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SARCOMAS OF SOFT TISSUES AND BONES IN ADEN, YEMEN

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ABSTRACT

Background: Very little epidemiological data regarding soft tissues and bone sarcomas from Yemen in general and Aden region in particular are available. **Objectives:** The aim of this study was to compile epidemiological data regarding age, gender and site distribution, and to correlate our findings with published western data in order to determine whether there were any significant differences in our population compared to the west. **Methods:** This cross sectional study was carried out at National Oncology Center, Aden, Yemen from 2009 to 2013. **Results:** Out of 107 newly diagnosed patients 54 belonged to bone sarcoma and 53 to soft tissue sarcoma group. The majority of STS in our study occurred in the second and third decades of life; they were common in females. The majority of bone sarcomas occurred in the first and second decades of life; they were common in males. The most common sites of origin was lower extremities in 55.1%, retroperitoneal 11.2%, trunk 10.3% and upper extremities 9.3%. **Conclusion:** The treatment of sarcoma is a multidisciplinary approach, and patients have benefited from multimodality treatment. In Aden region, Yemen, delayed and advanced stages of the disease are the rule. We recommend to establish good interdisciplinary relationships among the managing physicians and educate our patients on early presentation to the hospital.

KEYWORDS: Bone, soft tissue, Sarcoma, epidemiology, Yemen.

INTRODUCTION

Sarcomas, tumors of putative mesenchymal origin originate primarily from elements of the mesodermal embryonic layer. Soft tissue sarcomas often form at anywhere in the body and are classified according to the adult tissue that they resemble. Similarly, bone sarcomas are

usually classified according to the type of matrix production: osteoid producing sarcomas are classified as osteosarcomas and chondroid-producing sarcomas are classified as chondrosarcomas.

Soft tissue sarcomas make up 1% of all cancer types and have been estimated to occur approximately 3 cases among every 100,000 people. Liposarcoma, malignant fibrous histiocytoma (MFH), and leiomyosarcoma are the most common soft-tissue sarcomas. These tumors are anatomic site-dependent; in the extremities the common subtypes are MFH and liposarcoma, whereas liposarcomas and leiomyosarcoma are the common subtypes in the retroperitoneum and the abdominal cavity.

Bone and soft tissue sarcoma are uncommon group of cancer and the data is insufficient from Yemen. Bone sarcomas comprise only 0.2% of all malignant neoplasms.[1] and are one tenth as common as soft tissue sarcomas.[2] Soft tissue sarcomas can occur in the body's muscles, joints, fat, nerves, deep skin tissues, and blood vessels, but most originate in an extremity (59%), the trunk (19%).[3] The most common are malignant fibrous histiocytoma (28%), leomyosarcoma (12%), liposarcoma (10%) and malignant peripheral nerve sheath tumors (6%).[4]

In this study, the National Oncology Center in Aden shows that this tumors are in increasing. This study was conducted to show the distribution and pattern of sarcomas in Aden region and to correlate it with published western data in order to determine whether there were any significant differences in our population compared to the west.

MATERIAL AND METHODS

This study took place at National Oncology Center, Aden, Yemen. It serves the entire people of Aden region (Aden, Lahj, Abyan and Dala) an estimated population of 2.0 million people. We retrospectively reviewed 107 patients that were managed for bone and STS for a period of 5 years (January 2009 to December 2013). Materials for the study were obtained from the case notes as well as the histopathology reports. Patients variables analyzed included the age, sex, histology types, site of tumor and where available treatment given and outcome. Data collected was analyzed with a computer using SPSS® to generate frequency tables.

RESULTS

Over a 5 year period, 107 cases of bone and STS were reported. There were 59 male (55.1%) and 48 female (44.9%) patients with male: female is 1.2: 1. There mean age was 37.8 years ± 17 years (range 15-78 years).

Table 1: Histopathological distribution of bone and STS according to sex.

