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Original Article

Angiographic Classification and Embolotherapeutic Outcome of High-Flow Arteriovenous Malformations of the Body and Extremities: 4 Years Experience in Ramathibodi Hospital

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Abstract

Objective: To determine angiographic classification of the high-flow peripheral AVMs and to assess embolotherapeutic outcome in Ramathibodi Hospital.

Materials and Methods: Data from angiograms of the 16 consecutive patients with 18 AVMs were classified by two independent interventionists according to Cho et al classification into arteriovenous fistulae (type I), arteriolovenous fistulae (type II), arteriolovenulous fistulae with non-dilated fistula (type IIIa) and arteriolovenulous fistulae with dilated fistula (type IIIb). The embolotherapeutic outcome was assessed into cure, partial remission, no remission and aggravation. Complications were determined into major and minor conditions.

Result: For classification in 18 AVMs, 55.6% were type IIIb, 16.7% were IIIa, 16.7% were type II, 5.6% were combined type I and IIIb and 5.6% were combined type IIIa and IIIb. The embolotherapeutic outcome was assessed in 12 AVMs that 75 % were partial remission and 25 % had no remission. One patient had major complication (ulna artery dissection). Others had minor complications including pain, skin necrosis and transient erection of penis.

Conclusion: Type IIIb AVM was found as the most common type of high-flow peripheral AVMs. Transcatheter arterial embolization with/without superselection is a primary therapeutic modality in management of patients with peripheral AVM to improve, possible cure or as a presurgical intervention to reduce bleeding and maximize successful resection.

Keywords: Angiographic, Classification, Embolotherapeutic outcome, Arteriovenous malformation

Introduction

Vascular malformations are classified by Mulliken et al in 1982 according to endothelial characteristic and flow dynamic of the lesion into high-flow type such as arteriovenous malformation (AVM), and low-flow type, such as capillary, venous, lymphatic, and mixed malformation.[1-3] High-flow AVMs are rare and less common than low-flow malformations, but management of the high-flow AVMs is often more problematic.

Many patients remain asymptomatic through early life, becoming clinically significant at variable times dependent upon the location and size of the communicating arteries and veins. The symptoms include pain, local hyperhidrosis, ulceration, and bleeding. When massive, they may cause high-output cardiac failure.[4-9]

Currently, initial diagnostic modalities include Color Doppler Imaging (CDI) and Magnetic Resonance Imaging (MRI). CDI is an essential tool in the diagnostic work up of the AVMs. Accurate measurements of flow volumes and resistive indexes can be helpful in the initial evaluation and also are important noninvasive parameters for follow-up after therapy.[10-13] MRI has proven to be a mainstay in the initial diagnostic evaluation, as well as in assessing the efficacy of endovascular therapy.[14] Digital Subtraction angiography (DSA) is the best modality for diagnosis and follow-up, due to ability to assess hemodynamic flow and true comparison.

The angiographic description of AVMs is composed of feeding arteries, the complex network of arteriovenous shunts (referred to as the nidus), and draining veins.[5,15] A few reports have provided the descriptions or classifications of AVMs. In 1993, Houdart et al. classified intracranial AVMs into 3 types based on the morphology of the nidus: type I (arteriovenous), type II (arteriolovenous), or type III (arteriolovenulous).[5,8,16] In 2006, Cho et al.[5] modified this classic classification for use with peripheral AVMs into 4 types based on angiography [Fig.1] because the original system did not account for a unique feature of some AVMs - the blush type on angiography. Also some types of AVMs can be treated by direct puncture, whereas intracranial AVMs cannot. Type I is arteriovenous fistulae; type II, arteriolovenous fistulae; Type IIIa, arteriolovenulous fistulae with non-dilated fistula; and type IIIb, arteriolovenulous fistulae with dilated fistula.

Historically, surgery was the primary treatment for AVMs, but the cure rate was low. Recently, embolization with various embolic agents has been suggested as the primary therapeutic modality for improvement, cure or presurgical intervention to reduce bleeding and maximize successful resection.[3,4,17,18] In embolization, permanent occlusion of the nidus should be the goal. The technique can be applied though transarterial, transvenous or direct puncture approaches. Many endovascular occlusive agents are used including autologous clot, Gelfoam, Polyvinyl alcohol particles (PVA), various metallic coils with or without fibers, tissue adhesives glue (IBCA/NBCA), detachable balloons, Ethibloc, Sotradecol and ethyl alcohol.[3,13,18-21]

Due to the rarity of high-flow peripheral AVMs and embolotherapy is the primary standard treatment. The primary purpose of this study was to determine the angiographic classification of high-flow peripheral AVMs found in Ramathibodi Hospital. The secondary purpose was to assess embolotherapeutic outcome of peripheral high-flow AVMs in Ramathibodi Hospital.

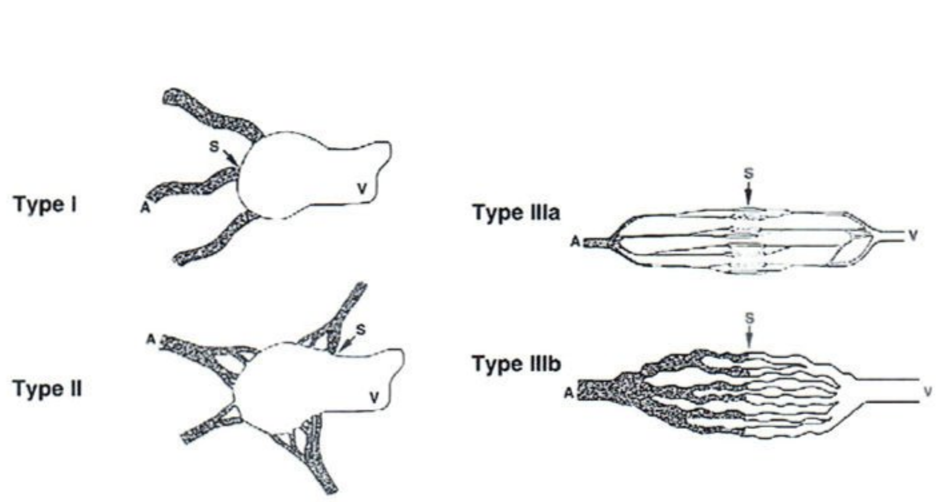

Fig.1 A diagram for the 4 types of AVMs based on angiogram of nidus morphology according to Cho et al. Type I (arteriovenous fistulae) AVMs: no more than 3 separate arteries shunt to the initial part of a single venous component. Type II (arteriolovenous fistulae): multiple arterioles shunt to the initial part of a single venous component, in which the arterial components show a plexiform appearance on angiography. Type IIIa (arteriolovenulous fistulae with non-dilated fistula): fine multiple shunts are present between arterioles and venules and appear as a blush or fine striation on angiography. Type IIIb (arteriolovenulous fistulae with dilated fistula): multiple shunts are present between arterioles and venules and appear as a complex vascular network on angiography. In types I and II, the first identifiable venous structure downstream of the shunt is the initial part of the draining vein. In types IIIa and IIIb, multiple venulous components of the fistula unit collect to a draining vein. A: arterial compartment of the fistula unit, V: venous compartment of the fistula unit, S: shunt

Materials and Methods

Patients

This was a retrospective study, approved by our institutional research ethic committee. We reviewed the angiographic data of patients who were diagnosed with body and/or extremity AVMs during a 4-years period from January 2004 to December 2007, searched from all databases at the Intervention Unit of the Department of Radiology, Ramathibodi Hospital.

The inclusion criteria included all patients, age 0-60 years, diagnosed with body and/or extremity AVMs by angiography at the Intervention Unit of the Department of Radiology, Ramathibodi Hospital. Patients with missing or inadequate data were excluded.

This study included 16 consecutive patients with 18 peripheral AVMs.

