Peripheral vascular malformations are now described according to some accepted guidelines, and the principle of proper treatment (nodus ablation) is becoming clear. An appropriate classification schema for vascular anomalies and definite indications for treatment are important to successful treatment overall. Non-invasive imaging (US, CT, and MRI) in association with clinical findings is critical in establishing the diagnosis, evaluating the extent of the malformation, and planning appropriate treatment. Direct nidus phlebography is useful not only in making a correct diagnosis but also in treating the lesion by sclerotherapy. When a patient suffers clinical complications, the nidus sclerotherapy becomes mandatory. If the vascular malformation remains bloodstream to a drainage vein during nidus opacification, flow control is necessary to achieve complete nidus ablation. A multidisciplinary approach is needed in the treatment of a high-flow lesion. A dedicated team approach is necessary for appropriate management in most cases.

Key words: arteriovenous malformations, ethanolamine oleate, sclerotherapy

INTRODUCTION

Vascular (arteriovenous) malformations (AVMs) constitute some of the most difficult diagnostic and therapeutic challenges. A classification system for vascular anomalies based on cellular features, flow characteristics, and clinical behavior was updated during the meeting of the International Society for the Study of Vascular Anomalies (Table 1). In 1996, Kawanabe et al. reported a system for practical classification of vascular lesions in which the treatment procedure was selected according to the vascular stasis within the lesion.

AVMs occur as a result of aberrant vessel angiogenesis. They are localized or generalized congenital vascular abnormalities composed of direct microscopic connections between arteries, veins, and lymphatic vessels without the normal capillary bed. AVMs have a high recurrence rate because they originate from the mesenchymal cells at an early stage of embryogenesis. These lesions may not be evident until additional growth or vascular engorgement manifests as a response to thrombosis, trauma, infection, or endocrine fluctuations. Unlike hemangiomas, AVMs generally increase proportionately in size as the child grows.

Some AVMs appear as part of a familial genetic disorder called angiomatous syndrome (Table 2). One manifestation of angiomatous syndrome, Rendu-Osler-Weber syndrome, is due to two genetic disorders, both of which result in the loss of function of cell receptors. Rendu-Osler-Weber syndrome usually presents with telangiectasia of the skin and mucous membranes, typically appears during puberty and causes bleeding.

SELECTION OF IMAGING MODALITIES

In the assessment of AVMs, ultrasound with color Doppler imaging, magnetic resonance imaging, and direct phlebography play the most parts in diagnosis, classification, and management. These imaging modalities should be used to evaluate the characteristics of the lesion, such as size, flow velocity, flow direction, relation to the surrounding structures (e.g., vessels, muscle, nerve, bone, skin), and lesion contents.

Ultrasound and color Doppler imaging

Ultrasongraphy is a non-invasive and essential tool that is widely used to examine superficial vascular lesions. It is easy to examine combined with the clinical examination. Color Doppler imaging permits real-time analysis of arterial and venous flow and measurement of flow velocities. It is useful in follow-up, enabling an assessment of the amount of the lesion that has been successfully occluded. It is limited in the assessment of deep lesions and lesions adjacent to interfering air or bone.
Computed tomography

The high temporal resolution and the easy with which findings can be interpreted are advantageous in evaluating vascular lesions. However, because CT involves significant exposure to ionizing radiation and provides less information about blood flow, MR imaging has replaced CT in the evaluation of vascular malformations.

Magnetic resonance imaging

MR imaging is the most valuable modality in the classification of vascular malformations. It depicts the anatomical relation between the vascular lesion and adjacent organs, nerves, tendons, and muscles. Slow-flow venous malformations are of high signal intensity on T2-weighted images, whereas high-flow AVMs and fistulas contain a signal void. Intravenous contrast is useful in evaluating the flow velocity (slow, intermediate, or high) of lesions and in monitoring patients who have undergone therapy.

Venography, arteriography, and phlebography

Venography and arteriography are not generally useful as the assessment of AVMs. With the use of direct nidus puncture phlebography, the nidus volume, flow pattern, extent and connection of vascular structures can be evaluated.

FIGURE 1
VASCULAR MALFORMATION
VASCULAR MALFORMATION IS UNRELATED TO ENDOTHELIAL PROLIFERATION; IT IS DUE TO FAILED FORMATION OF VESSELS DURING EMBRYOGENESIS. ARTERIOVENOUS MALFORMATION (AVM) IS A TYPE OF VASCULAR MALFORMATION IN WHICH ARTERIAL AND VEINOUS ELEMENTS PREDOMINATE. MOST OF AVMS ARE COMPRISING FEEDING ARTERIES, THE NIDUSES, AND DRAINAGE VEINS.

FIGURE 2
FLOW CHART FOR ETHANOLAMINE OLEATE SCLEROTHERAPY

FIGURE 3a, b, c, d
HEMANGIOMA, 33-YEAR-OLD MAN, ETHANOLAMINE OLEATE SCLEROTHERAPY VIA DIRECT PUNCTURE MAGNETIC RESONANCE IMAGE (MRI) SHOWS A SUBCUTANEOUS HEMANGIOMA IN THE LEFT PALM (3A, 3B). WITH A TOURNIQUET AROUND THE WRIST, CONTRAST MATERIAL WAS INJECTED PERCUTANEOUSLY (DIRECT NIDUS PUNCTURE) TO CONFIRM CONTRAST STASIS AND TO DETERMINE THE MAXIMUM DOSE OF CONTRAST (JUST BEFORE APPEARANCE OF THE DRAINAGE VEIN TO PREVENT OVERLOAD OF SCLEROSANT). SCLEROTHERAPY WAS PERFORMED BY GENTLE INJECTION OF ETHANOLAMINE OLEATE. AT LEAST 30 MIN OF ABLATION IS NECESSARY. IN THIS PATIENT, NO NEUROLOGICAL DEFICIT OR SKIN ULCERS WERE SEEN AFTER THE PROCEDURE. POST PROCEDURAL MRI SHOWS DECREASED LESION SIZE AND FLATTENED THE PALM SURFACE (3C). CONTRAST ENHANCEMENT EFFECT REMAINS WITHIN THE NIDUS (3D) AND CONSERVATIVE FOLLOW UP IS RECOMMENDED.
MATERIALS FOR ACLELOTHERAPY

Sclerotherapy is a new therapeutic modality that is accepted as independent therapy for AVM. It has helped improve surgical results and has expanded the role of surgical therapy.

