10 Angiographic Classification: Arteriovenous Malformation and Venous Malformation

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Introduction

Angiography is the gold standard of confirmative diagnosis as well as therapeutic tools in arteriovenous malformation (AVM). Typically, artery angiography in AVM shows feeding artery, nidus, and early draining veins. These malformed vascular structures can be imaged through the full-shot arteriography, but it is hard to separate individual vascular component only with a single overall angiographic image. Angiographic findings of AVM are totally different in every single patient, and the malformed vasculature is very complex to figure out the detailed vascular connection at a glance. Not infrequently, untrained physician cannot discriminate even between the artery and vein on angiography because the vessels are numerous, tortuous, and overlapped with each other. Therefore, systematized angiographic classification for the AVM seldom appears in the medical literatures. Complex vascular connection in AVM can be imaged better with selective arteriography, and sometimes direct puncture arteriography is helpful for understanding detailed vascular connection and hemodynamic status of AVM components vessel. Without understanding vascular anatomy and its hemodynamic interaction of the malformed vessels in the AVM, adequate treatment plan is difficult to make. As for the endovascular treatment, some type of AVM responds to treatment dramatically, but some AVM is hard to relieve even with repetitive procedures. Therefore, types and patterns of different AVMs have been required to be classified systematically to correlate with the treatment response.

For the intracranial AVM, several attempts were made to suggest systematized classification since 1977. Among them, Spetzler and Martin grading system [1] graded AVM with point scale considering the size of AVM nidus, cortical location, and the deep venous drainage. However, the number and connection pattern of feeding artery was not included in the classification system. Feeding artery in AVM can be single or multiple, and they usually show extremely tortuous appearance due to the increased arterial flow volume heading to arteriovenous shunt. Shi-Chen scale divided the size of AVM, the location and depth, arterial blood supply, and venous drainage into grades 1–4, and then the final grade was determined from each of four patterns of grades [2]. However, unlike the intracranial AVM, the importance of location factor is not so great in torso and extremity AVM. Furthermore, although the feeding artery, nidus, and draining vein can be seen on angiography, anatomical lesion location requires additional cross-sectional image data such as magnetic resonance imaging or

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computed tomography. Therefore, simplified angiographic classification of intracranial AVM proposed by Houdart et al. on 1993 considered to be an adequate model for application in peripheral AVM [3]. In this simplified classification system, AVM was divided into three different types, and the number and size of the feeding artery and vein as well as the vascular connection between the feeding artery and vein were the main consideration for the angiographic description and classification.

Based on these concept, Cho et al. proposed modified angiographic classification for the AVMs in the torso and extremity on 2006 [4]. Diagram in the Fig. 10.1 describes the vascular anatomical connection between feeding artery and draining veins. Type I was defined as an arteriovenous fistulae that consist with not more than three different feeding arteries shunt to a single draining vein which was a single direct arteriovenous fistula. Type II refers to arteriolovenous fistulae that consist with multiple arterioles shunt to

