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ORIGINAL ARTICLE

ENDOVASCULAR MANAGEMENT OF HEAD AND EXTREMITY VASCULAR MALFORMATIONS: A SINGLE CENTER EXPERIENCE

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Abstract

Aim: To report our experience in the management of head and extremity vascular malformations.

Methodology: From November 2010 to November 2012, the study included 20 patients. All AVM cases (11 patients, 55%) underwent trans-catheter embolisation using a mixture of N-butyl cyanoacrylate with lipidol. VM patients were allocated into either percutaneous injection of 99% ethanol under fluoroscopic guidance (3 cases, 15%) or percutaneous foam sclerotherapy using ethanol-amine oleate 5% under duplex guidance (6 cases, 30%).

Results: According to our scale evaluating the outcomes, Satisfying results were observed in 19 patients (95%) after this intervention protocol. Non-satisfying outcome was noticed in a single case (5%).

Conclusion: Detailed diagnostic imaging, arriving successfully to the nidus or the target lesion, delivering therapy in effective concentrations, completion angiography and the need for extra settings, all could accomplish the mission.

Keywords: malformations, endovascular, embolisation, sclerotherapy.

INTRODUCTION

Haemangioma was the most common terminology used for diagnosis of vascular anomalies.(1) Great progress in classifying and understanding vascular anomalies has been made on basis of cellular kinetic and clinical behavior. They are classified into two major categories: Haemangioma and vascular malformation.(2)

Vascular tumors, such as hemangiomas, have endothelial hyperplasia with increased endothelial turnover at histologic analysis. During early childhood, these tumors undergo an initial proliferative phase, and they finally involute with age, which usually makes invasive treatment unnecessary.(3)

Vascular malformations represent congenitally abnormal connections between arteries and veins devoid of the usual arteriolar resistance vessels or capillary beds. They are always present at birth and grow with the patient. Although this may not be clinically apparent in a child, a vascular malformation does not involute. They are frequently asymptomatic but can expand sporadically in puberty and are frequently exacerbated by pregnancy or after local trauma.(4)

Congenital vascular malformations are categorized further into high-flow lesions as arterio-venous malformations (AVM) and low-flow lesions as venous

malformations (VM), or combined vascular malformations. AVMs have been known as one of the most diagnostically difficult and therapeutically enigmatic illnesses in the practice of medicine. Complete surgical eradication of the nidus is rarely possible, except when the AVM is small, localized, and in a surgically accessible area. The ligation of feeding arteries and partial excision of the nidus is usually followed by recurrence, and the outcome is usually a worsened condition.[5,6] With improvement of catheter technology, superselective techniques, and the use of different embolic agents, embolotherapy targeted at the obliteration of the core nidus provide safe effective therapeutic option.[7]

Venous malformations (VMs) are the most prevalent vascular malformation. They have a propensity for the head and neck region, but they can be found anywhere in the body. They can be treated with sclerotherapy and/or surgical excision.[8]

We report our experience in the endovascular and percutaneous management of head and extremity vascular malformations.

PATIENTS AND METHODS

Our institutional review board approved the study design. Written consent for undergoing a procedure was obtained from all patients after a discussion about the advantages and risks associated with transcatheter and/or percutaneous embolotherapy.

From November 2010 to November 2012, 20 consecutive patients (mean age = 21.7 years) with peripheral vascular malformations were included in our series. The presence of peripheral vascular malformation was confirmed by combination of clinical examination and the use of imaging studies including magnetic resonance imaging (MRI) (9 patients), computed tomography angiography (CTA) (2 cases) and/or conventional angiography (9 patients). Doppler color flow imaging was performed for all patients.

AVMs were defined when multiple arteriolar components of the nidus shunted into a dilated venous component of the nidus. Venous malformations (VMs) are characterized by a soft, compressible, non-pulsatile tissue mass. Imaging studies demonstrated the absence of dilated feeding arteries. Doppler ultrasonography revealed compressible hypoechoic or anechoic lesions with monophasic low flow velocity. Various imaging modalities indicated the site and extension of the pathology.

Imaging studies of all patients in the current series were reviewed by consensus of a senior vascular surgeon and interventional radiologist. Patients were allocated to a specific treatment plan according to their diagnosis.

AVM patients were considered candidates for transcatheter embolotherapy. VM patients underwent percutaneous sclerotherapy.

Treatment of AVM: All trans-catheter embolisation procedures were performed under local anesthesia. Light sedation was required beside the conventional measures to avoid adverse reactions of the contrast. 9, 10, 11 General anesthesia was preferable in children. All procedures were performed using Integris H 5000; Philips machine.

