

Classification of Vascular Anomalies Using Clinico-Radiological Parameters – Is It Sufficient?

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ABSTRACT

Background: To classify vascular anomalies using clinico-radiological parameters. Settings: Government Medical College Srinagar and Army Hospital Research and Referral New Delhi. **Methods:** 129 cases of vascular anomalies were studied and classified using clinical and radiological methods from March 2011 to June 2018. Outcome: Vascular anomalies were broadly classified into vascular tumours and vascular malformations and these anomalies were further sub-classified based on clinical and radiological parameters. **Results:** Among 129 vascular anomalies studied there were 51 vascular tumours which included 50 hemangiomas (H) and 1 case of pyogenic granuloma (PG). Among Hemangiomas there were 34 Hemangiomas of Infancy (HOI) and 16 Congenital Hemangiomas (CH). Among HOI there were 26 Simple (Localized-17, Segmental-2, Indeterminate-6, Multifocal-1), 8 complex visceral hemangiomas (Liver-2, Subglottic-2, Parotid-4). Among Congenital Hemangiomas there were 4 Rapidly Involuting Congenital Hemangiomas (RICH) and 12 Non involuting congenital hemangiomas (NICH). There were 78 vascular malformations which included 51 slow flow malformations and 27 fast flow malformations based on colour doppler studies. Among slow flow there were 42 Simple {2 Capillary Malformations, 26 Venous Malformations (VM), 14 Lymphatic Malformations (LM), 4 Combined (2 each of Lympho-hemangiomas - LH and Lymphovenous Malformations – LVM)} and 5 Complex-combined (4 Klippel Trenaunay Syndrome – KTS and 1 case of Blue Rubber Bleb Nevus Syndrome - BRBNS). Among fast flow there were 6 simple (Nasopharyngeal Angiofibromas), 20 combined (Arteriovenous Malformations - AVM) and 1 Complex combined malformation (Sturge Weber Syndrome - SWS). **Conclusion:** Newborn with birthmark should always be documented by the attending health care provider at birth and referral to an expert for proper evaluation and careful parental/Guardian counseling. Doppler US should be the first line of investigation in broadly classifying vascular anomalies whereas CT Scan, MRI/MRA/MRV, Angiography, Venography help in further sub-classification. Some confusion still persists in classifying few lesions like Lympho-hemangioma (LH), Complex combined Malformations (Syndromes) like Sturge Weber Syndrome (SWS), Blue Rubber Bleb Nevus Syndrome (BRBNS), and Nasopharyngeal Angiofibroma (NPA).

Keywords: Classification, Hemangiomas, Vascular anomalies, Vascular malformations, Vascular Tumours,

INTRODUCTION

Classifying vascular anomalies has always troubled the clinicians over the ages because of overlapping characteristic among the different types of vascular lesions.^[1,6,7] Recently at the end of 20th century clinicians tried to classify these notorious lesions based on their clinical appearance, histopathological features, predictable biologic behaviour and radiological parameters but there are still some vascular anomalies not fitting into the laid criterions.^[2] Classifying them correctly is important because of the fact that improper classification may lead to inappropriate treatment and unfavourable results.^[3,4]

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MATERIALS AND METHODS

One hundred and twenty nine cases of vascular anomalies in the pediatric age group were studied and classified using clinical and radiological methods at Government Medical College Srinagar, Kashmir and Army Hospital (Research and Referral) Delhi from March 2011 to June 2018 in a prospective manner. All the patients in the paediatric age group younger than 14 years of age and diagnosed as vascular anomalies during the study period were included in the study. Written informed consent was taken from guardians/parents of all patients. Every new patient referred to these institutions was examined clinico-radiologically, to correctly diagnose the vascular anomaly. Thorough history and careful physical examination of the patient was recorded. It included details of the lesion/lesions location especially at trauma prone and potentially life threatening vital areas, size, presence at birth, age at appearance, progression of

the lesion, age at involution, pulsations, thrill, raised temperature, trophic changes, skeletal overgrowth, pain, discomfort, functional impairment and cosmetically severe deformity, and any symptoms and signs suggestive of visceral involvement. All lesions were photographed serially at 3 weekly intervals and change in the character of the lesions was noted. At the initial visit proper counselling of the parents with respect to the natural history of the lesions and the possible interventions required during the course of management was done.

Clinical examination was supplemented by following radiological examinations depending on the site and type of lesion and its symptoms.

- a. Non-invasive imaging studies such as ultrasonography (USG) of the lesion with high frequency probe followed by Colour Doppler USG, Contrast Enhanced Computed Tomography (CECT), and Magnetic Resonance Imaging (MRI) were done to further differentiate hemangiomas from vascular malformations. The parameters assessed on US with colour Doppler were volume, echogenicity, colour flow, high vessel density. On CECT density with contrast enhancement, feeders, efferent vessels and extension were noted to differentiate various lesions. On MRI the signal intensity during the sequential T1 and T2 weighted images, extent of the lesions and their relationship with the adjacent structures, and flow voids were noted.
- b. Invasive studies such as Magnetic Resonance Angiography (MRA), Arteriography, were done in cases requiring embolization like hepatic hemangioma, AVM, angiofibroma. The parameter assessed included early opacification (shunting), size, location and number of feeders and efferent veins and location of nidus. Biopsy was done in cases where surgical excision was performed. Relevant investigations like lesions involving visceral organs and associated with syndromes were done on individual case merit which included blood pool scan, UGI endoscopy, Colonoscopy, intraoperative enteroscopy for BRBNS, Ascending venography for vascular malformation lesions in KTS, and CECT brain for SWS. Radiological data was collected as recorded version of the diagnostic radiological procedures. Relevant routine haematological and biochemical investigations like complete blood counts, coagulation profile, kidney function tests with serum electrolytes, complete liver function tests were done as per the requirement of the individual case especially platelets count in cases of large vascular lesions.

