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## Sarcomas in Nigerian Children in Jos North Central Nigeria

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### Abstract

**Background**—There is a growing concern about childhood sarcomas, with recent studies suggesting an increase in the frequency of childhood sarcomas in sub-Saharan Africa. This study was carried out to determine the pattern of childhood sarcomas in Jos, North Central Nigeria and to compare the data obtained with other previous related studies.

**Methods**—Review of the Jos University Teaching Hospital cancer registry from January 2001 to December 2010. Data of all children (0-15 years) in the data base were retrieved for analysis.

**Results**—Two hundred and ten histological diagnosis of malignancies were made in children over the period, with 81 cases (39%) being childhood Sarcomas. The sarcomas occurred predominantly in males (54%) with male/female ratio of 2:1. The minimum age was 2 months and the maximum age was 15 years. Soft tissue sarcoma (STS) was the most predominant group which accounted for 73 cases (90%) of all sarcomas seen. Rhabdomyosarcoma (RMS) was the most common STS, it accounted for 65 cases (89%) of the STS and 80% of all the sarcomas. This is followed by Kaposi Sarcoma (KS) accounting for 6.9% of STS. There were 8 cases of Osteosarcoma which accounted for 10% of all the sarcomas. Embryonal RMS predominated in the very young children while all other sarcomas affected the older children. Extremities were the sites of predilection for most of the sarcomas (36%).

Seventeen (17) cases of the RMS were of superior prognostic group, 34 (54%) were of intermediate prognostic group while 24 cases (37%) were of poor prognostic group.

**Conclusion**—Childhood sarcomas are common in our environment and RMS is the single most common sarcoma while the non-rhabdomyosarcoma soft tissue sarcomas (NRSTS) are rare.

### Keywords

Childhood Sarcomas; Malignancies; Immunohistochemistry; Nigeria

### Introduction

There is growing interest in childhood mesenchymal malignancies particularly soft tissue sarcomas because they appear to be increasing in incidence in childhood and are associated with diagnostic challenges. Although most are aggressive tumours they are treatable to a large extent if accurately diagnosed and treated early.<sup>1,2</sup>

Tumours in children (aged 0-15 years) accounted for significant morbidity and mortality all over the world. Malignant childhood tumours accounted for more natural deaths than any other diseases within the first 15 years of life in developed countries, only second to accidental deaths and accounting for 9% of all natural deaths.<sup>1</sup>

In developing countries, infections and malnutrition were leading causes of death, however there appears to be increasing contribution from childhood malignancies particularly malignant mesenchymal tumours which are now recognized as a public health problem in developing countries of sub-Saharan Africa, including Nigeria.<sup>2,3,4</sup> For example in Ghana malignancies account for about 1.25% of clinical and 0.5% of autopsy causes of death in children.<sup>4</sup> Studies from developed and developing countries have revealed that most childhood tumours are mesenchymal in histogenesis in contrast with adult tumours that are largely epithelial. For instance Samaila et al in Zaria, north western Nigeria found out that 60.8% of the childhood solid malignant tumours reviewed over a period of ten years were mesenchymal in origin, while 39.2% were epithelial. The most common tumours in that study were rhabdomyosarcoma, Burkitt's lymphoma, retinoblastoma and nephroblastoma in that order. This trend is similar to most local data and even among western children.<sup>1-9</sup>

Are sarcomas really becoming commoner in our children? This trend could reflect a better understanding of the biology of sarcomas and improvement in their diagnosis particularly with the increasing use of immunohistochemistry and other molecular techniques. Kaposi sarcoma, an intermediate grade sarcoma has become prevalent among children because of the HIV/AIDS pandemic. In two separate studies (10 years review) four cases each were found in children in Zaria North Western Nigeria by Rafindadi<sup>10</sup> and Samaila.<sup>3</sup>

Primary sarcomas from bones are also seen in children in our environment, particularly osteogenic sarcoma.<sup>11</sup> Ewing's sarcoma is said to be the second most common sarcoma among western children, is rare among black children.<sup>11-17</sup> Infantile fibrosarcoma occurs exclusively in children but morphologically resembles the adult fibrosarcoma.<sup>15</sup> Childhood sarcomas are also been increasingly seen as second malignancies complicating radiation therapy and chemotherapy for first malignancies in early childhood.<sup>16,17,18</sup>

