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MAIN TOPIC

J. Zwerver · P. N. M. A. Rieu · R. J. J. Koopman
 P. H. M. Spauwen · F. G. M. Buskens · C. Boetes
 R. P. H. Veth · C. G. van Oostrom

Vascular malformations: a review of 10 years' management in a university hospital

Abstract In order to gain insight into the management of patients with vascular malformations (VM) in the University Hospital Nijmegen in the past 10 years, 151 cases managed by different specialists were reviewed. To avoid the usual confusion in terminology, all recorded diagnoses were reclassified according to the biological classification of Mulliken. The sex distribution was equal; 79% of the malformations were diagnosed at birth or in the 1st year of

life. The median time between presentation and consultation was 3 years. Sixty-two lymphatic, 26 venous, 24 capillary, 1 arterial, and 38 combined malformations (8 arteriovenous, 30 others) were found. The head and neck region was most frequently involved, followed by the lower and upper limbs and trunk. The pediatric surgeon was the most frequently consulted specialist. Confusing, mutually incompatible terminology and a wide variety of different diagnostic techniques and treatments had been used by the different specialists. To improve the management of patients with vascular malformations, the use of a uniform classification, an increase in basic investigations, and the development and evaluation of protocols for diagnosis and treatment by multidisciplinary teams are necessary.

J. Zwerver · P. N. M. A. Rieu (✉)

Department of Paediatric Surgery, HECOVAN, Multidisciplinary Team on Hemangiomas and Congenital Vascular Malformations Nijmegen, University Hospital St. Radboud, P.O. Box 9101, NL-6500 HB Nijmegen, The Netherlands

R. J. J. Koopman

Department of Dermatology, HECOVAN, Multidisciplinary Team on Hemangiomas and Congenital Vascular Malformations Nijmegen, University Hospital St. Radboud, Nijmegen, The Netherlands

P. H. M. Spauwen

Department of Plastic and Reconstructive Surgery, HECOVAN, Multidisciplinary Team on Hemangiomas and Congenital Vascular Malformations Nijmegen, University Hospital St. Radboud, P.O. Box 9101, NL-6500 HB Nijmegen, The Netherlands

F. G. M. Buskens

Department of Vascular Surgery, HECOVAN, Multidisciplinary Team of Hemangiomas and Congenital Vascular Malformations Nijmegen, University Hospital St. Radboud, P.O. Box 9101, NL-6500 HB Nijmegen, The Netherlands

C. Boetes

Department of Radiology, HECOVAN, Multidisciplinary Team on Hemangiomas and Congenital Vascular Malformations Nijmegen, University Hospital St. Radboud, P.O. Box 9101, NL-6500 HB Nijmegen, The Netherlands

R. P. H. Veth

Department of Orthopaedic Surgery, HECOVAN, Multidisciplinary Team on Hemangiomas and Congenital Vascular Malformations Nijmegen, University Hospital St. Radboud, P.O. Box 9101, NL-6500 HB Nijmegen, The Netherlands

C. G. van Oostrom

Department of Paediatrics, HECOVAN, Multidisciplinary Team on Hemangiomas and Congenital Vascular Malformations Nijmegen, University Hospital St. Radboud, P.O. Box 9101, NL-6500 HB Nijmegen, The Netherlands

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Introduction

In the preface to their book, Mulliken and Young described the difficulties encountered by patients with vascular birthmarks in trying to find a doctor who is able to understand and reassure them, so they can stop "shopping around" in the medical community [8]. Confusing nomenclature has always been an obstacle in the management of vascular lesions. To the despair of patients and their parents, consulted specialists use different terminologies and propose different diagnostic and therapeutic regimes.