For every case, we achieved access through a right femoral artery puncture and insertion of a 5 or 6 Fr introducer sheath. Complete selective and super-selective angiographic study was performed to delineate the AVM elements in order to assess the feeding arteries, the complexity of the nidus, the draining outflow vein and the flow across the fistula. (Figs. 1-5).

Case 1: (Occipital AVM)

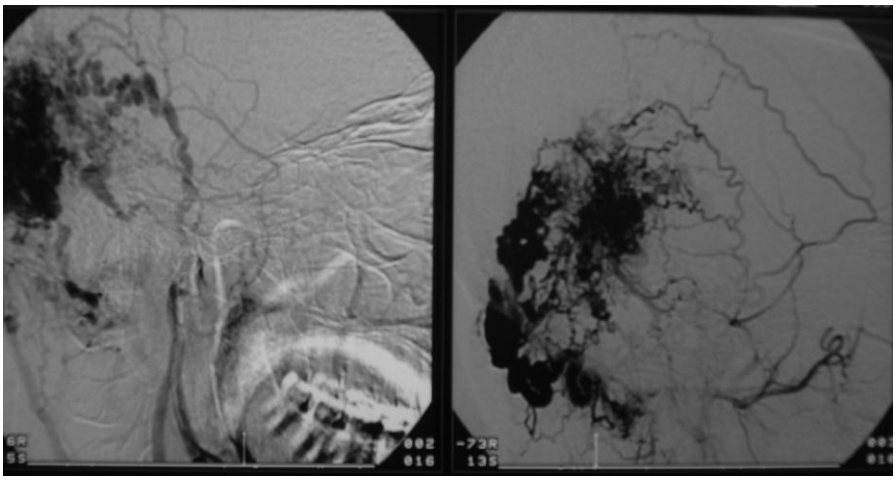


Fig 1. Selective right carotid angiogram revealed large occipital AVM supplied by the superficial temporal (STA) and occipital arteries.

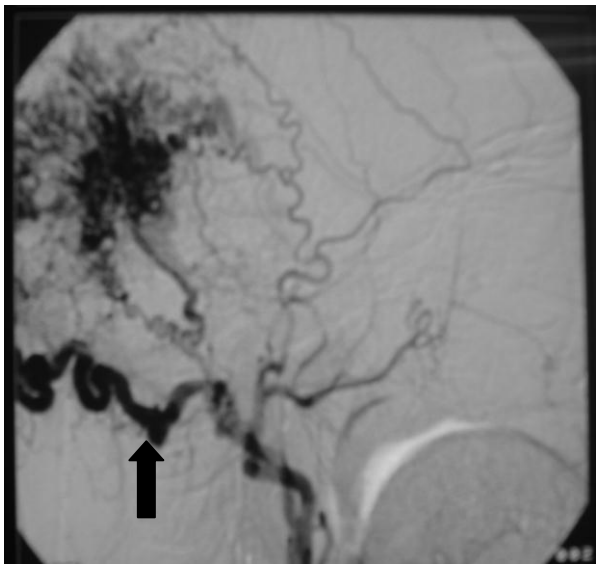


Fig 2. Selective right carotid angiogram, note the hypertrophied occipital artery (arrow).

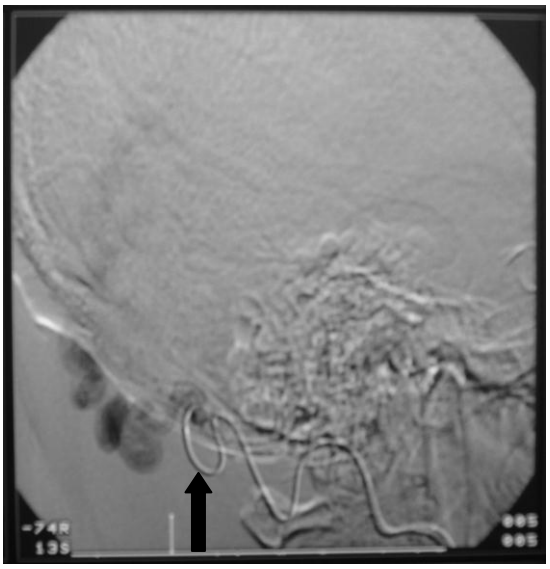
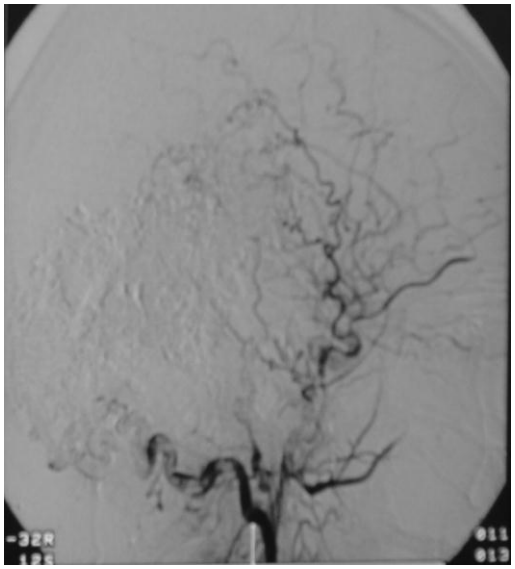
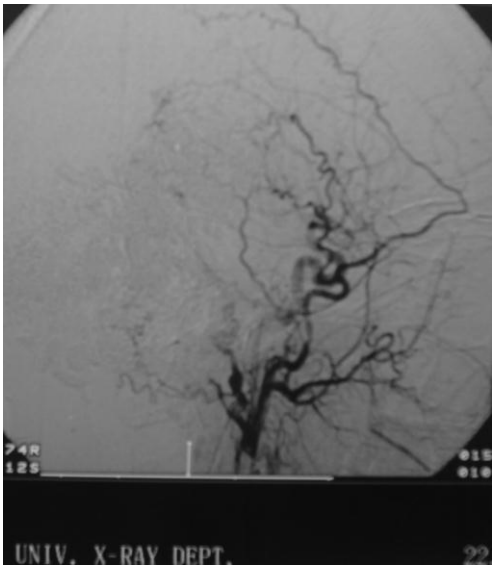


