

## Epidemiology of Soft Tissue Sarcomas in Jos, North Central Nigeria

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**Abstract****Background**

*Soft tissue sarcomas are a complex heterogeneous group of tumours that often cause a diagnostic problem. Special stains are often employed for further differentiation of these tumours. As a group, soft tissue sarcomas are not uncommon as previously believed.*

**Objective:** *To undertake a ten year retrospective study of soft tissue sarcomas in a Nigerian Teaching Hospital and to also discuss the pathology of the disease.*

**Methods:** *The study consisted of all records of histologically confirmed cases of soft tissue sarcomas from 1994–2003, seen at JUTH. The slides were retrieved and read separately by two pathologists. Special stain such as Reticulin, PTAH, Oilred O, were used to demonstrate basement membrane, muscle striation and lipids respectively. These aided confirmation of the diagnosis.*

*Information extracted from patients' records included age, site of lesion and sex.*

**Results:** *A total number of 266 cases of soft tissue sarcomas were recorded out of 2353 histologically confirmed cancers. These accounted for 11.3% of all cancers. One hundred and eighty three occurred in males and 83 in females, with male, female ratio of 2:1. Kaposi sarcoma (KS), Rhabdomyosarcoma (RMS), Malignant Fibrous Histiocytoma (MFH), and Fibrosarcoma (FS) were the commonest histological subtypes accounting for 69 (25.9%), 63 (23.7%), 56 (21.1%) and 49 (18.4%) respectively; with Rhabdomyosarcoma occurring more in paediatric and adolescent age groups, while the rest subtypes were more prevalent in adults. The commonest sites affected were leg/foot 71 (26.7%), head/neck 68 (25.6%) and thigh 50 (19%).*

**Conclusion:** *Soft tissue sarcomas constituted a significant proportion of cancers in our centre and are not uncommon as previously reported in the literature.*

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**INTRODUCTION**

Soft tissue tumours are traditionally defined as mesenchymal proliferations that occur in the extra-skeletal non-epithelial tissue. These diseases encompass more than 20 histological and biologically distinct subtypes<sup>1-3</sup>. It is reported to be rare in the developed nations of Europe and America accounting for 0.8-1% of all invasive cancers<sup>2,3</sup>. Survey of hospital based study in some African countries show higher proportion compared to what is seen in America and Europe.<sup>4-6</sup>

In most reported studies there is male preponderance. It is reported to be more common in Africans (Blacks) than Whites.<sup>7,8</sup>

The pathogenesis of soft tissue sarcoma is multifactorial and in most cases the cause is not certain. Documented risk factors include radiation therapy, history of trauma, chemical carcinogens and hereditary syndromes.<sup>1,3,4,6,8,9,10</sup>

Soft tissue sarcomas may arise from any part of the body, but they are more common in the subcutaneous tissue.

The purpose of this study is to describe the distribution of the various subtypes of soft tissue sarcomas in our environment and also determine their relative frequency in relation to other malignant tumours.

**MATERIALS AND METHODS**

All the records of soft tissue sarcomas seen in Histopathology Department were retrieved and fresh sections were cut and restained. The slides were examined independently by two pathologists.

Special stains such as reticulin, PTAH, Oilred O, were employed to demonstrate basement membrane, muscle striation and lipids where necessary to confirm diagnosis.

Confirmed cases were classified according to World Health Organization histological classification of soft tissue tumours.<sup>11</sup> Other information collected from patients records included age, sex and site of lesion.

**RESULTS**

There were a total of 2353 cancers registered within the

ten year study period giving an average annual rate of 236. A total number of 266 cases of soft tissue sarcomas were recorded out of the 2353 histologically confirmed cancers. These accounted for 11.3% of all cancers. One hundred and eighty three cases occurred in males and 83 in females, with male, female ratio of 2:1. KS, RMS, MFH and Fibrosarcoma were the commonest histological subtypes accounting for 69, 63, 56 and 49 cases respectively. RMS occurred more in the paediatric and adolescent age groups while the rest were seen more in the adults. The commonest sites affected were leg/foot (71), head/neck (68) and thigh (50), (Table I).

