Progress toward understanding vascular malformations
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Use of MRI for the Evaluation of Vascular Malformations of the Lower Extremity

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Introduction

With the biological classification of Glowacki and Mulliken it is possible to diagnose congenital vascular anomalies as being either hemangiomas or vascular malformations (1). Nearly 90% of lesions can be differentiated by history and clinical evaluation (2). Vascular malformations are the result of developmental errors of vascular morphogenesis. By definition vascular malformations are always present at birth, but often they present clinically later in life (1,2). They present with a wide range of abnormalities; from small and insignificant capillary nevi to large and hemodynamically important arteriovenous fistulae (2,3). Clinically lesions grow commensurately with the child. Trauma, sepsis or hormonal changes can exacerbate progression of the lesion (1,2). Vascular malformations are anatomically subdivided according to the predominant channel anomaly into either capillary, arterial, venous and lymphatic, or combinations (1-3). They can be further subdivided into high- or low-flow malformations. Any lesion that has an arterial component is considered a high-flow malformation.

Once the diagnosis of a vascular malformation is made it is of paramount importance to define not only the flow characteristics but also the full range of extension since the prognosis and appropriate treatment vary substantially for each type of anomaly. The two most useful non-invasive imaging techniques for assessing vascular malformations are Magnetic Resonance Imaging (MRI) and ultrasonography (4-16). MRI is a non-invasive method and at this moment it is the best single modality to demonstrate detailed information regarding the involved anatomic structures, extent and flow characteristics of vascular malformations (4-6,8,11). This information is vital to plan possible imaged-guided procedures or surgery. Although surgeons involved in treating patients with vascular malformations will have their requested MRI interpreted by a radiologist, it is imperative for these surgeons to also be able to understand and interpret the described MRI features before intervention is anticipated. The aim of this review is to give surgeons involved in treating patients with vascular malformations an opportunity to gain some background on MR images when assessing vascular malformations. Although MRI is a powerful modality for assessing vascular malformations, we will also discuss some of the limitations of MR imaging. We further suggest a diagnostic flow chart based on MRI features designed to help determine the composition of a vascular birthmark when intervention is anticipated. For the more thorough reviews on especially the radiographic features the reader is referred elsewhere (11-14).
### Low-flow vascular malformations

The diagnosis of venous malformations is strongly suspected by clinical characteristics: they are blue and are easily compressible and increase in size when venous pressure increases (2,13). Many venous malformations cause pain. Most of the venous malformations consist of spongy masses of sinusoidal spaces having variable communication with adjacent veins. The most common symptomatic vascular malformations of the extremities are venous (2,11). Venous malformations may further also consist of dysplasias of large and small venous channels. Lymphatic malformations are like venous malformations low-flow lesions. Lymphatic components of the malformation may contain cystic structures of various sizes varying from macrocystic to microcystic (2,11,13,19). Thoracic lesions are usually being macrocystic, and (the more common) cervicofacial lesions microcystic (14). Lymphatic malformations often have a rubbery or cystic component, but unlike venous malformations they cannot be manually compressed. Often the overlying skin contains small lymphatic vesicles or capillary malformations or both (14). Superficial vesicles are sometimes seen on the skin representing extensions of deeper lying lymphatic malformations (2).

Clinical features can often differentiate between high- and low flow lesions, but with ambiguous lesions, low-flow lesions can consistently be distinguished from high-flow lesions on the basis of MR findings (4-14) (Table A). Low-flow lesions are characterized on MRI by a serpentine pattern with internal striations and septations. Pathologically these findings correlate with fibrofatty septa between endothelium-lined vascular channels, or intervening muscle fiber remnants (4). The high signal intensity seen on the long TR/TE spin-echo (SE) sequences, is attributed to stagnant blood flow in these abnormally, dilated venous spaces. The hyperintens meshwork of low-flow malformations is also called “venous lakes”. These features though characteristic, are not pathognomonic of low-flow malformations, also occurring in rare tumors such as angiosarcomas or myxoid tumors (17,18).

