CHIRURGIE THORACIQUE / THORACIQUE SURGERY

PATTERN OF VASCULAR MALFORMATIONS IN A SUBSAHARAN AFRICA SETTING

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Summary

Congenital vascular malformations (VM) encompass a wide spectrum of lesions with varying degrees of severity. The confusing terminology in the literature has led to the development of many classifications systems. The International Society for the Study of Vascular Anomalies (ISSVA) classification is the mostly accepted and has been recently updated. In Africa in general and sub-Saharan Africa in particular, very few reports have written using those confusing terminologies thereby leading to poor description of the lesions. The aim of our study is to describe the patient's characteristic and pattern of VM in Tropical Africa in General and Yaoundé in particular outlining the importance of accurate description of the lesions in light of the current concepts, classifications and terminology. All the patients diagnosed of VM during a 10 years period (January 2007 to October 2016) are included. For each patient clinical examination, duplex ultrasound and if necessary X rays, Computer tomography, MRI. were performed. The following data were retrieved: epidemiological data, description of the malformation (the symptoms, the type, the location, the extend, the sizes), the presence of an associated non- vascular malformation. During the study period, 51 VM were diagnosed in 47 patients. The age was 2 weeks to 29 years. Thirty-six (76.59%) were male and 15 (23.40%) female. As far as the type of VM is concerned, the venous malformations were the most common followed by capillary and combined. 2 trinocular aortic malformation were seen. It was noticed that 8 of the 9 combined VM-Klippel Trenaunay syndrome (n=8) and Parkes-Weber syndrome (n=1) were found in male patients. The sizes of the VM ranged from spot like lesion to extensive lesion and huge mass. With regards to the location of the VM, the limbs were found to be most affected (58,82 %) with the lower limbs being more affected than the upper limbs.

Key words: Vascular, malformations, Africa

Introduction

Congenital vascular malformations (VM) arise due to an error in vessel development in the embryo They encompass a wide spectrum of lesions with varying degrees of severity, ranging from isolated and innocuous lesions, to those that are disfiguring and disabling, to those that signal the presence of a more complex syndrome[1] the confusing terminology in the literature has led to the development of many classifications systems. The International Society for the Study of Vascular Anomalies (ISSVA) classification is the mostly accepted and has been recently updated[1]. The ISSVA classification divides vascular anomalies into vascular tumours or Hemangiomas and VM; based on their endothelial cell characteristics, clinical presentation, natural history, and histopathological characteristics. VM are further classified according to the type of vessel involved into arterial, venous, lymphatic and arteriovenous malformations[1,2,3]. The VM are also divided into defects of the main vessel, also called truncular defect or defect of the major named vessels and area of displastic vessel in tissue which are called extraarticular. In some patients a more complex disease can be the result of a combination of malformations[1,2,3]. Though considered rare, VM are frequently encountered in vascular practice and are a source of morbidity and esthetic disturbances and in complex cases the management can be frustrating both for the patient and the management team. In Africa in general and sub-Saharan Africa in particular, very few reports have been written[4,5,6,7]. The limits of those previously published studies are the limited number of VM, the use of confusing terminology the-refore leading to poor description of the lesions and the mixing together of the Hemangiomas (vascular tumors) and VM which are different pathologies with different management approches, prognosis and evolution. The aim of our study is to describe the patient's characteristic and pattern of VM in Tropical Africa in General and Yaoundé in particular outlining the importance of accurate description of the lesions in light of the current concepts, classifications and terminologies.

