



Delayed Presentation of Bone and Soft Tissue Sarcoma of the Extremity in Makurdi: Do Traditional Bone Setters Play a Role?

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Authors' contributions

This work was carried out in collaboration between all authors. Author WTY designed the study, performed the statistical analysis, wrote the protocol and the first draft of the manuscript. Authors ICE, DDM, JNK and CTS managed the analyses of the study. Authors CNA, BAO and JAN managed the literature searches. All the authors read and approved the final manuscript.

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ABSTRACT

Background: Soft tissue and bone sarcomas are rare but are associated with high mortality rate when they occur. This is partly due to delay in presentation to a specialist center. Some guidelines have been introduced to aid early diagnosis of these tumours, however, these guidelines are not always observed as required. This study is aimed at highlighting the emerging role traditional bone setters may be playing in delaying presentation of patients with sarcomas of the extremity.

Methods: A 5 year retrospective review of histologically confirmed soft tissue and bone sarcoma patients was conducted at our hospital. Data obtained included socio-demographic characteristics, date symptoms were first noticed, date of first consultation with a specialist, date of first specific

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treatment and history of traditional intervention. Tumour size, location on the limb, fungation/ulceration at presentation as well as type of treatment were noted. Descriptive statistics was done to display frequency and measures of central tendency.

Results: There were 16 patients between ages 3 and 65 years. Fifteen had soft tissue sarcoma while one was an osteosarcoma. There were 10 males and 6 females giving a male to female ratio of 1.6:1. Mean Patient Delay was 56.4 weeks and Mean tumour size was 25.3 cm. There were 12(75%) rhabdomyosarcomas and 2(12.5%) Kaposi sarcomas. Seven of the tumours were located in the thigh (43.7%), 2(12.5%) each in the gluteal region, knee and leg. Out of the 6 that ulcerated, 5(83.3%) had traditional bone setters intervention with incisions made. Seven had Wide local excision and 2 had amputation after biopsy. Four patients had adjuvant chemotherapy with one year survival in one.

Conclusion: Delay in presentation of sarcoma is noticed more commonly among patients with small size tumours and those who have had traditional bone setters treatment.

Keywords: Sarcoma; delay; extremity; Nigeria.

1. INTRODUCTION

Current guidelines state that any patient with suspected cancer should be seen by a specialist within 2 weeks of referral [1]. However, a study conducted by Broun recommended 4 weeks for sarcomas [2]. A detailed history, examination and investigation including ultrasound, radiographs, CT scan or MRI and biopsy should be done to make diagnosis and determine extent of disease. Surgery is important in the treatment of most sarcomas [3]. Limb sparing surgery, as opposed to amputation, can now be used to save the limbs of patients in at least 90% of extremity tumor cases [4]. Additional treatments, including chemotherapy and radiation therapy, may be administered before and/or after surgery [5]. Chemotherapy significantly improves the prognosis for many sarcoma patients, especially those with bone sarcomas [5]. When there is delay, amputation becomes inevitable and outcomes are worse.

Some published articles have described delays in referral of patients with soft tissue sarcomas for treatment [6]. They found that general medical professionals were the source of the greatest delay. However, general practitioners and hospital consultants were not considered separately and both studies identified a time period which the authors thought constituted a delay only going on to analyse those patients with delay [6,7].

In our centre, we have observed an emerging trend where traditional bone setters(TBS) and some unorthodox healers are becoming an important cause of delay in presentation of patients with sarcoma of the extremities. These bone setters lack knowledge of guidelines and

international best practices. There are few studies that highlight the attempt by traditional bone setters to extend their activities to the treatment of malignancies [8]. The complications that result from the treatment of fractures by traditional bone setters are well known [9-13]. Based on this, a hypothesis that states that traditional bone setters contribute to patient delay in patients with extremity sarcoma was formulated.

This study was conducted to identify factors contributing to delay in presentation of sarcomas in our specialist centre.

2. MATERIALS AND METHODS

2.1 Setting

The study was conducted at Benue State University Teaching Hospital (BSUTH) Makurdi, Benue State, Nigeria. The state is located in the north central region of Nigeria. It has a population of 4.2 million. Benue State University Teaching Hospital is a relatively young State-owned Teaching Hospital that came into full operation in April 2012.