Fig 3. Super selective catheterization of the occipital artery using microcatheter (arrow).





Figs 4,5. Control angiogram after injection of the nidus by N-butyl cyacrylate mixed with lipidol (50%). Note complete obliteration of the nidus.

We replaced the diagnostic catheter with either 5 or 6 Fr guiding catheter. Navigation using a microcatheter with its corresponding guide wire (Ranegate microcatheter, Boston Scientific) was performed to reach the nidus. Reshaping of the guide wire tip was required to facilitate navigation through tortuous vessels. Tortuosity of the feeding arteries and complexity of the nidus determined to a large extent the duration of the procedure. Percutaneous needle insertion was recommended in difficult cases.

According to the fistula flow, we prepared the mixture of N-butyl cyanacrylate NBCA (glue) with lipidol. The concentration ratio in most of cases was 1 to 1; modified to double amount glue in high flow fistula and half amount glue in low flow lesions. (Figs. 8-10).

We inject the mixture guided by the road map. We stop injection when obliteration of the nidus is noticed with minimal reflux in the feeding artery. Upon completion of the procedure, control angiogram was done to assess the remaining part of the nidus. Multiple sessions with two-month interval were required in extensive lesions (n=6, 30%).

Analgesics, anti-inflammatory drugs and broad spectrum antibiotics were required two or three days after the procedure.

Treatment of Venous Malformations: We used either absolute alcohol or ethanolamineoleate 5% for the percutaneous sclerotherapy. The presence of ethanol in vessels causes endothelial damage, denaturation of blood proteins, thrombus formation, and vascular occlusion.[12]

Ethanolamineoleate 5%; a detergent sclerosing agent that causes injury by altering the surface tension surrounding endothelial cells. Compared with ethanol, it has less effect on deeper layers of the vascular wall, no penetrative effect, and is safer to use in situations where vascular structures are close to nerves.[13]

General anesthesia is usually required when direct percutaneous injection of absolute ethanol is targeted. Venous access is achieved with 22 gauge needle, venography was first performed using nonionic contrast media (Ultravist 370 mg) to assess correct placement of the needle, configuration and extension of the lesion. Injection of 99% ethanol after compression of the draining vein is usually needed to minimize the passage of the sclerosing agent into the systemic circulation.

The dose is calculated according to the size of the lesion; anyhow, we didn't exceed 1 ml / kg in a single setting according to guidelines. 14 Completion direct venography demonstrated how far the obliteration of the malformed vascular channels was achieved which determined the need for additional settings. Post procedural anti-inflammatory drugs and antibiotics were administrated for 3 days. (Figs. 6,7).

For the Ethanolamine oleate foam sclerotherapy, the Tessari method of mixing was our preferred way for micro-foam production. Two syringes, one containing sclerosant and the other filled with atmospheric air (usually in a ratio of 2:1 to 3:1), are attached to a three-way plastic connector. Foam is produced by passing sclerosant between the two syringes. 13 We didn't exceed 10 ml per setting as considered more safe according to guidelines.[15]

The treated extremity was elevated before sclerosant injection in order to empty the malformed venous channels. Injection was performed under Duplex guidance using linear probe L12-3 MHz (Envisor-Philips) in both transverse and longitudinal views.