Diagnostic Angiography and Embolotherapy Technique

- Written consent for the procedure was obtained from all patients after a discussion about the advantages and risks of the procedure.
- Risk of allergy to contrast media was evaluated. If the patient was at risk, premedication with oral and intravenous steroids was administered.
- Coagulogram (Partial thrombin time: PTT

and prothrombin time; PT), platelet count and serum creatinine were evaluated at the admitting ward. If a laboratory result was abnormal, it was corrected to normal value before the procedure.

- Patients were in supine position during the procedure under local or general anesthesia with monitoring of vital signs and oxygen saturation. Intravenous fluid was given via superficial vein of the left hand.

- Selective angiography was done using 4 or 5 F catheter via common femoral arterial approach. The anatomy of the feeding arteries to the lesions was identified and localized before embolotherapy. Transcatheter embolization was performed via 4 or 5 Fr catheter or coaxial micro- catheter technique in some patients with small-sized feeding arteries. The catheter was advanced as close as possible to the nidus of the AVMs. After that, pre-test embolization was performed by trial injection of contrast agent into the catheter during image acquisition. The volume of contrast material required to fill the nidus and just opacify the draining vein was noted. Embolization was done using a mixture of contrast media and embolic agent. Five to 10 minutes after embolization, an angiogram was performed to determine whether the AVMs had been embolized completely. Meticulous repetition of these techniques was required for complete embolization of at least 1 compartment of the AVM. All patients were followed for immediate or delayed complications.

- Additional embolization was performed if symptoms and signs remained or the AVMs persisted on follow-up imaging studies.

- Non-ionic contrast media used in the procedures included Hexabix 320, Xenetic 350, Ultravist 370 and Omnipaque 370. Dose of contrast media was 1-2 ml/kg.

- The angiogram machines used were Infinic VC DSA; (Toshiba, Japan) from 01 January 2004 to 30 September 2006 and Infinic VC-I flat panel DSA; (Toshiba, Japan) from 01 October 2006 to 31 December 2007.

Evaluation of Angiographic Studies and Clinical Data

Two reviewers independently interpreted the angiographic classification of the peripheral high-flow AVMs, one a board certified interventional radiologist (JT) and the other a second-year body interventional radiology fellow (KH). The interpretation of angiographic classification of the peripheral high-flow AVMs into types I, II, IIIa and IIIb was according to Cho et al.[5]

Data on patient demographics, clinical assessment, imaging studies, treatment, complications, and outcome were obtained from the medical records, as were the results of follow-up consultations, examinations, and imaging. Only the therapeutic and clinical outcomes of embolotherapy were included and not those of combined treatments. The final clinical data were updated by telephone if possible.

Definitions

The angiographic classification of the high-flow peripheral AVMs according to Cho et al.[5] modification are type I, II, IIIa and IIIb. Type I (arteriovenous fistulae) refers to at most 3 separate arterial shunts to a single draining vein. Type II (arteriolovenous fistulae) indicates multiple arterioles shunted into a single draining vein. Type IIIa (arteriolovenulous fistulae with non-dilated fistula) indicates fine multiple shunts between arterioles and venules which appear as a blush or fine striation on angiography. Type IIIb (arteriolovenulous fistulae with dilated fistula) refers to multiple shunts between arterioles and venules and appear as a complex vascular network on angiography.

The therapeutic outcomes are assessed according to clinical response and degree of devascularization at final angiography. Non-invasive imaging, including color doppler sonogram, CTA or MRA, are additional modalities used to evaluate residual or recurrent lesion. Cure is defined as complete resolution of the clinical symptoms and signs with 100 % devascularization of the AVMs. Partial remission is defined as complete resolution or an improvement in clinical symptoms and signs with 50% to 99% AVM devascularization. No remission is defined as an improvement or no change in clinical symptoms and signs with less than 50% devascularization. Aggravation is defined as a worsening of clinical symptoms and signs regardless of the degree of AVM devascularization. Treatment failure is defined as a procedure that resulted in amputation of the extremity at the lesion site or instances when the nidus of AVM could not be approached. Cure and partial remission are considered to be effective or successful therapeutic outcomes of embolization. Major complications are death, permanent adverse sequelae, need for major therapy or prolonged hospitalization (more than 48 hours). Minor complications are any component adverse sequelae, such as transient nerve injuries, a skin injury that spontaneously heals or pain that spontaneously resolves.

Statistical Analysis

The ordinal data, including age, sex, symptom and location of AVM, was presented as frequency and percentage. The consensus of angiographic classification between the two participants was evaluated using statistical methods (test of agreement, i.e. percent of agreement, Kappa analysis and p-value), and the outcome of embolotherapy was described.

Results

Patient characteristics

16 patients were enrolled, diagnosed with body and/or extremity AVMs by angiogram at the Intervention unit of Department of Radiology, Ramathibodi Hospital from January 2004 to December 2007. There were 6 males and 10 females. The mean age was 29 years (range 10-59 years). 13 patients had presenting symptoms, most common of which was mass (81%). One 10-year-old patient had congestive heart failure. 2 patients had no presenting symptoms, the AVM being an incidental finding from CT or MRI. Most common location of the AVM was the leg (22%). 2 patients had more than one location. One patient had three locations: the pelvic cavity, thigh and leg. Another one had a large AVM involving the forearm, hand and finger. Details of the patient data are shown in Table 1.

Angiographic findings

According to Cho et al[5] classification of high-flow peripheral AVMs, the most common type of AVM in this study was IIIb (n=10, 55.6%) [Table 2, Fig.1 and Fig.2A, 2B]. Type II and type IIIa AVMs were found in three patients (16.7%) [Fig.3 and Fig.2C, 2D]. Two AVMs were categorized as combined type; one type IIIa+IIIb and one type I+IIIb [Fig.4]. In the independent analysis of AVM type by two interventionists, JT and KH, 5 AVMs were classified differently. Two AVMs (II and IIIa) were classified as type IIIb [Fig.3B]. One type IIIb was regarded as type II. One combined type IIIa and IIIb was also

Table 1 Patient characteristic

Patient characteristics	Number (%)
Age (mean 29.5 ± 1.3 yrs, range 10-59 yrs)	
Sex (n=16)	
Male	6 (37.5)
Female	10 (62.5)
Symptom (n=16)	
Mass	13 (81.25)
Ulcer	2 (12.5)
Bleeding	2 (12.5)
Congestive heart failure	1 (6.25)
Pelvic discomfort	1 (6.25)
No symptom	2 (12.5)
Location (n=18)	
Trunk	2 (11.11)
Pelvic cavity	3 (16.67)
Arm	2 (11.11)
Forearm	3 (16.67)
Hand	2 (11.11)
Finger	2 (11.11)
Thigh	1 (5.56)
Leg	4 (22.22)
Foot	1 (5.56)

Table 2 Angiographic classification (n=18)

Angiographic classification	Number of lesion (%)
Type I	-
Type II	3 (16.67)
Type IIIa	3 (16.67)
Type IIIb	10 (55.56)
Type I+IIIb	1 (5.56)
Type IIIa+IIIb	1 (5.56)
Total	**18 (100)**

Type I (arteriovenous fistulae) AVMs: no more than 3 separate arteries shunt to the initial part of a single venous component.

Type II (arteriolovenous fistulae): multiple arterioles shunt to the initial part of a single venous component, in which the arterial components show a plexiform appearance on angiography.

Type IIIa (arteriolovenulous fistulae with non-dilated fistula): fine multiple shunts are present between arterioles and venules and appear as a blush or fine striation on angiography.

Type IIIb (arteriolovenulous fistulae with dilated fistula): multiple shunts are present between arterioles and venules and appear as a complex vascular network on angiography.

