Original Article ISSN (O):2395-2822; ISSN (P):2395-2814

A Clinical Study of Hemangiomas and Vascular Malformations.

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Received: March 2017
Accepted: March 2017

ABSTRACT

Background: A hemangioma is an abnormal proliferation of blood vessels that may occur in any vascularised tissue. Vascular malformations can be localised or diffuse errors of embryonic development. Aim: To study the incidence, age of onset, sex distribution, familial incidence, distribution of lesions, complications and associations of hemangiomas and vascular malformations. **Methods:** A total of 52,369 cases attended the dermatology OPD during the study period from august 2007 to September 2009 were studied. **Results:** Out of 52,369 cases, 56 cases (0.11%) were found to have vascular naevi. Among 56 cases, 36 of them were hemangiomas and 20 cases were port-wine stains. **Conclusion:** Among hemangiomas and vascular malformations, incidence of hemangiomas was twice as that of the other. There was a slight female preponderance of hemangiomas among females, in which head and neck was the commonest site and common complication was ulceration.

Keywords: Infantile hemangioma, congenital hemangiomas, vascular malformations, Port-wine stain.

INTRODUCTION

A hemangioma is an abnormal proliferation of blood vessels that may occur in any vascularised tissue. Mulliken strongly supports classification of hemangiomas as neoplasms.[1] Infantile hemangiomas are commonest tumors of infancy with prevalence of 4%-10% of full term neonates.[2] Infantile hemangiomas become apparent during the first month of life in about 90% cases where 65% are superficial, 15% deep, 20% mixed[3]. Spontaneous epithelial breakdown, crusting, ulceration, and necrosis complicate 5% of cutaneous hemangioma.[4] In the absence of complications, no treatment is indicated where a good cosmetic result can be predicted.[5]

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Vascular malformations can be localised or diffuse errors of embryonic development. The various types are due to faulty development during vasculogenesis and angiogenesis.[6] Capillary malformations are composed of dilated capillary to venule sized vessel in the superficial dermis. It includes salmon patch and Port-wine stain. Port-wine stain present as macular stains with a pink to dark red colour. It may progressively darken over many years, and occasional lesions develop secondary proliferative vascular blebs on their surface(cobble stone appearance).[7] PDL therapy is usually performed over several sessions, separated in time by 6-8 weeks, and can be quite effective in lightening these lesions.[8]

Aim

To study the incidence, age of onset, sex distribution, familial incidence, distribution of lesions, complications and associations of hemangiomas and vascular malformations.

MATERIALS AND METHODS

This is prospective observational study conducted in Department of Dermatology, Madras Medical College. Patients were screened for hemangiomas and vascular malformations. The clinical findings were recorded in a proforma. The parameters noted were age, sex, age of onset, reason for visiting the hospital, duration of lesion, site of involvement, morphology of lesions, number of lesions, and associated findings. Relevant investigations were carried out for any systemic association. The results obtained were tabulated and analysed.

RESULTS

Out of 52,369 cases, 56 cases (0.11%) were found to have vascular naevi. Among 56 cases, 36 of them

were hemangiomas and 20 cases were port-wine stains. Of the 36 cases of hemangioma, 30 cases had lesions at birth, 4 cases at 1st month, 1 case at 2nd month, 1 case by 3rd month. There was a female preponderance with female patients of 28 and males of 8. In our study 7 cases were born before 36 weeks and among 7 cases, 1 case was born before 32 weeks. The most common site of involvement is head and neck in 16 cases, 9 cases in trunk and 9 cases in extremities. 1 case had multifocal involvement. Out of 36 cases, 11 cases presented with various complications. 6 cases (16.67%)had ulceration, 3(8.33%)had bleeding, 2(5.56%) had feeding and breathing difficulty and 1 case had occlusion of eye. 1 case was associated with occlusion of vision and hydrocephalus and CT showed posterior fossa malformations in the form of Dandy Walker malformation. One case presented with hemangioma involving left cheek with stridor. Vascular malformations accounted for 20 cases of all the vascular anomalies. All had lesions since birth. 9 were males (45%) and 11 were females (55%). 15 cases had involvement of face, 4 cases (20%) had extremity involvement, 1 case had involvement of back. 5 cases (33.33%) had either ocular complication or CNS involvement or both. Klippel-Trenaunay Syndrome was noted in 2 patients. Phakomatosis pigmentovascularis was observed in 3 cases.