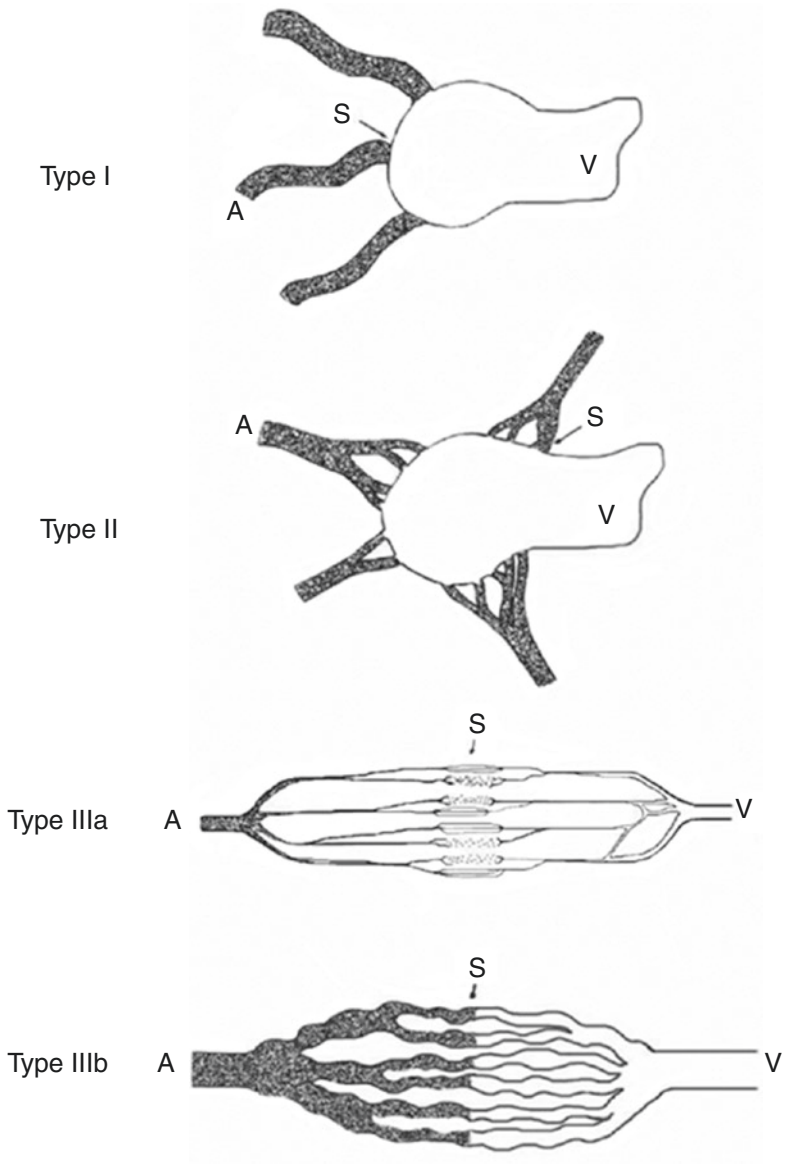

Fig. 10.1 Diagram for the angiographic classification of AVM

a single draining vein. Arterial components in type II AVM show a plexiform appearance on angiography. Ordinarily, both feeding arteries and draining vein are hypertrophied and tortuous, but the degree of vascular tortuosity does not influence on the angiographic classification in type II. Type III AVMs have multiple feeding arteries and multiple draining veins and subdivided into type IIIa which is arteriolovenulous fistulae with non-hypertrophied feeding artery and draining vein. Type IIIb is arteriolovenulous fistulae with hypertrophied feeding arteries and draining veins. Figure 10.2 shows typical angiographic appearance of AVM corresponding to each subtype of AVM. Response to embolization therapy is relatively good in types I and II. Both type I and II AVMs have dominant outflow vein, and the rate of cure in embolization was reported

