Actas Dermosifiliogr. 2007;98:219-35

REVIEW ARTICLE

Vascular malformations (II). Diagnosis, Pathology, and Treatment

P Redondo
Departamento de Dermatología, Clínica Universitaria de Navarra, Spain

Abstract. Diagnosis of vascular malformations is essentially clinical, based on the evolution and morphology of lesions. A biopsy is rarely needed to evaluate the nature of the vessels. Imaging examinations are necessary to assess the extension of malformations as well as the osteomuscular and visceral compromise. New techniques such as 3D angio-CT scan and angio-MRI improve the diagnosis of some vascular malformations, especially the large combined ones such as Klippel-Trénaunay syndrome, thus limiting the need for invasive procedures. On the other hand, the advances in laser technology, particularly pulsed dye laser for port-wine stains and Nd:YAG laser for superficial venous malformations constitute new alternatives for the management of these patients. Other emergent treatments include microfoam sclerotherapy for venous and combined, slow-flow malformations, and new embolizing materials associated to surgery for arteriovenous malformations. The second part of this review is focused on the complementary diagnosis (imaging exams, pathology and accessory tests) and multidisciplinary and specific treatment based on the different groups.

Key words: angio–CT scan, angio-MRI, pulsed dye laser, microfoam sclerotherapy, embolization, surgery.

MALFORMACIONES VASCULARES (II). DIAGNÓSTICO, HISTOPATOLOGÍA Y TRATAMIENTO

Resumen. El diagnóstico de las malformaciones vasculares es fundamentalmente clínico, y está basado en la evolución y morfología de las lesiones, siendo necesaria en muy raras ocasiones la realización de una biopsia para valorar histológicamente la naturaleza de los vasos. Para delimitar la extensión de las malformaciones, así como el compromiso músculo-esquelético y visceral, son necesarias las pruebas de imagen. La incorporación de nuevas técnicas como la angio-tomografía computarizada (TC) o la angio-resonancia magnética (RM) en 3D agilizan el diagnóstico de algunas malformaciones vasculares, especialmente las combinadas extensas tipo síndrome de Klippel-Trenaunay, limitando la necesidad de procedimientos invasivos. Por otra parte, los avances en tecnología láser, concretamente el láser de colorante pulsado para la mancha en vino de Oporto y el láser de Nd:YAG para las malformaciones venosas superficiales, la escleroterapia con microespuma en las malformaciones venosas y combinadas de bajo flujo, y los nuevos materiales embolizantes asociados con la cirugía en malformaciones arteriovenosas, son terapias emergentes para el seguimiento de los pacientes. La segunda parte de esta revisión está enfocada al diagnóstico complementario (pruebas de imagen, histología y pruebas accesorias) y al tratamiento multidisciplinar y específico según los diferentes grupos.

Palabras clave: angio-TC, angio-RM, láser de colorante pulsado, esclerosante en microespuma, embolización, cirugía.

Diagnostic Imaging

The diagnosis of cutaneous vascular malformations is based on the patient's medical history and a physical examination.[1-4] Imaging studies are used when there is doubt about the nature of the lesion and serve as a complementary tool used to clarify and confirm diagnosis. They also facilitate analysis of the extent of lesions and assessment of the nonvisible component. In certain situations, imaging techniques not only determine the optimum therapeutic approach but also form an integral part of treatment when this involves the application of embolic or sclerosing agents.

Plain radiography has today clearly been surpassed by other imaging techniques and is of only limited value even in demonstrating the degree of bone involvement and the presence of calcifications. Computed tomography (CT) is

Correspondence:
Pedro Redondo
Departamento de Dermatología. Clínica Universitaria de Navarra
Avda. Pío XII, s/n. Pamplona, Spain.
predondo@unav.es

Manuscript accepted for publication February 9, 2007.

a much more sensitive tool for these purposes and provides much more precise anatomic information.