The leg remained elevated for an additional 10 minutes after injection of the sclerosant to achieve more effective endoluminal obliteration. (Figs. 11-13).

Following injection, we placed compression bandages left in place 1 week. Patients were encouraged to ambulate and return to daily activities immediately after the procedure.

Interpretation of the results: We evaluated both radiological and clinical results following each procedure. Radiological outcome was evaluated according to the degree of devascularisation of the vascular lesion as less than 50%, more than 50% or 100%.

Clinical outcome was appreciated in the form of improvement of symptoms basically disfigurement and pain. Objective criteria were set as well in the form of reduction or disappearance of swelling, stop bleeding and/ or healed ulcers.

We defined our results in the current series into either satisfying or non-satisfying.

It was considered satisfying when we had more than 50% or 100% devascularisation, and complete or partial remission of symptoms and signs for the patient.

It was considered non-satisfying when we had minimal remission or aggravation of symptoms and signs regardless the radiological results.

Case 2: (hand venous malformation)

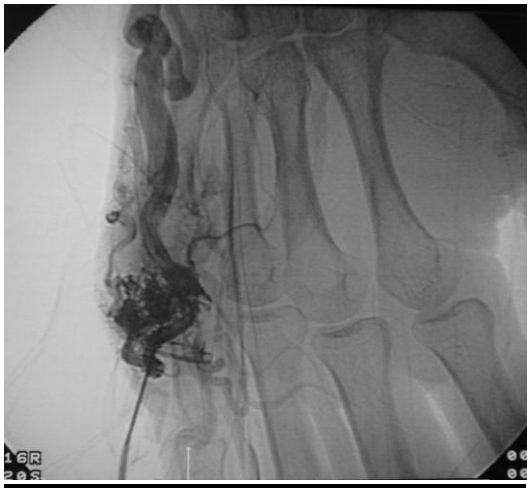


Fig 6. Percutaneous phlebography of venous malformation of the hand and injection of absolute ethanol.

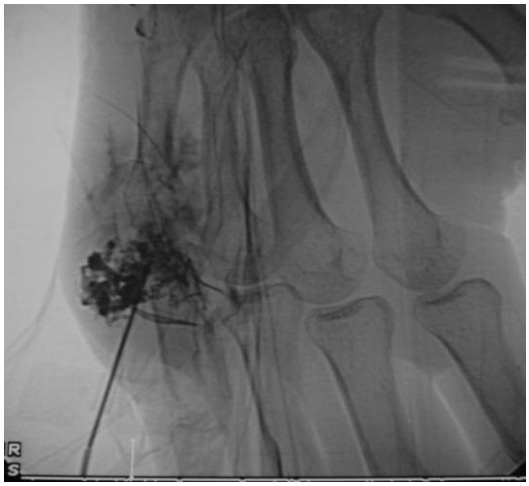


Fig 7. Control venography revealed partial devascularisation and occlusion of the outflow vein.

Case 3: (Thigh AVM)

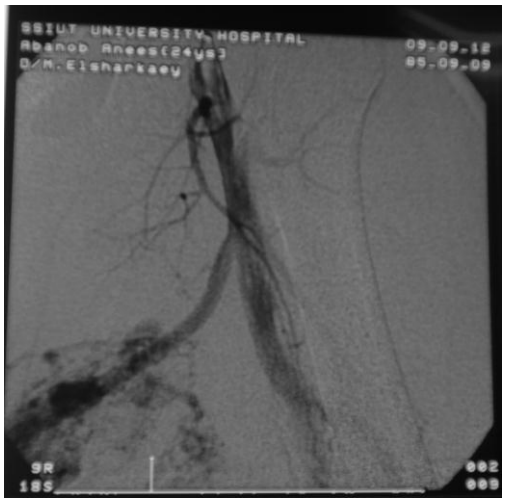


Fig 8. Selective angiography of the deep femoral artery with digital subtraction revealed AVM of the thigh supplied by hypertrophied muscular branches.

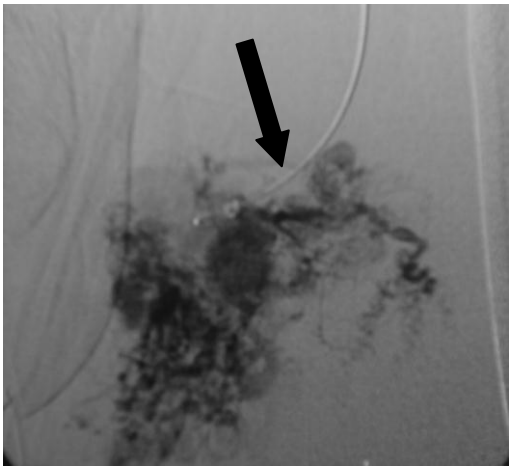


Fig 9. Digitral subtraction angiogram after superselective catheterization of the nidus using microcatheter (arrow).