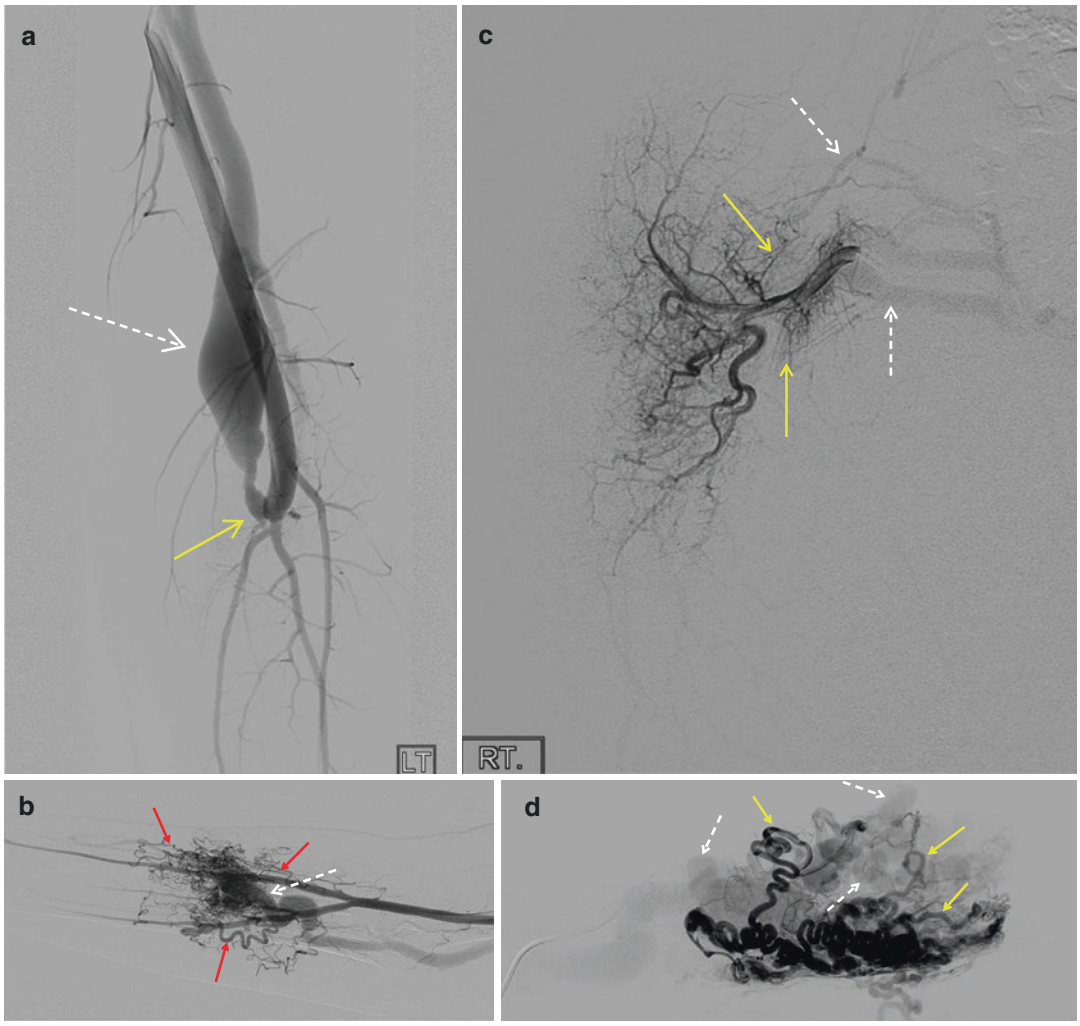

Fig. 10.2 Four subtypes of AVMs according to the angiographic classification. (**a**) Type I AVM with arteriovenous fistulae. Single feeding artery from tibioperoneal trunk (*arrow*) connected to single draining vein (*dotted arrow*). (**b**) Type II AVM with single early draining vein in the right forearm. Multiple feeding arteries (*red arrows*) are connected to single early draining vein (*white dotted arrow*). (**c**) Type IIIa AVM in the right buttock. Non-hypertrophied arterial feeders (*arrows*) are connected to multiple non-hypertrophied draining veins (*dotted arrows*). (**d**) Type IIIb AVM in the left upper arm. Multiple hypertrophied feeding arteries (*yellow arrows*) are connected to multiple hypertrophied draining veins (*white dotted arrows*)

up to 68 % [5]. On the other hand, types IIIa and IIIb show less therapeutic response to embolization. Although the modified angiographic classification system provided clear category between each type of AVM, not all the AVM falls into these four specific types completely. Twenty-four to thirty-two percent of patients are classified into complex AVM that shows more than two different types of AVM are combined [6]. Therefore, perfect angiographic classification is almost impossible to establish to stratify these complex vascular lesions clearly into individual patterns. One study showed that lesion extent is also related with treatment response [6]. Even with type IIIb AVM, localized lesion can show good result with repetitive treatment. However, extensive AVMs involving whole extremity are hard to show satisfactory result. Because recent angiographic classification system for AVM lacks the consideration of lesion extent, further research and treatment experience is required for revising the modified angiographic classification into more reliable and well correlated with therapeutic results.

Venous Malformation: Angiographic Classification

Venous malformations occupy the largest population in the congenital vascular anomaly and are responsible for more than 50 % of patient referrals to vascular anomaly centers [7, 8]. VMs are composed of abnormal veins that show variable luminal diameter and wall thickness, and they usually lack of venous valve [7]. Certain VMs consist with fine small diameter dysplastic veins but large proportion of VM consists with markedly ectatic and serpentine venous channels. Dysplastic veins are connected with each other in an irregular and disorganized pattern, and VMs have normal venous connection with superficial or deep venous system.