Subtype of Sarcoma	Female	Male	TOTAL	%
Osteosarcoma	13	12	25	23.4%
Ewing's Sarcoma & PENT	5	19	24	22.4%
Chondrosarcoma	2	3	5	4.7%
Liposarcoma	5	7	12	11.2%
Malignant Fibrous Histocytoma	6	5	11	10.3%
Synovial Sarcoma	3	4	7	6.5%
Leiomyosarcoma	6	0	6	5.6%
Rhabdomyosarcoma	2	4	6	5.6%
Neurogenic Sarcoma	4	0	4	3.7%
Kaposis sarcoma	0	3	3	2.8%
Undifferentiated STS	2	2	4	3.7%
TOTAL	48 44.9	59 55.1	107 100.0	100%
P 0.01				

Among the diagnosed patients bone sarcomas, 25 patients (23.3%) had osteosarcoma, 24 patients (22.4%) had Ewing's sarcoma and 5 patients (4.7%) had chondrosarcoma. The soft tissue sarcomas were found to be more frequent in this analysis; the liposarcoma was in 12 patients (11.2%) followed by malignant fibrous histiocytoma in 11 patients (10.3%) and synovial sarcoma in 7 patients (6.5%). The leiomyosarcoma and rhabdomyosarcoma were found in 6 patients each (5.6%).

Table 2: Age group distribution according to sex.

AGE	Female	%	Male	%	Total	%
10-20	13	27.1	16	27.1	29	27.1
21-30	8	16.7	14	23.7	22	20.6
31-40	11	22.9	10	16.9	21	19.6
41-50	4	8.3	9	15.3	13	12.1
51-60	5	10.4	3	5.1	8	7.5
61-70	3	6.3	6	10.2	9	8.4
71-80	4	8.3	1	1.7	5	4.6
Total	48	100	59	100	107	100

The large majority of sarcomas (>67%) occurred in the age groups below 40 years. (Table 2) 59 cases occurred in males, while 48 cases occurred in females. The frequency dropped after age 40 years.

Table 3. Site distribution of sarcomas.

Site	Frequency	%
Head & Neck	7	6.5
Lower extremity	59	55.1
Retroperitoneal	12	11.2
Trunk	11	10.3
Upper extremity.	10	9.3
Visceral	8	7.5
Total	107	100.0%

In table 3, lower extremity was the commonest in all sarcomas (55.1%) followed by the retroperitoneal and the trunk (11.2% and 10.3%), these three sites accounts for 76.6%. The involvement of upper extremity and visceral organs are less common.

Table 4: Stages of newly diagnosed Sarcoma Patients.

Stage	Frequency	%	
I	12	11.2%	
II	21	19.6%	
III	24	22.4%	
IV	50	46.7%	
Total	107	100.0%	

In table 4 about 69.1 % of sarcomas presented in advanced stages (stage III and IV) and the delay in presentation correlate directly with bad outcome.

Table 5: Outcome of sarcoma patients according to tumor burden.

Tumor size	Alive	Died	Loss of follow up	Total	P value
< 5 cm	14	0	3	17	0.01
5-8 cm	26	16	7	49	
>8 cm	8	29	4	41	
TOTAL	48 44.9	45 42.1	14 13.1	107 100.0	

In table 5 there is direct proportion between mortality and increase of tumor size > 8 cm and decrease number of death in patients with tumor size less than 5 cm.

Table 6: Outcome of Sarcoma Patients.

Outcome	Frequency	%	P value
Alive with recurrence	23	21.5	0.09
Alive without recurrence	25	23.4	
Died	45	42.1	
Loss of follow up	14	13.1	
Total	107	100	

Table 7: Outcome of sarcoma patients according to subtypes.