Patients and Methods

All the patients admitted in our clinic and diagnosed of VM during a 10 years period (January 2007 to 2016) have been considered. The Yaoundé General Hospital is the main referral Hospital in Cameroon and the only Vascular Center where most vascular patients of the country and neighboring countries are managed. Most modern vascular diagnostic tools (US, CTA…) and expert (Vascular surgeons, radiologist…) are available in this center. The diagnosis of VM is based on step by step procedure as follow: the clinical examination (the cornerstone of the diagnosis), duplex ultrasound and where necessary depending on the case; X rays, Computer tomography, MRI. For each patient, the following data were retrieved using a standardized case report form; 1-Epidemiological data (Age, sexes, age at onset, family history of VM, specific exposition of the mother during pregnancy (if known). 2- For the VM the symptoms, the location, the extend, the sizes, the type (venous arterial, lymphatic, combined, truncular or extratruncular) 3-the presence of an associated none vascular malformation. Hemangioma/vascular tumors as well as some specific malformations classified by the last ISSVA under complex malformations (as Proteus syndrome) are not part of this study .4 Data were entered into epi-info and analyzed as counts, frequency and percentages continuous data were

expressed as mean/standard deviation while categorical data were analyzed using Chi-square test with a Pvalue ≤0.05 regarded as statistically significant.

Results

During the study period, 51 VM were diagnosed in 47 patients. The age range was 2 weeks to 29 years. Thirty-six (76.59%) were males and 15 (23.40) females the lesions were present at birth in 44 (85 %) of the cases though not considered serious whereas it developed during the early years of life for the rest of the patients. For all the patients, no specific exposition of the mothers during pregnancy which may be considered as risk factor was declared. With regards to the type of VM, the venous malformations were the most common followed by capillary and combined (Table I).

Type of VM	N	%
Venous	25	49
Lymphatic	6	11.7
Capillary	9	17.64
Combined (Klippel Trenaunay)	8	15.68
Combined with arteriove nous fistula (Parkes Weber)	1	1.9
Arterial (aorta coarctation)	2	3.92
Extra vascular associated lesion (Overgrowth syndrome)	6	11.76

Table I: Distribution of the VM

Location of the malformation	N	%
Limbs Upper limbs Lower limbs	30 6 24	58.82
Trunk	13	25.49
Head and neck	6	11.76
Aorta	2	3.91
Genitalia	1	1.96

Table II: Location of the VM

The lesions encountered in VM were mostly unifocal (20 patients) and multifocal in 5 patients the limbs were the most involved location (Figure 1).

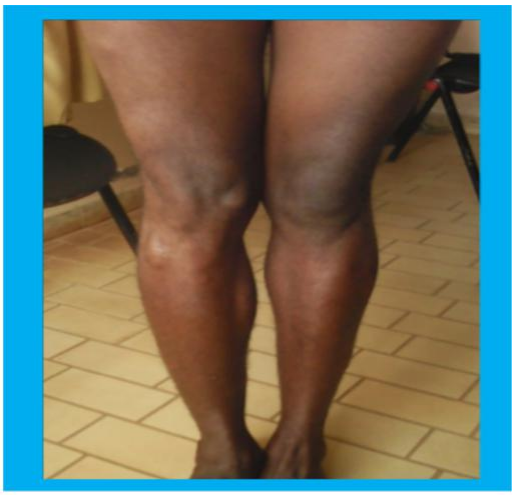

Figure 1: Venous malformation of the leg in a 15 years old boy

It was noticed that 8 of the 9 combined VM-[8 Klippel -Trenaunay Syndrom (KTS) and one Parkes- weber syndrom (PWS)] were found on male patients. The KTS which is a combination of Capillary, Venous (Varices) lymphatic malformations and associated limb overgrowth invarious proportion or discrepency were located on the lower limb for 7 patients and upper limb for one patient (Figure 2)

Figure 2: Klippel Trenaunay syndrome 21 years old male (right limb hypertrophy and non-systematic varicoces are

The patient with PWS had the classical presentation with its arteriovenous fistula in the mid-thigh region. The lymphatic malformations (Cystic hygroma/lymphangioma) were localized mainly around the neck region (Figure 3).