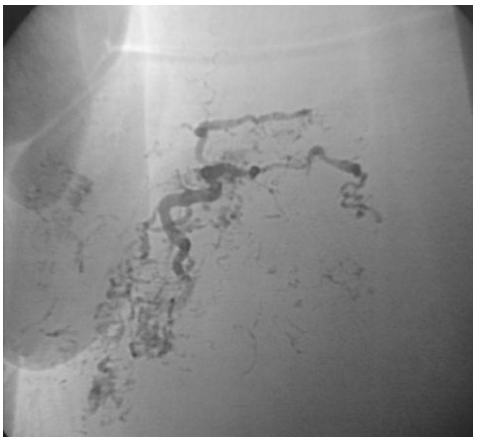
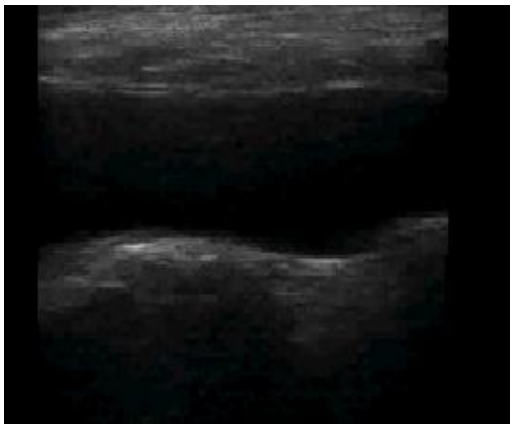
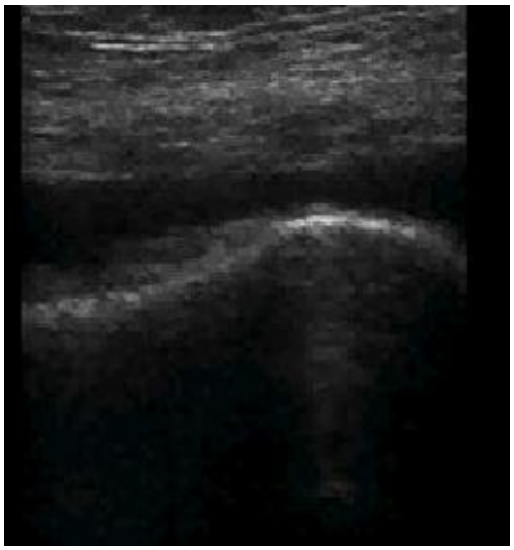


Fig 10. Completion angiography.

Case 4: Duplex guided foam sclerotherapy





Figs 11,12. Definite compressible vascular space as clearly seen by duplex.

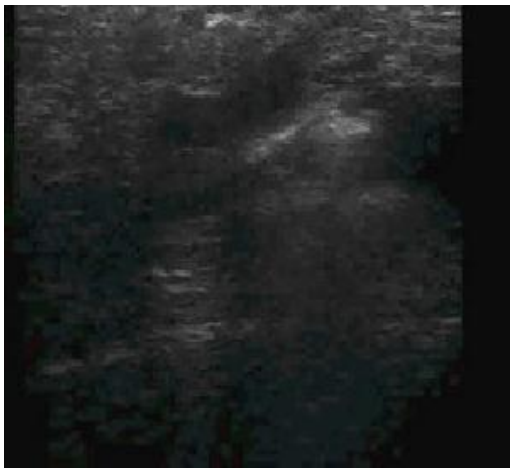


Fig 13. Injected micro-foam inside the dilated venous channel.

RESULTS

Twenty patients were included in the current study. Patient characteristics and diagnosis are presented in Table 1. Treated vascular malformations were mostly located in the extremities (14 cases, 70%), followed by the head region (6 cases, 30%). Table 2 demonstrates sites of encountered lesions.

Table 1. Patient characteristics and diagnosis.

Male	6 (30%)
Female	14 (70%)
AVM	11 (55%)
VM	9 (45%)

AVM: arterio-venous malformations.
VM: venous malformations.

Table 2. Site of lesion.

Site of lesion	
Occipital scalp	3 AVMs 1 VM
Temporal scalp	1 AVM
Face	1 AVM
Gluteal	1 AVM
Thigh	1 AVM 1 VM
knee	1 AVM 1 VM
Leg	2 AVM 1 VM
Foot	1 AVM 1 VM
Arm/Forearm	2 VM
Hand	1 VM
Wrist	1 VM
Total	20

All cases complained of disfigurement in the form of swelling, difference in limb circumference or bluish tints. They presented as well with: functional disturbance in cases of extremity vascular lesions (70%), pain in 3 cases (15%), bleeding in 2 patients (10%), ulcer in a single patient (5%) and symptoms of venous hypertension in 5 cases (25%).

