

SHORT COMMUNICATION

Prevalence of Malignant Soft Tissue Tumors in Extremities: An Epidemiological Study in Syria

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Abstract

Background: Although the majority of soft tissue masses are benign, it is important to consider malignancy in differential diagnoses. Because most soft tissue sarcomas present as a painless mass, clinicians must watch for signs suggestive of malignancy, including large size, rapid growth, and site deep into the deep fascia. The purpose of this study was to determine the relative prevalence according to sex and age, site of tumor, skeletal distribution, and treatment (surgery, chemotherapy and radiotherapy) before and after surgery, and ascertain the relative frequency of these tumors in specific anatomic sites and age groups based on pathological studies.

Methods: A total of 308 patients, with a musculoskeletal tumor were evaluated retrospectively. All of the patients enrolled into this study were referred to the Beirouni Hospital of Damascus University with a proven diagnosis of malignant soft tissue tumors from the beginning of January 2008 until the end of 2010. The prevalence of the malignant soft tissue tumors in these patients was analyzed. For purposes of analysis, all lesions were placed in 1 of 9 categories: hand and wrist, forearm, humerus (arm), proximal limb girdle (axilla and shoulder), foot and ankle, thigh, hip and buttocks region, trunk, and other lesions. Age and sex also were recorded.

Results: Malignant tumors consisted of seven diagnostic categories: malignant fibrous histiocytoma (23%), liposarcoma (22%), rhabdomyosarcoma (9%), leiomyosarcoma (8%), malignant schwannoma (5%), dermatofibrosarcoma protuberans (5%), synovial sarcoma (10%), fibrosarcoma (13%), extraskeletal chondrosarcoma (1%), and extraskeletal Ewing sarcoma (4%).

Conclusions: Despite the multitude of pathologic possibilities, most malignant soft-tissue tumors are classified into a small number of diagnoses. These may be further defined when the site of the lesion and the age of the patient are considered. Knowledge of tumor prevalence will assist radiologists in establishing a suitably ordered differential diagnosis when a soft-tissue tumor has a nonspecific radiologic appearance.

Key words: Oncology, Sarcoma, Syria

Introduction

Malignant soft tissue tumors, or soft tissue sarcomas, are more common than bone sarcomas. About 8,000 new tumor cases occur each year in the United States, which represents only about 1% of all malignant tumors (1). They usually occur in adults and present as a large painless or painful soft tissue mass. Because benign soft tissue "lumps" are far more common than malignant ones, the diagnosis of a soft tissue sarcoma may be made after excision of a presumed benign mass at a local hospital or in the doctor's office. Soft tissue tumors are also classified by their tissue

type and grade. Thus, the higher the grade, the greater the chance for aggressive growth at the original site, and this is true for metastases, at distant sites.

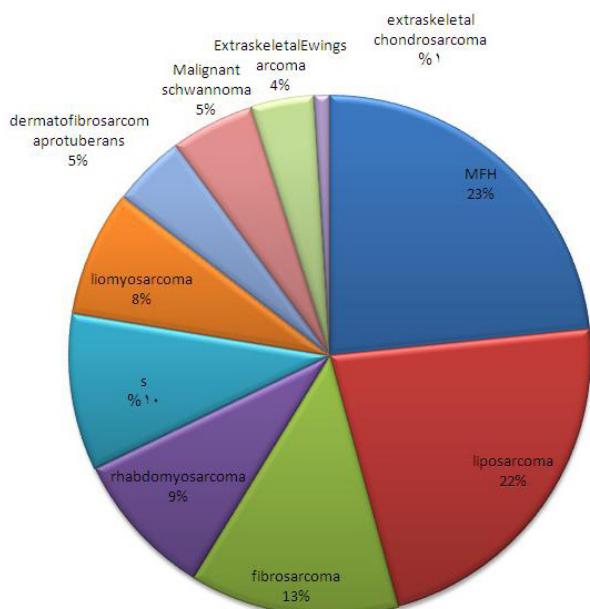
Staging a soft tissue tumor involves evaluating the histology (tissue grade) of the tumor, how extensive the tumor is at the primary site, and whether or not there are distant metastases. All of these characteristics help describe the tumor's behavior (biologic activity), the patient's prognosis, and the best treatment option.

MRI scanning is the best imaging study for soft tissue tumors because of its sensitivity in showing differences between normal and sarcoma tissue. It also clearly

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**Figure 1. Distribution of cases based on type of tumors.**

shows the relation of the tumor to nearby nerves and blood vessels. Moreover, a chest CT scan is always necessary to rule out lung metastases and a biopsy is done to determine diagnosis.