DISCUSSION

Infantile hemangiomas are the commonest tumours of infancy, with a prevalence of 1-3% after the first few days of life and 10% by the end of first year. In our study hemangiomas accounted for 36(64.29%)of all the 56 cases of vascular anomalies. 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. In the study of 327 patients of hemangioma by Chiller et al.[9] A total of 36% of patients had lesions at birth, whereas 40% developed within first month of life. In Senthilkumar et al.[10] study of 17 cases, 13(76.5%)cases were present at birth and 4(21.1%) appeared upto 1 year of age.In our study of 36 cases of hemangioma, 30(83.33%) cases had lesions at birth, 4(11.11%) cases had appearance of the lesion at 1st month, 1 case(2.78%) had appearance of lesion at 2nd month, 1 case(2.78%) had development of lesion by 3rd month. Thus in our study, almost all cases had their lesions within 3 months of life. In Chiller et al study, there were 79% girls and 21% boys with a female to male ratio of 3.7:1. In Senthilkumar et al study females (76.5%) and males (23.5%), with a sex ratio of 3.3:1.Similarly, we found that there was a female preponderance with female patients of 28(77.78%) and males of 8(22.22%) with a sex ratio of 3.5:1. Chiller et al documented a positive family history of hemangioma in 10% cases, whereas in our study there were 4 cases (11.11%) patients with family history of hemangioma. The prevalence of infantile hemangioma at 1 year in preterm infants was shown to be inversely related to gestational age at birth, and it occurs more commonly in infants born before 36 weeks.[11] In our study 7 cases (19.44%) were born before 36 weeks and among 7 cases, 1 case was born before 32 weeks. Hemangioma has inverse relationship to birth weight with increased prevalence in infants weighing less than 1500gm at birth. In our study 4(11.11%) cases were found to have LBW between 1500-2500 gm. Only 1 case (2.77%)hadbirth weight less than 1500gm. Mostly hemangiomas occur as a single lesion[12]. In Senthilkumar et al study only 8(47.1%) were solitary, the remaining 9(52.9%) were multiple. But in our study 32(88.89%) were solitary and 4(11.11%) were multiple. Chiller et al classified the lesions into localised, segmental, indeterminate and multifocal. Localised lesions have clear spatial containment, with involvement of 1 or 2 mapped sites. Segmental lesions demonstrated linear or geographical localisation over a specific cutaneous territory and were usually associated with at least some plaque like features. The indeterminate lesions were hemangiomas that could not be confidently categorised as either localised or segmental. Multifocal lesions had 8 or more individual non-contiguous lesions of any morphologic characteristic. In our study localised type was 24 cases (66.67%). Segmental type was in 5 cases (13.89%), indeterminate type was 6(16.67%) and 1 case (2.78%) had multifocal type. Of these, cases with segmental distribution had an increased incidence of complications which is in coherence with other studies. Although hemangiomas may occur onany part of the body, they demonstrate a striking predisposition for head and neck regions.[13] In our study hemangiomas were distributed in head and neck in 16(44.44%) cases, 9 cases (25%) in trunk and 9 cases(25%) in extremities. 1(2.78%) case had multifocal involvement. Thus the most common site of involvement was head and neck. The growth characteristics of hemangioma are often divided into 3 phases: proliferating, involuting and involuted. In our study of 36 cases, 4 cases (11.11%) were in involuting phase while 11 cases (30.56%) were in progressive phase and 21cases (58.33%) were in non-involuting phase. Of the 4 cases in involuting phase, involution started after 1 year of age in 3 cases and after the 2nd year in 1 case. Complications of hemangioma may result from their location, size, or rapid proliferating phase. In our study out of 36 cases, 11 (30.56%) cases presented with various complications. Out of which 6 cases (16.67%) had ulceration, 3(8.33%) had bleeding, feeding and breathing difficulty in 2(5.56%) and 1 case had occlusion of vision. Thus, ulceration was the most common complication in our study and it