RESULTS

Among various vascular anomalies studied there were 51 vascular tumours which included 50 hemangiomas (H) and 1 case of pyogenic granuloma (PG). Among Hemangiomas there were 34 hemangiomas of infancy (HOI) and 16 congenital

hemangiomas (CH). Among HOI there were 26 Simple (Localized-18, Segmental-2, Indeterminate-6), 8 complex visceral hemangiomas (Liver-2, Subglottic-2, Parotid-4). Among congenital Hemangiomas there were 4 Rapidly involuting congenital hemangiomas (RICH) and 12 Non involuting congenital hemangiomas (NICH). There were 78 vascular malformations which included 51 slow flow malformations and 27 fast flow malformations based on colour doppler studies. Among slow flow there were 42 simple malformations {2 Capillary Malformations (CM), 26 Venous Malformations (VM), 14 Lymphatic Malformations (LM), 4 Combined (2 each of Lympho-hemangiomas - LH and Lymphovenous Malformations - LVM)} and 5 Complex-Combined (4 Klippel Trenaunay Syndrome - KTS and 1 case of Blue Rubber Bleb Nevus Syndrome - BRBNS). Among fast flow there were 6 Simple (Nasopharyngeal Angiofibroma- NPA), 20 Combined (Arteriovenous Malformations - AVM) and one Complex combined Malformation (Sturge Weber Syndrome - SWS) as shown in [Figure 1 & Table 1] showing further sub-classification.

Clinical Parameters: [Photograph 1]

Age distribution with respect to appearance of lesions ranged from 36 weeks intra-gestation to 14 years with mean age of 85 months. Most of our patients were infants about 58% which included 76% of vascular tumours and 48% of vascular malformations. Children between 1-10 years accounted for 23% which included 20% of vascular tumours and 26% of vascular malformations. Children above 10 years accounted for 17% which included 4% of vascular tumours and 26% of vascular malformations as shown in [Figure 2]. Gender distribution: There were 58 (40%) male patients and 71 (60%) female patients. The male to female ratio was 1 : 1.22. Hemangioma group had 28 female and 22 male patients, CM included both females, VM had 18 females and 8 males, LM had 10 females and 4 males, AVM had 10 patients in each group. There was 1 female each with SWS, BRBNS, and Pyogenic granuloma. All cases with Angiofibroma, LVM, LH and KTS were males as shown in Figure No. 3. Appearance: In 55 (43%) cases lesions were present since birth whereas lesion appeared later in 74 (57%). Twenty four (48%) cases of hemangiomas, 10 (71%) of LM, 3 (50%) of syndromic patients and 2 patients each of LH and CM had lesion since birth. In 16 (61%) cases of venous malformation, 16 (80%) cases of AVM, 6 (100%) cases of NPA, 1 case of PG and 2 cases of LVM, there was history of appearance of lesion later in life. In 2 cases of KTS lesions appeared at 6 and 36 months of life and in a patient with BRBNS cutaneous lesions appeared by her second year of life. There was 1 (0.8%) case with antenatally detected lesion in the liver. Signs and symptoms:

Apart from presence of swelling seen in all but 3 patient, pain was the most common complaint seen in 40 (31%) followed by disfigurement in 37 (29%) patients, functional limitation in 34 (27%), skin discolouration in 2 and history of bleeding was present in 18 (14%) cases. There was no definite family history of similar lesions. Multiplicity and Anatomic distribution: There were total numbers of 166 lesions in 129 patients. Hemangioma accounted for 57 (34%), VM for 32 (19%), LM for 28 (17%), syndromes for 14 (8%), AVM for 22 (13%), AF for 6 (4%), and LH, LVM and CM accounted for 2 (1.5%) each and there was 1 case of PG. Eighty lesions (48%) involved head and neck region. 30 of 57 hemangioma lesions, 18 of 32 VM, 8 of 22 AVM, all 6 NPA, 6 of 28 LM and 2 each of LH, LVM and CM lesions, 6 of 14 in syndrome patients involved head and neck region. There were 75 (44%) lesions involving the trunk and extremities, majority 37 (50%) involving the lower limbs. Hemangioma accounted for 20, LM for 18, VM for 14, AVM for 14, syndromic patients for 8 (KTS) and pyogenic granuloma for 1 lesion. Eleven (8%) lesions involved various internal organs, 4 Parotid (2 each of Hemangioma and LM), 3 hepatic (All hemangiomas which included 1 lesion in a case of BRBNS), 2 subglottic hemangioma, 2 LM (mediastinum, mesentery of small intestine) as shown in [Figure 4]. Size of the lesions: Twenty out of 57 hemangioma lesions, 24 out of 28 LM lesions, 14 out of 22 AVM lesions, and 2 each of LH, LVM, CM, and a dermal lesion in a SWS patient measured greater than 20 cm² in size. Fourteen out of 32 VM lesions, and all 4 patients with KTS had lesions measuring more than 10 cm².

Radiological investigations : [Photograph 2]

US gray scale and Doppler evaluation: was done in 110 patients including 42 patients with hemangioma, 2 patients each with LH, LVM, CM and 1 case of PG, 14 LM, 18 AVM, 24 VM and 4 patients with KTS and one subcutaneous lesion in BRBNS. The parameters studied were echogenicity, vessel density, and flow characteristics. Majority (30/42) hemangiomas showed variegated echogenicity, (38/42) showed high vessel density (>5/ cm²) and all 42 cases had high flow rate. LM appeared mostly hypoechoic or hyperechoic with no flow on colour Doppler. VM mostly appear hypoechoic (20/24) with slow flow on Doppler in all cases of VM. AVM mostly appeared as variegated echogenicity with high vessel density and high flow on Doppler US. One case with KTS revealed presence of abnormal veins. Two patients diagnosed as LVM involving head and neck region on gray scale US revealed minor component of anechoic cystic cavity lymphatic lesion and associated hypoechoic venous component which on Doppler US revealed slow flow component. As shown in [Table 2]