In 1982, Mulliken and Glowacki introduced a biological classification for vascular lesions [7]. Based on cellular investigations and clinical observations, these lesions are classified as either hemangiomas, characterized by endothelial hyperplasia, or vascular malformations (VM), structural abnormalities due to errors in vascular morphogenesis. Hemangiomas show rapid postnatal growth followed by slow involution. Eponyms are no longer necessary because they have no relevance for describing the clinical behavior of the lesions. VMs, however, are present at birth, grow

Table 1 Reclassification of old nomenclature into Mulliken's biological classification

Old terminology	Biological classification
Capillary hemangioma	Hemangioma
Capillary-venous hemangioma	
Cherry angioma	
	Single malformation
Port-wine stain	→ Capillary malformation
Teleangiectatic anomalies	↗
Arterial abnormality	→ Arterial malformation
Cavernous hemangioma	Venous malformation
Venous abnormality	
Bockenheimer syndrome	
Lymphangioma	→ Lymphatic malformation
Cystic hygroma	↗
	Combined malformation
Arteriovenous abnormality	→ Arteriovenous malformation
Klippel-Trenaunay syndrome	→ Capillary-venous-lymphatic Malformation
Parkes Weber syndrome	→ Capillary-arteriovenous-lymphatic Malformation
Sturge Weber syndrome	→ combined malformation

commensurately with the child, and never regress spontaneously. They can be arterial, capillary, venous, lymphatic, or show any combinations of these components. In this study, the word hemangioma is thus restricted to a specific vascular lesion of childhood and is no longer used as a denominator for all kinds of different vascular lesions.

In order to gain insight into the management of patients with VMs, a retrospective study was carried out on all such patients treated in the University Hospital Nijmegen in the past 10 years.

Materials and methods

Using the hospital's computerized STARDO coding system, we generated a list of patients who had been seen and/or treated for a vascular lesion in the departments of dermatology, pediatrics, pediatric surgery, otolaryngology, orthopedics, plastic and reconstructive surgery, vascular surgery, and craniofacial surgery in the period 1983–1994. The search criteria included the following diagnosis: hemangioma, lymphangioma, congenital malformation of the peripheral vascular system, vascular hamartoma, and hamartosis. In the department of orthopedics, the search for patients who had been treated for vascular tumors was carried out through a special "tumor archive." In the department for non-invasive vascular investigations a list of patients who had been investigated for the Klippel-Trenaunay syndrome was available.

Although no uniform classification system for vascular lesions had been used during the reviewed period, we decided to use the biological classification of Mulliken in this study to obtain a better perspective of the different vascular lesions. After studying the records of the patients and, if available, the pathology reports, the original diagnosis was reclassified according to the Mulliken classification with the help of Table 1. In this way, hemangiomas were separated from VMs and excluded from the study. The VMs were subdivided into arterial, capillary (CM), lymphatic (LM), venous (VeM), and combined malformations. Furthermore, the age at presentation, the time between the first observation and first consultation of a specialist at this hospital, the kind of specialist who had been consulted, and the affected body part were recorded. All records were screened for the extra diagnostic

investigations that had been performed and, finally, all treatment modalities used were noted.

Results

From the 450 records initially found under the search criteria hemangioma, lymphangioma, congenital malformation of the peripheral vascular system, vascular hamartoma, and hamartosis, 151 patients (79 women, 72 men) had a VM according to the biological classification of Mulliken. The median age at presentation was 1 month; 79% of all VMs were noticed at birth or during the 1st year of life. The median time that passed between the first observation of the VM and the first consultation of a specialist at this hospital was 3 years (0–68 years).