Soft tissue sarcomas that develop in the limbs and the axial area, even those that are diagnosed early using appropriate methods and are treated adequately, have a 5-year survival rate of between 62 and 84% (2, 3).

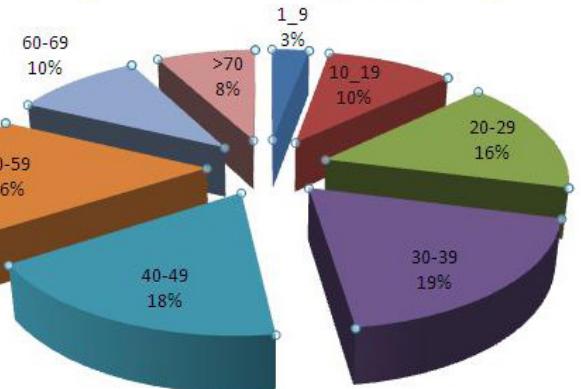
Materials and Methods

Of 378 patients with a diagnosis of malignant soft tissue sarcoma of the limb treated at Beirouni Hospital between 2008 to the end of 2010, we studied 308 cases based on our inclusion criteria and 70 cases were excluded.

Malignant soft tissue sarcoma was diagnosed based on entries in final pathology reports. Mesenchymal lesions that originated in soft tissue were included in the study.

The characteristics of the 308 cases of malignant soft

Sample distribution / age groups

**Figure 2. Frequency of cases with soft tissue sarcomas based on age.**

tissue sarcoma were analyzed based on tumor types determined at final diagnosis, the anatomical locations of tumors in limbs, for purposes of analyzing all lesions were placed in 1 of 9 categories: hand and wrist, forearm, humerous, proximal limb girdle (axilla and shoulder), foot and ankle, hip and buttocks region, thigh, trunk, and other lesions.

Malignant tumors were classified into seven diagnostic categories: malignant fibrous histiocytoma, liposarcoma, rhabdomyosarcoma, leiomyosarcoma, malignant schwannoma, dermatofibrosarcoma protuberans, synovial sarcoma, fibrosarcoma, extraskeletal chondrosarcoma and others.

Then the patients based on tumor type were grouped into the following age groups: 1-9 years, 10-19 years, 20-29 years, 30-39 years, 40-49 years, 50-59 years, 60-69 years and above 70 years old. In addition, patient's sex and place of living were recorded. Beirouni Hospital of Damascus University is the largest referral center for malignancies in Syria and the study was approved by research committee of our institution.

The treatment method of patients in the study was grouped into the following groups: neoadjuvant chemotherapy, adjuvant chemotherapy, neoadjuvant radiotherapy, adjuvant radiotherapy, wide resection surgery, radical surgery, amputations and no treatment.

Results

In this group of 308 cases, 143 (45%) were males and 175 (55%) were females, with a male:female ratio of 0.8:1. The distribution of cases was as follows: malignant fibrous histiocytoma (23%), liposarcoma (22%), rhabdomyosarcoma (9%), leiomyosarcoma (8%), malignant schwannoma (5%), dermatofibrosarcoma protuberans (5%), synovial sarcoma (10%), fibrosarcoma (13%), extraskeletal chondrosarcoma (1%), and extraskeletal Ewing sarcoma (4%) (Figure 1).

The frequency of cases with soft tissue sarcomas based on age was as follows: 1-9 years: 9 (3%) cases; 10-19 years: 31 (10%) cases; 20-29 years: 49 (16%) cases; 30-39 years: 58 (19%) cases; 40-49 years: 55 (18%); 50-59

Table 1. Frequency of malignant fibrous histiocytoma based on age

Percent	Frequency	MFH
0	0	9-9
7	5	19-10
18	13	29-20
10	7	39-30
18	13	49-40
18	13	59-50
18	13	69-60
11	8	<70
100	72	

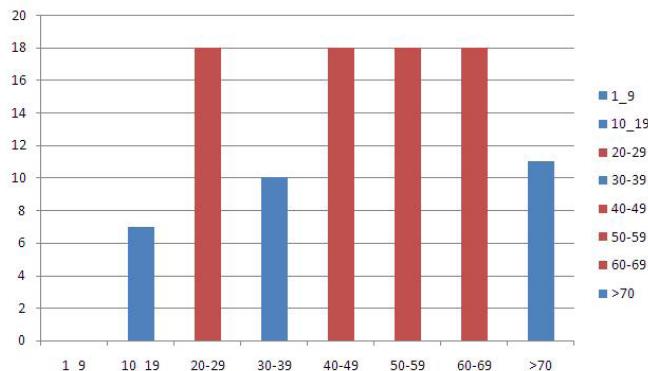


Figure 3. Frequency of malignant fibrous histiocytoma based on age.