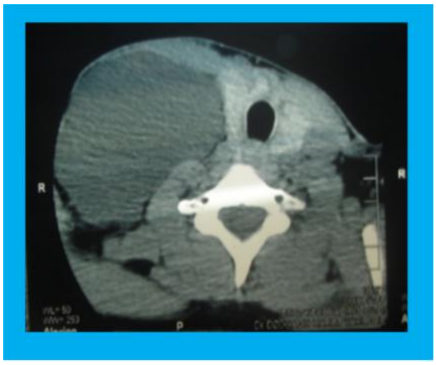

Figure 3: Cervical Cystic hygrom

Of the 2 aortic coarctations one was mid-aortic (Abdominal) and discovered during an US which depicted a left atrophic kidney. The other was a thoracic arch classical coarctation suspected by a blood pressure difference between the two arms. The sizes of the VM ranged from.

Discussion

VM remain a difficult diagnostic and therapeutic challenge due to the wide range of clinical presentations, unpredictable clinical course, erratic response to the treatment with high recurrence/ persistence rates, high morbidity following nonspecific conventional treatment, and confusing terminology. For the purpose of clarity and easy understanding, we have chosen the simplest form and terminology of the most recent ISSVA Consensus[1]. The aim of our study was specifically to describe the pattern of VM in tropical Africa and not the therapeutic approach or outcome. This is not the case with other reports from sub-Saharan Africa in which all the anomalies (hemangioma and VM) were considered together[4,5,6,7]. Those reports were emphasizing on surgical management and outcome without a clear description of the lesions[4,5,6]. The number of patients though small compared to what is seen in specialized vascular centers in Western countries[8] is probably the highest from a country of Sub-Saharan Africa and probably most parts of the developing world. This may be due the lack of vascular specialists. Actually, the field of vascular surgery is evolving in Sub-Saharan Africa but the practice is bedeviled by lack of skilled vascular surgeons and infrastructural challenges. The consequences are a low volume of operations and a dearth of data. Available data are not representative of the wider picture. In Senegal, among the 28 vascular anomalies presented by Diarra et al[20] were VM. In Tanzania only 33 among 134 cases of vascular anomalies were VM5 In Benin City (Nigeria), Oarsmen's and Evbuomwan published a report of 63 hemangiomas using this confusing terminology for every type of vascular anomalies6. In North Nigeria Dagbani et al[7] reported just 4 VM in their practice in 4 years in in a tertiary Hospital. Therefore, we strongly believed that our experience can be of help in contributing to the world data bank. As far as our patients are concerned, the long delay to seek vascular attention for most of the patients is probably due to the lack of vascular specialist. Therefore, some patients were correctly diagnosed only at adult age. The male to female ratio is not different from what is seen in most publications [4,5,6,8]. The isolated presentation and the sporadic pattern of VM is also commonly reported in the medico-surgical literature. Venous malformations are the most prevalent type of VM in our study; this is also the most reported type by the other authors[1,2]. It represents 59% in one of the renowned vascular mal-formation centers in Italy[8]. We can mention that simple cutaneous VM

(capillary) like « Port wine » may be difficult to see in some black patients and therefore they may not seek medical attention. Most of our capillary VM were seen in mucosa or in fair skin people. Combined VM are not rare in our patients (18%). In the literature, it represents about 5% of VM (8) . Diarra et al[4] reported 7 combined VM among their 20 VM in Senegal. We think that our higher proportion as was the case in Senegal[4] can be explained by the fact that they are the most serious and complicated cases and therefore people are more aware to seek a specialized attention. But it may also be a specific trend for our setting. We need more studies to clarify this issue. The lesions are very huge in many patients. In fact, as mentioned, most of our patients were diagnosed lately due the lack of vascular expertise. It is probably the case in most parts of the world. The commonest location is the extremities as is also the case in most reports[1,2,4,5,6,] though the lower extremity was mostly involved in our study unlike other reports[6]. This is probably because we have considered here purely VM and not all the vascular anomalies. It is well known that hemangiomas are more common at the cephalic extremity[1].

Conclusion

Vascular anomalies are the most common tumors, in infancy and childhood and pose major diagnostic and therapeutic challenges in our environment. Making an accurate clinical diagnosis is of critical importance. In particular it is important to correctly and precisely identify each type of lesion to reduce confusion. This is possible in our setting.

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