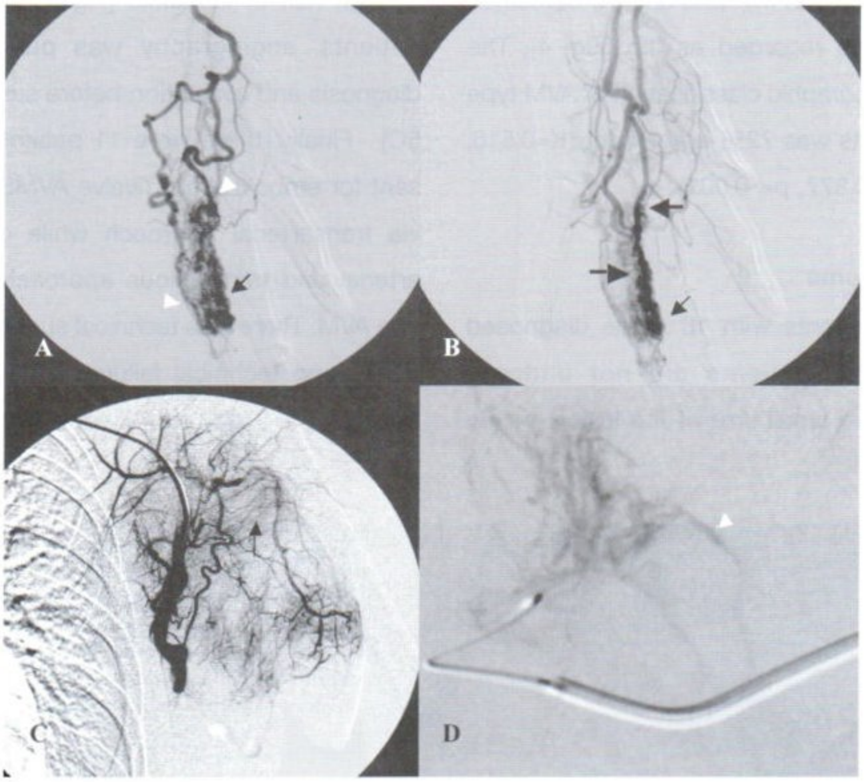

Fig.2 Angiogram of Type III AVM: (A): arterial and (B): venous phases of the type IIIb: complex arteriolar and venulous connection of the nidus (small arrows), arterial feeding (arrowheads) and early draining veins away from the nidus (large arrows). This AVM was not embolized, due to difficult treat safely and high risk for ischemia. The patient was treated by surgical excision. (C and D): two cases of type IIIa: nidus are seen as a blush or fine striation without vascular network (arrow). Note draining vein (arrowhead)

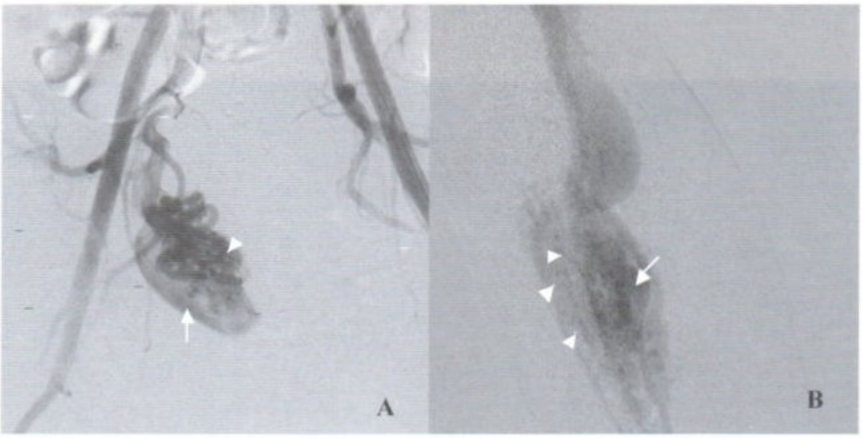

Fig.3 Angiogram of Type II AVM: (A): pelvic and (B): leg AVMs: complex network of arteriolar component and a large venous pouch, nidus (arrowheads) and venous pouch (arrows). The leg AVM (B): this case was classified as type IIIb when independent review because many components of the nidus (arrowhead).

classified as just type IIIa AVM, and another combined type I and IIIb was regarded as IIIb [Fig. 4]. The consensus of angiographic classification of AVM type by 2 interventionists was 72% agreement (K=0.516, 95% CI 0.156 to 0.877, p< 0.001).

Therapeutic outcome

Of the 16 patients with 18 AVMs diagnosed by angiography, 5 patients did not undergo embolization due to small size of the lesion, safety or high risk of ischemia [Fig.2A, 2B, 5A]. In some patients, angiography was performed just for diagnosis and evaluation before surgery [Fig.5B and 5C]. Finally, there were 11 patients with 13 AVMs sent for embolization. Twelve AVMS were accessed via transarterial approach while combined trans-arterial and transvenous approach was used with one AVM. There was technical success with 11 AVMs (85%) and technical failure in 2 patients (patients 10 and 11) [Fig.6 and 7] due to tortuous arterial

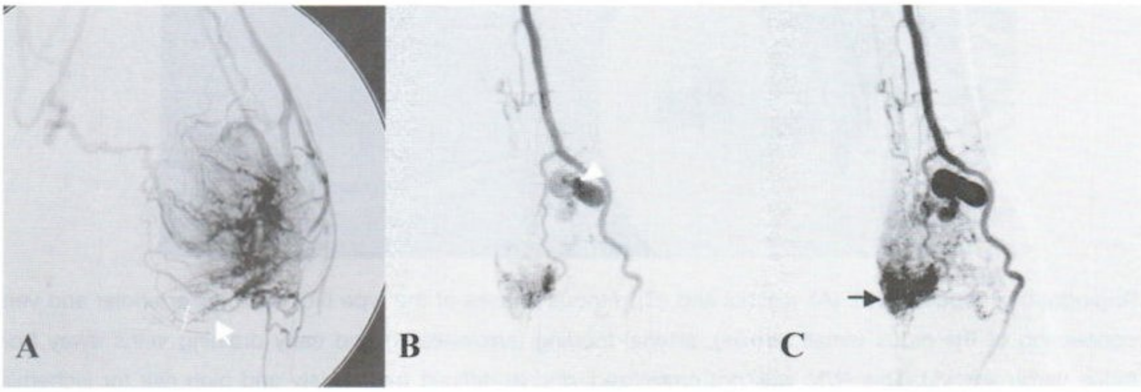

Fig.4 Angiogram of combine AVM type: (A): combine type IIIa and IIIb, which just classified as type IIIa because dominant of blush or fine striation and minimal complex network of nidus (arrow). (B and C): early and late arterial phases of combine type I and IIIb that just classified as type IIIb because dominant complex network of nidus (arrows). Note direct arterovenous fitula: type I AVM (arrowhead).

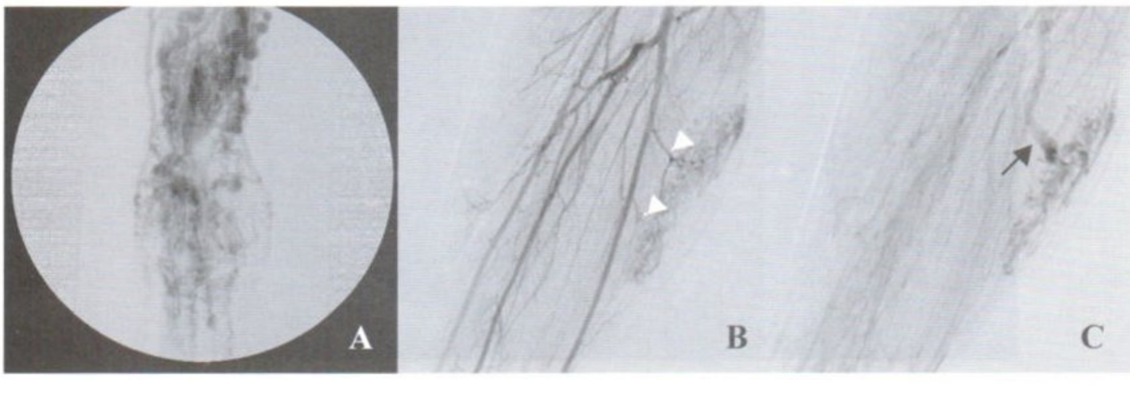

Fig.5 Angiogram of Type IIIb AVM: (A): Infiltrative AVM involving forearm and hand that cannot embolize, due to unsafe to treat. (B): arterial and (C): venous phases of another patient: localize small AVM at the forearm, arterial feeding (arrowheads) and early draining veins away from the nidus (arrow). The initial MRA from other institute showed localize AVM (not show). This AVM was consulted for pre-operative embolization.

feeders hindering superselection into the nidus. Effectiveness of embolotherapeutic outcome in this study was 9 of 12 AVMs (75%)[Table 3], all of which were considered to be in partial remission. No remission was found in 3 AVMs (25%) (patients 9,10 and 11). There was no cure or aggravation of AVM in this study. Embolotherapeutic outcome could not be evaluated in one patient (patient 4) due to consultation for pre-operative embolization 1 day before surgery.

