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Classification of venous malformations in children and implications for sclerotherapy

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Abstract *Objective*: The purpose of this work is to present a simple and descriptive classification system for venous malformations (VMs) that may serve as a basis for interventional therapy, and to test its usefulness in a sample of consecutively referred paediatric patients. *Materials and methods*: The classification system we developed includes four types: type I, isolated malformation without peripheral drainage; type II, malformation that drains into normal veins; type III, malformation that drains into dilated veins; and type IV, malformation that represents dysplastic venous ectasia. The system was prospectively tested using phlebography in a sample of 43 children and adolescents with VMs who were referred for treatment during a 10-month period. Our -hypothesis was that the type of VM would determine whether low-risk sclerotherapy was indicated. *Results*: Thirteen (30%) patients had a type-I VM, 16 (37%) had a type-II, 9 (21%) had a type-III, and 5 (12%) had a type-IV malformation. In more than 90% of patients with a type-I or type-II lesion, sclerotherapy could be performed without any problems. In one third of patients with a type-III VM, sclerotherapy had to be withheld and one of nine (11%) developed a severe complication after therapy. Of the five patients with type-IV lesions, three (60%) had to be excluded from sclerotherapy. *Conclusions*: Our initial results indicate that sclerotherapeutic intervention in patients with type-III and type-IV VMs must be carefully considered, while it can be safely performed in low-risk patients with type-I and type-II lesions.

Keywords Venous malformations · Classification system · Phlebography · Sclerotherapy · Children

Introduction

Congenital venous malformations (VMs) are a diagnostic and therapeutic challenge. Clinical presentations are variable, ranging from asymptomatic birthmarks to life-threatening conditions [1, 2]. They are present at birth and may grow slowly and steadily [3, 4]. VMs can be found anywhere, but mainly occur in the soft tissues. Treatment may be indicated because of appearance or for functional problems or cosmetic disturbances [5]. Symptoms are related to size and location, with pain being the most commonly reported symptom.

Sclerotherapy, the injection of an irritant solution, is suggested as the primary treatment for VMs [2]. Absolute ethanol is commonly used for percutaneous sclerotherapy of large VMs, and several authors have also suggested Ethibloc (Ethnor Laboratories/Ethicon, Neuilly, France) [6, 7]. Sodium-tetradecyl-sulfate (Thrombovar; Aventis Pharma France, Laboratoires Chiesi S.A., Courbevoie, France) is preferred for smaller lesions. Some groups have used absolute alcohol for lesions of all sizes [2, 8, 9, 10, 11, 12]. Regional complications of sclerotherapy include skin necrosis, ulcers or peripheral nerve damage [1, 12, 13]. In addition,

systemic effects such as pulmonary vasospasm, pulmonary embolism or direct cardiotoxicity may occur [12]. At least some of these complications are due to central embolisation of the sclerotic agent. Therefore, when planning sclerotherapy, clinical results and possible complications must be weighed against each other.

According to the biological classification of Mulliken and Glowacki [14] proposed in 1982, VMs are slow-flow malformations. This classification is based on cellular turnover, histology, natural history and physical examinations, and clearly separates haemangiomas, which are tumours with an early proliferative and a later involuting stage, from vascular malformations, which are capillary, lymphatic, venous, arterial or a combination of these types [2, 14, 15, 16]. In 1988, further refinements led to the Hamburg classification of congenital vascular defects [17, 18], where the malformation is first described by the predominant component of the vascular defect and then classed as truncular or extratruncular depending on the embryonal stage when developmental arrest occurred. Such classification systems are useful for making a precise clinico-anatomical diagnosis, serve as a basis for treatment choice, and facilitate communication among different specialists [17, 18].

VMs that are included in the above two classification systems have a wide range of anatomical appearances and haemodynamics. Specifically, the connection to the normal venous system varies and has not been taken into consideration by these systems. Therefore, we decided to develop a new classification system focusing on VMs and based on anatomical and haemodynamic features. The purpose of our study was to describe the development of this system and its empirical use in paediatric patients treated for VMs.

Materials and methods

Our aim was to create a classification system that encompasses all existing types of VMs and is as schematically simple as possible. The panel of physicians that met for this purpose decided to categorise the lesions according to anatomical and haemodynamic features, an approach that represented a synthesis of many years' experience in the treatment of VMs and an extensive review of the literature. The resulting classification system included four types of VMs:

- Type I – isolated malformation without peripheral drainage
- Type II – malformation that drains into normal veins
- Type III – malformation that drains into dysplastic veins
- Type IV – malformation that represents a dysplasia

To test the usefulness of this system empirically, we prospectively evaluated the VMs of 43 children and adolescents (mean age 6.7 years; range 3 weeks–17 years; 16 girls, 27 boys). These patients were referred for treatment at our department consecutively during a 10-month period. The VMs were localised in the head in 19 (44%) patients, the neck in 1 (2%), the trunk in 6 (14%), an upper extremity in 7 (16%), and a lower extremity in 10 (23%). Informed consent for therapy was given by parents. Approval of the institutional review board was not required for this type of study.