**TABLE I: Frequency, sex and site distribution of various types of soft tissue sarcomas**

Site	ASS		GCT		RMS		KS		FS		MFH		LMS		LS		TOTAL	
	m	f	M	F	m	f	m	f	m	f	M	F	M	F	M	F		
Head/neck	2	1	2	30	5	9	7	1	2	5	1					1	2	68
Trunk/back	2	1		3			3	15	2	11	5	-			3	2		47
Retroperitoneum	-		6	4	-	-	5	2	4	3	-	-	-	5	1			30
Thigh	-	1	-	-	3	3	2	10	5	13	7	4	2	-				50
Leg/foot					10	2	31	14	4	3	5	2	-	-	-	-	-	71
Total	4	2	1	2	49	14	43	26	35	14	38	18	4	2	9	5		266

**RMS- Rhabdomyosarcoma, KS-Kaposi Sarcoma**

**FS- Fibrosarcoma:** RMS- Rhabdomyosarcoma

KS - Kaposi sarcoma

FS- Fibrosarcoma

m- male; f- female

MFH-Malignant fibrous histiocytoma

LMS Leiomyosarcoma

LS - Liposarcoma

ASS-Alveolar soft part sarcoma

GCT- Granular cell tumour

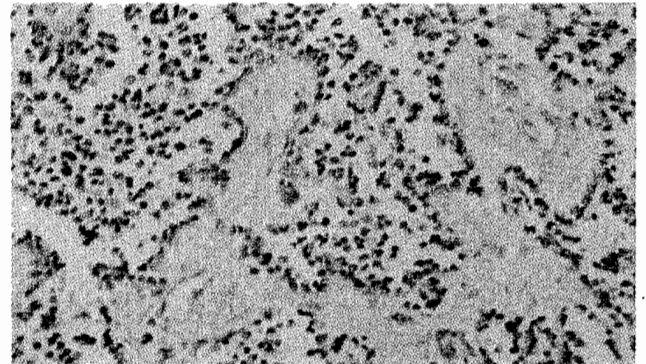
Majority (73%) occurred within ages 20 years to 60 years. Twenty five per cent of the cases were within ages zero to 19 years while less than one percent (0.06%) was seen within ages 60 years to 70 years, (Table II).

**TABLE II: Age Distribution of various soft tissue sarcomas**

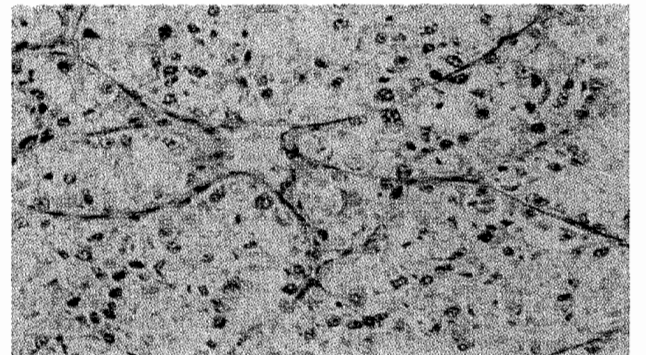
Age group in years	Histology								Total
	RMS	KS	FS	MFH	LMS	LS	ASS	GCT	
0-9	28	-	1	-	-	-	-	-	29
10-19	20	4	3	2	-	-	-	-	29
20-29	5	13	2	21	1	1	-	-	43
30-39	5	27	5	12	2	-	1	1	53
40-49	3	15	20	14	2	8	2	2	66
50-59	2	5	13	5	-	3	3	-	31
60-69	-	5	5	2	1	2	-	-	15
Total	63	69	49	56	6	14	6	3	266

**Keywords:** RMS- Rhabdomyosarcoma  
 KS - Kaposi sarcoma FS-Fibrosarcoma  
 MFH-Malignant fibrous histiocytoma  
 LMS Leiomyosarcoma  
 LS - Liposarcoma  
 ASS- Alveolar soft part sarcoma  
 GCT- Granular cell tumour

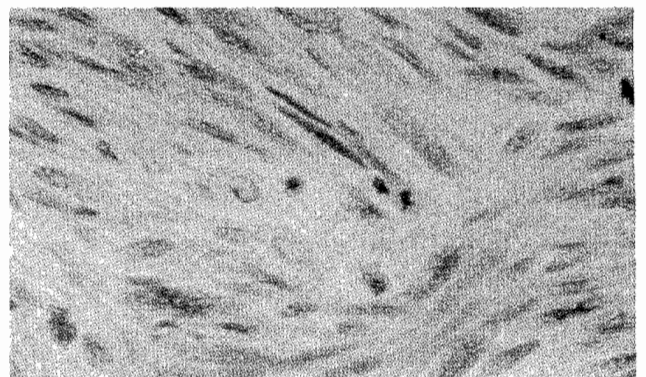
Figures 1, 2 and 3 show photomicrographs of alveolar rhabdomyosarcoma, alveolar soft part sarcoma, and fibrosarcoma respectively.