All AVM cases (11 patients, 55%) underwent trans-catheter embolisation using a mixture of N-butyl cyanoacrylate with lipidol. Three patients (15%) underwent combined intra-arterial embolisation and percutaneous injection of the nidus. VM patients were allocated into either percutaneous injection of 99% ethanol under fluoroscopic guidance (3 cases, 15%) or percutaneous foam sclerotherapy using ethanol amine oleate 5% under duplex guidance (6 cases, 30%).

A single setting was enough to achieve more than 50% radiological resolution of the vascular lesion in 6 AVM patients (30%). VM cases who underwent percutaneous injection using 99% ethanol were found to have a maximum of 2 settings while those who benefited from the foam sclerotherapy had 4 or more settings to achieve more than 50% devascularisation.

Evaluation of outcome is demonstrated in Table 3. Satisfying results were observed in 19 patients (95%) after this intervention protocol. Non satisfying outcome was noticed in a single case (5%). Surgical excision of the residual mass was required in 4 VM patients (25%) after the procedure.

Two AVM cases developed complications following the intervention. It was major in one case due to distal embolisation with forefoot gangrene and minor in the other patient where skin ulcer developed but healed with simple wound care.

Table 3. Evaluation of outcomes.

AVM	11 cases	-8 (more than 50% devascularisation) -3 (100% devascularisation)	-7 satisfying, 1 non-satisfying (aggravation of symptoms) - 3 satisfying
VMs	9 cases (3 fluorocopic guided, 6 duplex guided)	-all achieved more than 50 % devascularisation	-all satisfying -4 cases needed surgical excision of the residual mass in the duplex-guided group.

DISCUSSION

We're reporting our experience as regards the therapeutic strategy for Patients with high flow AVMs or low flow VMs at the Assiut University Hospital, EGYPT.

The treatment of vascular malformations is unsystematic with too little therapeutic evidence. As a result, it is commonly accepted that the therapeutic strategy should be established by a multidisciplinary medical, surgical and radiological team.[16,17]

Twenty patients presented with head or extremity vascular malformation and were included in our series. Diagnosis was based on different imaging modalities into either AVM in 11 patients and VMs in 9 cases.

Surgical removal of vascular malformations has been recommended only for selected patients due to the risk of massive hemorrhage during the operation and high recurrence and complication rates.[18]

Furthermore, it is reported that certain AVM lesions can be aggravated by incomplete surgical excision or feeding artery ligation due to rapid collateralization from adjacent arteries and further limit access to the nidus for future treatment.[19] Malan and Puglionisi proposed that pressure or flow changes in the vascular malformation lesion can reactivate dormant angiopoietic cells and stimulate endothelial cell growth.[20]

Growth factors, such as vascular endothelial growth factor (VEGF), angiopoietin-1 (Ang-1) and angiopoietin 2 (Ang-2), have been reported as the most potent regulators for neo-vascularisation.[21]

Accordingly, current trends in the treatment of vascular malformations largely depend on endovascular treatment.[22] Recent advances in techniques and materials supported such approach.

All AVM cases (11 patients, 55%) underwent trans-catheter embolisation using a mixture of N-butyl cyanoacrylate with lipidol. In all cases we aimed the nidus for obliteration. In 3 cases, it was the difficult arterial anatomy that made it impossible to reach the nidus and we shifted to percutaneous route to destroy the target.

Glue or tissue adhesive, such as N-butyl cyanoacrylate (NBCA) is a liquid embolic agent that polymerizes and hardens on contact with an ionic environment such as blood. It's very effective rapidly spreading through the nidus. However, during the procedure the catheter must be rapidly withdrawn after each injection to prevent catheter trapping, resulting in frequent, time-consuming catheter exchanges.[23]

When these agents are used, it is difficult to control the level of occlusion, and it has also been reported that lesions treated with nbutyl cyanoacrylate can recanalize.[24-26] Alcohol is the most definitive sclerosant agent and gives the best clinical results, although it may induce nerve injury and skin necrosis in 15% of the cases.[27]

In the current series, we achieved more than 50% devascularisation of the nidus in 8 cases and 100% devascularisation in 3 patients. Using this strategy, 10 patients out of eleven were satisfied in terms of clinical remission and no recurrence. We encountered a single complication in the form of distal arterial embolisation in a case of high flow AVM around the knee that needed trans-metatarsal amputation later on.

