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PATHOLOGIC FEATURES OF VASCULAR TUMOURS IN INFANTS AND CHILDREN IN LAGOS, NIGERIA

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ABSTRACT

Background. **This study was done to review the histopathologic characteristics of vascular tumours in infants and children in Lagos, Nigeria.**
Methods: **Fifty-two vascular tumours of children 0- 14 years of age that had been histologically diagnosed in the Department of Morbid Anatomy & Histopathology, LUTH, Lagos in the period January 1990 to December 1999 were reviewed. Clinical data on each of the cases was retrieved from the request cards. A minimum of two microscopic sections was examined for each surgically excised tumour in the study. The tumours were classified according to the latest WHO guidelines. The results are presented in the form of graphs, tables and analyzed using simple frequency distribution.**
Results: **Vascular tumours represented 27.3 % of soft tissue tumours in the period under review. The male to female ratio was 3.0: 1.0. Forty-eight of the tumors are benign in nature (92.2 %) of which the majority are haemangiomas (80.7 %). Intermediate and malignant vascular tumours accounted for 3.9 % each. However, there was no case of Kaposi's sarcoma. Most vascular tumours affected the head and neck region (40.4 %) followed by the trunk and extremities (15.4 % each). No multicentric tumours were found in the present study.**
Conclusions: **The study has shown that vascular tumours are the commonest soft tissue tumours of childhood in this environment. The histopathologic features of vascular tumours in infants and children in Lagos are similar to those described by earlier workers in Africa, Europe and the United States. However, Kaposi's sarcoma, a common tumour of children in East and Central Africa, is absent in this study as indeed in previous works on paediatric tumours from Jos and Zaria in Nigeria. There is the need for further studies to evaluate the factors responsible for these observed regional differences.**

Keywords: Histopathology; Vascular tumours; Children

INTRODUCTION

Vascular tumours are common in infancy and childhood. [1,2] The tumours are largely benign in nature and consist essentially of haemangiomas and lymphangiomas. However, arteriovenous malformations also commonly arise in

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also commonly arise in the vasoformative tissues in this age group. [3] These may be confused with certain neoplastic proliferations of the vasculature, for many of which new specific diagnostic and therapeutic measures have been developed in recent years.[4]

The true incidence and specific features of vascular tumours in infants and children in Nigeria is not known, because all the available studies are hospital-based surveys that often did not use standardized criteria. [5-7] However, the studies provide important data from this part of the world.

MATERIALS AND METHODS

The study was a retrospective histopathologic review of fifty-two vascular tumours that had been diagnosed in children aged 0-14 years old in the Department of Morbid Anatomy and Histopathology, LUTH, Lagos, in the ten-year period January 1990 to December 1999. The total number of histopathologic specimens of children received during the period was 2844 of which 190 were soft tissue tumours of various types including the 52 vascular tumours that are the subject of this study.

Clinical data on each case was extracted from the respective copies of the request cards. All tissues had been previously fixed in formalin. The haematoxylin and eosin (H and E) stained slides were retrieved in each case and reviewed microscopically by the authors. Where slides were missing or broken, fresh sections were taken from the paraffin-embedded tissue blocks of the specimens and stained. A minimum of two sections was examined for each surgically excised tumour in this study. To ensure adequate case selection, all previously unclassified soft tissue tumours of children were re-analyzed microscopically for vascular differentiation.

The tumours are classified into benign, borderline and malignant categories according to the latest WHO guidelines [7]. Cases in which the bio-data, clinical details, slides and / or tissue blocks were missing were excluded from the study. Special histochemical staining with Masson's trichrome method was used in establishing the true vascular nature of two lesions in addition to the routine H and E stains. The study was limited by the non-availability of immunohistochemical stains for the diagnostic confirmation of the vascular nature of the tumours. The results are presented in tabular form and analysed using simple frequency distribution.

RESULTS

Fifty-two vascular tumours of infants and children were analysed histopathologically. These constituted 1.8 % and 27.3 % of the surgical biopsies and the soft tissue tumours diagnosed in the age group 0-14 years respectively during the period reviewed (Figure 1). Thirty-nine of the tumours (75 %) occurred in males while thirteen (25 %) were diagnosed in females. The male to female ratio is therefore 3.0:1.0 (Table 1).

On microscopy, forty-eight of the tumours are benign in nature (92.2 %) while the intermediate and malignant types accounted for two cases each. Forty-two of the tumours are various types of haemangioma. These consisted of capillary haemangioma (26), pyogenic granuloma (10), cavernous haemangioma (3) and intramuscular hamangioma (3). There are six cases of lymphangioma and four of those are of the cystic hygroma

subtype. They showed proliferated, dilated lymphatic channels and occasional interstitial lymphocytic aggregates. Others (Table 2) were two haemangioendotheliomas, one angiosarcoma and one recurrent haemangiopericytoma with microscopic features of malignancy .