2.2 Study Population

The data was obtained from the records of patients who presented with extremity sarcoma in the hospital between April 2012 and March 2017. The data obtained included socio-demographic data, date symptoms were first noticed, date of first consultation with a specialist and date of first specific treatment (biopsy, chemotherapy or surgery). Evidence of traditional intervention and complications noticed were also documented. The size of the tumour at

presentation, location on the limb, presence or absence of pain, swelling, fever, scarification or ulceration were all documented as well as the type of treatment (surgery, chemotherapy and radiotherapy).

2.2.1 Delay

Absence of consultation by a specialist for more than four weeks in a patient with a diagnosis of sarcoma [2].

2.2.2 Patient delay

The time interval from first occurrence of symptoms of the tumour to the first medical consultation.

2.2.3 Professional delay

Time from first medical consultation to first specific treatment.

2.2.4 Symptom interval

Total period between presenting first symptoms and the first specific treatment.

A traditional bone setter is an unorthodox individual who treats fractures by manipulation, application of concoctions and tight splintage using wood and tree barks. Sometimes, they also make incantations and incisions under aseptic conditions when treating tumours.

Rural areas are sparse settlements located away from towns and cities. They usually have inadequate health and other infrastructural development.

Urban areas are dense settlements like towns and cities and usually have adequate health and other infrastructural developments.

Ethical clearance for study was obtained from the hospital research ethics committee (HREC).

2.3 Inclusion Criteria

All patients with a histological diagnosis of soft tissue or bone sarcoma of the extremity treated in the hospital.

2.4 Exclusion Criteria

Patients with histological diagnosis of other tumours were excluded.

2.5 Data Analysis

Descriptive statistics was used to display frequencies and measures of central tendency.

3. RESULTS

There were a total of 16 patients between the ages of 3 and 65 years presenting with extremity sarcomas during the period under review. Fifteen were soft tissue sarcomas while one was an osteosarcoma. There were 10 males and 6 females giving a male to female ratio of 1.6:1 (Table 1). The median age was 26.0 years and mean age was 28.6±18.8 years. Patient Delay ranged from 4 to 96 weeks an average of 56.4 weeks ±4.4 weeks (Table 2). Professional Delay ranged from 0.3 weeks to 6 weeks. Ten of them (62.5%) were from rural areas while 6(37.5%) were in Urban areas. There were 5(31.2%) children and 3(18.8%) farmers. The tumour sizes ranged from 4 to 72 centimeters in their greatest dimension with an average of 25.3± 2.8 cm. There were 12 (75%) rhabdomyosarcomas (Fig. 1), 2 (12.5%) Kaposi sarcomas, 1 (6.3%) fibrosarcoma and 1(6.3%) osteosarcoma (Table 3) Three patients were exposed to herbicides and 2 were HIV positive while 11 did not have a documented exposure to carcinogens. There was no family history of tumours documented. Seven of the tumours were located in the thigh (43.7%), 2(12.5%) each in the gluteal region, knee and leg while there was 1(6.2%) tumour each on the foot, elbow and clavicular region. Two had metastasis to chest and one to the skull before presentation. Six had incisional biopsy, 7 had Wide local excision, 2 had amputation after biopsy and 1 had trucut biopsy. Four patients had adjuvant chemotherapy using drug combinations like cyclophosphamide, Vincristine, Doxorubicin and Darcabazine for soft tissue sarcomas and Doxorubicin and Cisplatin for osteosarcoma. Three declined chemotherapy because of cost, 8 requested for referral to other centres for chemotherapy on account proximity. One patient died from complications arising from metastasis to the chest before chemotherapy could commence. They presented with a painful swelling in 15(93.7%), ulceration 6(37.5%), fever 3(18.8%), weight loss 11(68.8%), recurrence 5(31.3%) (Table 4). Out of the 6 that had ulceration, 5(83.3%) had traditional bone setters intervention during which incisions were made and tight splints were applied.