Although full-shot arteriography in AVM is easy to get an overall image for the vascular malformation, VM is hard to be imaged in a single injection phlebography through transvenous approach. Also, the routine ascending or descending phlebography cannot show the whole VM in a single image because the venous flow is coming out from the VM frequently. Therefore, direct puncture phlebography is useful to understand the dysplastic venous connection of the VM in detail, but even with direct puncture phlebography, whole VM cannot be filled with contrast media because the venous drainage of VM component that is apart from the needle insertion point passes through the different venous connection. Furthermore, direct puncture venography is an invasive procedure so that the exact phlebographic classification can be obtained with simultaneous sclerotherapy in many cases. Direct puncture phlebographic findings in VM are very complex in most of the patients; however, careful analysis of direct puncture phlebography provides detailed information for the patterns of normal venous connection and the appearances of dysplastic vein. Phlebographic evaluation sometimes requires manual compression or dispersion of contrast media to the rest of nonopacified VM component and nonopacified normal venous connection. Unless adequate phlebographic evaluation is done, VM cannot be characterized thoroughly. Former biological and clinical classification system like Hamburg classification or ISSVA classification did not reflect venous anatomical and hemodynamic factors [9].

Phlebographic classification for the VM has few data. In 1991, Dubois et al. suggested classification for the VM according to the type of venous drainage as type I was an excluded, well-circumscribed VM without visible draining veins; type II was venous lakes drained into a normal venous system; and type III was VM having ectatic abnormal draining veins [10]. In 2001, Dubois et al. described direct phlebographic findings into three simplified categories: cavitary, spongy, and dysmorphic veins [11]. However, this simple description is insufficient for stratifying the complex malformed venous structures in torso and extremity VMs. In 2003, Dr. Puig proposed modified phlebographic classification system [12]. Puig also considered the pattern of venous drainage and normal venous connection as a clue to stratify four different types of VMs. Figure 10.3 shows typical