occurred following trauma. Complications were common with children born either preterm or with LBW or both. Adjacent mucosal involvement was present in 4 cases (11.11%). But isolated mucosal involvement was not seen. Associations observed in our study were, 1 case with occlusion of vision and hydrocephalus and CT showed posterior fossa malformations in the form of Dandy Walker malformation. 1 case showed subglottic involvement in MRI.

Vascular malformations are localised or diffuse errors of embryonic development of which port-wine stain is a common vascular anomaly that is present at birth and persists throughout life. Port-wine stain accounted for 20 cases (35.71%) of all the vascular anomalies. All the 20 cases had lesions since birth. 9 were males (45%) and 11 were females (55%). In B.Tallman et al.[14] study of Port-wine stain in 310 patients, 68% had more than one dermatome involved. In our study 7 cases (35%) had unidermatomal involvement and 13 cases (65%) had more than multi dermatomal involvement. In the series of 121 cases by Bioxeda and colleagues, patients with bilateral distribution were 14%total population of those with capillary malformation.In our study,[15] 15 cases(75%) had involvement of face, 4 cases (20%)had extremity involvement, 1(5%) case had involvement of back. Of the facial involvement in 15 cases, extensive and bilateral involvement was present in 3 cases (20%), the rest 12 cases(80%)had unilateral involvement. The adjacent mucosal involvement was present in 6 cases (40%) out of 15 cases with facial involvement.

In Bioxeda et al study,[15] 88% had maxillary division of trigeminal nerve involvement. In our study also Port-wine stain had isolated involvement of maxillary distribution in 8 cases (53.33%) and ophthalmic branch was involved in 1 case (6.67%) and all the three branches were involved in 4 cases (26.67%). The patients with CNS or ocular complications or both had preferential distribution of the Port-wine stain over the maxillary branch of the trigeminal nerve. In our study of 15 cases with facial involvement 5 cases (33.33%) had either ocular complication or CNS involvement or both.

In our study out of the 3 cases with bilateral distribution, 2 cases (66.67%) and of the 12 cases with unilateral distribution, 1 case (8.33%) had glaucoma. Among the 3 cases with extensive & bilateral involvement, 2 cases (33.33%) and 1 case (8.33%) of unilateral distribution had a positive history for CNS involvement in the form of seizures. But all the three cases had normal CT finding.

In our study Klippel Trenaunay Syndrome was noted in 2 cases and both cases had limb length discrepancy and also an increased limb girth. Phakomatosis pigmentovascularis was observed in 3 cases of which 1 case was found to be associated with linear epidermal naevi while 1 was associated with both congenital melanocytic naevi and nevus of Ota and other case had an association with congenital melanocytic naevi alone.

CONCLUSION

Hemangiomas and vascular malformations are endothelial lesions of rare occurrence.Most of the hemangiomas had onset at birth and vascular malformations were present at birth. Hemangiomas had female predominance, while vascular malformations had equal sex distribution. The most common site for hemangioma was head and neck and Vascular malformation was in face. Hemangiomas commonly presented with single lesion, multi dermatomal involvement was in vascular malformations. Port-wine stain mostly occurred over distribution of maxillary division of trigeminal nerve. Most common complication of was ulceration in hemangiomas, seizures & glaucoma in vascular malformations.

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How to cite this article: Manjula J, Kumaravel S, Gokiladevi D, Lakshmi SV, Anandan H. A Clinical Study of Hemangiomas and Vascular Malformations. Ann. Int. Med. Den. Res. 2017; 3(3):DT05-DT08.

Source of Support: Nil, **Conflict of Interest:** None declared