Morphologic diagnosis of sarcomas in children is associated with some challenges as highlighted above. The tumours sometimes present as primitive small round blue cells that can be difficult to differentiate from other tumours of similar histological features in childhood. Therefore Hematoxylin & Eosin sections are sometimes not enough to fully categorise sarcomas in children, particularly undifferentiated primitive tumours will often require further ancillary investigation such as immunohistochemistry, cytogenetics and electron microscopy.<sup>5</sup> These special diagnostic techniques are rarely available in our tertiary health centres in the tropics. For instance a review of recent methods and techniques in histopathology in the tropics by Akang south western Nigeria revealed that in Nigeria only seven (7) public tertiary health centres have a functional immunohistochemistry unit. None has functional electron microscopy (EM) and other molecular methods in use are limited to research only.<sup>19</sup>

## Methods

The study was a 10 year hospital based retrospective study employing slides and tissue blocks of specimens from children aged 0-15 years diagnosed with sarcoma in the histopathology department of Jos university teaching hospital (JUTH) from January 2001 to December 2010. JUTH is referral tertiary health centre in north central Nigeria.

The materials used in the study included records from the cancer registry and archival slides and tissue blocks from the archives of the histopathology department of the hospital. The specimens studied included those obtained through excisional, incisional and tru-cut biopsies received in the department.

Records of previous diagnosis, specimen number, age, sex and site of biopsy was retrieved from the cancer registry of the hospital. Tissue blocks were traced and fresh sections 5um were cut from paraffin embedded formalin fixed blocks and stained with H/E for missing slides. The slides are read and the WHO classification was used to classify the sarcomas. The American national cancer institute grading (NCI) scheme favoured by the Childhood Onchology Groups (COG) was used to grade the childhood non-Rhabdomyosarcomas soft tissue sarcomas (NRSTS), while Rhabdomyosarcomas were graded based on the histologic subtypes into superior (botryoid and spindle cell RMS), intermediate (embryonal RMS) and low prognostic (alveolar RMS and undifferentiated sarcoma) groups. Where diagnosis was not definite based on H/E morphology immunohistochemistry was employed for proper categorization and classification. The choice of antibodies was based on the H and E morphologic impression, affordability and availability. Vimentin and desmin were used in this study to confirm undifferentiated suspected RMS. Where vimentin and desmin were negative other antibodies were used based on the differential diagnosis. A semi quantitative 4 point count scoring of immunoreactivity was used. All sarcomas histologically diagnosed between January 2001 and December 2010 from children aged 0-15 years in the histopathology department of JUTH with traceable archival slides or tissue blocks and clinical data including age, sex and site of biopsy were included in the study while cytologically diagnosed sarcomas were excluded.

The data was analyzed using EPI info statistical software. The data was displayed in histogram and tables.

## Ethical Considerations

The research was given due ethical consideration which was strictly adhered to. In line with this, the proposal was subjected to ethical review by the Hospital Ethical committee.

## Results

Within the period under review, a total of 210 childhood solid malignancies were recorded out of which 89 were sarcomas, 6 were excluded from the study for lack of complete information and/or tissue blocks, while 2 were recategorised as lymphomas. Therefore, a total of 81 cases were studied. These 81 cases accounted for 39% of childhood malignancies seen within the period. The male: female distribution was 54 males and 27 females giving a

male/female of 2:1. The mean age was 9.5 year, median age was 10 year and the modal age was 14 year. The minimum age was 2 months while the maximum age was 15 years. The standard deviation for the age was 4.5.

Soft tissue sarcomas (STS) were the predominant sarcomas seen accounting for 90% (73 cases) of all the sarcomas. Rhabdomyosarcoma (RMS) was the single most predominant type of sarcoma accounting for 80% of all the sarcomas seen and 65 (89%) of STS in particular. The RMS included the embryonal rhabdomyosarcoma (ERMS) 41 cases which accounted for 63% of RMS and 50.2% of all sarcomas, alveolar rhabdomyosarcoma (ARMS) accounted for 35% (23 cases) of RMS and 28.4% of all sarcomas while pleomorphic RMS accounted for 1.5% (1 case) of RMS and 1.2% of all sarcomas. Kaposi Sarcoma (KS) was the second most common STS accounting for 6.9% (5 cases) of STS and 6.2% of all sarcomas, while fibrosarcoma accounted for only 2.7% STS with only 2 cases and 2.5% of all sarcomas seen. One (1) case of malignant peripheral nerve sheath tumour (MPNST) was recorded accounting for 1.2% of all the sarcomas seen. Osteosarcoma was the only malignant bone tumour recorded accounting for 9.9% of all sarcomas seen.

