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Extremity Soft Tissue Sarcoma: A Review of 19 Cases.

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Summary

Background: Although soft tissue sarcoma is a rare tumour, it accounts for a significant proportion of malignancies seen in many orthopaedic practices. The objectives of this study are to evaluate the pattern of presentation of extremity soft tissue sarcoma and the treatment outcome in our patients.

Method: This is a 3 year prospective study in patients presenting with extremity soft tissue sarcoma at the oncology unit of the National Orthopedic Hospital Lagos. Data, such as age, sex, presenting complaints, anatomic location of lesions, histologic type of tumour, and treatment outcome were retrieved from prepared proforma. The average follow up period was 22.8 months.

Results: Nineteen patients were studied. The age range of the patients was 3 to 69 years, male to female ratio of 1.7: 1. Most tumours were located in the thigh and the leg, and malignant fibrous histiocytoma was the most common histologic diagnosis. Whilst all the patients presented with stages II and III diseases, limb salvage was only possible in 52.6% of these patients. Surgery was always combined with adjuvant radiotherapy. A survival rate of 69% and a recurrence rate of 16% were recorded over the period of the study.

Conclusion: Delayed presentation limited the scope of limb salvage surgery in centers such as ours, but appropriate tumour excision and adjuvant radiotherapy remains the mainstay of the management.

Introduction

Soft tissue sarcomas are the non epithelial extraskeletal tissue malignancies, exclusive of the reticuloendothelial system, glial and supporting tissues of various messenchymal organs.[1,2] Although rare tumors, incidence of 1 in 32771 population[1,3], there is an increased frequency in this center because it is an evolving oncology center with referral from institutions all over the west African sub-region..

The objective of this study is to evaluate the pattern of presentation and the management of soft tissue sarcomas of the extremities in this center from the year 2002 to 2005.

Methodology

A 3 year prospective study was conducted at the oncology unit of the National Orthopedic Hospital, Igbobi, Lagos. Data were retrieved from prepared proforma filled in the course of treatment and follow up of patients.

All patients were seen and examined on presentation by the leading author. Investigations such as plain radiographs of the chest and the extremity, as well as fine needle aspiration for cytology were performed. Computerized tomography scans were ordered in all cases but only 7 patients complied. In the same vein, only one patient could afford a magnetic resonance imaging.

Histologic diagnosis was obtained post operatively in all patients. Patients were given adjuvant radiotherapy at an affiliated radiotherapy unit in the same locality.

Information including age, gender, mode of presentation, anatomic location, histologic types, treatment and outcome of management were obtained, and data was analysed using the SPSS [version 11].

Results

A total of 19 patients were treated over this period. The age range of the patients was between 3 and 69 years [table 1], with a slight male preponderance (M: F 1.7:1).

While the locations of the tumours vary widely in the musculoskeletal system, most were located in the thigh (36%) and the leg (20%). The forearm, arm, foot and the shoulder region were the other common anatomic locations (table 2). Malignant fibrous histiocytoma was the most common histologic type (26.3%), other tumour types include liposarcoma, rhabdomyosarcoma, nerve sheath tumours, synovial sarcoma and Kaposi sarcoma (table 4).

There was no patient with stage I disease as most of the tumours were of the high grade, and were extracompartmental at the time of presentation (table3). Therefore, limb sparing procedure (wide local excision) was possible in 52.6% of the patients, while ablation was undertaken in 36.8% of cases. The ablative procedures consists of transmetatarsal amputation in one patient, shoulder disarticulation in 2 patients, above knee amputation in 2 patients and above elbow amputation in 2 patients as well. Surgery was followed by adjuvant radiotherapy (6300 -6400 cGy) in all the patients, and 4 patients had adjuvant doxorubicin based chemotherapy. Three patients presenting with advanced disease were offered only palliative radiotherapy. At an average follow up period of 22.8 months, 52.6% of the patients were well, while 15.8% had recurrence of the lesion.

