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Research Article

Clinical Characteristics and Management of Vascular Anomalies: Our Institutional Experience

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Abstract: The objective of the study was to assess the types of various vascular anomalies in terms of their diagnostic criteria, ages of presentation, location, sex distribution, and management in a government tertiary care setup. Relevant clinical data (demographic- age, sex, place, occupation) including history was obtained from the patient. A detailed clinical examination was performed. Design followed was case series in a multidisciplinary clinic in a medical college. Study involved sample population of 20 patients with vascular anomalies seen from January 2013 to December 2013.Main outcome measured was diagnosis before and after hospital visit, symptoms, and treatment recommendations, age of onset, age at clinic visit, location of lesion, sex, and type of referring physician. A total of 10% of patients had cutaneous involvement, 80% of patients had vascular malformations, and 10% had infantile haemangioma. Five percent of vascular malformations were first noted at birth and 80% were noted at later than 10 years of age. Eighty-percent of patients were symptomatic from their vascular lesion. . Further diagnostic workup was recommended in 90% of cases, and treatment recommendations were made in 80% of cases. Significant confusion still exists regarding the appropriate terminology, diagnosis, and management of vascular anomalies. Multidisciplinary clinics effectively address these complicated and troubling disorders by providing accurate diagnoses, clear treatment recommendations.
Keywords: Vascular anomalies, Haemangioma, AV malformations.

INTRODUCTION

Vascular anomalies 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. The classification system, originally proposed by Mulliken and Glowacki [1] and later updated by the International Society for the Study of Vascular Anomalies [2] divides vascular anomalies into malformations and vascular tumours, based on their endothelial cell characteristics, clinical presentation, natural history, and histopathological characteristics. Vascular malformations are congenital structural lesions composed of anomalous blood vessels and/or lymphatics lined with endothelium without cellular hyperplasia. In contrast, vascular tumours demonstrate endothelial cell hyperplasia and behave like neoplasms. Because vascular anomalies do not fit neatly into traditional medical and surgical specialties, and owing to the relative rarity of many types of vascular anomalies, patients are prone for misdiagnosis or mismanagement.

METHODOLOGY

A retrospective review of 20 patients seen during the 1-year study period conducted from Jan 2013 to December 2013 is included. 40 percent of patients (eight) were females and the rest were male.60% of all cases (12 patients) were children aged less than 18 years old. The mean (median) age at the clinic visit was 25 years.

Table 1: Figures show the sex incidence in the study population

Sex	Incidence	Percentage
Female	8	40
Male	12	60

Specific information extracted from each patient's included sex, age at presentation, age of onset, diagnosis, symptoms (pain, swelling, bleeding, functional compromise, disfigurement, infection, other), location of the lesion, treatment recommendations, number of physicians seen before coming to the

speciality clinic, and type of referring physician. Data were recorded and analysed using Excel software (Microsoft).

RESULTS AND OBSERVATIONS

The majority (45%) of vascular malformations were first noted at 10 -20 years of age.

Table 2: Age incidence of vascular anomalies in present study

Age group	Incidence	Percentage
<10	4	20
10-20	9	45
20-30	4	20
30-50	3	15

However, 7 vascular malformations (35%) were noted at later than 10 years of age and 2 cases were detected as late as 40 years of age. Of those vascular malformations diagnosed after 10 years of age, the majority 13 cases (65%) were venous malformations, followed by arteriovenous malformations in 2 cases (10%), mixed malformation in one patient (5%). 2 of infantile haemangioma were noted within the first month of life (2 at birth). No case of infantile haemangioma was first noticed after 6 months of life in this study.

Table 3:- Depiction of various types of lesions in the study group.

Type	Incidence	Percentage
Capillary	2	10
Venous	13	65
Lymphovenous	1	5
Arteriovenous	2	10
Haemangiomas	2	10

The anatomic distribution of lesions in our patient population was fairly, evenly divided between the head and neck, and the extremities and trunk.

Table 4: Showing region wise distribution of cases

Region wise distribution	Incidence	Percentage
Head and neck	9	45
Upper limb	7	35
Lower limb	1	5
Trunk	3	15

A large majority (90%) of patients were symptomatic from their vascular lesion. Pain was the primary symptom in 8 cases, functional compromise in 10 patients, swelling in 18 cases, and disfigurement in 17 cases. Interesting finding noticed was that lesions on the extremities were painful than lesions elsewhere (35% upper extremity, 5% lower extremity), head and neck in 9 cases 45% (Table 5). Ninety- percent of our patients had cutaneous and subcutaneous involvement.

Table 5: Showing symptom wise presentation of study cases

Symptoms	Incidence
Pain	8
Swelling	18
Functional compromise	10
Disfigurement	17
Infection	2
Bleeding	5
Asymptomatic	2

Management And Treatment Recommendation followed in this study were as follows-

Three cases were advised conservative management, two cases of AV malformation were studied with contrast CT and feeding artery and draining vein found, one case had mandible involved, one had facial artery involved .Three cases were given intralesional steroid injection. 8 cases of venous malformation were operated of whom there were two boys aged 12 years with finger involvement, a 11 year old male child with thenar affection, a 10 year old boy with foot involvement, a 21 year male with finger ,and 28 year male with dorsum of hand being involved , and an 50 year old female with forearm involvement , and two cases of burnt out lesions on face.

