissue in the periphery (yellow asterisks) and hypointense area on the medial side of the orbit (red asterisk). Note the anterosuperior protrusion of the eyeball (arrow). Right image: Axial T1 W MRI with gadolinium shows enhancement of the venous portion of the malformation (light blue arrows) and absence of enhancement of the lymphatic part (pink arrows).
Conclusion

In conclusion diagnosis of vascular malformations is usually based on medical history and physical examination. MRI features venous malformation including high signal intensity on T2 W, intermediate signal intensity on T1 W, presence of phlebolits and contrast enhancement. The presence of multicystic spaces with or without hemorrhage and absence of contrast enhancement (excepting in the septa) are characteristic of lymphatic malformations.

MR imaging can help characterization and diagnosis of the lesion, but its most important role is in the display of the extent and relationship of the lesion to surrounding structures.
References