Concerning clinical outcome, symptoms and signs were completely resolved in 1 of 10 AVMs (10%)(patient 1), improved in 7 AVMs (70%) and unchanged in 2 AVMs (20%). Clinical outcome could not be assessed in three patients. Patient 4 presenting with a mass could not be evaluated in an early post-embolization setting. Also two other patients (patients 5 and 6-1) were asymptomatic before embolization [Table 3]. The degree of devascularization was 76-99% in 7 of 13 AVMs (54%), 50-75% in 23% and less than 50% in 23% [Table 3]. Surgery was previously performed on five

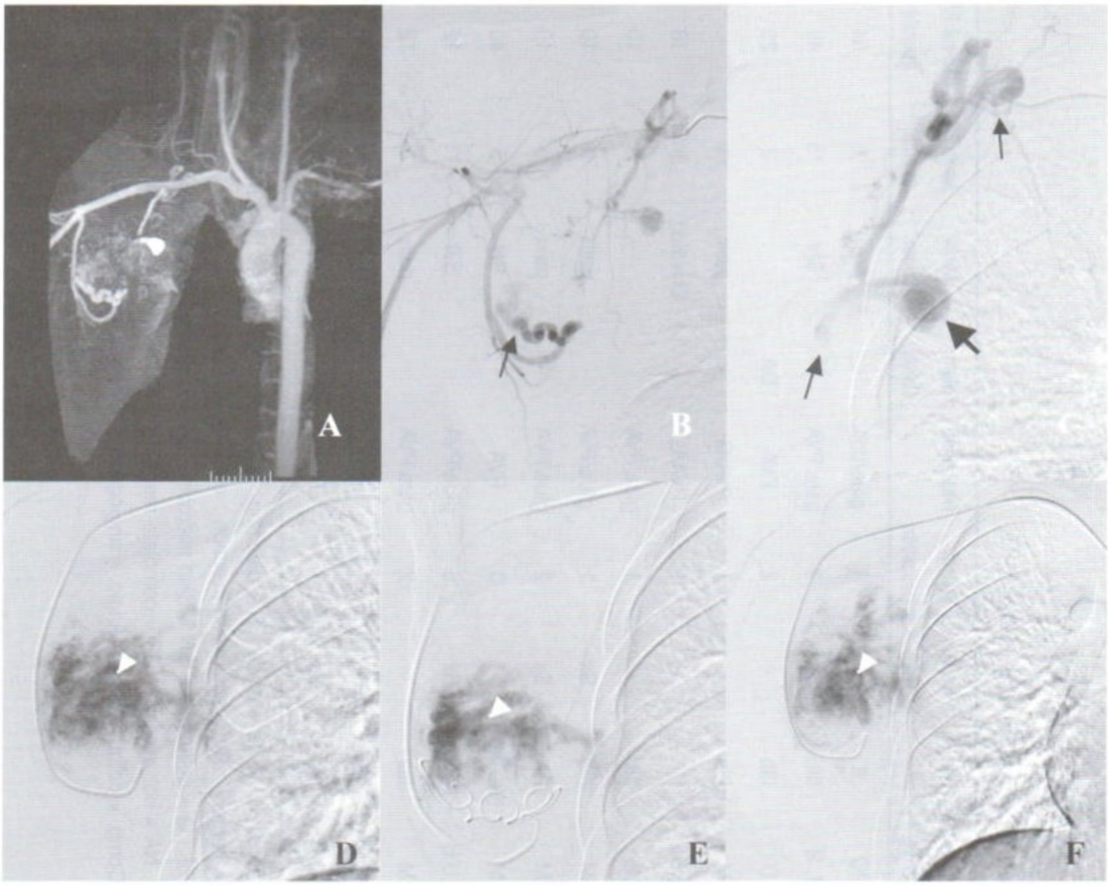

Fig.6 Angiogram of AVM at right-sided back in the patient 10: (A): 3D MRA showed AVM, (B): Early arterial phase showed tortuousity of the arterial feeding (thin arrow), (C): another arterial feeding (thin arrow) and aneurysm (large arrow), (D): late arterial phase showed complex network of the nidus (arrowhead), (E): Embolization using PVA by supperselective technique, (F): Post embolization angiogram showed less than 50% devascularization. Note failure to select the arterial feeding in C.

Table 3 Clinical data and outcome of embolotherapy

Patient No./ Sex/Age (y)	Location of AVMs	Symptoms and Signs[#]	Type	Previous Tx[$]	No. of embolization	Treatment[@]	Major Comp.	Minor Comp.[Ɛ]	Angio.F/U (month)[¥]	Clinical F/U (month)	Degree of Devas. (%)	Clinical Outcome[§]	Therapeutic Outcome[§]	Status of Therapy	Tx after em.[$]
1/F/33	pelvis	PD	II	-	3	glue/coil	-	pain	1	64	76-99	CR	PR	complete	-
2/M/24	leg	M, B	IIIb	Sx	3	glue/PVA	-	SN	15	49	76-99	improve	PR	complete	Scl
3/F/21	forearm	U	IIIb	-	2	PVA	DA	-	2	23	76-99	improve	PR	complete	Sx/Scl
4/F/11	back	M	IIIa	-	1	PVA/G	-	pain	Im	-	76-99	NA	NA	loss F/U	Sx
5/M/35	pelvis	No	II	-	1	PVA/G	-	Erection	Im	18	76-99	NA	PR	complete	-
6-1/M/30*	pelvis	No	IIIb	-	1	glue/PVA	-	-	6	10	76-99	NA	PR	complete	-
6-2/M/30*	thigh	M	IIIb	Sx	1	glue/PVA	-	pain	6	10	76-99	improve	PR	complete	-
6-3/M/30*	leg	M	II	Sx	2	glue/PVA	-	pain	6	10	50-75	improve	PR	ongoing	-
7/F/21	hand	M	IIIa+IIIb	-	3	PVA	-	SN	3	40	50-75	improve	PR	complete	Sx/Scl
8/M/55	leg	M,U,B	I+IIIb	Sx	6	glue/PVA	-	SN	11	12	50-75	improve	PR	refuse	-
9/F/10	arm	M	IIIa	-	2	glue/PVA	-	pain	6	12	<50	improve	NR	ongoing	-
10/M/26	back	M	IIIb	-	1	PVA	-	pain	Im	16	<50	no change	NR	ongoing	-
11/F/28	foot	M	IIIb	Sx	1	PVA	-	pain	Im	18	<50	no change	NR	complete	Sx

* Patient 6th has three lesions. # M = mass B = bleeding U = ulcer PD = pelvic discomfort No = no symptom

$ Tx = treatment Sx = surgery Scl = sclerosing injection em. = embolization

@ PVA = polyvinyl alcohol G = gelfoam Ɛ SN = Skin necrosis DA = dissected artery (ulnar artery)

¥ Im = immediate § NA = not assess CR = complete resolving PR = partial remission NR = no remission

Note: No. = number Comp. = complication Angio. = angiogram Devas. = devascularization