The non-rhabdomyosarcoma soft tissue sarcomas (NRSTS) as a group including KS, fibrosarcoma and MPNST seen in this study accounted for only 3.8% (8 cases) of all the 210 childhood malignancies seen within the period and 9.9% of all the 81 cases of sarcomas studied.

### Sex Distribution

The distribution of the sarcomas according to age shows 54 males and 27 females giving a male/female ratio of 2:1. All tumours show male predilection with embryonal RMS having M:F ratio of 2.2:1, ARMS 1.9:1, KS 1.5:1 and OS 1.7:1. All the two cases of infantile fibrosarcoma were seen in males, also the only case of pleomorphic RMS was seen in a male child while the only case of MPNST was seen in a female.

### Age Distribution

Generally, all the sarcomas occurred more in the older children than the very young, with a characteristic two peak age incidence. The age group 12-15 years had the most number of tumours accounting for 46% (38 cases) of all sarcomas seen followed by the 4-7 age groups accounting for 38.5%. These two peak age incidence reflects the incidence of ARMS in the 12-15 years and ERMS in the 4-7 years respectively. ERMS shows a bipolar peak age incidence 4-7 and 12-15 years see table I.

All the 8 cases of sarcomas occurring in the first 3 years of life were ERMS. ERMS and ARMS contributed equally to the 16 cases (19.8%, 8 cases each) of tumours in the age group 8-11 with contribution from the 2 cases of Osteosarcoma and 1 case of KS. The distribution of the individual sarcomas showed that 34% of ERMS occurred between 12-15 years, another 31% in the age group 4-7 years, 19.5% in the first 3 years of life while the remaining 14.6% occurred in the 8-11 years of life.

For ARMS, 56.5% of tumours occurred in the 12-15 years age group, 26.1% in the 8-11 years age group and 17.4% in the 4-7 year age group but non in the first 3 years of life. The only case of Pleomorphic RMS occurred in a 14 year old child.

For Karposi Sarcoma, the age distribution shows 40% each in the age groups 4-7 and 12-15 with the remaining 20% in the 8-11 years age group and non in the first three years. All the 2 cases of FS seen were seen in the age group 12-15 years, one at 14 years and the other at 15 years. The only case of MPNST occurred at the age of 14 years.

For osteosarcoma, 75% occurred between 12-15 years while the remaining 25% occurred between 8-11 years and non below 8 years.

### Anatomical Site Distribution

The anatomic distribution of the sarcomas were analysed based on 5 anatomical regions of the body: the head and neck, trunk, extremities (upper and lower extremities), the retroperitoneum and the genitourinary system (GUS).

Sarcomas from the extremities predominated accounting for 29 cases (36.6%) followed by the head and neck 23 cases (28%). The retroperitoneum was the third most commonly involved anatomical site with 11 cases (14%), the trunk 10 cases (12%) and the genitourinary system with 7 cases (10%). The head and neck region was the site of predilection for the STS (23%). ARMS shows site predilection for the head and neck region (34%), retroperitoneum (30.4%) and the extremities (21.7%). See Table II

Embryonal rhabdomyosarcoma also shows site predilection for head and neck region (13 cases, 31.7%), and extremities 10 cases (24%), while the trunk and the retroperitoneum were equally involved with 7 cases each (17%). All GUS tumours were RMS, 1 ARMS of the testis, 3 testicular ERMS, 2 ERMS from the urinary bladder and 2 from the vagina.

The only case of pleomorphic RMS seen was sited in the forehead of a 14 year old child. Of all the 5 cases of KS, 3 were from the lower limbs and 2 from the head and neck region (1 from the eye and 1 from the nasal cavity). The two cases of infantile fibrosarcoma occurred in the extremities. All the cases of osteosarcoma seen were from the long bones of the lower extremities, 6 from the lower limb involving the femur while the other 2 involved the upper limb (humerus).

### Tumour Grading

Using the COG (NCI) grading system for childhood sarcomas, RMS were classified into 3 prognostic groups based on the histologic subtypes while the other STS were graded based on the COG grading for NRSTS.

**RMS**—Eleven cases of Botryoid RMS were recorded and 6 cases of spindle cell subtypes were recorded which were classified as Superior Prognostic group (17). The remaining 24 conventional ERMS were classified into the intermediate prognostic group while the one case of Pleomorphic RMS together with the 23 cases of Alveolar RMS belongs to the Poor prognostic group (24).