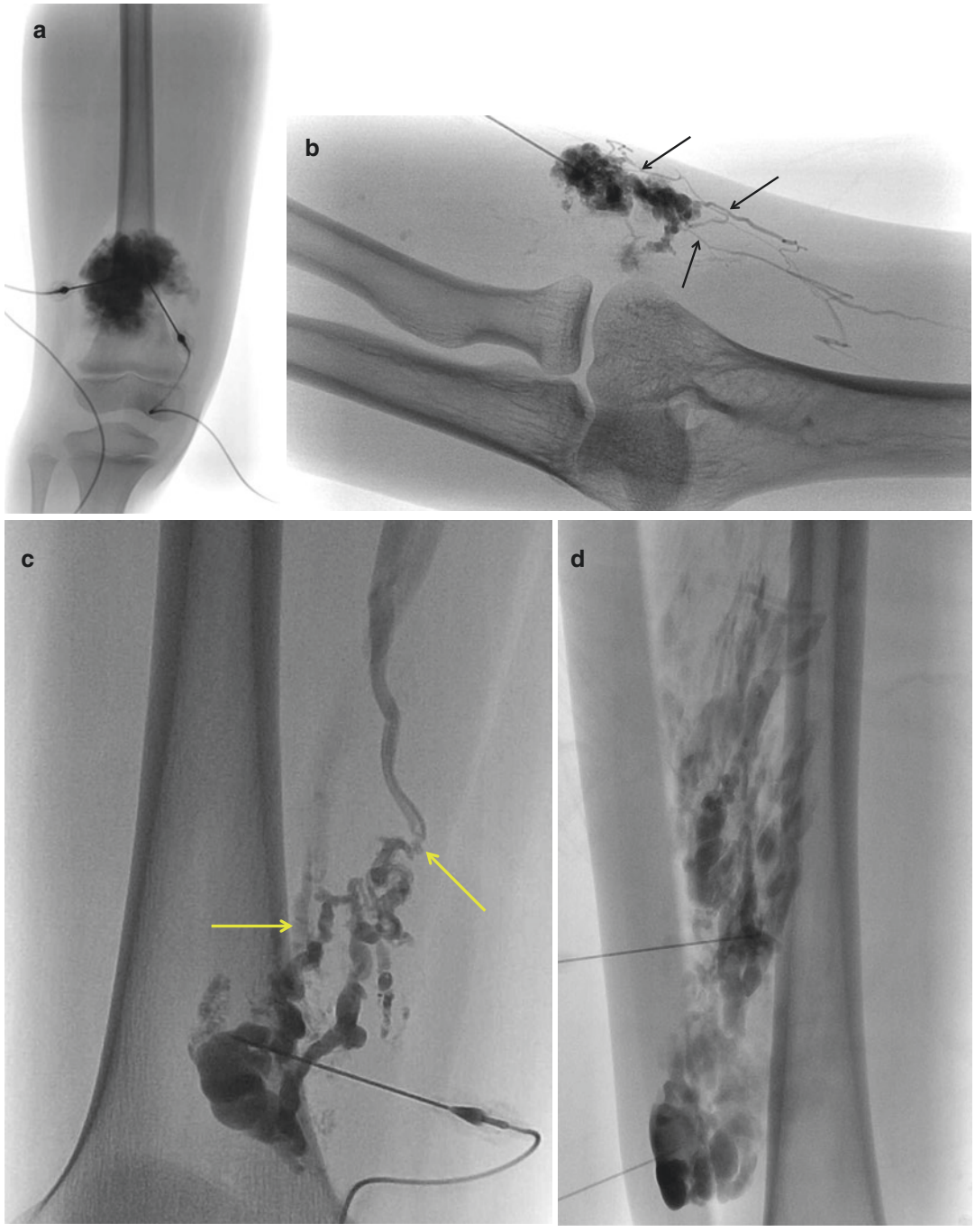

Fig. 10.3 Phlebographic classification of venous malformations. (**a**) Type I isolated VM without peripheral venous drainage. (**b**) Type II VM in the right elbow that drains into normal superficial veins (*arrows*). (**c**) Type III VM that drains into dysplastic veins (*arrows*). (**d**) Type IV VM with diffuse venous dysplasia. Complex, irregular, and ectatic veins are spread in the left thigh

phlebographic appearance of four types of VMs. Type I was defined as an isolated malformation without peripheral venous drainage (Fig. 10.3a). Type II was venous malformation that drains into normal veins (Fig. 10.3b). Type III was venous malformation that drains into dysplastic veins (Fig. 10.3c). Type IV was venous malformation that represents a venous dysplasia (Fig. 10.3d). Incidences of each type of VMs were reported as 30 % for type I, 37 % for type II, 21 % for type III, and 12 % for type IV [12]. However, these incidences can be varied from center to center because the data were derived from small population (43 patients) phlebographic study. The importance of Puig classification was that it emphasized venous anatomical factors including normal venous communication with dysplastic veins. Understanding the vascular anatomy of VM and the hemodynamic connection between dysplastic veins and normal venous channel is helpful to establish the treatment scheme. Puig classification (reference) is well correlated with percutaneous sclerotherapy results. Types I and II show good therapeutic response to sclerotherapy because the sclerosing agent can be confined within VM for sufficient time and sclerosant washout into normal venous system can be minimized. Type I and II VMs showed higher cure rate with lower number of treatment sessions [10]. However, sclerosing agent is hard to stay enough in type III and IV VMs that lead to less therapeutic response. Furthermore, type III and IV VMs have risks for embolic material spillage into the normal venous system that gives rise to systemic complication. Therefore, rate of exclusion from sclerotherapy is higher (up to half of patients) in type III and IV VMs [12]. This phlebographic classification system can be applied to simple VMs on ISSVA classification 2014 [9, 13], not for the combined types. Combined VMs are often more complex in phlebographic findings, and some of the lesions are difficult to get direct puncture venography because the dysplastic veins are too small. Current phlebographic classification lacks in consideration of lesion extent, location, and multiplicity. In a simple manner, phlebographically localized VMs are easy to treat, and the therapeutic response is better because the lesion extent is restricted, and sclerosing effect can be maximized. However, diffuse VMs are less likely to show good therapeutic response because the lesion extent is too broad. Diffuse VMs require too many number of treatment sessions to achieve satisfactory result or even impossible to treat with either sclerotherapy or surgery. The number of treatment sessions and the invasiveness of treatment are also related with the life quality of individual patient. There is a limitation that the phlebographic classification consider only the dysplastic vein, but some VMs have large proportion of solid or fibrous connective tissue stroma as a lesion component. The ratio of vascular component and solid component also affects the result of sclerotherapy [14]. Certain types of VM are hard to get direct puncture phlebography because of the abundant stroma with relative paucity of dysplastic venous channel. Therefore, further research, discussion, and agreement would be required to make improved, reliable, and outcome-related phlebographic classification system for VMs.

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