Magnetic resonance imaging: A total of 88 patients underwent magnetic resonance imaging study which included 26 VM, 20 AVM, 14 LM, 5 Syndromic patients (KTS-4, SWS-1), 18 hemangiomas, and 2 each of LH, LVM and 1 case of PG patient. The parameters studied were signal intensity during the sequential T1 and T2 weighted images, flow voids besides extent of the lesions and their relationship with the adjacent structures. The venous component in patient with lymphovenous malformation showed enhancement with gadopentate dimeglumine administration. All patients with hemangiomas and 1 patient with LH on MRI showed intermediate signal intensity on T1W and high signal intensity on T2W imaging. All patients with LM, LVM, one patient each with PG and LH and majority of AVM (18/20) showed low signal intensity on T1W whereas majority of lesions were high signal intensity on T2W imaging. All patients with AVM and 1 patient with KTS had presence of flow voids. Twenty three of 26 VM showed high signal intensity on T1W whereas majority of VM (24/26) were low signal intensity lesions on T2W. Three of the four patients with KTS showed intermediate signal intensity on T1W and high signal intensity in all on T2W. (as shown in [Table 3])

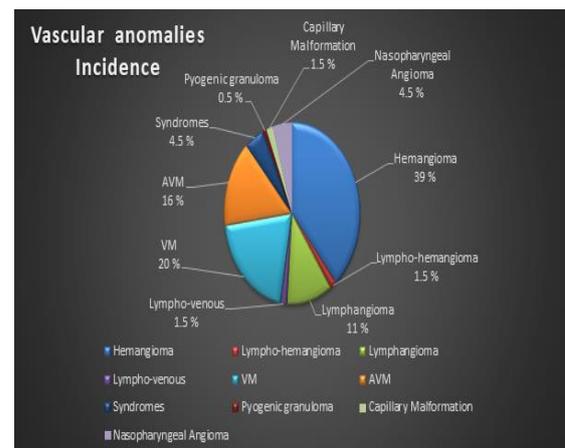


Figure 1: Incidence of various types of Vascular Anomalies

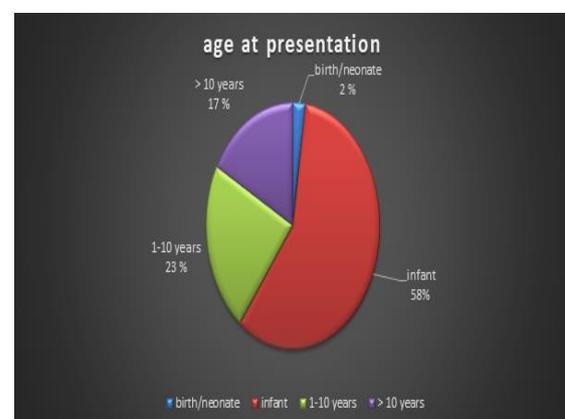


Figure 2: Pie chart showing age groups at 1st visit.

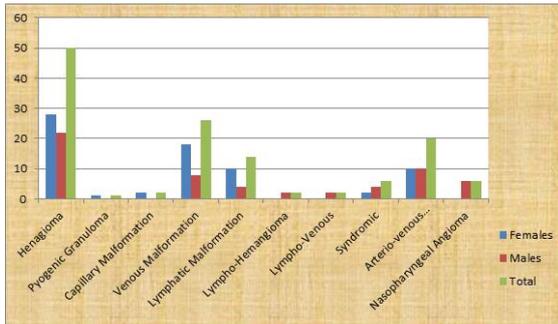


Figure 3: Sex ratio of patients with vascular anomalies.

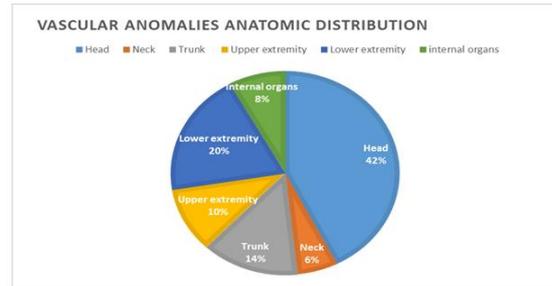
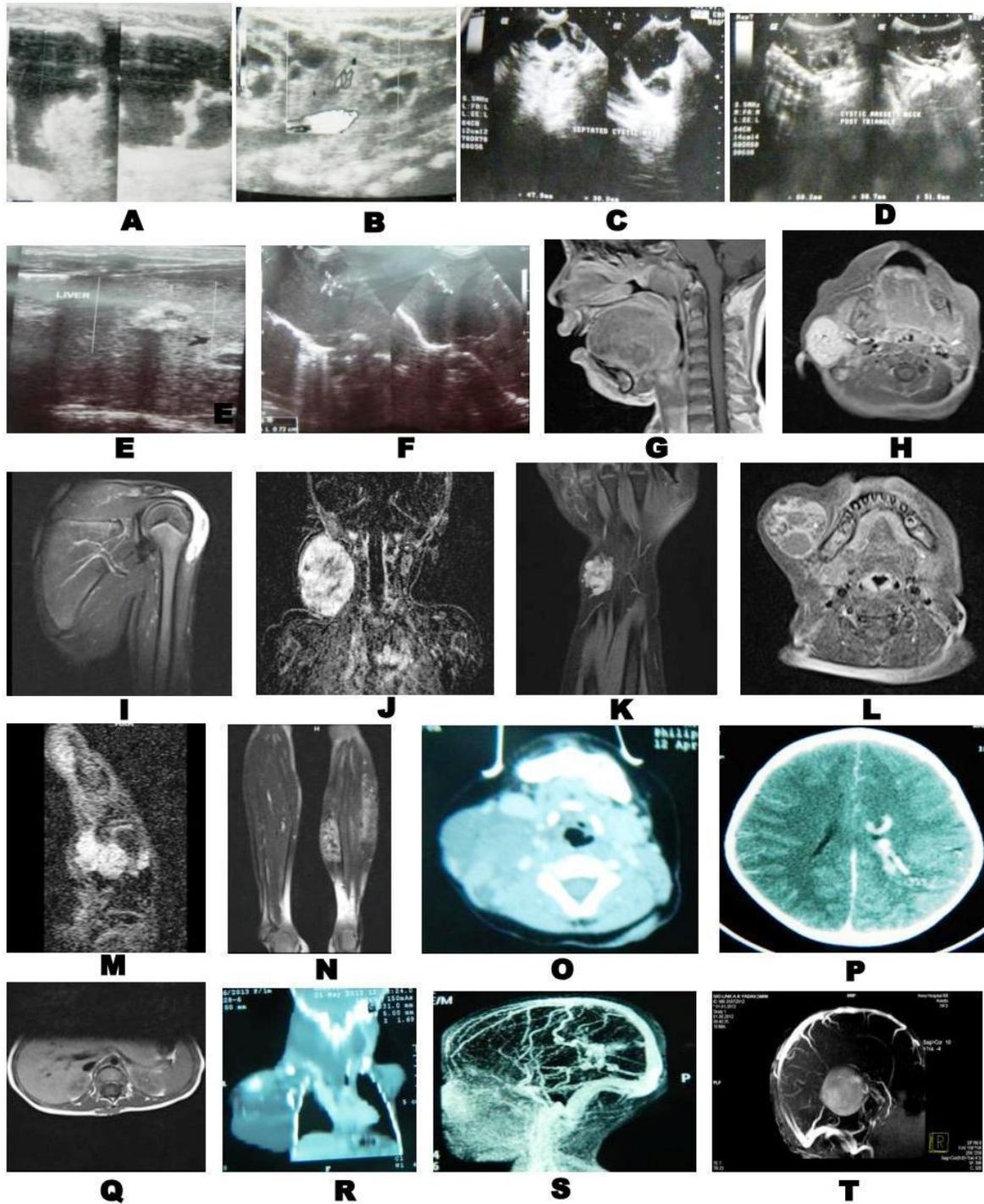


