

## Vascular nevi in children

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### ABSTRACT

**Background:** Vascular nevi are cutaneous anomalies of angiogenesis and vasculogenesis resulting in various different clinical presentations. **Aim:** The purpose of our descriptive study was to observe the various types of vascular nevi in children and their features. **Methods:** A total of 4256 pediatric cases attending the dermatology OPD during the study period from August 2002 to August 2004 were screened for vascular nevi. **Results:** Out of these, 19 children (0.44%) had vascular nevi—17 hemangiomas of infancy (HOIs) and 2 port-wine stains. The mean age of the affected children was 1.3 years (ranging from 2.5 months to 8 years). There were 13 girls and 6 boys. Seventeen (89.5%) patients had progressing lesions and two (10.5%) had non-involuting ones. A solitary lesion was seen in ten (52.6%) cases and two to five lesions were present in five (26.3%) cases. The nevi were distributed over multiple sites in seven (36.8%) cases, the head and neck in six (31.6%) cases, the chest and lower limb in two (10.5%) cases each, and the upper limbs and genitalia in one (5.3%) case each. Among the hemangiomas of infancy, 15 (88.2%) cases of superficial type and 2 (11.8%) cases of deep type were seen. The cutaneous complications included ulceration in four cases and infection in one. **Conclusions:** HOIs were the most common vascular nevi of childhood.

**Key Words:** Complications, Hemangioma of infancy, Vascular nevi

### INTRODUCTION

Vascular nevi are cutaneous anomalies of angiogenesis and vasculogenesis resulting in various different clinical presentations.<sup>[1]</sup> The incidence of all vascular nevi is probably approximately 6–25%.<sup>[2]</sup> Vascular nevi are classified into three major types—vascular malformations, hemangiomas, and angiokeratomas. Vascular malformations differ from hemangiomas pathologically by their lack of endothelial cell proliferation, and clinically by their presence at birth and their lack of any tendency to spontaneous resolution. They generally grow in proportion to the child, although they may become larger when

complicated by thrombosis, sepsis, or trauma.<sup>[3]</sup> Hemangiomas of infancy (HOIs) are benign, unique vascular tumors produced by proliferation of endothelial cells. They are the most common benign tumor of childhood.<sup>[1]</sup> Hemangiomas are often absent or small at birth and grow rapidly at a rate beyond the child's growth, followed by slow involution, often leading to complete regression.<sup>[4,5]</sup> Although most of these tumors are small and innocuous, some may be life- or function-threatening, or have associated structural congenital anomalies.

Frequently used modifiers of HOI such as strawberry, capillary, and cavernous are often ambiguous or

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confusing as they arise from faulty thinking regarding pathogenesis and should not be used.<sup>[6]</sup> Accepted modifiers for HOI refer to their clinical appearance, depending on where they arise in the cutis (superficial, deep, combined, or superficial and deep) or to their stage of growth (nascent, proliferating, involuting, or involuted). When larger numbers of infantile hemangiomas are seen in the neonate, the term benign neonatal hemangiomatosis is used. If they are associated with visceral hemangiomas, the term disseminated eruptive neonatal hemangiomatosis or diffuse neonatal hemangiomatosis is used. Multiple hemangiomas occur in as many as 25% of patients with hemangioma.<sup>[6]</sup> The purpose of our study was to determine the various types of vascular nevi in children and their features.

## METHODS

Children (aged below 14 years) attending the skin OPD of a tertiary care center were screened for vascular nevi and those with vascular nevi were the subjects of a descriptive study. The clinical findings were recorded in a proforma. The parameters noted were age, sex, age of onset, reason for visiting the hospital, family history, duration of nevi, site of involvement, morphology of lesions, number of lesions, and associated findings. In addition to the presenting symptoms, a skin biopsy was done wherever necessary. Relevant investigations were carried out for any systemic association. The results obtained were tabulated and analyzed.

## RESULTS

A total of 4256 pediatric cases attended the dermatology OPD during the study period from August 2002 to August 2004. Out of these, 19 cases (0.44%) were found to have vascular nevi. Seventeen of them were HOIs and the remaining were port-wine stains. The mean age of these cases was 1.3 years (ranging from 2.5 months to 8 years). There were 13 females and 6 males with a sex ratio of 2.2:1. No family history could be elicited in any of these cases. Fifteen (78.9%) cases were present since birth and remaining four (21.1%) cases appeared by the first year of life.