In Table 2, the original and revised diagnoses according to the biological classification are shown. In 17 patients with a recorded diagnosis of hemangioma the biological diagnosis appeared to be a VeM (16) or arteriovenous (1) malformation (AVM). Most lymphangiomas, hemangio-lymphangiomas, and cystic hygromas were translated into LMs except in 4 cases, which were reclassified as VMs or venous-lymphatic malformations. A wide variety of names (Table 2) was used to describe CMs. We classified the Rendu-Osler syndrome as a CM. The two patients with Bockenheimer syndrome appeared to have predominantly venous anomalies, and were renamed as VeMs. The original record diagnoses of AVMs complied with the revised diagnoses, while 1 hemangioma turned out to be an AVM. The Klippel-Trenaunay syndrome (port-wine stain, venous lesions, and hypertrophy) was translated into a combined malformation or, more specifically, capillary-venous-lymphatic malformation. The other combined malformations (8) were originally diagnosed as Parkes-Weber syndrome (4) (port-wine stain, multiple AV fistulae, and hypertrophy) or Sturge-Weber syndrome (4) (port-wine stain of the face and vascular lesions of leptomeningeal vessels leading to neurological problems).

The head and neck region was affected in 75 patients (face 35, neck 32, oropharynx 8), the lower limb in 62, the upper limb in 18, and the trunk in 15. (Note: since in 17 patients two or more body parts were involved, the total number of affected body parts is more than 151.) The pediatric surgeon was the most frequently consulted specialist, followed by the dermatologist, orthopedist, and plastic surgeon. Thirty patients (20%) consulted two or more specialists. Table 3 shows the consulted specialists in relation to the different kinds of VMs.

Forty-eight patients did not undergo additional diagnostic investigations. The majority were diagnosed as having a LM (22) or CM (20). No further diagnostic investigations were done in 5 patients with a combined malformation and 1 with a VeM. Of the 103 patients who underwent extra diagnostic procedures, 68 were subjected to two or more investigations. The total number of diagnostic procedures was 191. Table 4 shows which non-invasive and invasive diagnostic procedures were applied in relation to the

Table 2 Original and revised diagnoses (*art* arterial, *cap* capillary, *lymph* lymphatic, *ven* venous)

Original diagnosis	Diagnosis according to biological classification									Total
	Single				Combined					
	Art	Cap	Lymph	Ven	Art-ven	Ven-lymph	Cap-ven-lymph	Cap-art-ven-lymph	Other	
Lymphangioma	—	—	45	3	—	1	—	—	—	49
Klippel-Trenaunay syndrome	—	—	—	1	—	—	21	—	—	22
Hemangioma	—	—	—	16	1	—	—	—	—	17
Port-wine stain	—	16	—	—	—	—	—	—	—	16
Cystic hygroma	—	—	15	—	—	—	—	—	—	15
Arteriovenous malformation	—	—	—	—	7	—	—	—	—	7
Parkes-Weber syndrome	—	—	—	—	—	—	—	4	—	4
Sturge-Weber syndrome	—	—	—	—	—	—	—	—	4	4
Venous anomaly	—	—	—	4	—	—	—	—	—	4
Angioma serpiginosum	—	2	—	—	—	—	—	—	—	2
Bockenheimer syndrome	—	—	—	2	—	—	—	—	—	2
Hemangiolymphangioma	—	—	2	—	—	—	—	—	—	2
Nevus vascularis	—	2	—	—	—	—	—	—	—	2
Couperosis	—	1	—	—	—	—	—	—	—	1
Granuloma telangiectatica	—	1	—	—	—	—	—	—	—	1
Hypoplastic vascular system	1	—	—	—	—	—	—	—	—	1
Nevus telangiectaticus	—	1	—	—	—	—	—	—	—	1
Rendu-Osler syndrome	—	1	—	—	—	—	—	—	—	1
Total	1	24	62	26	8	1	21	4	4	151

Table 3 Consulted specialists for different vascular malformations (Total > 151 because of multidisciplinary treatment in 30 patients)