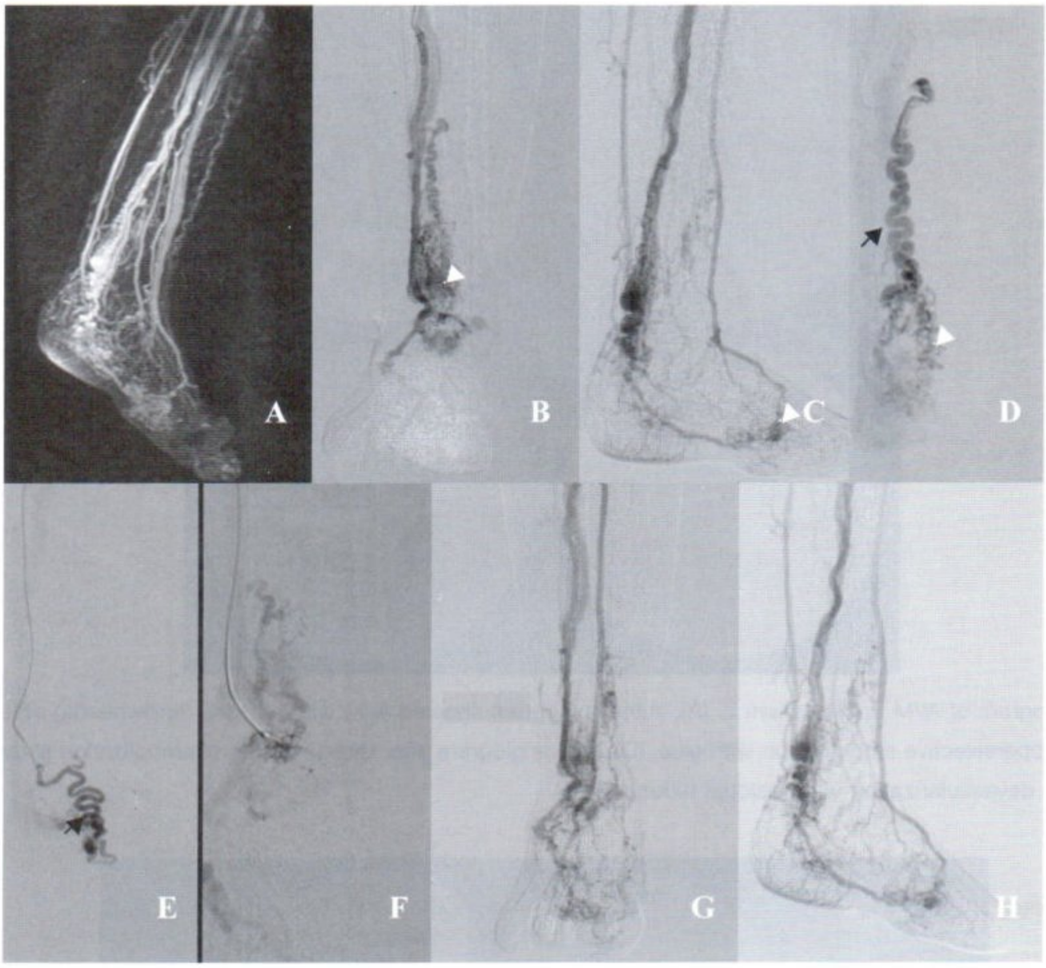

Fig.7 Angiogram of AVM at left foot in the patient 11: (A): 3D MRA and (B, C): arterial phase of PA and lateral angiogram showed infiltrative AVM with nidus (arrowhead). (D, E and F): superselective technique that the microcatheter just located into tortuous arterial feeding (arrows). (G and H): Post embolization angiogram showed less than 50% devascularization.

AVMs (38%). Number of embolizations in this study ranged from 1 to 6 settings (mean = 2.1±1.4 settings). The most number of embolizations was performed in patient 8. The time of follow-up angiography ranged from 0 to 15 months (mean = 4.3±4.7 months), the longest interval found in patient 2. The clinical follow-up time ranged from 10 to 64 months (mean = 23.5±17.8 months). All patients had no symptoms or signs of recurrence. The most used embolic agent was polyvinyl alcohol (PVA) in 12 AVMs. Other embolic agents included glue, coil and gelfoam [Table 3].

In the group of partial remission of therapeutic outcome, the degree of devascuralization was 76-99% in 7 AVMs (70%) [Fig.8] and 50-75 % in 3 AVMs (30%)[Fig.9 and 10]. The best outcome of embolization was found in 3 AVMs (patients 1, 5 and 6-1). These AVMs were decreased in degree of

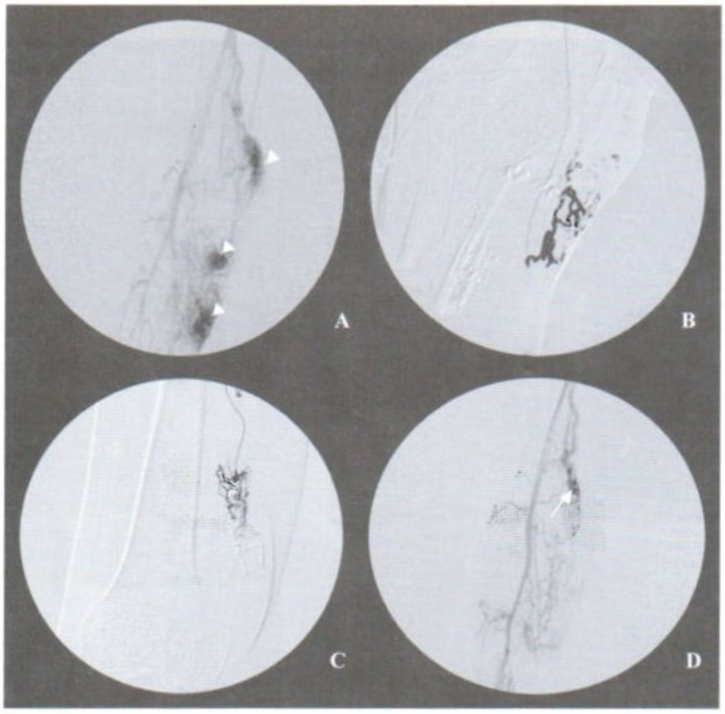

Fig.8 Angiogram of AVM in the patient 2: (A): initial angiogram showed AVM with niduses (arrowheads) at leg. (B and C): superselective embolization with glue. (D): final angiogram after three settings of embolization showed about 80% devascularization with residual nidus (arrow).

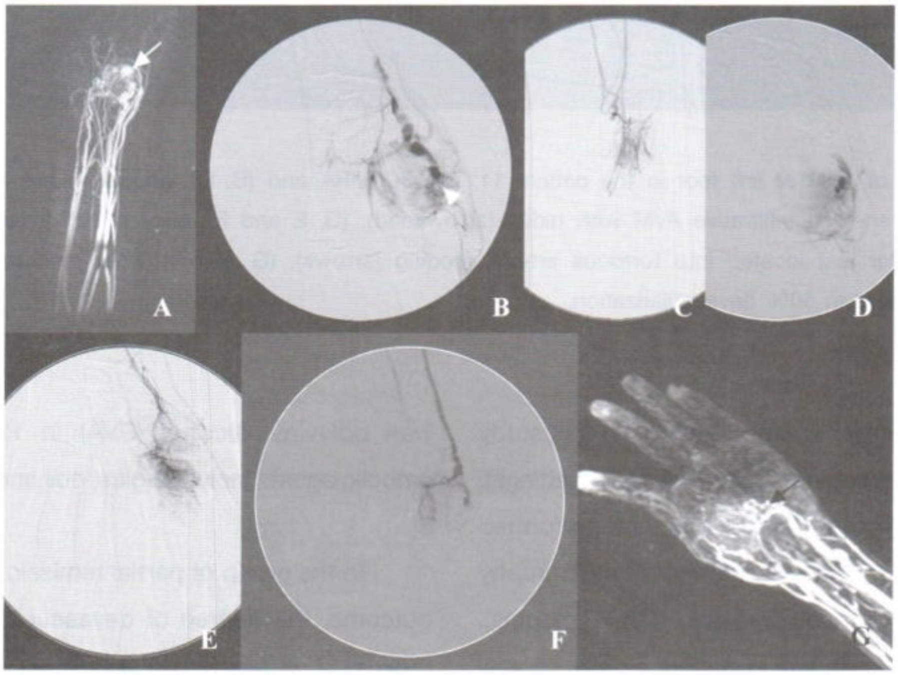

Fig.9 Angiogram of AVM in the patient 7: (A): 3D MRA and (B): late arterial phase of angiogram showed AVM (arrow) and nidus (arrowhead). (C, D and E): superselective technique with PVA embolization. (F): final angiogram after three times of embolization showed about 70% devascularization. (G): 3D MRA after two years of embolization showed small residual AVM (arrow).