Table 1. Showing Socio demographic characteristics of patients presenting with sarcomas of the extremity in Makurdi

	Frequency	Percentage
Sex		
Male	10	62.5%
Female	6	37.5%
Age		
≤10	3	18.3%
11-20	5	31.2%
21-30	2	12.5%
31-40	2	12.5%
41-50	2	12.5%
51-60	1	6.2%
61-70	1	6.2%
Residence		
Rural	10	62.5%
Urban	6	37.5%
Profession		
Child	5	31.3%
Farmer	3	18.8%
Business	2	12.5%
Student	2	12.5%
Teachers	1	6.3%
Driver	1	6.3%
Civil servant	1	6.3%
Tumour size(cm)		
1-20	10	62.6%
21-40	4	25.0%
41-60	1	6.2%
61-70	1	6.2%



Fig. 1. Showing a 20 year old man with rhabdomyosarcoma of the left thigh

4. DISCUSSION

Our study suggests that soft tissue sarcomas of the extremity are more common among young males. This is a pattern observed in many other studies in Nigeria [14,15,4]. The commonest type of soft tissue sarcoma was rhabdomyosarcoma followed by Kaposi Sarcoma

[14,15], however one study in Jos had Kaposi Sarcoma as the leading type followed by Soft tissue sarcoma [16]. This may be due to the high prevalence of immune suppressive conditions like retroviral disease in our region and exposure to herbicides without proper personal protective equipment among our farmers. Metaanalytic studies show a weak association between soft tissue sarcoma and exposure to herbicides [17]. There was delay in presentation of all our patients with the mean patient delay of 54 weeks. This is much higher than the recommended 2 weeks in UK guidelines, 4 weeks recommended by Broun 2 [6] or 12 weeks recommended by a study in Uganda [18]. Other studies have also recorded patient delays of 56 weeks and 40 weeks [19,20]. The reasons for the delay were attributed to general practitioners in some patients and multi factorial in others. Soft tissue sarcomas are rare. As a result of this, patients and physicians often assume more frequent diseases like tendinitis, sciatic pain or osteoarthritis to be the cause of these symptoms [21]. These reasons are similar to the ones we observed in our study. However, the emergence

Table 2. Showing the association between socio demographic factors of patients with extremity sarcomas and mean delay in presentation in weeks

	Patient Delay(wks)	Professional Delay(wks)	Symptom Interval(wks)
Sex			
Male	45.6	2.7	48.3
Female	57.0	2.2	59.2
Age			
0-29	36.5	2.6	39.1
30-59	61.3	2.0	63.3
60-89	17.5	3.5	21.0
Residence			
Rural	59.2	2.7	61.9
Urban	34.6	2.2	36.8
TBS			
Yes	60.3	9.2	69.5
No	46.3	15.0	61.3
Profession			
Child	38.4	2.2	40.6
Farmer	78.0	4.0	82.0
Business	72.0	2.5	74.5
Student	26.0	3.1	29.1
Teacher	48	1	49
Driver	48	3	51
Civil servant	96	2	98
Tumor size(cm)			
1-20	62.8	3.4	66.2
21-40	50	1.1	51.1
41-60	32	1	33
61-80	36	1	37
Histology			
Rhabdomyosarcoma	46	2.8	48.8
Fibrosarcoma	96	1	97
Osteosarcoma	32	1	33
Kaposisarcoma	60	2	62

_ TBS = Traditional Bone Setters

Cm = Centimeters

Patient Delay +professional delay=Symptom interval

Table 3. Showing the histological pattern of sarcomas presenting in Makurdi

Type	Frequency	Percentage
Rhabdomyosarcoma	14	87.5
Kaposi Saracom	2	12.5
Fibrosarcoma	1	6.3
osteosarcoma	1	6.3
	16	100

of traditional bone setters in cancer treatment in our region has constituted a new and worrisome source of delay. Patients who went for traditional treatment before presentation to our unit were observed to have a longer patient delay(60.3 weeks) compared to those who did not (46.3 weeks). Some of our patients were kept for several months where this unorthodox treatment

was carried out but to no avail until they developed distant metastasis. Furthermore, most of those presenting with ulceration/fungation weight loss and fever had prior TBS intervention. During the intervention, incisions are made aseptically and tight splints are applied which may predispose to infection .The presence of AIDS-related complications of Kaposi sarcoma