Figure 4: Pie chart showing anatomic areas involved by various vascular anomalies.



Photograph 1: Photograph showing A). Hemangioma of Infancy cheek B). NICH lesions . C). RICH lesion involving Pubic region D). Hemangioma of Infancy lesion on Forehead E). NICH lesion on trunk F). Large HOI with sudden increase in size G). Segmental HOI involving oral cavity and extending into neck and chest H). Ulcerated HOI Scalp I). Leaking LM involving leg J). Cystic Hygroma ia a Newborne K). Capillary Malformation face L). AVM left cheek M). VM Lower lip and adjacent cheek N). VM involving right ring finger O). Diffuse hemangiomatosis of lower limb P). Intra oral Hemangioma Q). Infantile Hemangioma in an Infant interfering with line of vision R). Large AVM involving Lower Lip, tongue, Floor of Mouth n both cheeks S). Large subcutaneous Hemangioma in an Infant.



Photograph 2 : Photograph A & B): Showing cystic lesion on US, anechoic without flow on Doppler most likely Lymphangioma in an 8 months old girl with right preauricular swelling. Photograph C & D): Showing lymphovenous malformation on US. Photograph E & F): Showing antenatally diagnosed SOL liver confirmed by CD post birth. On followup ultrasound at 6 months the lesion had disappeared (resolved completely). Alpha fetoprotein was within normal range. Photograph G): showing T1W Gadolinium Tongue hemangioma. Photograph H & I): Showing T2W imaging in various hemangiomatous lesions in the face, neck and shoulder. Photograph J): MRI Showing a Hemangiomatous lesion involving neck. Photograph K): MRI Sagittal view VM right wrist. Photograph L): MRI showing a Lympho-Hemangioma lesion. Photograph M): T2W coronal view AVM. Photograph N): T2W COR dark image of a LM involving leg in a 7 year old child. Photograph O): showing CT scan of 8 months old girl with right preauricular swelling, most likely Lymphangioma extending into neck. Photograph P): showing left parieto-occipital AVM evident on CECT head in a 4 years old girl with Sturge Weber Syndrome. Photograph Q): showing CT scan of hemangioma at D 11-12 region, with no intraspinal extension. Photograph R): showing a large Cystic Hygroma in a neonate with intra thoracic component pressing on trachea. Photograph S): showing left parieto-occipital vascular malformation(AVM) evident on Angiography in a 4 years old girl with Sturge Weber Syndrome). Photograph T): showing Vein of Galen malformation, child presented with seizures.

Table 1: Classification of vascular anomalies based on clinico-radiological parameters (no. of cases in parenthesis):

Vascular Anomalies (129)									
Vascular Tumours (51)			Vascular Malformations (78)						
Hemangiomas (50)		Congenital (16)	Other (1) Pyogenic Granuloma (1)	Slow Flow (51)			Fast Flow (27)		
Hemangioma of Infancy (34)				Simple (42)	Combined (4)	Complex Combined (5)	Simple (6)	Combined (20)	Complex Combined (1)
Simple (26)	Complex (8)	RICH - 4 NICH - 12	Capillary - 2	LVM - 2	CLVM - 4 (KTS) ?BRBNS - 1		AVM - 20 Stage - I - 4 Stage - II - 9 Stage - III - 6 Stage - IV - 1	?SWS - 1	
Localized - 17 Segmental - 2 Indeterminate - 6 Multifocal - 1	Visceral (8) - Liver - 2 - Subglottic - 2 - Parotid - 4		Type I - 6 Type II - 15 Type III - 4 Type IV - 1 Lymphatic - 14 (All ET form) 1. Superficial > Simplex - 8 (micro-2, macro-4, mixed-2) > LC(micro) - 2 2. Cavernous > Loose - 1 > Compact - 1 (macro) 3. CH (macro) - 2 ?4. LH - 2				?AF - 6		

HOI - Hemangioma of Infancy, RICH - Rapidly Involuting congenital hemangioma, NICH - Non Involuting congenital hemangioma, LH - Lymphohemangioma, LV- Lymphatico-venous Malformation, AVM- Arteriovenous Malformation, AVF - Arteriovenous Fistula, BRBNS - Blue Rubber Bleb Nevus Syndrome, SWS - Sturge Weber Syndrome, CLVM - Congenital Lymphatico-venous Malformation, KTS - Klippel Trenaunay Syndrome, CAVM - Congenital Arterio-venous Malformation, PWS - Parkes Weber Syndrome, SWS - Sturge Weber Syndrome, AF- Angiofibroma, ET – Extra truncular, ? confusion still persist