Table 6: Different management strategies adopted in our study group

Management	Incidence	Percentage
No intervention	3	15
Surgery	8	40
Sclerotherapy	3	15
Palliative	1	5
Multiple modalities	5	25

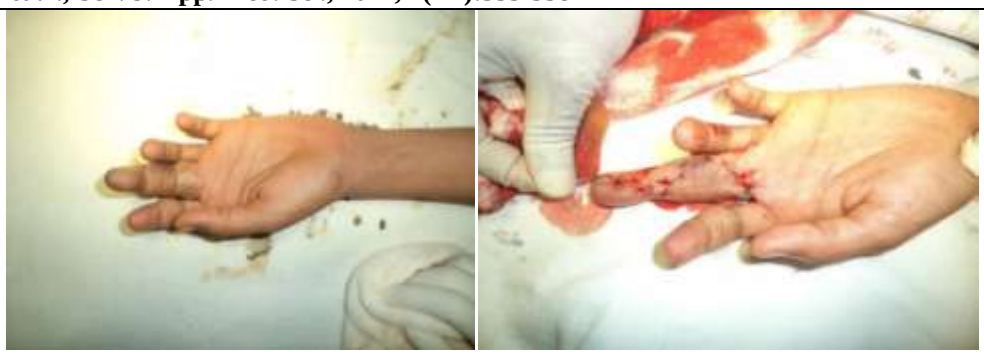

Fig. 1: Haemangioma over the middle finger of left hand before and after surgery

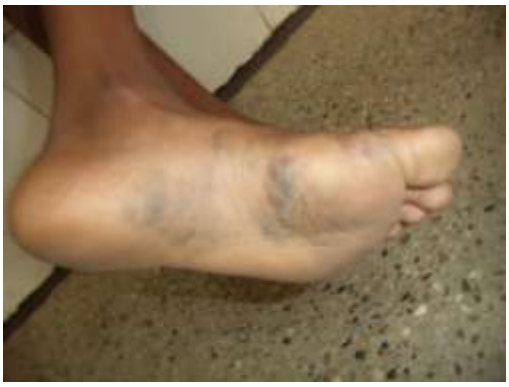

Fig. 2: Vascular malformation over the sole in an adult male

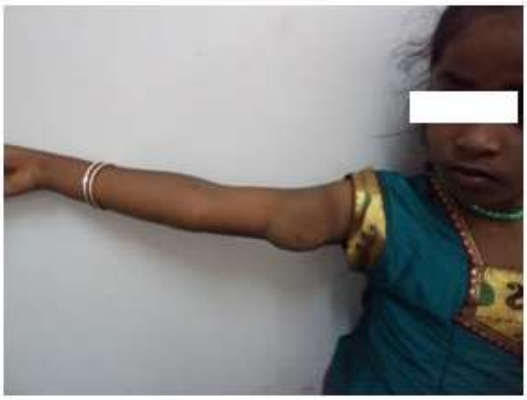

Fig. 3: Lymphovenous malformation in right arm of a girl aged 7 years

DISCUSSION

Although the classification of vascular anomalies has been well established in the literature over the past 2 decades, our study demonstrates that significant confusion still exists regarding the appropriate management.

The most common treatment methods employed were interventions such as sclerotherapy for venous and macro cystic lymphatic malformations, surgical excision, and palliative therapies such as low-dose aspirin and compression stockings. In 10% of patients for whom no treatment recommendation was made, no intervention was believed to be the most appropriate course of action. Such patients had uncomplicated infantile haemangioma. The knowledge transfer and extensive counselling that take place for each patient is of equal importance in treatment recommendation. The most common diagnosis seen was venous malformation, followed by infantile haemangioma and mixed vascular malformations. This distribution is similar to what has been reported in other series, yet clearly does not reflect the true incidence of these diagnoses in the population. The vast majority of infantile hemangiomas reported in literature were seen in the Paediatric Dermatology Faculty Practice.

The age of onset of the lesions in our study was also somewhat different from what was initially reported by Finn *et al.* [3]. Interestingly, in our population only 10% of the vascular malformations were apparent at birth, as opposed to 99% in study by Finn et al. Our finding is similar to that of several more recent publications that report a significant minority of patients with venous and arteriovenous malformations presenting either in childhood or adulthood [4, 5]. The difference in the age of onset reported in more recent studies as compared to older studies by Finn et al could be mainly due to better understanding and classification of vascular lesions in the last 2 decades.

The symptoms that our patients report are similar to those reported elsewhere [6]. A large majority (90%) of our patients are symptomatic from their vascular lesion. Lower extremity lesions are complicated by pain, which often leads to decreased functionality. A significant proportion of oral lesions are complicated by swelling and bleeding. Clearly, many vascular anomalies are very troubling to patients and can have a profound impact on therapy.

CONCLUSION

Many of the vascular lesions appear few years after birth though congenital in nature.

Lesions in extremities are often painful and impair functionality. Lesions in mucosa require a multimodality approach.

The location of the vascular anomaly also plays an important role in predicting subsequent complications.

Dermatologists should be aware of the clinical manifestations and management of this group of diseases and should play an important role in multidisciplinary vascular anomalies clinics.

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