All the 2 cases of Fibrosarcoma, the 5 cases of KS and one case of MPNST were well differentiated tumours with no necrotic foci seen and very scanty mitotic figures in some and all were placed as grade 1 tumours. Osteosarcomas were not graded based on the NCI/POG for NRSTS but all were well differentiated tumours and were graded as such (well differentiated osteosarcomas).

### Immunohistochemistry

Ten undifferentiated round blue cells tumours all previously diagnosed as RMS were immunostained with Vimentin and Desmin. Immunoreactivity was scored using a semiquantitative 4 point count as stated in the methodology. (see table III for immunohistochemistry result)

### Discussion

The study showed that childhood sarcomas accounted for 39% of childhood solid malignancies in Jos, Central Nigeria suggesting that non-lymphoid mesenchymal malignancies are common among children in our environment. Data from western societies shows that soft tissue sarcomas accounted for only 7% of childhood malignancies<sup>2,5,6,20</sup>. Previous studies by Tanko et al in Jos central Nigeria<sup>2</sup> identified STS particularly RMS as the most frequent childhood malignancy as opposed to a decline in the frequency of Burkitt's lymphoma previously reported to be the most frequent childhood malignancy in Africa<sup>3,4</sup>. This trend has also been demonstrated by Ocheni et al and Ojesina et al in Eastern and Western Nigeria respectively<sup>7,9</sup>. The pathogenic factors responsible for this high frequency of childhood sarcomas in our environment as revealed by this study and other similar studies calls for further research tailored at elucidating the possible factors responsible for this changing trend of rising incidence of childhood sarcomas.

The male dominance of all tumours in this studies is comparable with most local studies<sup>2,3,7</sup>. The study also showed that soft tissue sarcomas are the predominant sarcomas in childhood. Sarcomas from bones constitute the second category in childhood. In this study, STS accounted for 90% (73 cases) of childhood sarcomas out of which 89% (65 cases) were RMS. Osteosarcoma was the only primary malignant bone sarcoma seen in this study and it accounted for 9.9% (8 cases) of all the sarcomas recorded. In western societies, soft tissue sarcomas are the most common group of sarcomas seen in childhood, particularly RMS which is said to account for 40% of STS among western children<sup>5,6</sup>. Our study shows a higher relative frequency of RMS of 89% among black children doubling that of western societies. However, the non rhabdomyosarcoma STS (NRSTS) account for only 11% of STS showing they are rare tumours in children in Africa. Most local studies have consistently shown that childhood STS in Africa is predominated by RMS<sup>2, 4, 6, 19-26</sup>. However, extending the age to adolescents and young adults, the NRSTS will begin to appear as they are common in that age group.

The two cases of fibrosarcoma seen in this study occurred in the older children within the 12-15 years age group which is in sharp contrast with the early age of onset of congenital, infantile fibrosarcoma that occur almost at birth. The adult and infantile FS have similar morphology, but can be differentiated by cytogenetic abnormalities.<sup>27</sup> Infantile

fibrosarcoma is one of the most common NRSTS, several case reports of congenital fibrosarcoma have been published<sup>28,30</sup> The study has suggested some aetiological factors that may play a significant role in the pathogenesis of paediatric sarcomas. All the 5 cases of KS seen in this study were HIV seropositive suggesting the role of immunosuppression in the pathogenesis. This is similar to all the 4 cases recorded by Samaila in Zaria and 3 cases by Pindiga et al in Maiduguri, all of which were HIV seropositive<sup>3,25</sup>. Also, the appearance of MPNST in a child suggests the role of neurofibromatosis as an inherited cancer predisposing genetic syndrome recognized by some authors<sup>20</sup>. The early onset of ERMS and some other sarcomas in this study (table I) suggested that some genetic syndromes and in-utero factors may contribute in the aetio- pathogenesis of these tumours in our environment. Again, childhood osteosarcoma is the most common Second Malignant Neoplasm (SMN) complicating chemotherapy and/or radiation therapy for primary malignancy in early childhood<sup>16,17</sup>. A cohort follow up study of children treated for malignancy early in childhood may suggest SMN. Most literature have shown that Ewing's sarcoma/PNET are uncommon among black children<sup>17,11,12</sup> In this study, no case of Ewing's Sarcoma/PNET was recorded. In western countries, it is the second most common sarcoma of childhood<sup>13,17,39,40</sup>