To evaluate the anatomy and haemodynamics of the VM, phlebography was performed on all patients. For this, the lesions were punctured percutaneously and nonionic contrast medium (Omnipaque 300 mg I/ml, Nycomed Amersham, Paris, France) directly administered via a 20-G catheter. According to the characteristics of the VM visible with phlebography, two radiologists classified the lesions by consensus. In no cases were these two radiologists the same as the radiologist who performed all the phlebographies and subsequent interventions.

The decision to perform sclerotherapy was made by the interventional radiologist, regardless of the classification of the lesion. Our hypothesis was that in type-I and type-II lesions, an intervention could be performed without risk, while in VMs of type III and IV a risk of central embolisation was to be expected due to the wide vascular or dysplastic venous channels, and thus the decision for sclerotherapy had to be carefully considered. For sclerotherapy, one of the following sclerosing solutions was used: pure ethanol, a mixture of ethanol and Ethibloc, or Thrombovar. The interventions were performed under fluoroscopic control. For the treatment of VMs localised in an extremity, tourniquets were used. All procedures required general anaesthesia.

Results

The results of our efforts to classify the VMs are displayed in Table 1. There were 13 of 43 (30%) children with a type-I VM, 16 of 43 (37%) with a type-II, 9/43(21%) with a type-III, and 5 of 43 (12%) with a type-IV malformation. Examples of the different types are shown in Figs. 1, 2, 3, 4.

Of the patients with a type-I VM, all experienced complication-free sclerotherapy, except for one who was not treated interventionally because the VM was located in the orbit with an attendant risk of loss of vision due to embolisation of the ophthalmic vein. Thus, the VM classification had a predictive value of 92.3% (i.e. in 12 of 13 patients, sclerotherapy could be performed without risk or complications).

Of the 16 patients with a type-II VM, 15 received sclerotherapy without any problems, resulting in a

Table 1 Classification of venous malformations according to anatomy and haemodynamics

Type	Description	*n* (%)
Type I	Isolated malformation without peripheral drainage	13 (30%)
Type II	Malformation that drains into normal veins	16 (37%)
Type III	Malformation that drains into dysplastic veins	9 (21%)
Type IV	Venous ectasia	5 (12%)

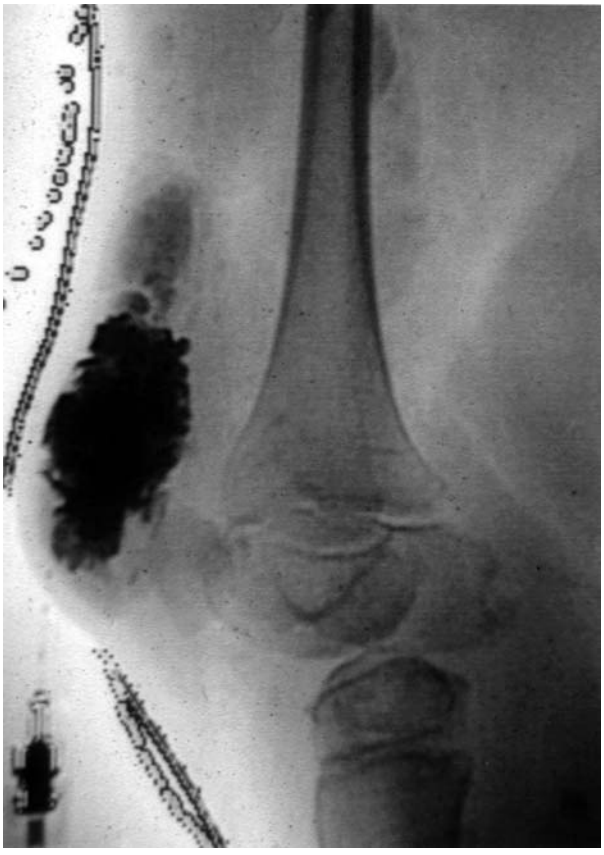

Fig. 1 After injection of contrast medium, no communication with the adjacent venous system is visible in this venous malformation (VM) at the level of the distal femur, making it a type-I VM