Specialist	Vascular malformation									Total
	Single				Combined					
	Art	Cap	Lymph	Ven	Art-ven	Ven-lymph	Cap-ven-lymph	Cap-art-ven-lymph	Other	
Pediatric surgeon	1	2	58	8	3	—	2	—	—	74
Dermatologist	—	20	1	4	1	—	20	4	—	50
Orthopedic surgeon	1	—	1	13	2	—	2	—	—	19
Plastic surgeon	1	2	11	2	—	1	—	—	—	17
Pediatrician	—	3	3	2	1	—	1	—	3	13
Otolaryngologist	—	—	5	—	3	—	—	—	—	8
Craniofacial surgeon	—	—	2	—	1	—	—	—	1	4
Vascular surgeon	—	—	—	—	1	—	—	2	—	3

Table 4 Diagnostic techniques (Total > 151 because of multiple procedures in 68 patients. *US* ultrasound, *ED* echo-Doppler, *X* plain X-ray, *CT* computed tomography, *MRI* magnetic resonance imaging, *SCIN* scintigraphy, *DSA* digital subtraction angiography, *PHL* phlebography, *END* endoscopy, *BIOP* biopsy)

Malformation	Diagnostic technique										Total
	Non-invasive					Invasive					
	US	ED	X	CT	MRI	SCIN	DSA	PHL	END	BIOP	
Single											
Arterial	—	1	—	—	—	—	1	—	—	—	2
Capillary	1	1	1	1	—	—	1	—	1	1	7
Lymphatic	17	2	24	3	4	1	1	—	1	3	56
Venous	4	7	18	7	9	5	13	1	2	2	68
Combined											
Arterio-venous	—	2	4	3	2	2	6	1	—	3	23
Other	1	22	5	—	—	—	5	2	—	—	35
Total	23	35	52	14	15	8	27	4	4	9	191

Table 5 Treatment modalities (Total >151 because of multiple procedures in 21 patients. *Corticost* corticosteroids, *cryo* cryotherapy, *emboli* embolization)

Malformation	Treatment									Total	
	None	Non-invasive				Invasive					
		Compression	Corticost	Laser	Cryo	Sclerotherapy OK432	Emboli	Excision	Amputation		
Single											
Arterial	—	—	—	—	—	—	—	—	1	—	1
Capillary	10	1	—	10	—	—	—	1	2	—	24
Lymphatic	11	1	—	—	—	—	1	—	49	—	62
Venous	3	2	2	1	—	—	—	—	19	—	27
Combined											
Arterio-venous	1	—	—	—	1	1	—	3	5	—	11
Combined other	11	15	—	2	1	—	—	3	3	2	37
Total	36	19	2	13	2	1	1	7	79	2	162

different VMs. Retrospectively, it was impossible to determine whether diagnostic procedures were used to come to a final diagnosis or whether they were performed to obtain preoperative information about anatomic borders of the lesion or underlying anomalies.

In 36 patients no treatment was used; in the other 115 one or more methods were employed. Table 5 shows the distribution of the different VMs and the various non-invasive (36) and invasive (90) treatments used. Two patients with the reclassified diagnosis of a VeM were treated with corticosteroids, presumably because their lesions were considered to be hemangiomas. Compression garments were mainly used in patients with combined or VeMs, whereas laser therapy was performed for the treatment of CMs. In 1 patient with a LM OK 432 was injected into the lesion [9]. Embolization was primarily used for the treatment of AVMs and combined malformations. Since the majority of malformations were lymphatic or venous and since the main treatment for these malformations was excision, excision was by far the most employed treatment.

Discussion

Confusing terminology has always been an obstacle in the management of patients with vascular lesions. The word hemangioma is used by pathologists and clinicians to describe a variety of VMs of different etiologies. Mulliken and Glowacki proposed a biological classification for vascular lesions based on both cellular and clinical studies [7]. In this useful classification, the word hemangioma is reserved to describe a rapidly growing vascular tumor of childhood that is characterized by endothelial hyperplasia. All other vascular lesions caused by errors in vascular morphogenesis are called malformations. Dependent on the kind of abnormal vascular structure, the malformations are subdivided into arterial, capillary, lymphatic, venous, or combined malformations. VMs have a relatively low incidence, and the different types of malformation are treated by various specialists.