Twenty-one of the tumours (40.4 %)occurred in the the head and neck regio and involved the scalp, eyelids, lips, pinnae, oral mucosa and orbit. Lower and upper extremities and trunk are the next commonest areas involved, accounting for eight (15.4 %) of cases each. The neck was affected in 6 cases (11.5 %). Other sites were muscle (3 cases) and perineum (2 cases).

Figure 1: Comparison of vascular and other soft tissue tumours

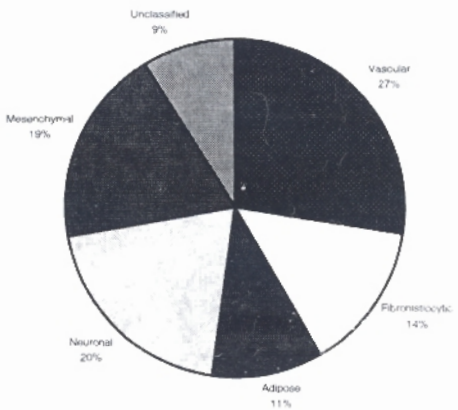

Table 1: Prevalence of vascular tumours in children (0-14 years)

Histological type	Male (%)	Female (%)	Total	M: F ratio
Benign				
Capillary haemangioma	20 (53.9)	6 (53.8)	26	3.0:1.0
Cavernous haemangioma	3 (7.7)	0 (0)	3	-
Granuloma pyogenicum	6 (15.4)	4 (30.8) 10	10	
Deep haemangioma	1 (2.5)	2 (15.4) 3	3	
Lymphangioma	6 (15.4)	0 (0)	6	-
Intermediate				
Haemangioendothelioma	1 (2.5)	1 (2.5)	2	1.0:1.0
Malignant				
Angiosarcoma	1 (2.5)	0 (0)	1	-
Malignant haemangiopericytoma	1 (2.5)	0 (0)	1	-
Total (%)	39 (100)	13 (100)	52	3.0:1.0

Table 2: Anatomical distribution of vascular tumours in children

Site	CH	CVH	PG	DH	LS	C	HME	MHP	ANG	Total (%)
Oral cavity	4	-	5	-	-	-	-	-	-	9(17.3)
Extremities	6	-	1	-	-	-	-	1	-	8(15.4)
Neck	-	-	-	-	1	4	-	-	1	6(11.5)
Trunk	6	1	-	-	-	-	1	-	-	8(15.4)
Scalp	4	-	3	-	-	-	1	-	-	8(15.4)
Intra-muscular	-	-	-	3	-	-	-	-	-	3(5.8)
Perineum	-	1	-	-	1	-	-	-	-	2(3.8)
Eye & Orbit	2	1	1	-	-	-	-	-	-	4(7.7)
Skin (Others)	3	-	-	-	-	-	-	-	-	3(5.8)
Not Stated	1	-	-	-	-	-	-	-	-	1(1.9)
Total	26	3	10	3	2	4	2	1	1	52(100.0)

CH – Capillary haemangioma; CVH- Carvenous haemangioma: PG – Pyogenic granuloma: DH – Deep haemangioma: LS – Lymphangioma simplex; C – Cystic hygroma; HME – Haemangioendothelioma; MHP – Malignant haemangiopericytoma, ANG - Angiosarcoma

Table 3: Age and type of vascular tumours in infants and children in Lagos

Age (Yrs)	HAEM. (%)	LS + CH (%)	HME (%)	MHP (%)	ANG (%)	Total (%)
0 – 2	21 (40.4)	3 (5.7)	-	-	-	24 (46.1)
2 – 4	4 (7.7)	2 (3.9)	-	-	-	6 (11.6)
4 – 6	9 (17.3)	1 (1.9)	-	-	-	10 (19.2)
6 – 8	1 (1.9)	-	-	-	-	1 (1.9)
8 – 10	2 (3.9)	-	2 (3.9)	-	-	4 (7.7)
10 – 12	2 (3.9)	-	-	1 (1.9)	-	3 (5.8)
12 – 14	3 (5.7)	-	-	-	1 (1.9)	4 (7.7)
Total	42 (80.7)	6 (11.6)	2 (3.9)	1 (1.9)	1 (1.9)	52 (100.0)

HAEM. –Haemangioma; LS + CH – Lymphangioma simplex & Cystic hygroma; HME – Haemangioendothelioma; MHP – Malignant haemangiopericytoma; ANG – Angiosarcoma