Absolute ethanol is one of the main agents used in treatment of surgically inaccessible lesions. It is administered via transarterial, transvenous, or direct-puncture injection. The presence of ethanol in vessels causes endothelial damage, denaturation of blood proteins, thrombus formation, and vascular occlusion.

A 64-96% response rate, defined as an improvement in symptoms or a reduction of the lesion, has been reported after ethanol sclerotherapy of venous malformations.

Ethanolamine oleate (5-10%) is also one of the main agents used for sclerotherapy. It is a salt of an unsaturated fatty acid and is used as a sclerosing agent because it has excellent thrombosing properties. Injection into varices leads to thrombogenesis as a result of chemical damage to the vascular wall. A hemolytic effect occurs if the sclerosing agent leaks into the systemic circulation.

The prophylactic hepatoglobin (2000-4000 U/hr) and albumin adjustment (3.0 g/dl) should be used prior to injection to prevent permanent renal insufficiency. In comparison to ethanol, ethanolamine oleate has less effect on the deep vascular layer (and no penetrative effect), so ethanolamine oleate is not associated with neurological side effects despite the proximity of the nervous system to the vascular system.

Because there is no convincing evidence of permanent damage to the endothelium, embolic agents (e.g., coil, N-butyl-cyanoacrylate, small particles) should be used for subsequent multi-faceted treatment of AVMs that includes, for example, flow reduction and control of bleeding.
**PREOPERATIVE ANESTHESIA**

Local or general anesthesia is necessary because of the severe pain that occurs during the sclerotherapy with absolute ethanol or ethanolamine oleate. Local anesthesia, narcotic, or non-narcotic anti-inflammatory drug treatment is needed to relieve the pain for a few days after the procedure.

**PRINCIPLES OF TREATMENT**

AVMs are responsive to various stimulations and improper treatment often makes the condition worse. Therefore, complete destruction of the nidus of an AVM is the only potential cure. Decreasing the clinical symptoms can be another goal in treating problematic AVMs. Clinical improvement can be achieved with several courses of sclerotherapy.

The key concept of sclerotherapy is the achievement of nidus flow stasis. The physician should prepare the nidus flow stasis (using tourniquet, balloon catheter, and/or coiling) and ablate nidus itself using sclerotic agents (such as absolute ethanol and/or ethanolamine oleate).

**INJECTION PROCEDURE**

The sclerosant injection should be in the nidus or as near to the nidus as possible. It is necessary to reach the nidus, a transarterial, transvenous, or transosseous approach is recommended if the percutaneous (direct) approach is difficult in some reason.

If the approach route is free from severe complication, direct puncture with an 18-22-gauge needle is recommended. For a small lesion, only one session is enough to complete the procedure. When treating a non-flow AVM, careful and gradual injection is needed to prevent overdose and overflow into the drainage vein.

A high-flow AVM is usually difficult to ablate with sclerotic agents without any flow control and it has a relatively high recurrence rate. A multidisciplinary approach is strongly recommended.

**FOLLOW-UP**

The patients’ symptoms should be carefully monitored during the follow-up period. After the procedure, patients complain of swelling and discomfort in the area of the treated lesion. At least 2 years of follow-up is recommended to detect any recurrence and determine whether any complications have worsened (or resolved).

Using image modalities, for objective observation, MR T2-weighted image is highly recommended. When the sclerotherapy is achieved successfully, the nidus may decrease in volume and in signal intensity. T1-weighted image with contrast administration is useful in evaluating the vascularity of lesions and reactive changes of the surrounding tissue.
CONCLUSION

Vascular malformations are the one of the most difficult challenging field of interventional radiology. The best way to manage these patients is to treat in regular basis and delicate team approach (various surgeries, internal medicine, interventional radiology, and psychiatrist) is necessary to appropriate management.

SUMMARY

Klinička definicija i klasifikacija vaskularnih anomalija definiše i indikacije za lečenje koje su od značaja za uspešnost celokupnog tretmana. Ne invanzivni imid'ing tehnike (US, CT i MRI) u medjusobnoj korelaciji su odgovorne za preciznu dijagnostiku, planiranje procedure i evaluaciju rezultata tretmana. Direktna nodusna punkcija (hemangiografija i slično) nemaju dijagnostičkog značaja ali su praktično metode pod kojima se izvodi skleroterapija. Kod pacijenata se komplikovanom i teškom kliničkom slikom skleroemboloterapija je metoda izbora za rešavanje simptomatskih arterio-venskih malformacija. Procedura se izvodi u lokalnoj ili generalnoj anesteziji i embolizaciono sredstvo koje koristimo je apsolutni alkohol ili etanolamin oleat. Lokalna anestezija se može biti neophodna nekoliko dana postproceduralno zbog bola. Najbolji terapijski pristup jeste timski rad vaskularnog hirurga, interventnog radiologa i interniste.

Ključne reči: malformacije, sklerozacija, interventna radiologija
REFERENCES