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SURGICALLY TREATED HEMANGIOMAS: CLINICAL- PATHOLOGICAL ASPECT

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Background: The Vascular anomalies are a complex pathological group of lesions with different modalities of treatment. The menagement of these lesions depends on a multidisiplinary approach.
Objective: To have an epidemiological aspect of these lesions in our centrefor building later a standartised way of treatment for them.
Method:A retrospective data analysis of all patients surgically treated between 2007 and 2012 in the different clinics of Universitary Hospital centre "Mother Theresa" was performed. The data was collected by the archive of Pathological Anatomy and included demographics, clincal diagnosis, correlation with pathological one and dispertion based on location of lesions and treatment information were obtained from the clinic's database.
Results: Of 220 patients diagnosed histologically with hemagioma 69 of them were surgically treated for another lesion, 215 hemangiomas (capillary or cavernous) and 5 patients with vascular malformations were found in the database. 15.3% (34 cases) belonged to the age range from 0-12 months; 12.6% from 1-9 yrs and 21.8% in the age range from 10-20 yrs old and the rest (9%;12.7%,12.7%.9% 6.3%) for each one decade after 20yrs old and the last, > 60yrs old. Hemangioma was the terminology used more by the clinicians and pathologist in their diagnosis (67% and 98%). 52% were females and 59% (130 cases) were located in the head and neck region, 32.7% in trunk and extremities and 8.3 % in other organs. 38% (13 cases) of infantile vascular lesions have been excised during the first 3 months and 41% during 3-6 mo. 12% underwent diagnostic imaging investigation and no data for treatment with laser or sclerotherapy
Conclusions: Hemangiomas and vascular malformations can occur at any anatomical site but mostly head and neck region. The approach toward infantile hemagiomas is complex and with diverese modalities of treatment. Surgical excision is made for small lesions and multimodalities of treatmet are in way for larger and more complex lesions.
Key words: Hemangioma, infantile heangioma, surgical treatment.

Introduction

Vascular anomalies are a complex group of pathologies with different pathological and clinical, aspects, complications and associated syndromes. One infant in three is a portant of a vascular sign , red, blue or purple [1]. For more than centuries it has been used a confusonal terminology which has led to inadequate diagnosis, treatments and researches not good oriented [2]. *Angioma* is still used as a generic terminology to decribe the vascular tumours and vascuar malformations. In 1982 it has been done a publication by Glowacki et Mulliken [1]. which proposed a biological classification clear and without ambiguities of vascular anomalies based on clinical aspects, natural history of evolution and cellular activity of the lesion. They proposed to devide the vascular anomalies in two categories: *vascular tumours* and *vascular malformations (VM)*. Their classification is actually widely accepted. The vascular tumours are composed by a proliferation of endothelial cells which are not present in birth and have a charasteristic natural evolution. The vascular malformations are result of an anomaly of development of vascular elements during embirogenesis. They are present during birth and they grow up in a proprortional manner together with the growth of the child. They are described in accordance to the predominant vessel that composes the lesion as: capillary malformations, venous, lymphatique arterial and aterio-venous malformations [3]. Sugery is one of the tretment modalities, especially of the hemangiomas which have not changed during their natural history. Frequently surgery treats the sequela of angiomatous lesions. In other cases the surgery can be done more eralier especially if there

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are complications and urgency: vital distress, complications realted to function of the organ etc [4.5] .

It can be done earlier even in the lesions that are estetically very large or have low possibilities of involution (median or paramedian hemangimas or hemangiomas with an important subcutaneous component). Here we are presenting our data on surgically treated hemangiomas for a 5-year period discussing the epidemiology, the terminology used and different possibilities of treatment.

Materials and Method

A retrospective data analysis of all patients surgically treated between 2007 and 2012 in the different clinics of Universitary Hospital Centre "Mother Theresa" was performed. The data was collected by the archive of Pathological Anatomy and included demographics, clinical diagnosis, correlation with pathological one and dispertion based on anatomic location of lesions. Treatment information were obtained from the clinic's database. The analysis of the data was done using microssoft excel to calculate the number and percentages of histopathological diagnosis, clinical diagnosis, anatomical location of the lesion, the size of the lesion, refferral of the surgical margins. The pathology reports were cross- checked for consistency with the clinical diagnosis.

