HISTOPATHOLOGIST'S OUTLOOK OF VASCULAR LESIONS IN CHILDREN - A 2 YEAR STUDY

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ABSTRACT: BACKGROUND: Traditionally there has been much debate about vascular lesions being developmental malformations or types of neoplasia. Vascular tumours are believed to result from developmental errors that occur at 4-10 weeks of gestation. Most cases are sporadic. Vascular malformations result from abnormal sized or abnormal number of vascular structures. These malformations usually manifest as cutaneous birthmarks and have had a number of classifications, producing a range of confusing terms. **AIMS AND OBJECTIVES:** To study the incidence, classification and spectrum of various vascular lesions in children with a clinicopathological perspective. **MATERIAL AND METHODS:** The present retrospective study was undertaken at the Department of Pathology, Paediatric Referral Hospital, during the period from June 2009 to May 2011. About 44 patients of the paediatric age group were analysed for clinical details and pathological diagnosis. Vascular lesions were classified histopathologically according to International Society for the Study of Vascular Anomalies (ISSVA) classification broadly into two groups, mainly vascular tumours and malformations. **RESULTS:** Of the 44 cases reported in our study, vascular lesions were commonly seen in infants. The male to female ratio was 1:2.1. Most common site involved was head and neck region (73%). 39 cases were classified as vascular tumours which included haemangiomas and 5 cases classified as vascular malformations which included lymphangiomas. **CONCLUSION:** Vascular lesions though clinically diagnosed only as haemangiomas, represent pathologically varied entities. These lesions when present in infants and in functional areas, their precise diagnosis by the pathologists makes a significant difference to the management of these simple looking vascular lesions of the children.
KEYWORDS: International Society for the Study of Vascular Anomalies (ISSVA) classification, Haemangiomas, Lymphangiomas.

INTRODUCTION: Vascular lesions are abnormalities of vascular elements that presents at birth or during infancy. The estimated prevalence of these vascular anomalies is 4.5% [1]. Traditionally there has been much debate about vascular lesions being either developmental malformations or types of neoplasia [2].

Vascular tumours are said to result from developmental errors that occur at 4-10 weeks gestation. Most of the cases are sporadic and are sometimes occasionally inherited in an autosomal dominant fashion. Vascular malformations result from abnormal sized or abnormal number of vascular structures. They usually manifest as cutaneous birthmarks and have had a number of classifications, producing an array of confusing terms [3].

Mulliken and Glowacki (1982) suggested a dualistic classification for vascular anomalies based on pathological features. International Society for the Study of Vascular Anomalies (ISSVA)

adopted this classification in 1996 and expanded it. Since then this classification is widely accepted as it shows a systematic approach towards vascular lesions that correlates predictably with clinical history, course of the disease and treatment decisions.

For the diagnosis of vascular lesions in children, medical history and physical examination are important. Emphasis has to be laid on precise histopathological report, meticulous clinical and radiological assessment [5]. Histopathology helps in confirming the clinical diagnosis, while imaging aids in diagnosis, estimating the extent of the lesion, and determining the feasibility of surgical resection.

The present study evaluates the incidence, classification and spectrum of various vascular lesions in children with a clinicopathological perspective.

MATERIAL AND METHODS: The present study of vascular lesions in children is a 2 year retrospective study done at Department of Pathology, Paediatric Referral Hospital from June 2009 to May 2011. The study group consisted of 44 patients of the paediatric age group, 14 of them being boys and 30 girls. They were analysed for clinical details and pathological diagnosis. The materials for the study comprised of specimens received at the Department of Pathology. Biopsies of surgical specimens were formalin fixed, paraffin embedded and sectioned. The slides were stained with Haematoxylin and Eosin and were examined under light microscopy. Vascular lesions were histopathologically classified according to ISSVA classification into two major categories which included vascular tumours and malformations.

RESULTS: In the present study, we reviewed 44 cases of vascular lesions occurring in children. Vascular lesions were commonly encountered in infants. There were 30 girls and 14 boys. Sex incidence clearly showed female preponderance with Male: Female ratio of 1: 2.1. Table 1 and 2 shows age and sex incidence of all the vascular lesions.