Lee B.B. in his clinical series described excellent interim results with no recurrence in all 9 cases underwent NBCA embolotherapy. He reported a single patient who had pulmonary embolism following the use of NBCA glue.[17]

White et al reported long-term follow-up results for palliation of AVMs with the use of cyanoacrylate. Durable long-term (mean, 7.4 years) relief of symptoms was achieved in 11 patients with localized upper extremity AVMs, but there was high incidence of amputation in five (55.6%) of nine patients with lower extremity AVMs due to distal embolisation.[27]

In the current series, we reported 9 cases with head or extremity venous malformation. Three patients were treated with percutaneous injection of 99% ethanol under fluoroscopic guidance while the other 6 cases benefited from subcutaneous sclerotherapy using ethanolamineoleate 5% under duplex guidance.

We achieved more than 50% devascularisation and partial clinical remission in all VM patients after 99% ethanol sclerotherapy. They were satisfied and didn't need further surgical intervention. On the other hand, following sclerotherapy with ethanol amineoleate, we needed 4 settings or more in all patients to achieve more than 50% devascularisation.

Four patients out of six needed surgical excision of residual mass after sclerotherapy. Comparable results were reported by different series describing treatment of low flow venous malformations by ultrasound guided foam sclerotherapy.[28-30]

Although, we achieved the target with absolute ethanol sclerotherapy in 2 settings or less and no further surgical intervention, we couldn't perform this strategy in cases when malformations were in close vicinity to the skin or nerve structures to avoid inevitable damage. On the contrary, ethanolamineoleate foam sclerotherapy was feasible in such group of patients. It was performed in the out-patient clinic, without anesthesia. Blood loss during surgical excision in the 4 VM cases was minimal.

Absolute ethanol remains the most reliable substance for permanently occluding peripheral arteries and veins although the risk of major adverse effects (e.g. skin and soft tissue necrosis, nerve damage, deep vein thrombosis) is significant. It should be used only by experienced personnel.[31-33]

S. Blaise et al in their clinical series on 24 patients, they found that ultrasound guided sclerotherapy is less effective than alcohol but has many advantages as a result of being well tolerated. This technique can be performed as an outpatient procedure, while more powerful sclerosing agents such as alcohol cannot. The procedure requires no anesthesia. Post-intervention

surveillance only lasts few hours, and is not conditioned by the same requirements as when general anesthesia is used after alcohol injection.[34]

In conclusion Endovascular and direct percutaneous approaches in management of vascular malformations has become the mainstay for treatment of such difficult disease category in the face of surgical incompetence in management of those cases. Different materials have been used for embolisation or sclerotherapy. All future researchers should investigate the embolo-therapeutic agents of choice. Our single center experience could assume that detailed diagnostic imaging, arriving successfully to the nidus or the target lesion, delivering therapy in effective concentrations with least possible complications, completion angiography and the need for extra settings, all could accomplish the mission.

REFERENCES

1. Upton J, Mulliken JB, Murray JE. Classification and rationale for treatment of vascular anomalies in the upper extremity. J Hand Surg. 1985;10A:970-75.

2. Mulliken JB. Cutaneous vascular anomalies. Semin Vasc Surg. 1993;204-15.

3. Mulliken JB. Classification of vascular birthmarks. In: Mulliken JB, Young AE, eds. Vascular birthmarks: hemangiomas and malformations – Philadelphia: WB Saunders. 1988:24-37.

4. Belov S. Anatomopathological classification of congenital vascular defects. Semin Vasc Surg. 1993;6:219-24.

5. Szilagyi DE, Smith RF, Elliott JP, Hageman JH. Congenital arteriovenous anomalies of the limbs. Arch Surg. 1976;111:423–9.

6. Flye MW, Jordan BP, Schwartz MZ. Management of congenital arteriovenous malformations. Surgery. 1983;94:740–7.

7. Mourao GS, Hodes JE, Gobin YP, Casasco A, Aymard A, Merland JJ. Curative treatment of scalp arteriovenous fistulas by direct puncture and embolization with absolute alcohol. J Neurosurg. 1991;75:634–7.

8. Abernethy LJ. Classification and imaging of vascular malformations in children. Eur Radiol. 2003;13:2483-97.

9. Kaufman J, Michael L. Vascular and interventional radiology: the requisites. Philadelphia: Mosby. 2004.

10. Pannu N, Wiebe N, Tonelli M. Prophylaxis strategies for contrastinduced nephropathy. JAMA. 2006;295:2765-79.

11. Merten GJ, Burgess WP, Gray LV. Prevention of contrast-induced nephropathy with sodium bicarbonate. JAMA. 2004;291:2328-34.