Results

220 patients were surgically treated with the pathological diagnosis of a vascular lesion (hemangioma and malformation) during a 5 year period from 2007-2012 in the service of Plastic Surgery and Pediatry of the Universitary Hospital Centre " Mother Theresa". 69 cases (33%) came with a clinical diagnosis other than vascular lesion (tab 1). The histopathological diagnosis were 215 hemangiomas and 5 vascular malformations. 64.5% were referred as capillary hemangioma and 2.2% as vascular malformation with no further subdivision. Hemangioma was the terminology used more by the clinicians and pathologist in their diagnosis (67% and 98%).

15.3% (34 cases) belonged to the age range from 0-12 months; 12.6% from 1-9 yrs and 21.8% in the age range from 10-20 yrs old and the rest (9%;12.7%,12.7%.9% 6.3%) for each one decade after 20yrs old and the last more than 60yrs old (tab 2).

Table 1. Correlation of clinical diagnosis with histopathologic diagnosis

	Histopathologic diagnosis			
Clinical diagnosis	Capillary hemangioma	Cavernous hemangioma	AVM	Percentage
Suspect hemangioma	56	35	0	41.3
Hemangioma	40	7	0	21.3
Other	46	26	0	32.7
AVM	0	5	5	2.3
	142	73	5	

52% were females and 59% (130 cases) were located in the head and neck region, 32.7% in trunk and extremities and 8.3 % in other organs. 96.1% (125 cases) of the head and neck region were less than 3 cm but the report of free surgical margin was done only in 26% of the cases (tab 3).

38% (13 cases) of infantile vascular lesions have been excised during the first 3 months and 41% during 3-6 mo. 12% underwent diagnostic imaging investigation and no data for treatment with laser or sclerotherapy were collected becouse of not performing this kind of treatment in both clinics.

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Table 2. Distribution of histopathologic diagnosis by gender and age

Age	Gender	Capillary hemangioma	Cavernous hemangioma	AVM**	%
0-3m*	M	8	1	0	5.9
	F	4	0	0	
4-7m	M	2	2	0	6.3
	F	9	1	0	
8-12m	M	3	0	0	3.1
	F	4	0	0	
1-5	M	4	0	0	4.5
	F	6	0	0	
6-9	M	6	2	0	8.1
	F	6	4	0	
10-20	M	19	10	0	21.8
	F	12	6	1	
21-30	M	6	5	0	9
	F	2	6	1	
31-40	M	4	7	1	12.7
	F	12	4	0	
41-50	M	10	3	0	12.7
	F	9	6	0	
51-60	M	4	4	1	9
	F	6	4	1	
> 60	M	2	5	0	6.3
	F	4	3	0	
		142	73	5	99.56

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Table 3. Anatomic distribution of vascular lesions, and their size and excisional margins

			Excisional margins		
Organ distribution		**Size (cm)**	**No pathology**	**With pathology**	**No referral**
Face	mucosa	<0.5	7	2	17
		0.5-1	0	4	7
		1-3	1	3	6
		3-7	0	0	2
		7-10	0	0	0
		>10	0	0	0
	skin	<0.5	5	6	23
		0.5-1	5	2	9
		1-3	3	4	9
		3-7	0	0	3
		7-10	0	0	0
		>10	0	0	0
	periorbital region	<0.5	0	0	6
		0.5-1	0	0	6
		1-3	0	0	0
		3-7	0	0	0
		7-10	0	0	0
		>10	0	0	0
Trunk	skin	<0.5	4	2	13
		0.5-1	0	0	11
		1-3	1	0	12
		3-7	0	0	2
		7-10	0	0	1
		>10	0	0	0
Extremities	skin	<0.5	0	0	8
		0.5-1	0	0	5
		1-3	0	0	10
		3-7	0	0	3
		7-10	0	0	0
		>10	0	0	0
Visceral organs	Brain, liver etc	<0.5	0	5	3
		0.5-1	0	2	2
		1-3	0	0	3
		3-7	0	0	3
		7-10	0	0	0
		>10	0	0	0

90.7% of the head and neck region were located on skin face and mucosa and the rest in the periorbital region.