Table 1. Age Distribution (years)

Age	No
0-4	1
5-9	1
10-14	1
15-19	1
20-24	3
25-29	2
30-34	4
35-39	1
40-44	1
45-49	0
50-54	1
55-59	1
60-64	0
65-69	2
Total	**19**

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Table 2. Anatomic Location of tumours

Site	No	%
Thigh	7	36%
Leg	4	20%
Foot/ ankle	2	11%
Shoulder	2	11%
Upper arm	2	11%
Fore arm	2	11%
Total	**19**	**100%**

Table 3. Surgical Stages of soft tissue sarcoma

	A	B
Stage I	0	0
Stage II	6 (32%)	5(26%)
Stage III	2(11%)	6(32%)

Table 4. Histologic Types of soft tissue sarcoma

Malignant fibrous histiocytoma	5 (26%)
Liposarcoma	3 (16%)
Fibrosarcoma	3 (16%)
Rhabdomyosarcoma	2 (11%)
Nerve sheath tumours	2 (11%)
Undifferentiated	2 (11%)
Kaposi sarcoma	1 (5%)
Synovial sarcoma	1 (5%)

Table 5. Outcome of Treatment of soft tissue sarcoma

Recurrence	3 (16%)
Well	10 (53%)
Deceased	2 (11%)
Indeterminate*	4 (21%)

* Patients lost to follow up within the first six months.

Discussion

Soft tissue sarcoma though rare, accounts for 50% of extremity malignancies and 26.7% of all the musculoskeletal tumors seen in this center. There is a slight male preponderance from this study, and this has been documented by workers in the south-eastern Nigeria and worldwide[3,4,5]. Various reasons have been adduced for the increase incidence in the male gender, the most prominent being the increase exposure to environmental hazards from chemical and related industries[3]. Many of these industries have existing regulations against the employment of female technicians and engineers in factories and oil mining installations.

The most common histologic type in this study was malignant fibrous histiocytoma. This is widely believed to be the commonest extremity soft tissue sarcoma in the adult [1,6]. Since a large proportion of our patients were adults, this study further confirms that this histologic type is the most common extremity soft tissue sarcoma in the African adult patients. The thigh is the commonest anatomic location which is in agreement with previous studies on this disease condition.[1,3,4.] The shear volume of the thigh in comparison to the other parts of the extremity, and the abundance of non epithelial extraskeletal tissues in this region of the musculoskeletal system could be responsible for the increase incidence at this location.

Most of the patients in this center presented at the late stages of the disease[7], limiting the scope of limb salvage procedures. This pattern of presentation is common in the developing countries such as ours where there is widespread poverty and limited access to specialized medical care. Therefore, ablative procedures were the surgical options in a significant proportion of our patients. The indications for ablation include, encasement of major extremity nerve tissue, multiple skip lesions preventing enbloc resection, and major infection complicating biopsy. The proportion of limb salvage procedures performed is lower compared to reports from other centers where limb salvage was possible in almost 90% of patients with soft tissue sarcoma [2,8,9,10]. These centers were mostly located in more advanced societies where there are structured referral systems, and patients were seen at the early stage of illness. In some centers, procedures such as in situ preparation of the neurovascular tissue using vinyl sheet barriers, and subsequent pasteurization, alcohol soaking, or distilled water soaking, have further improved limb salvage.[9]

The follow up care in these patients also posed serious management problems, and four patients [21%] were lost to follow up within the first 6 months post surgery. The oncologic outcome in this group of patients is not certain after the recorded period of initial outpatient care. This hindered correct estimation of 2 and 5 year disease specific survival rates. However, previous studies have documented 77% survival at 24 months, which is still higher than the rate of 69% [at 22.8 months mean follow up period] in our patients. Late presentation of patients, financial constraints and lack of advanced facilities for procedures such as 'in situ preparation'[9] could be responsible for this lower survival rates.

We conclude that appropriate tumor excision and adjuvant therapy still remains the mainstay of treatment of extremity soft tissue sarcoma. However, early presentation of patients and early intervention in the course of the disease will no doubt increase the functional and oncologic outcome in these patients.

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