Table 2: showing radiological findings on US/Colour Doppler US

Lesion	No. of patients	Gray scale/ Doppler US	Echogenicity				Vessel Density			Flow	
			V	An	Hypo	Hyper	H	L	F	S	
H	50	42	30	4	6	2	38	04	42	0	
LH	2	2	2	0	0	0	2	0	2	0	
LM	14	14	2	0	6	6	-	-	no flow		
LVM	2	2	0	0	2	0	-	-	0	2	
CM	2	2	0	0	2	0	-	-	0	2	
VM	26	24	2	0	20	2	-	-	0	24	
AVM	20	18	18	0	0	0	18	0	18	0	
KTS	4	4	Abnormal veins- 1, Visceral involvement- 0								
BRBNS	1	1	-	-	1	-	-	-	no flow		
PG	1	1	0	1	0	0	-	-	0	1	
NPA	6	0	-----								
SWS	1	0	-----								
Total	129	110									

H- Hemangioma, LH- Lympho-Hemangioma, LM- Lymphatic Malformation, LVM- Lympho-Venous Malformation, VM- Venous Malformation, AVM- Arterio-Venous Malformation, KTS- Klippel Trenaunay Syndrome, BRBNS-Blue Rubber Bleb Nevus Syndrome, SWS- Sturge Weber Syndrome, PG- Pyogenic Granuloma, NPA- NasoPharyngeal Angioma, CM- Capillary Malformation, US-Ultrasound, V-Veriegated, An-Anechoic, H-High, L-Low, F-Fast, S-Slow.

Contrast Enhanced Computed Tomography: was performed in 22 patients including all 6 NPA, 7 hemangiomas, 5 LM, 3 VM and 1 syndromic patient (SWS). The parameter studied were density, contrast enhancement, feeders, efferent vessels and extent. Hemangiomas were found to be hypodense (7/7)

with peripheral enhancement in all cases on contrast injection. LMs were hypodense (5/5) without peripheral enhancement on contrast. All 6 patients with NPA showed peripheral enhancement on contrast. Three patients with VM subjected to computed tomography showed hyperdense lesion

with peripheral enhancement on contrast injection. One patient with large CM of face (SWS) underwent computed tomography head which revealed hyperdense lesion with enhancement. (as shown in [Table 4])

Angiography/Venography: Ten patients which included 5 cases with AVM, 3 NPA, one case of

liver hemangioma and a case of SWS underwent angiography studying parameters like early opacification (shunting), size, location and number of feeders and efferent veins and location of nidus. One patient with Klippel-Trenaunay Syndrome had ascending venography done. Five patients with VM were subjected to Venography. (as shown in [Table 5])

Table 3: showing MRI findings in various vascular anomalies.

Lesion	No. of patients	MRI/MRA/MRV	T1W Signal intensity			T2W Signal intensity		Flow voids	
			High	Inter	Low	High	Low	Present	Absent
H	50	18	0	18	0	18	0		
LH	2	2	0	1	1	2	0		
LM	14	14	0	0	14	14	0	-	-
LVM	2	2	0	0	2	2	0	-	-
VM	26	26	23	3	0	2	24	-	-
AVM	20	20	0	1	19	20	0	20	0
KTS	4	4	0	3	1	4	0	1	0
SWS	1	1		1	0	1	0	0	0
PG	1	1	0	0	1	1	0	0	0
NPA	6	0	-----						
CM	2	0	-----						
BRBNS	1	0	-----						
Total	129	88							

H- Hemangioma, LH- Lympho-Hemangioma, LM- Lymphatic Malformation, LVM- Lympho-Venous Malformation, VM- Venous Malformation, AVM- Arterio-Venous Malformation, KTS- Klippel Trenaunay Syndrome, SWS- Sturge Weber Syndrome, PG- Pyogenic Granuloma, NPA- NasoPharyngeal Angioma, CM- Capillary Malformation, BRBNS- Blue Rubber Bleb Nevus Syndrome, MRI-Magnetic Resonance Imaging, MRA- Magnetic Resonance Angiography, MRV- Magnetic Resonance Venography, T1W-T1 Weighted, T2W-T2 Weighted

Table 4: showing Computed Tomography findings in various vascular anomalies

Lesion	No. of patients	CECT done in	Density		Enhancement
			Hypo	Hyper	
Hemangioma	50	7	7	0	7
Lymphatic malformations	14	5	5	0	0
Venous Malformation	26	3	3	0	1
Sturge Weber Syndrome	1	1	0	1	1
Nasopharyngeal Angioma	6	6	0	0	6
Total		22			

CECT-Contrast Enhanced Computed Tomography

Table 5: showing findings on Angiography/Venography

Lesion	Patient No.	Venography	Angiography	Opac Feeder Nidus		
Venous Malformation	26	5	-	5	0	0
Arteriovenous Malformation	20	-	5	5	5	5
Hepatic Hemangioma	1	-	1	0	1	0
Nasopharyngeal angiofibroma	6	-	3	0	3	0
Klippel-Trenaunay Syndrome	4	1	-	Absent deep veins		
Sturge Weber Syndrome	1	-	1	1	1	0
Total		6	10			

Opac - Opacification

DISCUSSION

Vascular anomalies comprise a heterogeneous group of anomalies divided into two broad categories, tumours and malformations, on the basis of clinical behaviour and cellular kinetics. Some of these lesions are relatively common whereas others are rare. A classification system for vascular anomalies has already been developed some thirty years ago which provides the framework for understanding the pathophysiology, diagnosis, and prognosis of these anomalies but there are few notorious complex

lesions which does not fit into the criterion laid for distinguishing these lesions.^[5,6] The present study involved 129 patients referred to our institutes which were classified as per the previously laid down criterions for further sub-classifying vascular anomalies using the clinical and radiological parameters as shown in [Table 1].