Discussion

Hemangioma continues to be used as a common terminology for any kind of vascular anomaly. This use is common in many medical fields as among surgeons, pathologists and pediatricians. In our cases 67% of the clinical diagnosis came for hemangioma and 98% of the pathologic diagnosis were hemangiomas. In a study of Hassanein et al "Hemangioma" was used incorrectly in 71.3 percent of publications. The erroneous use of hemangioma was independent of the authors' discipline: pediatrics (60.0 %), internal medicine (61.4 %), surgery (68.9 %), and obstetrics/ gynecology (70.0 %) [6]. In another study of Hand et al it was seen a variety of terms used to describe vascular anomalies. The degree of agreement with the accepted classification of International Society for Study of Vascular Anomalies (ISSVA) varied

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both within and among texts with an agreement that ranges from 22% to up 75% [7.8]. Even in textbooks of pathology the terminology "hemangioma" is largely used to describe all the vascular anomalies: tumours and vascular malformations [3]. For certain entities it is even difficult to distinguish on histopathological examination if the lesion is a malformation or a tumour, as for example lymphatic malformations of capillary type. In the same manner the terminology "cavernous hemangioma" is used to describe a large number of cases which were composed of dilated vessels. In our study 33% of histopathological diagnosis was cavernous hemangioma and vascular malformations composed only 2.2% of the diagnosis. Based only in the caliber of the vessel observed on microscopic examination the lesion can be a venous malformation, arterio-venous malformation and even infantile hemangioma in the stage of involution where the vessels are better seen and especially in their subcutaneous or submucous forms. As seen by the results the vessel caliber is used as criteria for the diagnosis especially when this is not associated by a special clinical indication. That is a possible explanation why in our cases the malformations are as low as 2.2% . In a cohort study of 5621 patients 35.2% had a tumor and 64.8% had a malformation. Types of tumors included infantile hemangioma (85.9%), hemangioendothelioma (7.8%), congenital hemangioma (5.4%), and pyogenic granuloma (0.9%). Malformations consisted of venous (36.8%), lymphatic (28.3%), arteriovenous (14.3%), capillary (11.0%), and combined slow-flow (9.6%) lesions [9]. Confusion over the classification of vascular malformations can lead to inappropriate management, such as waiting for 'haemangiomas' to regress spontaneously [10]. In another study done in a multidisciplinary clinic for vascular lesions 54% of the vascular malformations diagnosed at this centre were previously diagnosed as haemangiomas [9].

In the old nomenclature the terminology mostly used for hemangiomas as vascular tumors were: capillary hemangioma, cavernous hemangioma and capillary –cavernous hemangioma versus current terminology as superficial hemangioma, deep hemangioma and compound hemangioma [11]. The multidisciplinary approach for those pathologies and the work done by ISSVA brought to a classification of vascular tumours and vascular malformations which improved the usage of different terminologies to a common language by different specialities, and to a better understanding of different anatomo- clinique aspects of those pathologies [3]. (tab4).

Table 4 The scheme of ISSVA classification

Vacular Anomalies		
Vascular Tumours	**Vascular Malformations (VM)**	
Benign vascular tumours Infantile hemangioma Congenital hemangioma other	Simple VM arterial Venous lymphatic	Combined and complex malformations
Vascular tumours of uncertain malignancy		
Malignant vascular tumours		

Hemangiomas are categorized into two types: "infantile" and "congenital." The rare "congenital" hemangioma is less understood and present at birth. Congenital hemangiomas either rapidly involute (rapidly involuting congenital hemangioma (RICH)[12.13] over a very brief period in infancy or never involute (noninvoluting congenital hemangioma; (NICH) [14].