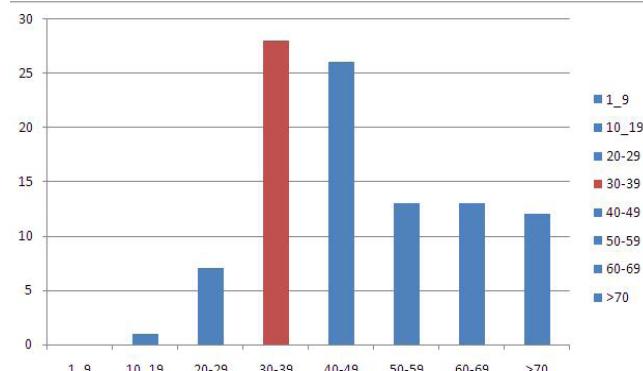


Figure 4. Frequency of fibrosarcoma based on age.

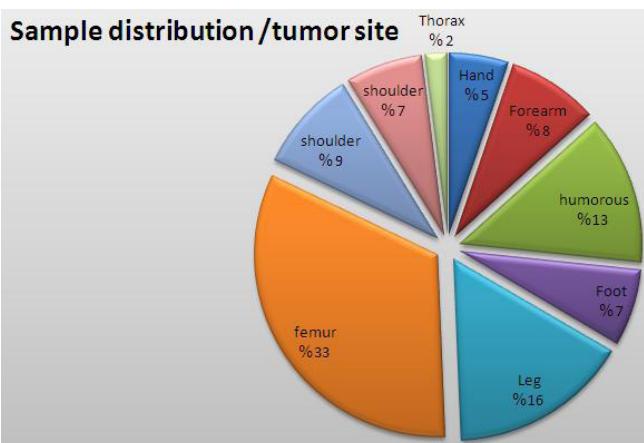


Figure 5. Distribution of tumors based on every part of the body.

years: 50 (16%) cases; 60-69 years: 31cases (10%) and more than 70 years: 24 cases (8%) (Figure 2).

Malignant fibrous histiocytoma was the most frequent tumors and frequently occurred in patients > 40 years old (Table 1 and Figure 3), while 46% of fibrosarcoma cases involved adults between 40 and 60 years of age (Figure 4).

Of the 308 cases of soft tissue sarcomas, the tumor was observed involving every part of the body as follows: hand and wrist: 16 cases (5%); forearm: 25 cases (8%); humerus: 41 cases (13%); proximal limb girdle (axilla and shoulder): 28 cases (9%); foot and ankle: 21 cases (7%); leg: 49 cases (16%); hip and buttocks region: 21 cases (7%); femur:101 cases (33%); and the trunk: 6 cases (2%) (Figure 5).

Wide resection was performed in most cases, which is the preferred treatment in most cancer referral centers to salvage the limb. Radical resection was performed in a few cases that were mostly malignant fibrous histiocytoma (MFH). Most studies have focused on survival after surgical treatment and chemoradiotherapy, which is the most important and perhaps only indicator for evaluation of treatment modalities. Surgery was performed in 92% of cases and wide resection was used for 73% of patients (Figure 6).

Fourteen percent of patients undergoing neoadjuvant

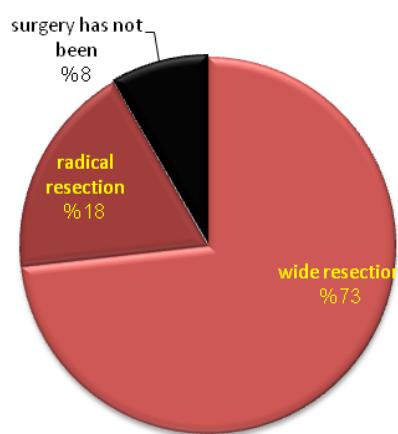


Figure 6. Distribution of cases based on surgical techniques ratio / %.

chemotherapy and adjuvant chemotherapy were performed in 55% of patients. Radiotherapy was performed in 11% of cases before surgery and 64% after surgery (Figure 7).