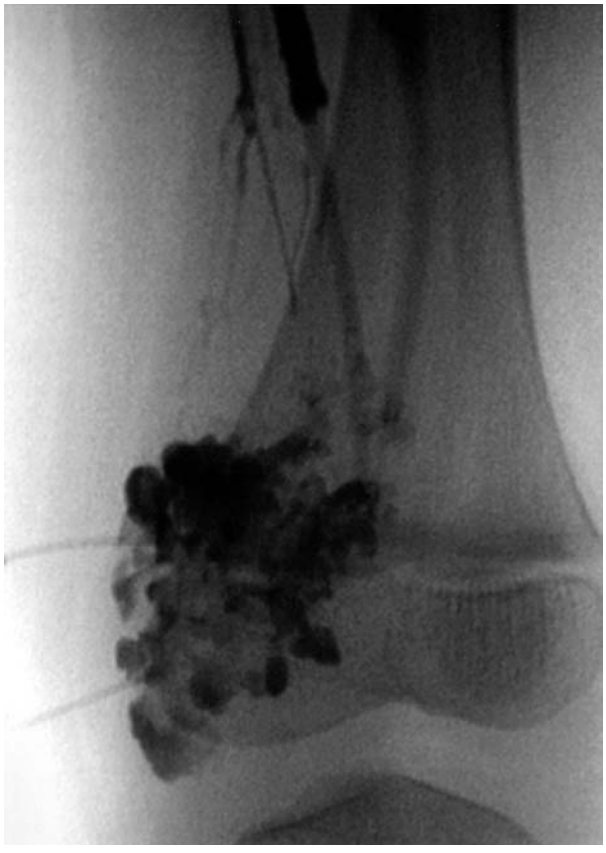

Fig. 2 A 7-year-old boy with a type-II venous lesion of the left knee. The lesion is drained by regular veins, making it a type-II VM

predictive value of the VM classification of 93.8%. The one patient who was excluded had already been scheduled for surgical resection of a lesion of the right hand.

In three of nine patients with a type-III lesion, no sclerotherapy was performed because the VM was located on the head or trunk, and the risk of symptomatic systemic embolisation of sclerosing material was thought to be too great. One of the six patients in this group who were treated interventionally was a 13-year-old child with a VM of the right knee, in which paralysis of the sciatic nerve at the level of the knee occurred after therapy. Considering the three patients who were excluded from sclerotherapy and the one patient with the major complication, our hypothesis that sclerotherapy should not be performed in type-III VMs was confirmed in 44.4%.

Finally, the rate of exclusion from sclerotherapy in patients with type-IV lesions was 60% (three of five patients). Of the five patients, two had VMs at the more distal parts of the extremities, and sclerotherapy did not result in any complications.

For the 35 performed sclerotherapies, pure ethanol was used in 33 of 35 cases (94%), a combination of Ethibloc and ethanol was used in one case (3%), and Thrombovar was used in one case (3%).

Discussion

VMs are often asymptomatic, but if accompanied by pain and functional or cosmetic defects, require treatment. In many cases, primary surgical treatment of VMs is difficult, impossible, or provides an unsatisfactory result [1]. Furthermore, the haemorrhagic risk is high because haemostasis is impossible except in limb malformations. Percutaneous injection of liquid agents is the preferred approach in many institutions and has a high reported success rate [1, 8, 9, 12, 19, 20, 21, 22, 23]. Local infiltration or intravascular injection of ethanol causes cell death by lysis of the cell membrane and, in addition, endothelial damage, denaturation of blood proteins, thrombus formation and vascular occlusion [9, 21, 24, 25]. After a review of the literature, O'Donovan et al. [12] reported the incidence of complications for ethanol embolisation to range from 7.5% to 23%. Major complications include local tissue injuries such as skin necrosis, peripheral nerve palsies and hypotensive