Subtype of Sarcoma	Alive	Died	Loss of follow up	TOTAL
Osteosarcoma	11	10	4	25
Ewing's Sarcoma & PENT	13	9	2	24
Chondrosarcoma	1	4	0	5
Liposarcoma	2	8	2	12
Malignant Fibrous Histocytoma	6	4	1	11
Synovial Sarcoma	3	3	1	7
Leiomyosarcoma	4	1	1	6
Rhabdomyosarcoma	3	2	1	6
Neurogenic Sarcoma	1	2	1	4
Kaposis sarcoma	2	0	1	3
Without differentiation	2	2	0	4
TOTAL	48 44.9	45 42.1	14 13.1	107 100.0
P value 0.59				

DISCUSSION

Sarcomas represent the heterogeneous group of cancer with diverse tumor biology. Chemotherapy, being the main stay of treatment for certain sub-types of bone sarcoma e.g. Ewing's sarcoma, has proved to improve the recurrence free survival in adjuvant setting in Osteogenic sarcoma.[5] but has a controversial role in soft tissue sarcomas. Wide adequate surgical resection with pathologically proven clear margins is the most effective therapeutic approach for management of soft tissue sarcoma.

The most common bone sarcomas in our study are osteosarcoma, Ewing's sarcoma and chondrosarcoma, Although soft-tissue sarcomas can arise anywhere in the body, the lower extremities are the most common site. Incidence is as follows: lower extremities (55.1 %); retroperitoneum (11.2%); trunk (10.3%) and upper extremities (9.3%).

In general The majority of sarcomas occurred in the first three decades of life and dropped after the 40 years of age. Western studies show that the sarcomas are common at the age above 50 years and the peak incidence is in the fifth to seventh decades of life.[6] Osteosarcoma in our study are equal in male and female. In western data the osteosarcoma is more common in male than female in a ratio of 3:2 especially in young patients.[7]

Ewing's sarcoma is an aggressive tumor that is most commonly seen under age 20 similar to studies reported by Saeter et al (2007) and Jurgens and Dirksen (2011).[8-9] The large majority of Ewing's sarcomas, occurred in the first three decades of life, especially in the second and third decades. Incidence above age 30 was extremely rare. Our study show that this tumor is the second commonest bone sarcoma. Peak incidence is in the second decade of life. In other similar study the Ewing's sarcoma is extremely uncommon above the age of 30 years.[6, 10-12]

Many factors such as delayed presentation, advanced stages, age, gender, tumor size, and localization have been investigated as prognostic factors, but the results are conflicting.[13-15] The prognostic factor accepted by all authors is the presence of metastatic disease.[16-17] In our study, there is some correlation between gender, tumor size, disease progression and overall outcome similar to studies reported by Jurgens et al 1988 and Hayes et al 1989.[18-19] Up to 46.7% of patients have clinically metastatic disease at diagnosis and their outcome remain poor, despite treatment with intensive chemotherapy protocols. The presence of metastasis at diagnosis considered one of the most bad prognostic factors for sarcoma.

Baldini et al., (1999) observed in his study, the non-metastatic patients younger than 30 years have better prognosis and overall survival than patients age 30 years and older. This difference has not been seen in metastatic patients. some studies concluded that patients older than 30 years have a worse survival rate than those younger than 30.[20] Similarly, in Grier et al.'s study, the survival rate worsened with advanced age.[14]

In this study, the bulky tumors had a significant bad prognostic outcome. Tumors with large tumoral masses reflect advanced stages and higher possibility of organ, bone and nerve invasion with delay of disease regression.[21-22]

The disease recurrence was significantly observed in our study in comparison to the reported literature but with significant lost to follow rate. Further large scale and prospective studies are needed to have a more comprehensive understanding of the behavior and outcome of this heterogeneous disease in our population.

The management of sarcomas in Yemen is still a serious topic because of the late presentation of our patients and also because of nonavailability of some modern chemotherapy agents, target therapy, radiotherapy and appropriately manage these patients. However, within the limit of the available resources, we need to establish a good interdisciplinary relationship among managing physicians and to educate our patients on early presentation to the hospital.

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