Clinical Parameters:

Age: Vascular tumours: Majority of vascular tumours are present at the time of birth or appear during infancy (76% in the present study). Out of

these, 16 patients had lesions present at the time of birth and were grouped as congenital hemangiomas. Only 4 cases were considered as RICH and rest 12 as NICH according to their clinical behaviour.^[7] Rest Thirty four cases were considered as hemangioma of infancy. Vascular malformations: Majority of vascular malformations are noted at birth or within 2 years of life. 48% lesions were present at the time of the birth or infancy. In 26% patients the lesions were noticed around puberty due to increase in size because of hormonal disturbances.^[8,9] Incidence: Vascular tumours: The incidence of hemangiomas among vascular anomalies varies extremely ranging from 30% to 80%.^[1, 6] In a study done by Finn MC et al hemangiomas formed a group of 271 lesions out of 337 (80%).^[10] In the present study vascular tumours formed approximately 39% of the total cases which included 50 hemangiomas. Overall it is very difficult to ascertain the true proportion of vascular tumours among vascular anomalies. Since our institutes are a tertiary care referral centre, lesions which presented with symptomatology were generally referred. Vascular malformations: on the other hand have a lower incidence of occurrence than hemangiomas.^[10] VM, AVM and LM formed approximately 20%, 16% and 11% of the cases respectively in our study. Sex ratio: Jackson I.T et al from London, Fishman SJ et al and Finn MC et al from Boston, while studying vascular anomalies, found a sex ratio of 2-5:1 with female predominance.^[10, 11] In our study overall sex ratio was found to be 1: 1.22 with female predominance, 1: 1.27 in favour of females for hemangiomas, which is far less than the earlier published studies. The sex ratio among vascular malformations in our study was found to be 1: 1.22, slightly more towards females. This was found to be at par with the earlier studies.^[1, 6, 11] Clinical presentation: In general there are two major categories of vascular anomalies: tumours and malformations. Medical history and physical examination can distinguish between vascular tumour and vascular malformations with a diagnostic accuracy of over 90%.^[1, 10] Appearance of lesion: The majority of vascular tumours (70 to 90 percent) appear between the first and fourth week of life.^[6] Twenty four out of the total 50 hemangiomas and 10 out of 14 cases with LM had lesions present at birth. Contrary to the global trends, half of the vascular malformations (52%) were seen to be noticed later in life in the present study.^[1, 8] The larger numbers in this group may be because of hormonal influences at puberty and referrals to our tertiary care institutes from peripheral institutes. Though these patients came into contact with the treating institute at older age, most of them had developed smaller lesions earlier in life. Among syndromes lesions maybe present since birth (2 cases of KTS with limb hypertrophy and 1 case of SWS with capillary malformation of face) or may develop later in life (cutaneous lesions in BRBNS and 2

cases of KTS).^[12, 13] With the protocol of Antenatal sonograms in the current era lesions can be detected antenatally as incidental findings (1 case of hepatic hemangioma detected antenatally). Symptoms and Signs: In the year 2005, Jennifer J M and Mulliken JB from Children's hospital and Harvard Medical school Boston studied vascular anomalies in children. They found these anomalies to be mostly asymptomatic.^[8] Vascular tumours: in addition to cosmetic disfigurement may cause major skeletal distortion or hypertrophy. Sometimes hemangiomas can produce a mass effect on local facial skeleton.^[14] One patient with oral hemangioma in our study caused dental malocclusion requiring wiring earlier in life. Bleeding, ulceration and infection may complicate the lesion which was seen in 18 patients.^[15] Some symptoms are site specific like seen as stridor in 2 cases of subglottic hemangioma and pain and lump abdomen in a case with hepatic hemangioma.^[16, 17] Vascular malformations: clinical presentations include varicose veins, limb edema or overgrowth, and port-wine stain. In a Mayo Clinic study of 185 patients with lesions of the extremities and pelvis, the most frequent clinical sign was skin discoloration (43%), and the most frequent symptom was pain (37%).^[18] In our study disfigurement due to swelling was the most common complaint seen in 27 (40%) patients followed by pain in 20 (29%), functional limitation in 17 (25%) and history of bleeding in 18 (14%) cases and all of these are the definitive indications for treatment.^[8] Lymphatic malformations are difficult to distinguish from subcutaneous hemangiomas clinically when present since birth but failure to involute over a period of time may allow clinical difference. Cystic hygromas in addition to swelling may present with respiratory distress if there is extension into mediastinum.^[1] A thrill and bruit are the most common clinical characteristics in high flow vascular malformations like AVM.^[19] The patient is often distressed by the buzzing sound, pain, and elevated local temperature. In our study we found raised local temperature and pain as presenting complaint along with swelling in 5 patients (50%) of arteriovenous malformation. Vascular malformation with associated syndromes may present with seizures in SWS because of AVM of brain, severe anemia with melena in BRBNS and limb length and size discrepancy in KTS. Multiplicity of lesions and Anatomic area involved: Multiplicity does not have much significance except that patients with more than 5 hemangiomas have a strong possibility of visceral hemangiomas, particularly the liver and gastrointestinal tract.^[1] In our study 1 patient of multiple subcutaneous lesions had multiple gastrointestinal venous malformations (> 100 lesions). The same child had a hepatic hemangioma confirmed on histopathology examination. There was total number of 166 lesions seen in 129 patients (excluding lesions in a case of BRBNS). Eighty percent hemangiomas present as a

single lesion and only 20% of hemangiomas are multiple.[20] In our study too, majority of hemangioma had single lesion. Classifying HOI according to areas involved into Localized, indeterminate, Segmental and Multi-focal is beneficial as Segmental variety of such lesions tend to be more aggressive and difficult to manage. Vascular malformation tends to be solitary except when associated with syndromes. Mulliken JB and Young et al of Boston in their study found hemangiomas to be most commonly located in the head and neck region (60%), followed in frequency by the trunk (25%) and extremities (15%).^[20] In the present study 80 lesions (48%) involved head and neck region which included 52% hemangiomas. Seventy five (44%) lesions involved extremities and trunk. Eleven (8%) lesions involved various internal organs including 3 hepatic and 2 subglottic hemangioma 4 parotid lesions. Lymphatic Malformations have strong predilection for cervicofacial region followed by axilla, mediastinum and extremities, in that order of prevalence. In our study 32% LM lesions (including LH and LVM) involved cervicofacial region and two LM lesions involved mediastinum and mesentery of small intestine, and 1 involved the gastrointestinal tract. VM lesions principally occur in the skin and subcutaneous tissues but can also involve muscle viscera, joint structure and central nervous system.^[1,10] In our study, 56% VM lesions involved head and neck region. Size of the lesion: Vascular tumours: Majority of the lesions in the hemangioma group were less than 20 square centimetres (37/57) and can be followed with observation. Vascular Malformations: Size of the lesions may vary from few millimetres to very large lesions. Majority of the vascular malformations excluding venous malformations were greater than 20 square centimetres in the present study (largest being approximately 300 square centimetres). Size of the lesion commensurate the growth of the child or may increase with infection, trauma or due to hormonal effects during puberty.^[9]