| Table A: MRI characteristics of vascular malformations |
|---------------------------------|-----------------|-----------------|
|                                 | Low-flow anomalies                          | High-flow anomalies                         |
| **T1-weighted Image**           | 1. Increased signal intensity when compared to fat and skeletal muscle | 1. Signal voids                |
|                                 |                                               | 2. Small amount of tissue matrix              |
| **T2-weighted Image**           | 1. Decreased signal intensity when compared to fat | 1. Signal voids                |
|                                 | 2. Increased signal intensity when compared with with skeletal muscle | 2. Small amount of tissue matrix              |
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Figure 1:
Suggested decision tree to help determine the composition of vascular birthmarks based on MRI characteristics. * Hemangiomas with a clear history of rapid enlargement followed by involution are not included. ** Capillary malformations have only minor skin thickening on MRI. *** Contrast medium plays an important role in the evaluation of vascular malformations. Some authors suggest giving contrast to all vascular malformations. We only administer a contrast medium when there is a good indication as mentioned in the text i.e. clinical suspicion for a lymphatic malformation.
With administration of a contrast medium, e.g., gadolinium, septal enhancement may be visualized in lymphatic malformations, defining the septations or cysts of macrocystic lymphatic malformations (11). Microcystic lymphatic malformations may appear quite homogeneously in signal intensity. The contents of the lymphatic cyst are not enhanced with the administration of intravenous gadolinium. Fluid levels seen with macrocystic lymphatic malformations suggest hemorrhage or proteinaceous contents (19,20). T2 weighted MRI of venous malformations shows a hyperintense mass, that enhances diffusely with intravenous contrast. Fat suppression on the T2 weighted image is recommended so that venous malformations can be separated from the high signal of subcutaneous fat (4-6). Signal voids in venous malformations on spin-echo sequences, have been explained to possibly be thrombosed vessels, phleboliths, linear, fibrous striations cut in cross section or small AVF’s (4,6,10,11). Contrast administration results in variable enhancement and is important to document veins with extremely slow flow that may not be seen on MR venography (14). The addition of gradient-recall-echo (GRE) sequences in imaging vascular malformations, may show high signal intensity within areas of void on SE images, indicating that these areas are flow related (10,11). A drawback of GRE is that the difference between high-flow and low-flow lesions is not so well described on GRE images as seen on spin-echo sequence (4). A previous, recent X-ray or CT scan may also suffice in diagnosing a phlebolith. The presence of

Figure 2.
A T2-weighted transverse image 5-cm above the ankle joint taken of a female patient with a pulsatile mass located just above her ankle on the dorsal side of the leg. Figure 2(a) is an axial image taken 5 cm above the ankle joint, with the arrow demonstrating a signal void. The high-flow malformation is located primarily in the posterior compartment of the leg. This compartment is largely replaced by fibrofatty tissue/muscle atrophy. Figure 2(b) is a sagittal image demonstrating large signal voids.
A 14-year-old boy was referred with pain in his right upper leg. He had a lateral venous anomaly that was resected four years earlier, but now he has increasing pain in this leg. Figure 3(a) is a T-2 weighted axial image 10-cm above the knee showing a hyperintense image in the subcutaneous tissue, but also infiltrating the muscle and the femur. The white arrow shows a fluid level (in the quadriceps muscles), sometimes seen in a slow-flow malformation. Figure 3(b) is a coronal image indicating the wide area of infiltration. Above the knee some intra-muscular venous convolutes are clearly visible (arrow).

Phleboliths has been described as being pathognomonic of venous malformations (21).