Fifteen (78.9%) cases were asymptomatic and four (21.1%) cases were painful. Seventeen (89.5%) cases had progressing lesions and two (10.5%) cases had noninvoluting ones. Out of the 17 cases of progressing lesions, 13 (68.4%) were gradually progressive and remaining 4 (21.1%) were rapidly progressive. Solitary lesion was seen in 10 (52.6%) cases, 2–5 lesions were present in 5 (26.3%) cases, 6–10 and multiple (>10) lesions were present in 2 (10.5%) cases each. Six (31.6%) cases were on the right side, 6 (31.6%) on the left side, 4 (21.1%) were bilateral, and 3 (15.8%) were central. The nevi were distributed over multiple sites in seven



Figure 1: Early telangectatic macular lesions (scalp) of hemangiomas of infancy



Figure 2: Proliferative phase of hemangiomas of infancy having ulceration (nape of neck)

(36.8%) cases, head (Figure 1) and neck in six (31.6%) cases, chest and lower limb in two (10.5%) cases each, and upper limb and genitalia one (5.3%) case each (Table 1).

A single case of HOI had oral mucosal involvement. Among the HOIs, 15 (88.2%) cases were of the localized type; the remaining two (11.8%) were multifocal (Table 2).

Among the HOIs, 15 (88.2%) cases of superficial type and 2 (11.8%) cases of deep type were seen (Table 2).

The cutaneous complications included ulceration (Figure 2) in four cases and infection in a solitary case.

## DISCUSSION

Infantile hemangiomas are the commonest tumors of infancy, with a prevalence of approximately 1–3% after the first few days of life and approximately 10% by the end of the first year.<sup>[3]</sup> In a study of pattern of pediatric dermatoses in a referral center in South India, out of 2144 dermatoses recorded, 11 cases (0.5%) of hemangioma were seen.<sup>[7]</sup> In our study, hemangiomas were seen in 17 cases.

Early on, the clinical appearance of HOI varies, depending on the location of the tumor within the skin.<sup>[4]</sup> The lesion in the superficial dermis is raised, lobulated, and bright red. Deep hemangiomas arise in the reticular dermis or subcutis and appear as soft

masses, often with a bluish cast. The overlying epidermis usually appears normal. Not uncommonly, HOI will have both superficial and deep features, named by one author as a “poached egg.” In one series, 62% of observed HOIs were superficial, 15% were deep, and 22% were both superficial and deep.<sup>[4]</sup> In our study, among the HOIs, 15 (88.2%) cases of superficial type and 2 (11.8%) cases of deep type were seen. Thus, the superficial type was much more common than the other types. In a study conducted by Chiller *et al.*<sup>[6]</sup> the mean age of HOI at the first visit was 1 year. Similarly, in our study, the mean age of HOI at the first visit was 0.8 years. Girls are more frequently affected than boys, with the ratio ranging from 5:1 to 2:1, except in small preterm infants where it is closer to 1:1.<sup>[8]</sup> In a study of 327 patients with HOI, there were 257 (79%) girls and 70 (21%) boys with a female to male ratio of 3.7:1.<sup>[6]</sup> Similarly, we found females (76.5%) to be much more frequently affected than males (23.5%), with a sex ratio of 3.3:1. Chiller *et al.*<sup>[6]</sup> documented a positive family history of vascular birthmarks in 10% cases, but there was no family history of vascular birthmarks in our patients.

Hemangiomas are often absent or small at birth and grow rapidly at a rate beyond the child’s growth followed by slow involution, often leading to complete regression.<sup>[4,5]</sup> In the study of Chiller *et al.*<sup>[6]</sup> a total of 36% of patients with HOIs had lesions at birth, whereas 40% developed lesions within the first month of life. In our study of 17 cases of HOI, 13 (76.5%) cases were present at birth and remaining 4 (21.1%) cases appeared up to 1 year of age. Thus in our study, almost all cases developed their lesions within the first year of life, which is comparable to other studies in which cases also there was an early onset of HOI.

The growth characteristics of HOI are often divided into phases—nascent, proliferating, involuting, and involuted.<sup>[4]</sup> In approximately 50% of neonates a premonitory mark may be evident at birth. These nascent lesions may appear as a telangiectatic macule surrounded by a pale halo, a pale macule, an erythematous macule, or a scratch. Most HOIs begin their growth phase in the first few weeks of life. The phase of rapid growth is usually most pronounced during the first 3–6 months of life but may be followed

**Table 1: Site distribution of vascular nevi and hemangiomas of infancy**

	Vascular nevi	Hemangioma of infancy
Multiple sites	7 (36.8%)	7 (41.2%)
Head and neck	6 (31.6%)	4 (23.5%)
Chest	2 (10.5%)	2 (11.8%)
Lower limb	2 (10.5%)	2 (11.8%)
Upper limb	1 (5.3%)	1 (5.9%)
Genitalia	1 (5.3%)	1 (5.9%)