One of the most common vascular tumours is infantile hemangioma (IH) present in 5-10% childrens before one year old [3]. 15.3% of our cases belonged to the age group from 0-12 months. IH appears shortly after birth and has an evolution in phases: proliferation, quiescence followed by spontaneous involution during a period of several years. In the stage of proliferation they may be confused with other red lesions of birth, but rapid proliferation and vertical growth will help in diagnosis. In most IH-s eighty percent of proliferation occurs by three months of life but may last longer [3]. 38% of infantile vascular lesions in our cases have been excised during the first 3 months and 41% during 3-6 months. During proliferation and rapid growth may occur ishcemia, ulceration, bleeding which brings in most of cases parents to ask for surgical removal of the lesion of their child. After proliferation IH enters the phase of quiescence that lasts from 9 to 12 months of age. During involution, the skin that lyies over becomes gray and the deeper components shrink. Different reports suggest that involution in 50%, 70%, and 90% of the hemangioma occurs by 5, 7, and 9 years of age with some variability [9]. At the final stages of involution, a fibrofatty protuberance may remain. Surgical excision is one of the modalities of treatment for IH of any age. In our cases nearly half of angiomatous lesions have been excised during infancy, adolescence and early adult years. If there are no indications for surgery of urgence the assessment of extent of textural change, scarring and distortion of the anatomic place can be better done until regression has occured. The surgical excision is better to be done before the school age,

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because of the possible implications in the pshyco-social life of the child, especially for lesions of the head and neck [15.16]. Complete surgical excision with immediate reconstruction is accepted as the gold-standard treatment for AVMs and is reported to be the most successful for small, localised lesions [17]. Medical and surgical options are available for the treatment of hemangiomas depending on the clinical scenario. Medical treatments include systemic therapies ,topical and intralesional one. Surgical management involves excision, laser treatment or both. It is the clinicians that must carefully weigh the risks and benefits for each treatment [18.19]. Excision is the appropriate for localized lesions and the fibrofatty remnants of involuted hemangiomas. Elective subtotal excision of massive protuberant proliferating hemangiomas can be employed in order to maintain aesthetic facial boundaries. Small remnants of disease are then left for involution. In our cases 26% of the excised lesions had no residual disease in the surgical excisional margins. Residual erythema and telangiectasias frequently remain in involuted hemangiomas and are best treated by selective photothermolysis using the flash pulse
dye laser (FPDL). FPDL can be used also for ulcerative lesions during proliferation to induce healing and new epidermal growth. [20]. Surgery remains one of the most superior treatment options and may offer a cure for localized VM. However, the excision of complex lesions remains difficult secondary to intraoperative bleeding but preoperative sclerosant can be used prior to excisionn(24–48 hours) to decrease surgical risk [21.22] . In our clinics neither laser therapy nor sclerotherapy couldn't be performed, a thing that should be changed as soon as possible. Patients with extensive disease will often require combined modality therapy for disease control over many years.

It has been done now for several years a practice the creation of multidisciplinary clinics dedicated to the diagnosis and management of such vascular lesions [9]. Those multidisiplinary centres regroup together different specialists like surgeons, dermatologists, pediatrician, raiologists and pathologists which meet in a regular basis to discus the therapeutical strategy especially for complex lesions like venous an arterio-venous malformations. A multidisciplinary team facilitates accurate diagnoses, and helps guide imaging and treatment decisions. Joint clinics eliminate also the need for multiple visits to see different specialists, thus avoiding conflicting diagnoses and treatment plans [9].

Conclusion: More than two thirds of excised vascular lesions are considered hemangiomas clinically and pathologically. Vascular lesions are a complex group of lesions presenting as tumours and malformations. Correct diagnosis is an important clue for the appropriate treatment.

The most common vascular anomalies in order of presentation include hemangiomas, lymphatic malformations, capillary malformations, venous malformations, and arteriovenous malformations. Treatment of vascular anomalies is complex and involvement of multiple disciplines and therapeutic options has become a need even in our country, especially for complex and "problematic" hemangiomas and VM. Confusion exists between surgeons, pediatriciens and pathologists in the nosology of such lesions. This reinforces the need for more education regarding early accurate diagnoses of vascular lesions and proper treatment of them.

Disclosure

The authors report no conflict of interest in this work.

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