Radiological Parameters :

1. US Doppler: Hemangiomas: Doppler US show a characteristic high flow pattern, variable echogenic mass and high vessel density especially during the proliferative phase. During the involuting phase, the lesions decreases in volume with a reduced number of vessels per square centimetres but flow remains high in the remaining vessels.^[21] In our study majority (30/42) hemangiomas showed variegated echogenicity, (38/42) showed high vessel density (>5/ cm²) and all 42 cases had high flow rate. High-resolution prenatal ultrasonography can detect these lesions during intrauterine life as seen in a case of hepatic hemangioma in our study. Vascular malformations: Trop I and Dubious J et al tried to describe the diagnostic features, appearance, and vascularisation pattern of venous malformations using 7-10 MHz linear transducers Doppler ultrasound with low pulse repetition frequency (mean 1680 Hz) and the lowest wall filter of 25-50 Hz. They studied 51 subjects from 1991 to 1997 and found that in paediatric patients, doppler US is a non-invasive, easily available, and rapid mode of investigation of vascular lesions and can help confirm the diagnosis of vascular malformations. As seen in our study, VM appear as slow flow, hypoechoic lesions or as mixed echogenicity lesions suggestive of phleboliths seen in 16% patients. AVM appear as heterogeneous lesions with feeding vessels and are high flow lesions with arterialisation of the veins. Ultrasound in macrocystic LM shows a multi-loculated cystic mass whereas microcystic LM are hyperechoic (because of numerous interfaces) without any lesional flow at Doppler.^[21,22] US doppler can be used to assess the venous system involved within an affected extremity in case of KTS.^[23] Sonography is also helpful in delineating subcutaneous lesions in BRBNS as seen in our study.
2. Magnetic Resonance Imaging: MRI is a reliable technique for diagnosing vascular anomalies in children and in evaluating the extent of the disease and its relationship to adjacent structures. MRI offers the additional advantage of demonstrating the flow dynamics of the lesion and its distribution and has now become the initial examination in majority of the lesions. The criterion of hypersensitivity relative to normal liver parenchyma on T2 weighted sequences as studied by Rios PR and Lubbers PR et al was also evident in our study.^[24,25] Hemangiomas and malformations with arterial components like arteriovenous malformation are high flow lesions, former appears bright on T2-weighted sequences, and appears solid whereas latter has presence of flow voids on presaturation images. Lymphatic and venous malformations manifest slow flow as evident in our study. All VM appear slightly more intense than muscle on T1-weighted presaturation images and lymphatic lesions appears to be low intense to muscle on T1-weighted presaturation images and do not enhance with gadopentetate dimeglumine.^[26,27] In patients with syndromes, MRI is a valuable diagnostic modality to define both the type and extent of vascular malformations especially in KTS. The venous malformations associated with KTS are hyperintense on T2W images and lack flow voids. Further MRI is helpful in determining extremity hypertrophy in these patients.^[28]
3. Computed Tomography: Patients were subjected to CT scans in limited number of patients in our study especially where there was diagnostic dilemma to categorize the patient. CT scan shows size, outline, positional relationship, and tissue contents. In CT, proliferative hemangiomas appear as homogenous masses with intense, persistent enhancement, usually organized in a lobular pattern.^[29] In vascular

malformations CT demonstrates the extent of the lesion, its characterization (as hypodense or hyperdense), and heterogeneity before contrast and after contrast injection and help in differentiating different types of malformations.^[30]

- Selective angiography and venography: Selective angiography demonstrates the extent of the vascular involvement, the feeder and draining vessels, and also provides the guidelines for selection of the initial treatment. Venous and arteriovenous malformations are identified by early opacification. Presence of a nidus is an important component for arteriovenous malformation.^[5] Highly selective angiography and embolization plays a very important role in the diagnosis of high flow vascular malformations. Angiography may demonstrate shunting. Arterial and venous phases are present in the same arteriographic film if there is a significant degree of shunting.^[31] Microshunts which are not delineated by angiography become apparent shortly after ligation, incomplete embolization, or partial resection because of reorientation in blood supply and changed hemodynamics.
- Biopsy: is advocated for the lesions suspicious of malignancy.^[1] There was no case in our study which suggested malignancy on clinico-radiological examination. However, thirty one resected lesions were sent for histopathologic examination post operatively which helped to correlate the clinico-radiological findings.

Based on these clinical and radiological parameters these notorious lesions can be further sub-classified like Venous malformations into Types I-IV,^[32] Arterio-Venous Malformation grouped into Clinical Stages I-IV,^[33] HOI into Localised, Indeterminate, Segmental, Multi-focal, Lymphatic Malformations into microcystic, macrocystic or mixed varieties but in spite of all the investigative armamentarium available and previous classification methods applied in the past some lesions still overlap among the different categories like Lympho-hemangioma (LH),^[1,2,6,7] Complex combined Malformations (Syndromes) like Sturge Weber Syndrome (SWS), Blue Rubber Bleb Nevus Syndrome (BRBNS), and Nasopharyngeal Angiofibroma (NPA) as shown in [Table 1]. It also appears that all of the above classifications seem to be inadequate when it comes to the complex congenital vascular lesions and insights into the genetics and molecular level might offer novel results for better understanding these notorious vascular anomalies in near future.

CONCLUSION

Newborn with birthmark should always be documented by the attending health care provider at birth and referral to an expert for proper evaluation and careful parental/Guardian counseling. Doppler US should be the first line of investigation in broadly classifying vascular anomalies whereas CT Scan,

MRI/MRA/MRV, Angiography, Venography help in further sub-classification. Some confusion still persists in classifying few lesions like Lympho-hemangioma (LH), Complex combined Malformations (Syndromes) like Sturge Weber Syndrome (SWS), Blue Rubber Bleb Nevus Syndrome (BRBNS), and Nasopharyngeal Angiofibroma (NPA).