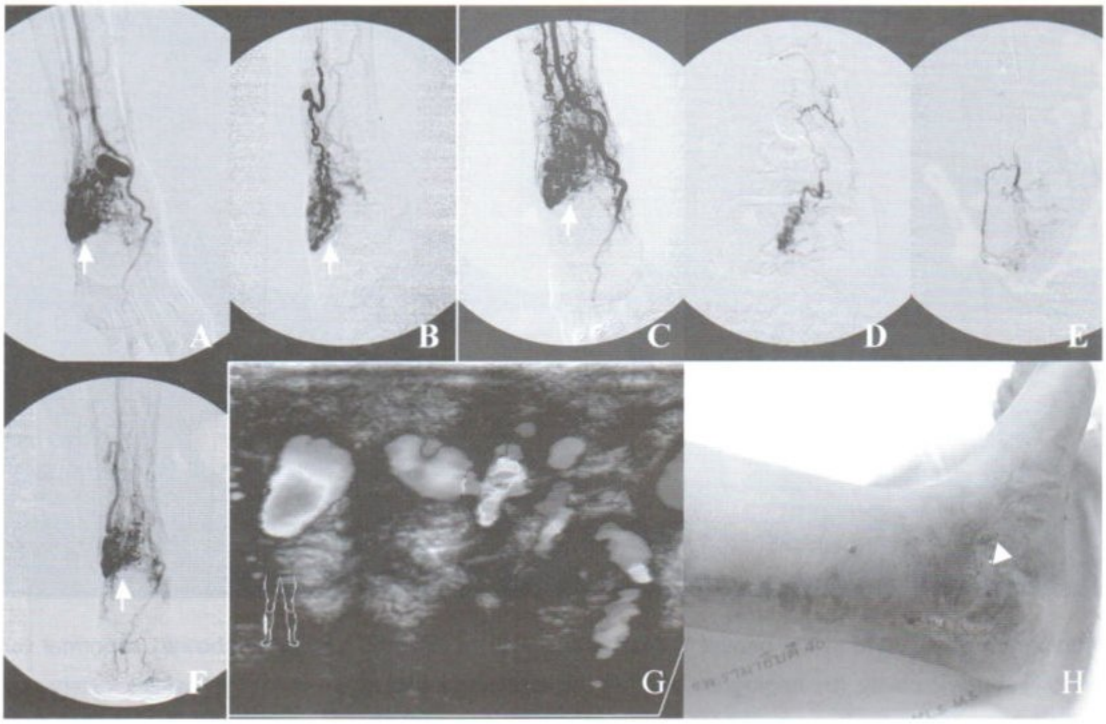

Fig.10 Angiogram of AVM in the patient 8: (A): arterial phase of left foot angiogram showed AVM with nidus. (B): superselective with PVA and glue embolizations. (C): angiogram after three settings of embolization showed 20% devascularization. (D and E): superselective of 4th and 5th embolizations. (F): Post final embolization angiogram showed about 60% devascularization. (G): Doppler US after six years of embolization showed residual AVM. (H): scar after treatment of skin necrosis (arrowhead). Note nidus (arrow).

vascularity more than 90% [Fig.11 and 12] with complete resolution of symptoms in patient 1. Additional treatment was found in three AVMs because of difficult access and high risk of limb ischemia. One AVM (patient 2) was treated by sclerosing agent injection (3% ethoxysclerol), and two other AVMs (patients 3 and 7) were treated by combined excision and sclerosing agent injection after embolization. Seven AVMs underwent complete treatment. One patient (patient 8) refused next embolization or additional treatment after the sixth embolization. Another AVM (patient 6-3) is awaiting further sessions of embolization.

In the group of no remission, all AVMs (patients 9,10 and 11) were less than 50 % devascuralized [Table 3]. One patient (patient 9) was clinically improved and awaits next embolization. There was technical failure in two patients, which did not change clinical outcome after embolization. One patient (patient 10) awaits surgery. Another AVM (patient 11) was excised after failed embolization.

There was one major complication in this study. In one patient (patient 3), ulnar artery dissection occurred during embolization with no symptoms or signs of distal ischemia due to multiple collateral arterial supply from the radial artery and partial recanalization on follow-up imaging[Fig.13]. Five patients (7 AVMs, 53.8%) experienced pain. Skin

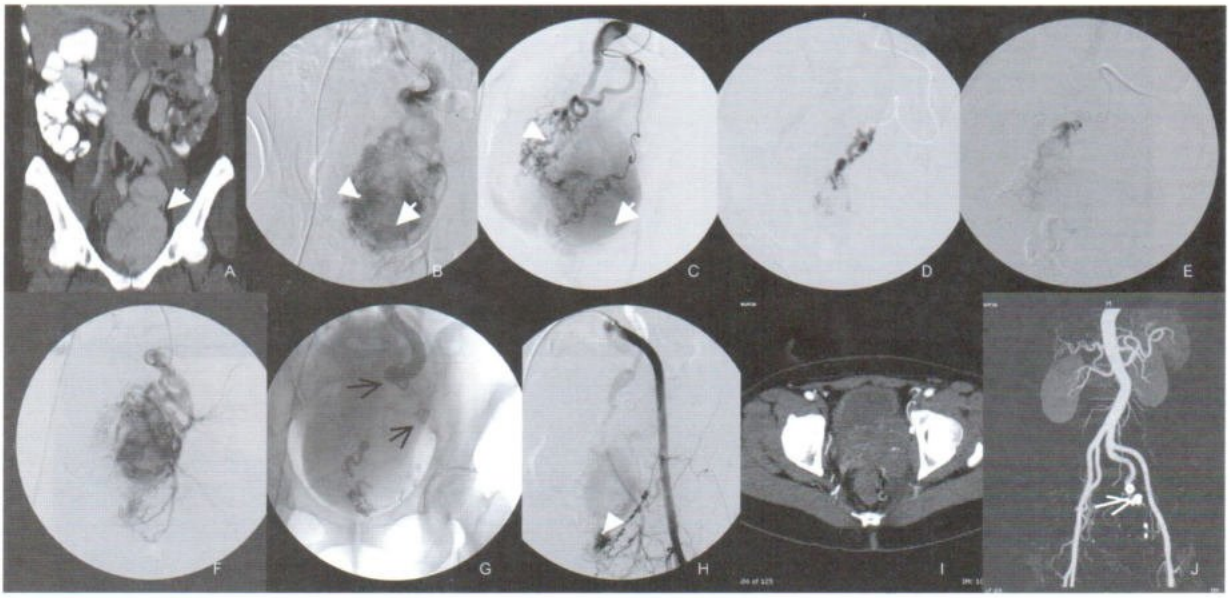

Fig.11 Angiogram of the AVM in the patient 1: (A): coronal MIP of initial CT abdomen showed abnormal venous lake (arrow) in pelvic cavity, (B): angiogram and (C): superselective angiogram of left internal iliac artery showed type AVM with nidus (arrowheads) and large venous pouch (arrows), (D, E): superselective embolization with glue, (F): pre-embolization of final angiogram showed 50% devascularization, (G): multiple coil embolizations with complete occlusion of the left internal iliac (thin arrows), (H): 90 % devascularization with minimal residual nidus (arrowhead) and arterial feeding from branch of left external iliac artery, (I) axial and (J) 3D reformat of the CT angiogram after six years of embolization showed total thrombosis of venous pouch and no recurrence of AVM. Note coils (thin arrow).

necrosis occurred in three patients (23.1%) [Fig.10H]. Another patient (patient 5) had transient frequent erection of the penis. No complications occurred in one lesion (patient 6-1).