**High-flow vascular malformations**

Most arterio-venous malformations become symptomatic during puberty, except for the extremely high-flow lesions, where cardiac overload may present in infancy (14). Clinical features may include local hyperthermia, pulsations, thrill and a bruit. Extremity high-flow lesions are often associated with overgrowth. Flow voids seen in high-flow lesions are attributed to a “time-of-flight phenomena” or “turbulence-related dephasing” (6). Additional characteristics used to diagnose high-flow lesions are the visible feeding arteries and draining veins (hypertrophic high-flow vessels), the small amount of tissue matrix and absence of venous lakes in comparison to low-flow vascular malformations (4). High-flow lesions are known to have a variety of MRI findings, including some with hyperintense components on T2-weighted imaging sequences, focal accumulations of fatty tissue, hypertrophy of muscle and bone changes which include sclerosis or lytic defects (14). Hemangiomas consistently have high-flow signal voids within the lesion seen on spin-echo sequences during their proliferative phase (5,7). Proliferating-phase hemangiomas are distinguished from high-flow vascular lesions in that the later has no parenchymal component (13,14,18).
Hemangiomas are usually focal, well-marginated soft tissue masses of low T-1 weighted and high T-2 weighted signal intensity. During the involuted phase, hemangiomas have signs of a low-flow lesion (5,8). Involuting hemangiomas retain their specific MRI appearances until they are replaced by fat. Congenital fibrosarcoma is an important differential diagnosis in infant's (11). It is most common in the lower limbs and is also characterized by rapid growth (22). This lesion shows less homogeneous signal characteristics after contrast administration, while hemangiomas show diffuse enhancement with contrast (11). MRI does not distinguish between benign and malignant forms but can differentiate necrosis from solid tissue which can then guide the biopsy to make a definite diagnosis (22). It is best to always perform a biopsy of any atypical lesions when the clinical features, imaging results, or both are equivocal.

Imaging of endothelial lesions

The value of the MRI as initial diagnostic investigation in vascular malformations was already proposed by Pearce et al. in 1988 (23). We do not feel that MRI warrants to be the initial investigation in each patient. If only a diagnosis is needed and no immediate intervention is anticipated like so often in children, then grayscale ultrasonography coupled with color Doppler flow imaging has the advantage of providing a
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Figure 4.
A twenty-year old male presented with a swelling on the medial side of his left upper leg extending down to his knee. Although being visible for years, he only recently developed severe complaints of pain. Figure 5 shows a T-1 weighted spin echo (TR 850 / TE 12) coronal image of a low-flow malformation located subcutaneously with no muscle or intra-articular infiltration.

rapid, relatively inexpensive and non-invasive assessment of the vascular lesions (15,16). If the lesion is clinically a vascular malformation and intervention is anticipated, MRI is our initial diagnostic investigation. In that case it is of paramount importance to have an accurate estimate of infiltration. Although ultrasonography is notoriously operator dependent, other limitations of ultrasonography include the small field of view, restricted depth of penetration, especially with high-frequency transducers; difficulty in depicting flat superficial lesions; and detecting tiny vessels with low flow (15). Ultrasonography does not only have an important role in diagnosing vascular malformations, but some feel it also has an important place in assessing lesions during follow-up and also in interventional radiology (13-16).

MRI is a non-invasive and non-ionizing method, with up-to-date no harmful side effects described (6). T1-weighted images are obtained to delineate the anatomy, while T2-weighted sequences were obtained to demonstrate the flow characteristics/pathology. For effective evaluation sequences are obtained in at least the axial and coronal planes, incorporating both extremities for comparison. Although there is no signal modality providing more information about the flow characteristics and internal structure as MRI, there are some limitations (4,5,7-9,17). MRI is a relatively expensive imaging modality and requires sedation in infants. It is further not possible to accurately visualize capillary malformations with MRI, except for the minor skin thickening (6,9). Sometimes increases in subcutaneous thickness or prominent veins are seen with capillary malformations (11). Although the different MRI characteristics of venous- and lymphatic malformations have been mentioned before, they can occur together. The administration of a contrast medium is important to separate the two components. Although MRI can diagnose high-flow lesions by the presence of dilated feeding arteries and draining veins, it cannot differentiate arteries from veins (4,7,8). If embolization is considered, angiography is necessary to delineate the nidus and supporting vessels in high-flow
### Table 5.2