Commonest site involved by vascular lesions was head and neck (32/44, 73%), followed by upper extremity (4/44, 9%) and trunk (4/44, 9%). The other sites were lower extremity (3/44, 7%) and anogenital region (1/44, 2%). (Table 3)

Histopathological assessment was done according to ISSVA classification. Vascular lesions in our study were broadly classified into vascular tumours and malformations. Increased endothelial cell turnover was witnessed in vascular tumours and was not appreciated in vascular malformations. Increased endothelial cell turnover was assessed histopathologically under light microscopy by documenting increased mitoses. Of the 44 cases we reported, 39 cases were classified as vascular tumours which included haemangiomas (Figure 3A, 4A, 4B, 3B) and 5 cases were classified as vascular malformations which included lymphangiomas (Figure 5A, 5B). Figure 1 Shows bar diagram with vascular lesions.

In the present study, vascular tumours which included haemangiomas occurred predominantly in girls. 28 of 44 girls presented with haemangiomas. (Figure 2)

Complications were seen in 4 cases (2.95%). 2 cases presented with ulcer, 1 with abscess and 1 with bloody discharge. The site of the lesion was scalp in 2 cases, arm in 1 and chest in 1 case. All 4 patients were girls with haemangiomas.

DISCUSSION: Vascular lesions have been designated a variety of names since nineteenth century. They were thought to be formed due to longing of the mother for particular things or her dislike to them, hence these marks resembled mulberries, strawberries, grapes, pines or bacon. Virchow regarded them as vascular tumours and classified them based on channel architecture into angioma simplex, cavernosum and racemosum [1].

ISSVA classification system divides vascular anomalies into two categories namely vascular tumours and malformations. The major difference between two categories is based on increased endothelial cell turnover. Vascular tumours have increased endothelial cell turnover while vascular malformations do not. Vascular malformations are structural abnormalities of the capillary, venous, lymphatic and arterial system that grow in proportion to the growth of child [4].

In the present study, vascular lesions were commonly encountered in infancy. Sex incidence clearly showed female preponderance with Male: Female ratio of 1: 2.1. These findings were consistent with other studies [6, 7, 8, 9, 10].

The most common site involved by the vascular lesions in our series was head and neck (73%) followed by trunk (9%), upper extremity (9%), lower extremity (7%) and anogenital region (1/44, 2%). In one series about 60% of vascular lesions occurred on the head and the neck, 25% on the trunk, and 15% on the extremities [11]. Studies by other authors showed similar findings [9, 10]. Our findings are consistent with the available literature.

Mulliken et al. (1982) reviewed 49 specimens from variety of vascular lesions for their cellular features. There were two major categories of lesions that emerged from this review which included vascular tumours like haemangiomas (26 cases) and vascular malformations (26 cases) [2].
In one large series, 1127 cases of vascular lesions were evaluated and classified into haemangiomas (969 cases) and malformations (158 cases) [12].

Al-Adnani et al.[13] (2006) evaluated 144 cases of paediatric vascular anomalies which included 70% classified as vascular tumours and 30% classified as vascular malformations according to International Society for the Study of Vascular Anomalies (ISSVA) classification.
According to another study, vascular lesions were classified into haemangiomas (84%) and vascular malformations (16%) [10].

In this present study, of the 44 cases reported, 39 cases were classified as vascular tumours which included haemangiomas and 5 cases as vascular malformations which included lymphangiomas. Our results were consistent with available literature.

Haggstrom et al. (2006) reported complications in cases with haemangiomas. Ulceration was noted in 23.2 % of the cases, visual compromise in 6.9 %, airway obstruction in 1.8%, auditory canal obstruction in 1.1% and cardiac compromise was seen in 0.4% [14]. In our study, complications were seen in 4 cases of haemangiomas. Of these, 2 presented with ulcer, 1 with abscess and 1 with bloody discharge.

CONCLUSION: Vascular lesions although clinically diagnosed only as haemangiomas, represent pathologically diverse entities. Haemangiomas do not appear as simple as they sound and present with wide range of diagnostic dilemmas. Morphologically they exhibit varied pictures. It is important for pathologist to differentiate them into vascular tumours and malformations. These lesions when present in infants and in functional areas, their precise diagnosis by the

pathologists makes a significant difference to the management of these simple looking vascular lesions of the children.