Discussion

In this study, we calculated population-based incidence rates of soft tissue. This report is the largest study of soft tissue tumors in the Syrian population to date. We used the recent 2002 WHO criteria to define histological subtypes of soft tissue sarcoma and included all cases, regardless of site of origin except bones and joints. This approach provided a valuable opportunity to determine accurate and clinically applicable incidence rates by histological type and subtypes.

By comparing the result of different studies, the most common soft tissue tumor was MFH. In 2002, WHO declassified MFH as a formal diagnostic entity and renamed it as an undifferentiated pleomorphic sarcoma as the most common followed by liposarcoma. Other similar results have been obtained in other studies (4). Also, Gustafson al. reported that MFH and grade IV were the most common soft tissue tumor (5). Similarly the most common soft tissue tumor was MFH in our patients (Figure 1).

However, in another study conducted by Toro et al, the

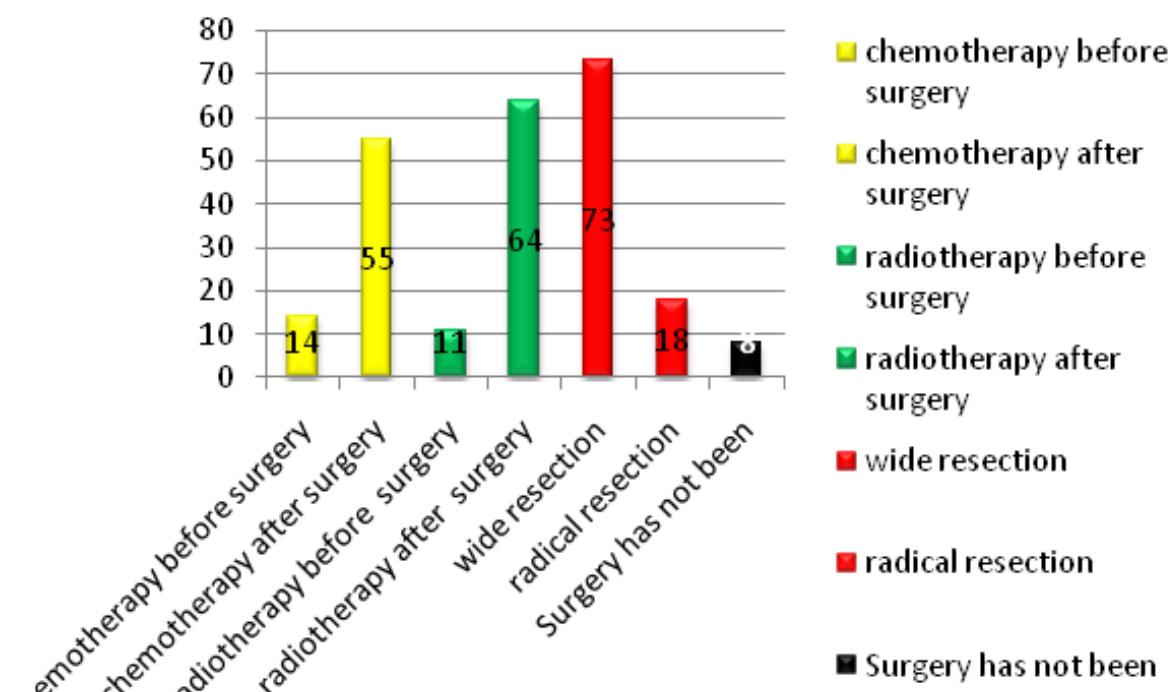


Figure 7. Distribution of cases based on how to treatment.

most common soft tissue tumor was leiomyosarcoma (23.9%) (6).

In our series, female prevalence (55%) was slightly higher, which is consistent with most previous studies, although some previous studies reported higher incidence in males (1,4,5,7-11). Thus with this inconsistency in the results of different studies, no clear trend in gender predilection can be stated.

Age-specific rates for all soft tissue tumors were elevated by increased age (especially between 30-59 years). The peak incidence was about 30-39 years, with marginal decreases in 40-49 years. Gustafson et al. reported that the median age was 64 years (5). Wibmer et al. found that the highest peak was around the age of 60 years (7). The most common anatomic site of involvement was the thigh (33%), followed by the leg and arm.

The main surgical procedure in most cases was wide local excision (margins of at least 1 cm).

Radical excisions were applied to the small number of

patients and the majority of those cases were MFH. For 8% of the patients surgery was not performed due to the patient's refusal of surgery or poor medical conditions.

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