Discussion

Congenital AVMs are present at birth, though they may remain clinically silent until later in life. Embryologically, the vascular system evolves through three stages. In the first stage, called the stage of minimal differentiation, blood lakes organize into capillary networks. The second stage is the retiform stage in which capillaries form larger conduits, but no vessel wall differentiation is seen. The final stage of adult maturation begins with the completion of vessel wall maturation. It is felt that the majority of these congenital lesions evolve if a vessel fails to reach the latter stages of differentiation.

According to Cho et al. modified classification[5] of the peripheral AVM type [Fig 1.], the angiographic findings in this study showed that 55.6 % were type IIIb, 16.7% type II, 16.7 % type IIIa, 5.7 % mixed type I and IIIb and 5.7 % mixed type IIIa and IIIb. The original Cho et al. modified classification that reviewed angiographic findings of peripheral AVMs in 66 patients from December 1996-December 2004 showed 45% type IIIb, 20% type II, 18% mixed type IIIa and IIIb, 14 % mixed type II and IIIb and 3% type IIIa. When compared, two studies showed the same results, that is type IIIb [Fig.5,7,8 and 13]

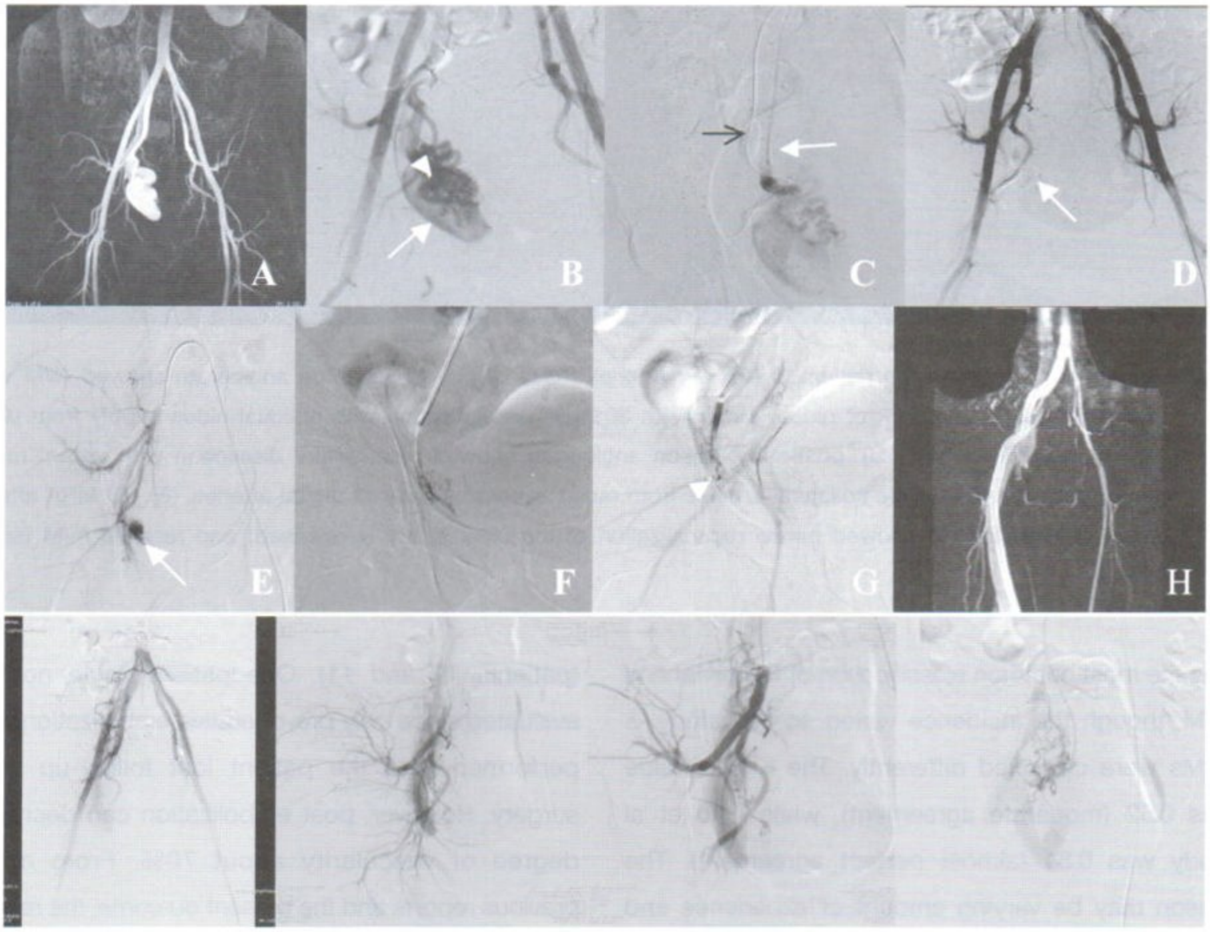

Fig.12 Angiogram of AVM in the patient 5 (A-D): (A): 3D MRA showed AVM in pelvic cavity. (B): late arterial angiogram showed type II AVM with nidus (arrowhead) and venous pouch (large arrow). (C): embolization with PVA by combine transarterial (large arrow) and temporary transvenous balloon occlusion techniques (small arrow). (D): post-embolization angiogram showed nearly complete occlusion of AVM (large arrow). Another angiogram of small AVM in patient 6-1 (E-H): (E): arterial phase angiogram showed small type IVb AVM (large arrow). (F): superselective glue embolization. (G): post-embolization angiogram showed 95% devascularization with retained glue material in arterial feeding (arrowhead). (H): 3D MRA after 1 month of embolization showed no residual or recurrent AVM.

The patent comes back for clinical of recurrent disease, seen from follow up MRA 2 years later. The angiogram (four pictures of the third row of fig.12) showed multiple fine vascular feeding to AVM without demonstrable AVM nidus or significant early draining vein. However, there is still seen large draining vein from the right sided of pelvic cavity. The second partial embolization of the arterial feeding was done with residual some fine feeding artery.

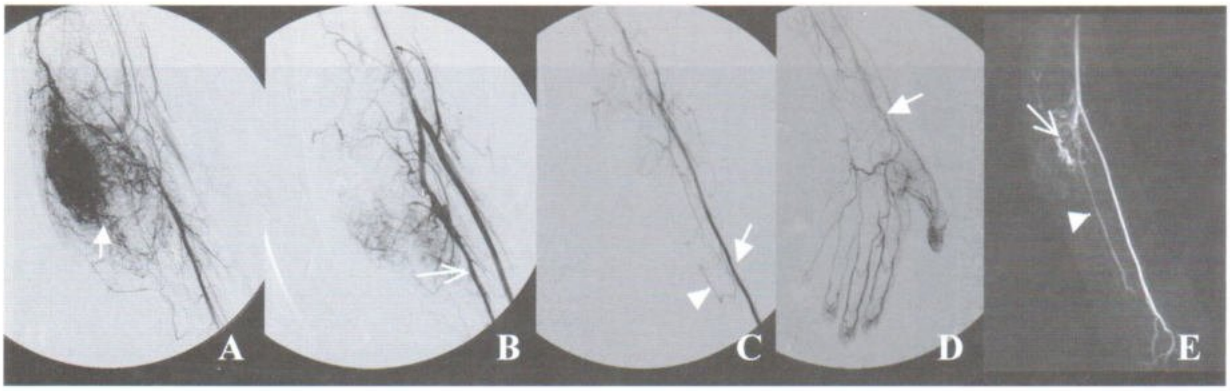

Fig.13 Angiogram of AVM in the patient 3: (A): late arterial phase of pre-embolization angiogram showed AVM with complex vascular network of nidus (arrow), (B): 80% devascularization with residual nidus supply from ulnar artery (small arrow), (C, D): post-embolization angiogram showed ulnar artery dissection with patent radial artery (arrow) and multiple collateral arteries from radial artery to supply all digital arteries, (E): 3D MRA after 3 years of embolization showed partial recanalization of the ulnar artery (arrowhead) and residual AVM (small arrow).

was the most common classification of the peripheral AVM, though the incidence varied. In this study, 5 AVMs were classified differently. The Kappa value was 0.52 (moderate agreement), while Cho et al study was 0.81 (almost perfect agreement). The reason may be varying amount of experience and the small number of the AVMs in this study.