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Table 1: Age Distribution.

VASCULAR LESION	< 6 MONTHS	> 6 MONTHS -1 YEAR	1-5 YEARS	> 5 YEARS	
Vascular tumours	7	17	6	9	39(89%)
Vascular malformations	1	0	4	0	5 (11%)
	8 (18%)	17 (39%)	10 (23%)	9 (20%)	44 (100%)

Table 2: Sex Distribution

VASCULAR LESION	MALES	FEMALES
Vascular tumours	11	28
Vascular malformations	3	2
Total	14 (32%)	30 (68%)

Table 3: Distribution of Lesions at different locations.

VASCULAR LESION	HEAD AND NECK	UPPER EXTREMITY	TRUNK	LOWER EXTREMITY	ANOGENITAL REGION
Vascular tumours	27	4	4	3	1
Vascular malformations	5	0	0	0	0
	32 (73 %)	4 (9%)	4 (9%)	3 (7%)	1 (2%)

FIGURES:

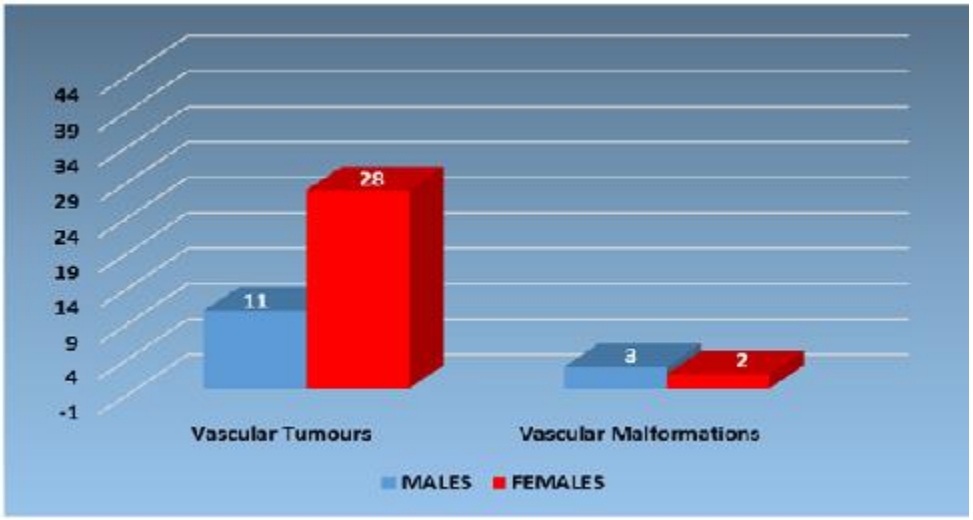

Figure 1: Bar diagram showing the percentage of different vascular lesions.

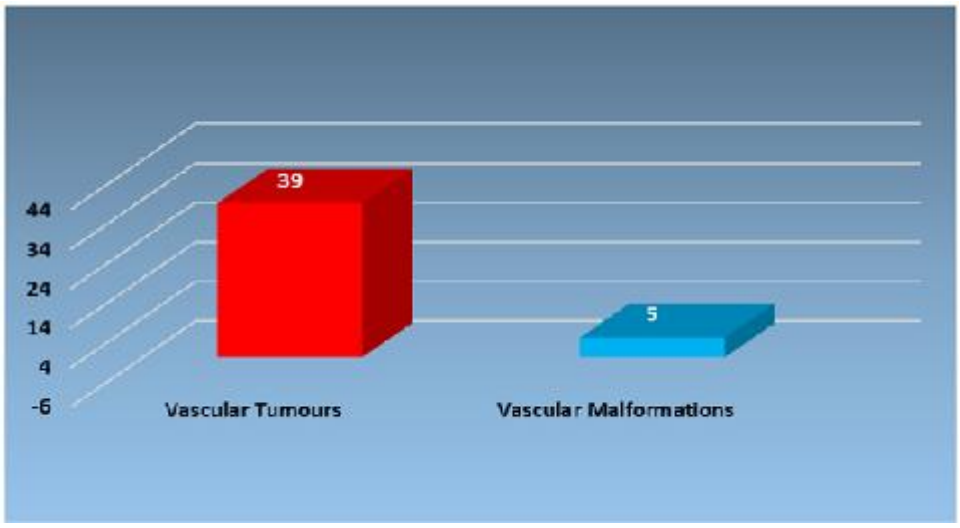

Figure 2: Bar diagram showing the sex incidence in different vascular lesions.