DISCUSSION

Among the commonest neoplasms diagnosed at birth or soon thereafter in childhood are those that arise in the vasoformative tissues. They are reported in 1.1 to 2.6 percent of all newborns.[1] They also account for between 3.2 to 25 percent of soft tissue tumours of children in reported series.[3,9]

A total number of fifty-two vascular tumours from this center recorded over a ten-year period is a rather low figure. This may be explained by the fact that the study is essentially a hospital-based survey. The figure, however, correlates with fifty-eight cases of vascular tumours in children previously reported from Zaria over a similar period.[5] A substantial number of the fifty-one vascular tumours reported from Enugu also occurred in childhood.[6] Coffin and Dehner encountered 228 vascular tumours in a clinicopathologic study of

cases in 222 children and adolescents. [9]

Majority of the vascular tumours in the study are benign (92.2 percent). Earlier workers have highlighted the preponderance of benign vascular tumours in this age group. [5, 6, 9] Haemangiomas alone account for a huge proportion of all the tumours here as in many reports in the literature where various incidences of between 2 – 10 % are frequently quoted. [6,9,10] A previous study of vascular tumours in this center had concentrated on haemangiomas alone. [11] By far the commonest indication for surgical intervention among those patients was cosmetic. The other common presenting complaints are ulceration with or without superimposed infection, bleeding, and also muscular swelling and pain. Giant haemangiomas may selectively trap circulating platelets and lead to severe thrombocytopenia often with a fatal outcome (Kasabach-Merritt syndrome). [12] Much importance tends to be attached to some other equally rare, complex syndromes associated with haemangiomas that are often of little clinical significance. [13, 14] Nonetheless, it is well to remember that a child with a known haemangioma of one site may have other undetected lesions involving a distant organ. [10] A search may therefore be quite rewarding. Wherever haemangiomas are found in a patient with multiple enchondromatosis there is a serious risk of malignant transformation of the mesenchymal component (chondrosarcoma) in 23 percent of cases. [12]

Rao, and also Coffin and Dehner, had noted multicentricity in fifty-one and nine patients with haemangiomas respectively. [9, 14] Despite this well recognized tendency, multiple haemangiomas are not found in the present report. Intramuscular or deep haemangiomas are uncommon here (only 3 cases) as indeed in many previous studies. [3,14] They are often discovered within the muscular planes and become apparent because of recurrent pains and swelling. [14]

Lymphangiomas are second only to haemangiomas in number and account for 11.5 percent of the vascular tumours in this study. They are comparatively rare tumours. [5, 9, 15] The cystic hygroma variant is said to occur in 1 in 12,000 live births. [16] The sex incidence is roughly equal, though a slight male predominance is often recorded [20] It is estimated that 50-65 percent of these tumours are present at birth, and as many as 90 percent may be clinically apparent by the end of the first year of life. The tumour almost never becomes malignant and therapy is largely dictated by their location and clinical extent. [16] Lymphangioma may cause serious respiratory embarrassment by impinging on the structures in the neck. In rare instances *in utero* foetal death has been experienced with these tumours in association with chromosomal abnormalities. [3]

The intermediate and malignant vascular tumours are rare tumours in children. [9, 17, 18] Two cases of haemangioendothelioma and one case each of malignant haemangiopericytoma and angiosaracoma are recorded in this series. Malignant haemangiopericytoma of infancy differs in several respects from the adult counterparts. [19] It tends to have histological features strongly suggestive of malignancy but it is noted to run a generally benign course and may even regress spontaneously. Dutz and Stout made a specially study of Kaposi's sarcoma in children and observed that it affects 4 percent of children less than 16 years of age in the United States. [20] AIDS produces profound immunodeficiency and susceptibility to opportunistic infections and various tumours, notably Kaposi's sarcoma. A remarkable aspect of the

review, therefore, was the complete absence of Kaposi's sarcoma. This finding correlates with recent works in Nigerian children done in the era of AIDS that have also failed to demonstrate the influence of the epidemic on the incidences of Kaposi's sarcoma. [5,18] In other African centers there has been noted an upsurge of the tumour and it is also noticed to run an aggressive and frequently fatal course particularly in association with HIV/AIDS. [21] In the industrialized nations, however, AIDS-associated Kaposi's sarcoma is known to preferentially affect a young adult population (mean age, 39 years). [19] These observed regional differences merit further investigation.

It is concluded that vascular tumours of infants and children are common soft tissue tumours in Lagos, Nigeria. The histopathological features of these tumours are basically same with those reported from the rest of the world, but Kaposi's sarcoma is absent in this age group here as opposed to the experience in East and Central Africa.

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