In this study, we tried to obtain uniformity in the nomenclature by reclassification of the originally recorded diagnoses. In this way, only 151 true VMs could be selected out of 450 patients with a vascular lesion. The remainder consisted of hemangiomas (according to the biological classification) and some non-vascular lesions. The results concerning sex distribution, affected body part, and presentation of the selected group of VMs proved to be in correspondence with other reports on this subject [1–3, 5, 6]. VMs were diagnosed in male and female patients in an almost equal distribution (72:79). The head and neck region was most frequently affected, followed in decreasing order by the lower and upper limbs and trunk. A majority of the VMs (79%) had already been noticed at birth or during the 1st year of life. We did not find a second incidence peak, which has been described to occur around puberty or pregnancy due to enlargement of the still-undiscovered malformation from hormonal changes [3, 11].

The delay between manifestation of the VM and consultation of a specialist (median: 3 years) can be partly explained by the fact that some patients consulted other specialists before they were referred to this hospital. Some visited this hospital on their own initiative after failure of treatment elsewhere. No complete data were available on these prior treatments. On the other hand, this delay indicates that VMs can exist for a period of time without causing problems that require medical attention [3, 11]. The number and distribution of VMs shown in Table 2 should not be considered to be the incidence of VMs in the normal population. Since not all VMs require medical attention and not all patients with VMs were sent to this hospital, the figures in Table 2 merely demonstrate the kinds and numbers of VMs that were treated in this hospital.

A wide variety of non-invasive and invasive diagnostic techniques has been used for investigation of the different VMs. Echo-Doppler and magnetic resonance imaging/angiography have played an increasing role in the diagnostic work-up of vascular lesions during the past few years [3, 4]. However, their exact position in the work-up of VMs still has to be determined. In our opinion, invasive diagnostic techniques like angiography should only be used if relevant

information for diagnosis, prognosis, or further treatment can be obtained or if embolization is considered the most appropriate therapy for a particular malformation.

During the past 10 years excision has been the most frequently used treatment for VMs in this hospital. This is primarily due to the predominance of LMs, for which excision is an established treatment modality. In contrast, sclerotherapy, and embolization have only occasionally been employed. These techniques have, however, been applied more successfully for special forms of VMs during the past 10 years [10]. In this study no data were collected on postoperative complications, recurrence rates, and the patient's and clinician's satisfaction with the result of the treatment. Perhaps a more conservative approach to some malformations or more aggressive treatment of others is indicated, but this can only be proven by new investigations in which the management of patients with VMs according to previously determined protocols is prospectively evaluated.

To avoid the problems of confusing terminology and interdisciplinary differences in management in the future, we believe a team approach to most VMs will be beneficial. Various specialists, all using the same nomenclature, have to share their experience in this difficult area of vascular pathology to arrive at a rational diagnostic strategy and treatment plan for VMs. Furthermore, standardized protocols for diagnosis and treatment with long-term follow-up should be developed in national and international organizations like those in oncology. Basic research in pathology and embryology in order to understand the underlying causes of VMs, which could have implications for new treatments and even prevention in the future, is essential. Because VMs have a relatively low incidence, cooperation between national and international multidisciplinary teams is desirable [1, 6].

We conclude that the presentation of VMs in the University Hospital Nijmegen during the past 10 years has been in accordance with other reports in the literature. We also found that a large gamut of confusing terminology as well as a wide range of diagnostic and treatment modalities had been used by the different specialists. In order to improve

the management of patients with VMs, the use of a uniform classification should be encouraged, basic research has to be stimulated, and standardized protocols for diagnosis and treatment have to be developed and evaluated. National and international cooperation in this field must be promoted. The formation of multidisciplinary teams in which, due to the age at presentation of most VMs, a pediatrician and pediatric surgeon are represented, seems to be a prerequisite to achieve these goals.

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