Conventional radiography can be used to reveal the presence of phleboliths in venous malformations and to detect the rare calcifications that occasionally occur in some lymphatic malformations.[5,6] While venous and arteriovenous malformations are not visible as such on a plain radiograph, their repercussions on adjacent bone structures are visible (for example, asymmetric hypertrophy or atrophy, osteoporosis, or lytic lesions).[7-9]

In addition to anatomic information, Doppler ultrasound also provides hemodynamic data, such as the velocity and direction of flow, a contribution of considerable value in both high flow (arteriovenous) and low flow (venous) malformations.[10-12] Ultrasound is a harmless noninvasive technique that does not involve exposure to ionizing radiation. Moreover it is very accessible, economical, and particularly effective in children because it does not require a great deal of cooperation on the part of the patient.

The principal limitation of ultrasound is that it is an operator-dependent technique, making good reproducibility difficult to achieve. Venous malformations are hypoechoic and their appearance is similar to that of cysts, although a Doppler system, unlike the techniques discussed above, will demonstrate venous flow, particularly when compression maneuvers have been carried out. Lymphatic macrocystic malformations appear as hypoechoic or anechoic multiloculated masses with septa of varying thickness. In the case of arteriovenous malformations, the role of Doppler ultrasound is restricted to confirming the vascular nature of the lesion, which will present both arterial and venous waves.

The definition of soft tissues obtained with CT is better than that of conventional plain radiography but far inferior to that obtained with magnetic resonance imaging (MRI). The great advantage of MRI is its excellent demonstration of bone structures and calcifications, while one of the disadvantages of this technique is that it is based on the use of ionizing radiation and that a contrast media is nearly always necessary. These media are associated with risks because of their nephrotoxicity and the adverse reactions they may cause. In addition, sedation is necessary in pediatric patients because image quality is impaired by movement.[13-16]

MRI provides excellent tissue differentiation and this, together with its capacity to acquire images in multiple spatial planes, makes it the best radiologic technique for demonstrating anatomic relationships and studying the tissue adjacent to the vascular malformation. MRI provides both anatomic and hemodynamic data. The presence of a fast or turbulent flow decreases the intensity of the signal, and when the flow is slow or thrombosis is present, the intensity of the signal increases.[17-19] Like ultrasound, MRI does not use ionizing radiation, so that it is of great use not only in diagnosis but also for monitoring disease progression and for post-treatment follow-up. The gadolinium used as a contrast media is extremely safe and does not in general provoke any clinically significant side effects, although in patients with nephropathy a possible pathogenic relationship has been shown with fibrosing dermopathy.

The chief limitation of MRI is that it requires cooperation on the part of the patient and sedation is necessary in claustrophobic patients and children. The use of scanners with open architecture can reduce this problem, although these devices usually have a lower magnetic field strength, a characteristic that affects the quality of the image and the scan duration when multiple sequences are required.

MRI is the technique of choice for initial assessment of venous malformations because it is noninvasive and can define the complete extension of the lesion on various anatomic planes.[20,21] These malformations produce a lower signal intensity than adjacent fatty tissue in T1-weighted sequences and a higher signal intensity in T2-weighted images.

In the case of lymphatic malformations, MRI can enhance the contrast between the lesion and adjacent tissue and delineate this border anatomically.[20,21] In T1-weighted sequences, the intensity of the signal is similar or slightly lower than that produced by muscle, while in T2-weighted sequences there is a marked increase in the signal, which is of even higher intensity than that of adjacent fat and muscle.

Phlebography is indicated in the case of low-flow or venous malformations.[22] Phlebography by direct puncture of the malformation complemented by the use of tourniquets to redirect blood flow facilitates anatomical delimitation of the extension and components of the malformation as well as assessment of the volume of the venous compartments. Traditionally, phlebography has been the procedure of choice for studying the deep venous system and its patency in the limbs of patients with large venous malformations or the combined malformations of the Klippel-Trenaunay syndrome.

As mentioned earlier, in the study of arteriovenous malformations MRI images do not define afferent and efferent vessels, the nidus, patterns, or flow velocities with sufficient precision. Consequently, despite advances in 3-dimensional MRI technology, angiography remains the procedure of choice for evaluating the angioarchitecture of these lesions. It is, therefore, an indispensable prior requisite for embolization treatment and, in this context, can play a therapeutic as well as a diagnostic role.

Today, new MRI and multi-ring CT scanners can acquire images with great speed and produce high quality multiplanar reconstructions and noninvasive angiographic studies.[13,23] We recently studied 16 patients with Klippel-Trenaunay syndrome using multidetector CT venography and 3-