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REFERENCES

- Fishman SJ, Mulliken JB. Vascular Anomalies – A primer for Pediatricians. *Pediatric Clinics of North America*, 1998; 45(6): 1455-77.
- Enjolras O., Mulliken J.B. Vascular tumors and vascular malformations (new issues). *Adv Dermatol* 1997; 13:375-423
- Lee BB. Congenital venous malformations: new approach to old problem with multidisciplinary approach. In: Wang, ZG, editor. *Vascular Surgery Proceedings of 3rd Congress of Asian Vascular Society*. Beijing: International Academic Publishers, 1998, pp 252-54.
- Lee BB and Do YS et al. Management of arteriovenous malformations: A multidisciplinary approach. *J Vasc Surg*, 2004; 39: 590-600.
- Mulliken J.B., Fishman S.J., Burrows P.E. Vascularanomalies. *Current Problems Surgery* 2000; 37:517-584
- Mulliken J.B., Glowacki J. Hemangioma and vascular malformations in infants and children: a classification based on endothelial characteristics. *Plast Reconstr Surg*, 1982; 69: 412-20
- Mulliken J.B., Enjolras O. Congenital hemangiomas and infantile hemangiomas: missing links. *J Am Acad Dermatol* 2004; 50: 875-82
- Marler JJ, Mulliken JB. Current management of hemangiomas and vascular malformations. *Clin Plastic Surg* 2005; 32: 99-116
- Perksy MS. Congenital vascular lesions of the head and neck. *Laryngoscope* 1986; 96: 1002
- Finn MC, Glowacki J and Mulliken JB: Congenital vascular lesions: Clinical applications of a new classification. *Journal of Pediatric Surgery*, 1983; 18(6): 894-900.
- Jackson IT and Carreno R. Hemangiomas, Vascular Malformations, and Lymphovenous Malformations: Classification and methods of treatment. *Plastic and Reconstructive Surgery*, 1992; 91(7): 1216-29.
- Samuel M, Spitz Z et al. Klippel Trenaunay syndrome: clinical features, complications and management in children. *British J of Surgery*, 1995; 82: 757-61
- Fishman S.J, Smithers C.J, Folkman J, et al. Blue rubber bleb nevus syndrome: Surgical eradication of gastrointestinal bleeding. *Ann Surg* 2005; 241: 523-28
- Murphey MD, Fairbairn KJ, Parman LM, Baxter KG, Parsa MB, Smith WS. Musculoskeletal angiomatous lesions: radiologic-pathologic correlation. *RadioGraphics* 1995; 15: 893-917
- Drolet BA, Esterly NB, Frieden IJ. Hemangiomas in children. *N Engl J Med* 1999; 341: 173-81
- Powers C, Ros PR, Stoupis C, Johnson WK, Segel KH. Primary liver neoplasms: MR imaging with pathological correlation. *Radiographics* 1994s; 14: 459-82
- Orlow SJ, Isakoff MS, Blei F. Increased risk of symptomatic hemangiomas of the airway in association with cutaneous

- hemangiomas in a “beard” distribution. *J Pediatr* 1997; 131: 643–6
18. Pritchard DA, Maloney JD, et al. Surgical treatment of congenital pelvic arteriovenous malformation. *Mayo Clin Proc*, 1978; 53:607
 19. Boyd JB, Mulliken JB, Kaban LB et al. Skeletal changes associated with vascular malformations. *Plastic Reconstr Surg* 1984; 74: 789
 20. Mulliken JB, Young A. *Vascular birthmarks: hemangiomas and malformations*. Philadelphia: WB Saunders; 1988
 21. Dubois J., Garel L. Imaging and therapeutic approach of hemangiomas and vascular malformations in the pediatric age group. *Pediatr Radiol* 1999; 29: 879-93
 22. Trop I and Dubious J et al. Soft tissue venous malformations in pediatric and young adult patients: Diagnosis with Doppler US. *Radiology*, 1999;212(3): 841-45
 23. Elsayes KM and Menias CO et al. Vascular Malformation and Hemangiomas syndromes: Spectrum of imaging manifestations. *Am J of Radiology*, May 2008; 190: 12 91-98
 24. Cohen MD, Edward MK. *Magnetic resonance imaging of children*. Decker 1990; Philadelphia: 399
 25. Rios PR, Lubbers PR, Olsted WW, Morillo G. Hemangioma of the liver: heterogenous appearance on T2-weighted images. *AJR* 1987; 149: 1167-70
 26. Cohen JM, Weinreb JC, Redman HC. Arteriovenous malformations of the extremities: MR imaging. *Radiology* 1986; 158: 475-79
 27. Siegel MJ, Glazer HS, St. Amour TE, Rosenthal DD. Lymphangiomas in children: MR imaging. *Radiology* 1989; 170: 467-70
 28. Vilanova JC, Barcelo J, Smirniotopoulos JG, Andres RP, Villalon M, Miro J et al. Hemangioma from head to toe: MR imaging with pathologic correlation. *Radiographics* 2004; 24: 367-85
 29. Levine E, Wetzel LH, Neff JR. MR imaging and CT of extrahepatic cavernous hemangiomas. *AJR Am J Roentgenol* 1986; 147: 1299–1304
 30. Martin J. Fine, Roy A. Holliday, and J. Thomas Roland. Clinically Unsuspected Venous Malformations Limited to the submandibular Triangle: CT Findings. *AJNR Am J Neuroradiol* 16; March 1995: 491–94
 31. Withers EH, Franklin JD and Lynch JB. Resection of a massive arteriovenous malformation of the head and neck. *Plast. Reconstr. Surg.* 1979; 63: 566
 32. Puig S, Aref H et al. Classification of venous malformations in children and implications for sclerotherapy. *Pediatr Radiol* 2003; 33: 99-103.
 33. Kohout M.P., Hansen M., Pribaz J.J., Mulliken J.B.: Arteriovenous malformations of the head and neck: Natural history and management. *Plast Reconstr Surg* 1998; 102: 643-54.

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