The anatomical distribution of the sarcomas follow the adult pattern with tumours of the extremities being more predominant particularly when STS are combined with malignant bone tumours. The head and neck and retroperitoneal regions were also frequent anatomical sites seen in this study. Most botryoid ERMS occurred in the hollow organs of the head and GUS (eye, nasal cavity, oral cavities, vagina, cervix and urinary bladder). All RMS of the GUS except 1 case of testicular ARMS were ERMS mostly of the botryoid subtype with superior prognosis involving the vagina, cervix, the urinary bladder and paratesticular sites.

Most grading schemes for STS are based on adult data, however, the POG adopted NCI is favoured in paediatric NRSTS while the international classification of RMS emphasizes histologic prognostic and correlation therefore RMS is prognostically classified based on the histologic subtypes.

The study showed that all the NRSTS were of grade 1 POG/NCI grading which confers good prognosis as exemplified by the reported good prognosis of infantile fibrosarcoma and Kaposi Sarcoma. Only the 23 cases of ARMS were classified as poor prognostic tumours, 24 conventional RMS were of intermediate prognosis while the remaining 17 cases of ERMS were of botryoid and spindle cell variants and were classified as superior prognostic tumours. This prognostic grouping of RMS has implication in planning for treatment, allocation of resources and health planning.

Though the immunohistochemical antibodies used in this study (Vimentin and Desmin) were rather non specific for RMS compared to more specific markers like MYODI and Myogenin, this study has shown that Vimentin and Desmin if used carefully in childhood, can be useful adjuncts in confirming childhood RMS. Particularly, Desmin has been shown to be a very sensitive Myogenic marker and is positive even in an undifferentiated RMS<sup>41</sup>. This study has attested to this claim as all the RMS stained gave a minimum of ++ degree of immunoreactivity except the two cases that were negative and were recategorised as

lymphomas based on reactivity with LCA. Vimentin aside being a general mesenchymal marker is also recognized as a marker of tissue preservation, all our tumours had some degree of positivity for Vimentin suggesting good preservation.

Though Myogenin is specific for RMS, ERMS is reported to show less expression of Myogenin and may even lack Myogenin reactivity and can be supported with a more sensitive Desmin in a panel<sup>41</sup>.

MyoD1 & Myogenin are often negative in pleomorphic RMS<sup>41</sup>. Therefore, though Desmin is less specific, is a sensitive Myogenic marker, if used in combination with Vimentin, they can be reliable in confirmation of childhood RMS in resource poor settings.

## Conclusion

Childhood sarcomas are common in our environment accounting for about 2 of every 5 childhood malignancies. RMS is the single most common variety while the non-rhabdomyosarcoma soft tissue sarcomas (NRSTS) are rare. The vast majority of tumors affect the extremities

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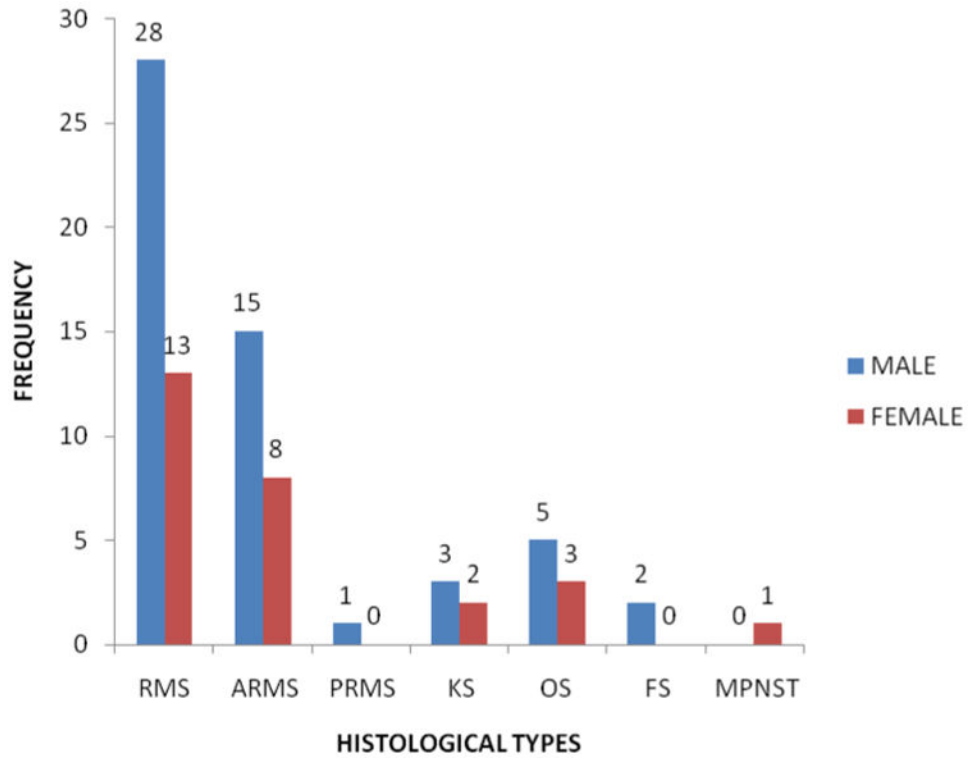
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## FREQUENCY AND SEX DISTRIBUTION OF CHILDHOOD SARCOMA