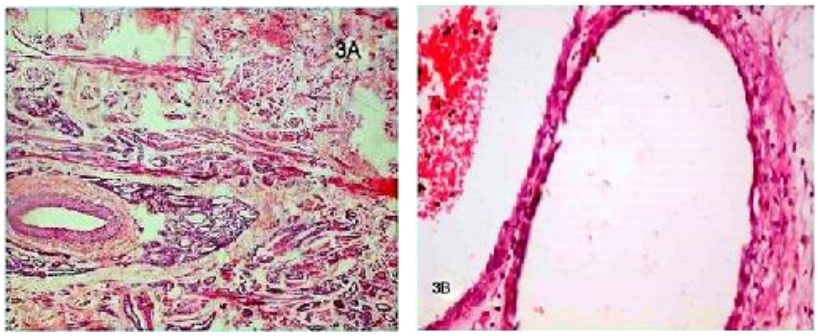

Figure 3: (A) Photomicrograph of haemangioma (Haematoxylin and Eosin, 10X) (B) Photomicrograph of haemangioma (Haematoxylin and Eosin, 10X)

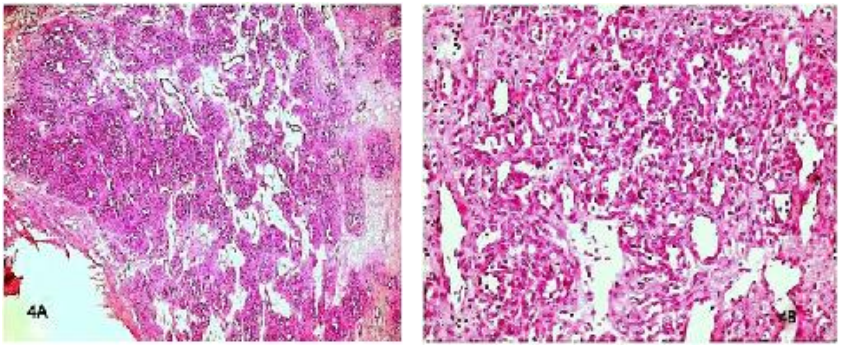

Figure 4: (A) Photomicrograph of lobular capillary haemangioma (Haematoxylin and Eosin, 4X) (B) Photomicrograph of lobular capillary haemangioma (Haematoxylin and Eosin, 40X)

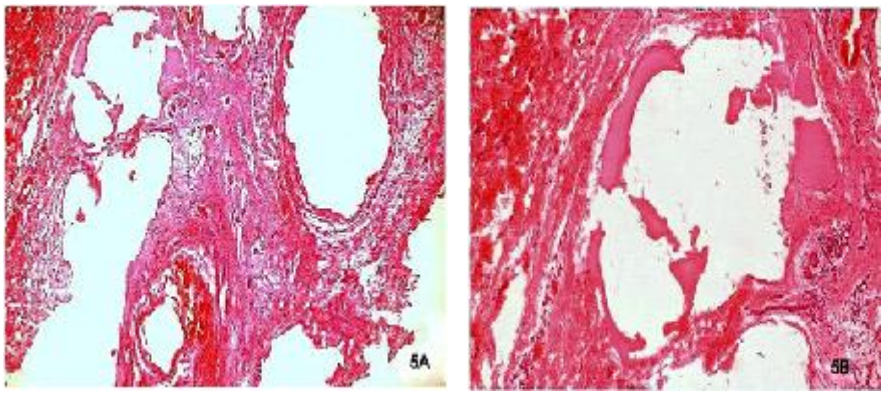

Figure 5: (A) Photomicrograph of lymphangioma (Haematoxylin and Eosin, 10X) (B) Photomicrograph of lymphangioma (Haematoxylin and Eosin, 10X)

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