12. Legiehn GM, Heran MK. Classification, diagnosis, and interventional radiologic management of vascular malformations. Orthop Clin North Am. 2006;37:435-74, vii-viii.

13. Mitsuzaki K, Yamashita Y, Utsunomiya D. Balloon-occluded retrograde transvenous embolization of a pelvic arteriovenous malformation. Cardiovasc Intervent Radiol. 1999;22:518-20.

14. Yakes WF, Rossi P, Odink H. How I do it: arteriovenous malformation management. Cardiovasc Intervent Radiol. 1996;19:65-71.

15. Choi YH, Han MH, O-Ki K. Craniofacial cavernous venous malformations: percutaneous sclerotherapy with use of ethanolamine oleate. J Vasc Interv Radiol. 2002;13:475-82.

16. Blaise S., Charavin-Cocuzza M., Riomet H. Treatment of Low-flow Vascular Malformations by Ultrasound-guided Sclerotherapy with Polidocanol Foam: 24 Cases and Literature Review. Eur J Vasc Endovasc Surg. 2011;41:412e417.

17. Lee B.B. New Approaches to the Treatment of Congenital Vascular Malformations (CVMs), A Single Centre Experience. Eur J Vasc Endovasc Surg. 2005;30:184–97.

18. Trout 3rd HH, McAllister Jr HA, Giordano JM, Rich NM. Vascular malformations. Surgery. 1985;97:36e41.

19. Arneja JS, Gosain AK. Vascular malformations. Plast Reconstr Surg. 2008;121:195ee206e.

20. Malan E, Puglionisi A. Congenital angiodysplasias of the extremities. I. Generalities and classification; venous dysplasias. J Cardiovasc Surg (Torino). 1964;5:87e130.

21. Lobov IB, Brooks PC, Lang RA. Angiopoietin-2 displays VEGF-dependent modulation of capillary structure and endothelial cell survival in vivo. Proc Natl Acad Sci U S A. 2002;99:11205e10.

22. Kim Y.-W. Proper Selection of Patients for Percutaneous Embolo-Sclerotherapy in Patients with Congenital Vascular Malformations (CVMs). Eur J Vasc Endovasc Surg. 2010;39:S49eS54.

23. Wei Zhou, Peter H., Eraso A. Embolotherapy of peripheral arteriovenous malformations. Advances in embolic agents and microcatheter techniques have allowed more successful treatment of this uncommon clinical entity. Endovascular today. 2005:85-91.

24. Rao VR, Mandalam KR, Gupta AK. Dissolution of isobutyl 2-cyanoarylate on long-term follow-up. AJNR Am J Neuroradiol. 1989;10:135–41.

25. Schweitzer JS, Chang BS, Madsen P. The pathology of arteriovenous malformations of the brain treated by embolotherapy. II. Results of embolization with multiple agents. Neuroradiology. 1993;35:468–74.

26. Do YS, Yakes WF, Shin SW. Ethanol embolization of arteriovenous malformations: interim results. Radiology. 2005;235:674-682.28.

27. White RI Jr, Pollak J, Persing J, Henderson KJ, Thomson JG, Burdge CM. Long-term outcome of embolotherapy and surgery for high-flow extremity arteriovenous malformations. J Vasc Interv Radiol. 2000;11:1285–95.

28. Pascarella L, Bergan JJ, Yamada C, Mekenas L. Venous angiomata: treatment with sclerosant foam. Ann Vasc Surg. 2005;19:457e64.

29. Bergan J, Pascarella L, Mekenas L. Venous disorders: treatment with sclerosant foam. J Cardiovasc Surg. 2006;47:9e18.

30. Nitecki S, Bass A. Ultrasound-guided foam sclerotherapy in patients with Klippel-Trenaunay syndrome. Isr Med Assoc J. 2007;9:72e5.

31. Lee BB, Kim DI, Huh S, Kim HH, Choo IW, Byun HS. New experiences with absolute ethanol sclerotherapy in the management of a complex form of congenital venous malformation. J Vasc Surg. 2001;33:764–772.

32. Lee BB, Do YS, Byun HS, Choo IW, Kim DI, Huh SH. Advanced management of venous malformation with ethanol sclerotherapy: mid-term results. J Vasc Surg. 2003;37:533–8.

33. Marrocco-Trischitta MM, Nicodemi EM, Nater C. Management of congenital venous malformations of the vulva. Obstet Gynecol. 2001;98:789–93.

34. Blaise S, Bosson JL, Diamand JM. Ultrasound-guided sclerotherapy of the great saphenous vein with 1% vs. 3% polidocanol foam: a multicentre double-blind randomised trial with 3-year follow-up. Eur J Vasc Endovasc Surg. 2010;39:779e86.