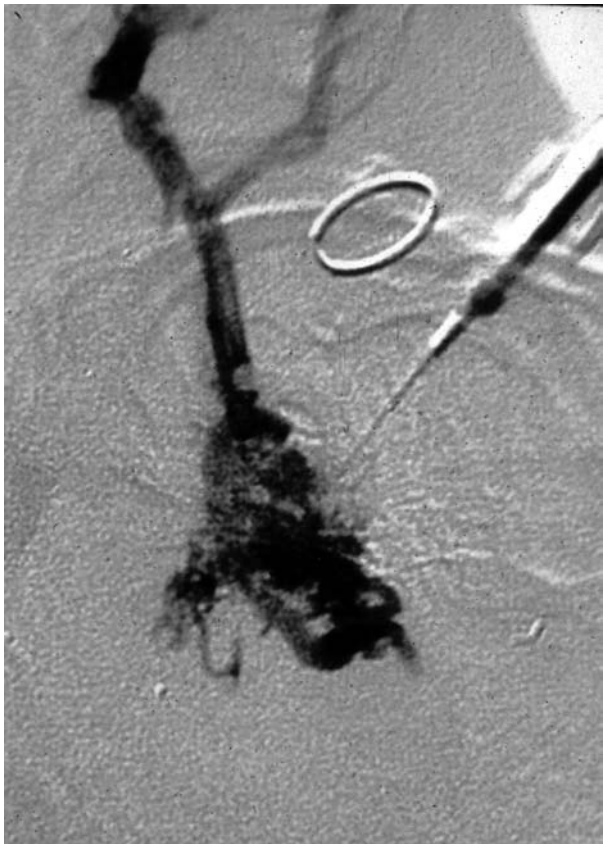

Fig. 3 After injection of contrast medium in the VM, it drains into dysplastic veins, making it a type-III VM

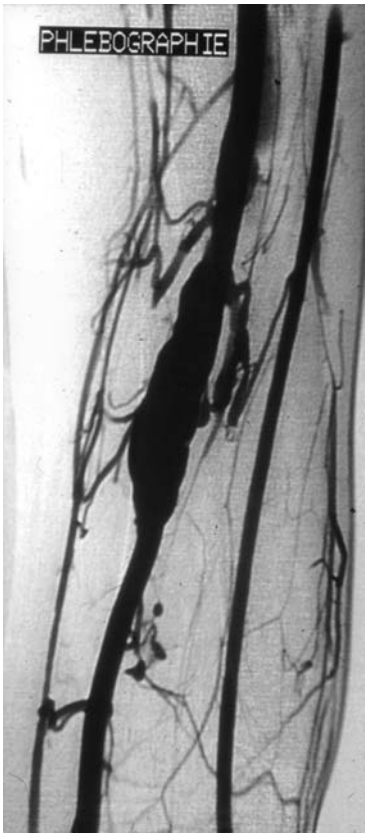

Fig. 4 Digital subtraction phlebography of the upper extremity demonstrates venous ectasia, making it a type-IV VM

crises, as well as fatalities such as cardiac arrest and pulmonary emboli [8, 9, 24, 26, 27, 28, 29, 30, 31]. Although the exact mechanism of systemic toxicity from ethanol injection is not completely understood, it is assumed that the most likely mechanisms are pulmonary vasospasm leading to pulmonary embolism, and direct cardiotoxicity [12]. Local anatomy of the malformation and the overall volume of liquid are factors that contribute to complications. Reflux from superficial venous channels can flow into skin capillaries and lead to skin necrosis [3]. Large and/or dysplastic vessels that drain the malformation support the possibility of central embolisation of sclerosing liquids.

The classification system that we developed includes four types of VMs. Our study showed that each of these four previously defined types was present in a sample of consecutively referred patients. The categorisation was quite easy with the use of phlebography, which demonstrates the extension of the malformation and its association with venous dysplasia. In future research, this system may be useful for the evaluation of outcome or to compare different techniques. Further, it can serve as a guideline for the interventional radiologist. In our experience, percutaneous sclerotherapy in type-I and type-II lesions carries almost no risk for central embolisation. In fact, in more than 90% of these patients, sclerotherapy could be performed without problems or major complications. Although a type-III or type-IV lesion is not an absolute contraindication for percutaneous sclerotherapy, the risk of central embolisation is thought to be much greater because of the wide vascular or dysplastic venous channels. Therefore, intervention in these patients must be carefully considered by taking into account numerous factors, such as localisation or size of the lesion. In our sample, sclerotherapy had to be withheld from approximately 50% of patients with type-III or type-IV VMs.

The lesions were classified by two radiologists in consensus. Ideally, there should have been independent assessment and adjudication of disagreement as a measure of interobserver variability. This should be considered a limitation of our study. Furthermore, we did not document the detailed rationale of the interventional specialist for each individual case of type-III and type-IV lesions regarding the decision to perform sclerotherapy.

Despite these limitations, we could demonstrate that there is an association between the type of VM and the possibility of performing complication-free sclerotherapy. Our categorisation offers a clear and simple

descriptive system that can serve as a basis for treatment choice and offers the possibility of uniform analysis and comparative reporting between scientific investigators. It may help to ensure maximal clinical efficacy while minimising the incidence of complications. Future research is required that would test the system in other samples and address its usefulness for the evaluation of outcome and to compare different techniques.

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