<table>
<thead>
<tr>
<th>Condition</th>
<th>Fluid/Fixed Levels</th>
<th>Enhancement</th>
<th>Signal</th>
<th>Low Signal Mass</th>
<th>Lymphatic Malformation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Endothelial (microvascular)</td>
<td>No high flow vessels</td>
<td>High signal mass</td>
<td>No or no</td>
<td>No</td>
<td>Venous malformation</td>
</tr>
<tr>
<td>Exophytic</td>
<td>High flow vessels</td>
<td>High signal mass</td>
<td>Inhomogeneous</td>
<td>diffuse or mass (f/e)</td>
<td>Involved hemangioma</td>
</tr>
<tr>
<td>Soft tissue mass</td>
<td>No high-flow vessels</td>
<td>Decreased signal</td>
<td>No</td>
<td>No</td>
<td>Low-flow</td>
</tr>
<tr>
<td>Endothelial</td>
<td>No high-flow vessels</td>
<td>High signal mass</td>
<td>No</td>
<td>No</td>
<td>Low-flow</td>
</tr>
<tr>
<td>Heterogeneous</td>
<td>Abnormal tissue</td>
<td>High signal mass</td>
<td>Decreased</td>
<td>No</td>
<td>Low-flow</td>
</tr>
<tr>
<td>High-flow vessels</td>
<td>High-flow vessels</td>
<td>High signal mass</td>
<td>No</td>
<td>No</td>
<td>Low-flow</td>
</tr>
<tr>
<td>Soft tissue mass</td>
<td>Soft tissue mass</td>
<td>High signal mass</td>
<td>High signal</td>
<td>Uniform intensity</td>
<td>Low signal mass</td>
</tr>
<tr>
<td>Containing</td>
<td>High-flow vessels</td>
<td>High signal mass</td>
<td>No</td>
<td>No</td>
<td>Low-flow</td>
</tr>
<tr>
<td>Diffuse</td>
<td>Low signal mass</td>
<td>High signal mass</td>
<td>Uniform signal</td>
<td>Uniform intensity</td>
<td>Low signal mass</td>
</tr>
<tr>
<td>MRI</td>
<td>Lesion</td>
<td>Gradient echo</td>
<td>T1 weighted</td>
<td>MRI T1 contrast</td>
<td>MRI T2 weighted</td>
</tr>
</tbody>
</table>
lesions (5,24,25). The angiographic characteristics of high-flow lesions are dilated and lengthened afferent arteries, with early opacification of the enlarged efferent veins (21). Angiography entails an invasive intervention requiring use of ionizing radiation and contrast material with the associated sequella (26-29). In low-flow malformations no additional information is obtained with angiography. In the literature the results of MR angiography (MRA) are promising to delineate the nidus, and in the future MRA will undoubtedly have an important place (6,12). MRA can also be used to differentiate between the draining veins and feeding arteries. Currently in our center angiography is still performed more commonly then MRA. Angiography is only performed when a radiological intervention is suspected, and then often as a combination of angiography/embolization. Intracortical invasion of the vascular malformation is not well evaluated on MRI and when intraosseous vascular malformations and secondary bone changes are evaluated, it is better to perform a computed tomography scan (30). A disadvantage of a CT scan is that lesion size is often underestimated, and that it is impossible to determine flow characteristics.

In conclusion the treatment of vascular malformations still remains a great challenge to treat in modern medicine, even to the most experienced clinicians. Attributing to this problem is the extreme rarity of these vascular lesions (2,3). In our unit MRI has become the golden standard when confronted with a vascular malformation that warrants accurate evaluation for possible therapeutic intervention. Although no single imaging technique answers to all our questions, the exact reason for requesting an investigation should determine the choice of investigation requested. We suggest a diagnostic flowchart based on MRI features designed to help determine the composition of a vascular birthmark when intervention is anticipated.

References