In fact, the aim of AVM treatment outcome is to improve clinical signs and symptoms as well as partially decrease degree of vascularization. Complete devascularization is quite difficult; however, it should be done if possible. Of the many previous studies on embolotherapeutic treatment of congenital AVMs, all reported improved clinical outcome (complete and partial improvement).[5,13,14,17] Recently, Do YS et al (2005)[6] reported the therapeutic outcome of AVM embolization in 40 patients using ethanol from 1996-2003. The results showed 40% cure, 27.5% partial remission, 17.5% no remission, 2.5% aggravated and 12.5% failed. This study showed 75% partial remission and 25% no remission (patients 10 and 11). One patient could not be evaluated since only pre-operative embolization was performed, and the patient lost follow-up after surgery. However, post embolization can decrease degree of vascularity about 70%. From many previous reports and the present outcome, the results showed overall improvement of clinical and therapeutic outcomes. Therefore, embolotherapeutic modalities have advantages in the treatment of high-flow peripheral AVMs. In addition the patient's compliance is an important factor because many high-flow AVMs cannot be treated with good outcome during a short period of embolization.

Historically, surgical ligation of the feeding arteries to the AVM was commonly performed to decrease the blood supply to the lesions. However, this treatment was often unsuccessful as a result of AVM nidus revascularization by microscopic shunts that were not apparent before surgical ligation. Moreover, these shunts are often numerous, small, tortuous, and difficult to access, making subsequent transcatheter treatments more difficult and often impossible.[3] Recently, embolization has been suggested as the primary therapeutic modality for cure, improvement or presurgical intervention to reduce bleeding and maximize successful resection. There were several studies that mentioned unsuccessful surgical treatment.[3,5,6,11,15] In this study, there were 5 AVMs that recurred after surgery [Table 3]. Four AVMs were in partial remission after embolization while there was no remission of one AVM due to difficult access [Fig.7]. However, surgical treatment has a role in small or localized lesions, embolization failure or safety concerns. In this study, there were 5 non-embolized AVMs that were sent for surgery. 4 other AVMs were also sent for surgery after embolization. Further embolization was deemed unsafe in three lesions, and there was technical failure in one lesion (patient 11) [Table 3]. Additional treatment by sclerosing agent injection was also found in three AVMs for the same reasons as surgical treatment.

In general, transarterial embolization has been preferentially used to embolize AVMs. Recently, transvenous or direct puncture embolization has been used as an alternative technique when important normal arterial branches arise in very close proximity to a malformation or when extreme arterial tortuosity or previous treatment (including surgical ligation and embolization of the feeding artery) preclude successful transarterial catheterization.[5,8] These techniques will increase chance of cure. In our institute, the interventionists still prefer transarterial approach with or without coaxial microcatheter technique for embolization [Fig.6E, 11D and 11E] because of their experience. For this reason, the number of embolizations and angiographic follow-up time were quite less than other previous reports (6) [Table 3], and there was technical failure from transarterial approach in two AVMs [Fig.6 and 7]. However, in one AVM, temporary transvenous balloon occlusion technique was used to decrease flow before transaterial embolization by PVA, which resulted in about 95 % devascularization [Fig.12A-D].

From years past to currently, many endovascular occlusive agents have been used including autologous clot, Gelfoam, Polyvinyl alcohol particle (PVA), various metallic coils with or without fibers, tissue adhesives (IBCA/NBCA), detachable balloons, Ethibloc, Sotradecol and ethyl alcohol.[3,13,18-20] The technique can be applied though transarterial, transvenous or direct puncture approaches to permanently occlude the nidus. Gelfoam, PVA, coils, or detachable balloons rarely cure peripheral AVMs.[3,7,13,18] Tissue adhesives (IBCA/NBCA) were initially thought to be permanent occluding agents. However, their use is difficult, and it is now well-documented that recanalization does occur. In this study, combined embolic agents were more commonly used than single embolic agents. The most common embolic agent was PVA (12 from 13 AVMs), [Fig.9C-E] and second most common was glue (7 from 13 AVMs) [Fig.8B and 8C] [Table 3]. In one pelvic AVM (patient 1) receiving arterial supply from the left internal iliac artery, coil and glue was used, resulting in excellent outcome (90% devascularization and complete resolution of symptoms) [Table 3, Fig.11]. In general, proximal embolization of the feeding artery should be avoided because the lesion recruits collateral flow through more complex.[12] However, in the last embolization of patient 1, coil was use to occlude the left internal iliac artery [Fig.11G] because of near complete occlusion of the nidus from two previous procedures and the clinician's requirement of cure as the outcome. No serious complications occurred. From review of medical records, this patient underwent a normal pregnancy and labor without complications. The last CT follow-up (December 2008) showed no residual, recurrence or aggravation of disease [Fig.11I and J].

Recently, various reports document that the use of ethanol embolization has the potential for cure in management of AVMs of the body and extremities. However, this technique has great risks, including injury to adjacent normal tissues, particularly the mucosal surface, skin and neurologic tissue. Cardiopulmonary collapse may also be caused by ethanol escape to the right-side heart and pulmonary bed so that close monitoring by the interventionist team is required during the procedure.[3,13,20,21] In our opinion, the selection of embolization technique and embolic agent depends on the availability of materials and instruments in that institute. Moreover, the experience of the interventionists is important. However, in the future, we will try and apply alternative techniques for increased effective outcome in our institute.

One major complication, ulnar artery dissection, occurred in this study, but there were no symptoms and signs of distal ischemia due to multiple collateral arteries from the radial artery and partial recanalization on follow-up imaging [Fig.13B-E]. The rest were minor complications [Table 3]. All cases of skin necrosis healed with wound dressing, and no skin graft was needed [Fig.10H]. Transient frequent erection of penis in patient 5 resolved two days after embolization. The cause may have been increased arterial supply to the penis after AVM was occluded. Pain was resolved a few days to weeks after embolization. Other studies reported complications after embolotherapy. Tan KT. et al[3] in 2004 found 2 major complications including tibial plateau fracture and L5 neuropathy. Cho SK. et al[5] in 2006 reported embolism, brain infarction, bladder necrosis, infection, permanent nerve injury and acute renal failure. Occurrence of reported complications might be dependent on technical approach, embolic material used, location and aggressiveness of the AVM. In this study, there were no serious complications, probably due to aim of palliative embolization. Also, lesions that could not be embolized without placing normal tissue at undue risk were not treated.

There are some limitations in this study. First, the number of AVMs is small due to rarity of the disease, which is appropriate with 4 periods of collected data. Second, the evaluation of AVM type is based on angiographic morphology alone without pathologic confirmation. Third, because of varying embolotherapeutic techniques and embolic materials used in many studies, it may be difficult to compare our results. Fourth, due to the invasive nature of angiography, a routine follow-up angiogram cannot be performed. We compared post-embolization images of the last angiogram with the first angiogram before embolization in almost all cases. Non-invasive imaging, including color doppler sonogram and MRI were just indirect modalities for follow-up. Fifth, clinical follow-up time may not have been enough to assess recurrence.

Conclusion

Congenital arteriovenous malformation (AVM) is a rare disease. Type IIIb (arteriolovenulous fistulae with dilated fistula) was found to be the most common type of peripheral AVM. Transcatheter arterial embolization with/without superselection is the primary therapeutic modality in the management of patients with peripheral AVMs for improvement, possible cure or presurgical intervention to reduce bleeding and maximize successful resection. The morbidity is also less than primitive surgical excision. Further long-term assessment is necessary to evaluate efficacy of this treatment and recurrence.

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