**Table 2: Characteristics of hemangiomas of infancy**

Characteristics of hemangioma	No. of cases (%)
Pattern of distribution	
Localized	15 (88.2)
Multifocal	2 (11.8)
Segmental/ indeterminate	0
Depth of hemangioma	
Superficial	15 (88.2)
Deep	2 (11.8)
Mixed	0

by a period of slower growth. Some may not have proliferative growth and some may continue to grow for more than 1 year. Similarly, the time of onset of involution is somewhat unpredictable, in some cases beginning at a few months, and in most cases by 12–18 months. Complete involution of HOI occurs at an estimated rate of 10%/year.<sup>[4]</sup> In our study of 17 cases of HOI, 15 (88.2%) were progressive and two (11.8%) were non-involuting. Out of these 17 progressive cases, 11 (64.7%) were progressing gradually and remaining 4 (21.1%) were progressing rapidly. Thus in our study of 17 cases of HOI with a mean age of 0.8 years, majority of the cases were in the proliferative phase and only two (11.8%) were non-involuting. Also, no cases were in the involuting phase or completely involuted phase in our study.

In 80% of cases of HOI, only a single lesion is present, but it is not uncommon for them to be several, and occasionally, large numbers may occur.<sup>[3]</sup> Approximately 15–30% of infants with hemangiomas have multiple lesions; however, they are usually few in number.<sup>[8]</sup> In our study of 17 cases of HOI, only 8 (47.1%) were solitary, the remaining 9 (52.9%) were multiple. Out of these nine cases with more than one lesion, five (29.4%) had two to five lesions, whereas two (11.8%) had six to ten and multiple (>10) lesions. Thus, in our study, cases with solitary and multiple lesions were in almost equal percentage in contrast to the earlier studies.

Although HOIs may occur on any part of the body, they demonstrate a striking predisposition for the head and neck regions.<sup>[4]</sup> In a large series, 60% of HOI occurred on the head and neck, followed by 25% on the trunk, and 15% on extremities.<sup>[9]</sup> In another series, the most commonly involved anatomic segment (6% of the lesions) was the anterior cheek, followed in order of decreasing frequency by the forehead and the preauricular area. Thus, the majority of lesions were on the head and neck.<sup>[6]</sup> We found that hemangiomas were distributed over multiple sites in seven cases, head and neck in four cases, chest and lower limb in two cases each, and upper limb and genitalia in one case each. Thus, as far as single site of involvement is concerned; the most commonly involved site was the head and neck,

followed by the chest and extremities, which is comparable to earlier studies. In a study of 472 lesions of HOI in 327 patients, the mucous membrane was involved in 48 (10%) lesions.<sup>[6]</sup> In our study, a solitary case of HOI had oral mucosal involvement.

In their study of HOI, Chiller *et al.* made a diagram of the human body and gave a numerical value to each anatomic location.<sup>[6]</sup> With the help of photographs of individual lesions or written descriptions, they mapped the individual lesions. Finally, they classified the lesions into localized, segmental, indeterminate, and multifocal. Localized lesions demonstrated clear spatial containment, usually with involvement of only 1 or 2 mapped sites. Segmental lesions demonstrated linear and/or geographical localization over a specific cutaneous territory and were usually associated with at least some plaque-like features. They were often unilateral and usually sharply demarcated at the midline, but there were exceptions, particularly nasal and lip lesions. The indeterminate lesions were hemangiomas that could not be confidently categorized as either localized or segmental. Hemangiomas were considered multifocal if the infant had eight or more individual noncontiguous lesions of any morphologic characteristic.<sup>[6]</sup> In the study of 472 (327 cases) lesions of HOI, 339 (227 cases) were of the localized type, 84 (78 cases) were of the segmental type, 37 (34 cases) were of the indeterminate type, and 12 (12 cases) were of the multifocal type. Chiller *et al.* also found that segmental hemangiomas had a significantly higher rate of complications and higher incidence of developmental anomalies.<sup>[6]</sup> In our study of 17 cases of HOIs, the localized type was seen in 15 (88.2%) cases and the multifocal type in two (11.8%) cases, but there was no case with the segmental or indeterminate type. Thus, the majority of our cases was of the localized type, as also noted in the above study.<sup>[6]</sup>

Complications of HOI may result from their location, size, or rapid, proliferating phase. They include lesional ulceration, bleeding, pain, infection, etc.<sup>[3,4]</sup> Ulceration is the most frequent complication, affecting 5–13% of all lesions. Ulceration does not herald involution but occurs during the proliferative phase.<sup>[4]</sup> In the study by

Chiller *et al.*<sup>[6]</sup> 40% of the lesions had some type of complication. Ulceration was observed in 75 (21%) lesions along with bleeding (usually minor) in 27, cutaneous infection in 14, and significant pain in 12. In our study, four (23.5%) cases had some type of complication. Those complications included ulceration in all four cases and associated infection in a solitary case. Thus, ulceration was the most common complication in our study as in other studies.

To conclude, HOIs are one of the most common vascular nevi of childhood. The superficial type of HOI occurs more often than the deep type. The head and neck are the frequently affected sites. Moreover, they frequently occur as multifocal lesions. Ulceration is the most common complication.

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