Table 4. Showing Complications of Traditional Bone Setters Intervention on Presentation of Sarcomas in Makurdi

Effect on presentation	Traditional bone setters intervention		Total
	Yes	No	
Ulceration			
Yes	5	1	6(37.5)
No	6	4	10(62.5)
Weight loss			
Yes	8	3	11(68.8)
No	3	2	5(31.3)
fever			
Yes	3	-	3(18.8)
No	8	5	13(81.3)

could also explain the the fever and weight loss seen in some of the patients.

Similarly the average size of the sarcomas was 25.3 cm. This is much higher than the recommended 5 cm by UK guidelines [1] and 9.9 cm recorded in a study by Smith [22]. The larger tumours tended to present earlier while there was more delay by patients with smaller size tumours. This earlier presentation of large tumours may be due to social embarrassment associated with a huge swelling on a conspicuous part of the body Most (62.5%) of the patients were from rural areas and had the longest mean patient delay (59.2 weeks). This may be due to ignorance and poor access to health care. It has also been shown that rural practice physicians experience stronger barriers to referral [23]. Furthermore, the physicians themselves may be lacking in knowledge gained at continuous medical education and cancer conferences enjoyed by their urban counterparts [24]. This may adversely affect their management of sarcomas since it is not a diagnosis they encounter frequently.

Most of the tumours (43.7%) occurred in the thigh. This may have contributed to patient delay because the muscle bulk in the thigh can mask a swelling for a long time until it becomes large and obvious. A similar study done in Jos found the leg and foot to be the commonest site of occurrence [16]. However, in that study soft tissue sarcomas of the whole body were considered.

Females had a relatively longer patient delay in our study compared to males. Other studies have recorded similar findings [25-28]. This has been explained by the hypothesis that women more often cited competing priorities of work and family over their own health [22] Others have opined that this difference might be due to the

different way in which men and women recognise abnormalities, attribute body changes to illness and assess the seriousness of their condition [25-28]. United Nations Development Programme has reported higher gender inequality index among low human development countries and in 2015, it reported a labour force participation rate of 48.4 for females compared to 64.0 for males in Nigeria [29]. This relative economic disadvantage female folks have in the developing world may also be responsible for delay in seeking medical attention from experts because of the cost implication.

Pupils were found to have the shortest patient delay. This is probably due to the fact that children are not likely to be able to endure the discomfort and pain of sarcoma for long without crying for attention. This will likely make their parents and care givers to seek expert treatment earlier. Also due to the small body size of children, even a relatively small tumour in an adult body will appear large in a child. The longest patient delay of 78 weeks and 96 weeks were observed in farmers and a civil servant respectively. This may be related to the lack of access to health care by farmers since most of them also live in rural areas. The current economic recession in Nigeria has adversely affected the economic fortunes of civil servants and hence may have affected their health seeking behavior as well. A similar work done in Germany showed that employees had the shortest patient delay of 8.7 weeks while unemployed patients had the longest delay of 12.7 weeks [30].

Delay in presentation have been associated with worse clinical course and lower survival rates [31]. Delay of greater than three months has been found to have a detrimental effect on treatment options and outcomes in one fifth of patients [32]. Studies have shown a correlation

between the duration of symptoms prior to treatment and disease relapse, distant metastases, survival as well as chemoresponse [32-34].

Findings in our study agree with other Nigerian and Caucasian studies however, our sample size is rather small for inferential statistics. Further prospective studies will need to be conducted to identify risk factors associated with development of sarcomas in our environment as well as the impact of delay on patient outcomes.

5. CONCLUSION

In conclusion, there was delay in presentation of most of the sarcoma patients according current guidelines. Patient delays were more among rural dwellers, farmers, patients with smaller tumour size and those who had traditional bone setters intervention.

CONSENT

All authors declare that informed written consent was obtained from the patient for publication of this paper and accompanying images.

ETHICAL APPROVAL

As per international standard or university standard, written approval of Ethics committee has been collected and preserved by the authors.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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