**Figure 1: Photomicrograph of Alveolar Rhabdomyosarcoma; H&E stain (x40 Objective)**



**Figure 2: Photomicrograph of Alveolar Soft Part Sarcoma; H&E stain (x40 Objective)**



**Figure 3: Photomicrograph of Fibrosarcoma; H&E stain (x40 Objective)**

## Discussion

Soft tissue sarcomas accounted for 11.3% of all cancers in our Hospital. This is higher than 0.8-1% recorded in Britain, and U S A.<sup>1,2</sup> Records from Europe showed that soft tissue sarcomas account for more than 1% of annual cancer rate.<sup>1,3,4</sup> Being a hospital based study and in the absence of national data on cancer, these figures might not be the true reflection of actual incidence of soft tissue sarcoma.

Most soft tissue sarcomas in our study occurred in the region of the lower limbs and head/neck. Several studies in Connecticut, U S A showed similar results.<sup>1,3,7</sup> This study also showed male preponderance in the ratio of 2:1. This same ratio of 2:1 was reported in Europe and America<sup>2</sup>.

Kaposi sarcoma and Rhabdomyosarcoma accounted for the highest histological subtypes. These two tumours have been reported to be four times more common in males than females.<sup>2,3</sup> In this study they were 4-5 times more common in males than females. In a review of tumours in Connecticut U S A, annual increase in soft tissue sarcoma was higher in males than females<sup>1</sup>.

The pathogenesis of soft tissue sarcoma is centred mainly on environmental and host factors. Radiation therapy is a well established cause of soft tissue sarcoma; several studies of patients with ionizing radiation have observed an excess risk of soft tissue sarcoma with tumour arising from within the field of radiation. Similar study conducted by Armed Forces Institute of Pathology showed that there is a latent period for post radiation which varies from 2-8 years with a median age of 8 years. Similar reports have been published by British scientists.<sup>6,8</sup>

Occupational exposure to agricultural chemicals has been identified as a risk factor for soft tissue sarcoma.<sup>12</sup> A six fold increase in risk of soft tissue sarcoma associated

with agricultural or forestry chemicals such as phenoxyacetic acid and chlorophenol have been reported by Swedish scientists.<sup>12,13</sup>

This occupational hazard might have contributed to the high incidence of soft tissue sarcoma in our environment where most of the people use these chemicals for pest and weeds control in their subsistence farming which is the major means of livelihood.

Kaposi sarcoma is exceptionally high among persons infected by HIV.

The diagnosis of Kaposi sarcoma is more frequent in homosexual and heterosexual than those with intravenous drug abuse. Several candidate viruses have been evaluated but current suspicion is directed at herpes virus 8. The risk is also increased in transplant Recipient<sup>5,6,14,15</sup>.

It has been suggested that trauma or chronic inflammatory lesions may increase the risk of soft tissue sarcoma although it is possible that local injury simply calls attention to a pre-existing tumour or perhaps accelerated its growth.

Soft tissue sarcomas had been reported at the site of surgical scars, burns scar or chronic skin ulcers.<sup>5,6,9</sup> Previous study of skin cancer in this centre showed that soft tissue sarcoma arose from skin ulcers.<sup>14</sup> Other risk factors for the pathogenesis of soft tissue sarcoma include genetic and immunological factors. Li-Fraumeni syndrome is a well documented hereditary defect associated with Soft Tissues Sarcoma and wide spread cancers. Most families have shown germ line mutation of tumour suppressor gene (P53) which is located on the chromosome 17p13.<sup>10,16</sup>

## Conclusion

This study shows that soft tissue sarcomas are not uncommon as was previously thought, and that they are more frequent in males than females.

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