**Figures 1.**

**Key,**ERMS, Embryonal Rhabdomyosarcoma; **RMS**, Rhabdomyosarcoma; **ARMS**, Alveolar Rhabdomyosarcoma; **PRMS**, Pleomorphic Rhabdomyosarcoma; **OS**, Osteosarcoma; **FS**, Fibrosarcoma; **MPNST**, Malignant Peripheral Nerve Sheath Tumour;

**Table I**

**Relative Age Incidence of Childhood Sarcomas in Jos**

	ERMS	ARMS	PRMS	KS	OS	FS	MPNST	%
0-3	8 (19.5%)	-	-	-	-	-	-	8 (9.9%)
4-7	13 (31.7%)	4 (17.4%)	-	2 (40%)	-	-	-	19 (25.5%)
8-11	6 (14.6%)	6 (26.1%)	-	1 (20%)	2 (25%)	-	-	15 (18.5%)
12-15	14 (34.2%)	13 (56.5%)	1 (100%)	2 (40%)	6 (75%)	2 (100%)	1 (100%)	39 (48.1%)
<b>Total</b>	<b>41 (50.6%)</b>	<b>23 (28.4%)</b>	<b>1 (1.2%)</b>	<b>5 (6.2%)</b>	<b>8 (9.9%)</b>	<b>2 (2.5%)</b>	<b>1 (1.2%)</b>	<b>100</b>

**Key:**ERMS, Embryonal Rhabdomyosarcoma; ARMS, Alveolar Rhabdomyosarcoma; PRMS, Pleomorphic Rhabdomyosarcoma; OS, Osteosarcoma; FS, Fibrosarcoma; MPNST, Malignant Peripheral Nerve Sheath Tumour;

() = % of tumour in the age group

**Table II**  
**Distribution of Childhood Sarcoma According to Anatomical Regions**

	ARMS	ERMS	PRMS	KS	FS	MPNST	OS	Total	%
Extremities	5	10	1	3	2	-	8	29	36
Head & Neck	8	13	-	2	-	-	-	23	28
Retropertoneum	7	4	-	-	-	-	-	11	11
Trunk	2	7	-	-	-	1	-	10	12
GUS	1	7	-	-	-	-	-	8	10
<b>Total</b>	<b>23</b>	<b>42</b>	<b>1</b>	<b>5</b>	<b>2</b>	<b>1</b>	<b>8</b>	<b>81</b>	<b>100</b>

**Table III**  
**Immunohistochemical profile of childhood Sarcomas in Jos**

Age	Sex	Site	Previous H/E Diagnosis	LCA	Vimentin	Desmin	Final Diagnosis
1	F	Retroperitoneum	Alv RMS	-	++	++	RMS
2	M	Retroperitoneum	Alv RMS	-	++	+++	RMS
3	F	Neck	Emb RMS	-	++	+	RMS
4	M	Nose	Emb RMS	++	-	-	Lymphoma
5	F	Supraclavicular	Alv RMS	-	-	+++	RMS
6	M	Retroperitoneum	Alv RMS	-	++	+++	RMS
7	M	Back	Alv RMS	-	+	+++	RMS
8	F	Jaw	Alv RMS	+++	-	-	Lymphoma
9	F	Vaginum	Emb RMS	-	+	+++	RMS
10	M	Retroperitoneum	Alv RMS	-	+	++	RMS