Prevalence of Head and Neck Sarcomas in the Main Health Centers in Yazd from 1994 to 2014

Original Article

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Received: Jun 23, 2015
Accepted: Aug 17, 2015

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Abstract

Introduction:
Head and neck sarcomas involve a group of rare malignant diseases with a high histological variability involving various anatomical sites that can lead to under-reporting of the true incidence of these neoplasms. This study aimed to epidemiologically investigate the occurrence of sarcomas of the head and neck within the past 20 years in Yazd, Iran (1994–2014).

Materials and methods:
In this descriptive, cross-sectional and retrospective study, 16114 patient’s records with malignant tumors were examined via the census method, which were available in the archives of 8 main treatment centers in Yazd, Iran within a 20-year period. Age, sex, occupation, habitat, type of sarcoma, tumor location and grade, metastasis, recurrence, and history of head and neck irradiation were recorded. Data were analyzed in SPSS software version 17.

Results:
Among 586 cases of sarcomas, 59 cases (10.06%) were identified with head and neck sarcomas. The mean age of the patients was 32.22 ± 8.31 years, of which 26 (44.01%) patients were males and 33 (55.9%) were females. Soft tissue sarcomas were noted in 41 cases (69.5%); rhabdomyosarcoma was the most common (27.1%). Eighteen (30.5%) patients had hard tissue sarcomas; osteosarcoma (15.3%) was the most common. Soft tissues of the head and neck were the most (49.20%) involved sites. Most sarcomas were low grade. In 5 patients (8.5%), metastases occurred to the head and neck, and the tumor relapsed in 16 patients (27.1%).

Conclusion:
The findings of the current study were in agreement with those of other reports referred to in different studies. This suggests that the epidemiology of head and neck sarcomas in Yazd, Iran is similar to other geographical regions.

Key words:
•Head and Neck Neoplasms •Sarcoma •Yazd
Introduction

Sarcomas involve a rare and heterogeneous variety of malignant tumors of mesenchymal origin with a specific and distinct histopathology\(^1\). The mesenchymal cells can develop into tumors affecting the soft tissues of muscle, fat, and fibrous tissue. Bone and nerves can also be involved.\(^2\) Occasionally, these tumors are associated with trauma, genetic syndromes as well as exposure to previous radiation, though there is mostly no apparent cause. Pathological classification is most valuable in the treatment and prognosis of head and neck sarcomas.\(^3\)

The incidence of sarcoma is more prevalent in children than adults. Approximately 1% of all adult cancers\(^2-4\) and 10%–20% of pediatric cancers are sarcomas\(^2\). About 5%–15%\(^2,3\) of adult sarcomas are in the head and neck region; 35%\(^3,5\) of children are diagnosed with head and neck sarcomas.\(^3\)

Common soft tissue sarcomas in order of frequency include liposarcoma, malignant fibrous histiocytoma, fibrosarcoma, rhabdomyosarcoma, leiomyosarcoma, synovial sarcoma, malignant peripheral nerve sheath tumors, angio-sarcoma and kaposi sarcoma, whereas hard tissue sarcomas in order of frequency entail osteosarcoma, chondrosarcoma, as well as Ewing’s sarcoma\(^12\).

The results of some studies conducted in Iran demonstrated that 60% of sarcomas occurred in males with a mean age of 36 years, that the most common sarcomas in adults were malignant fibrous histiocytoma and synovial sarcoma and osteosarcoma, whereas hard tissue sarcomas in order of frequency entail osteosarcoma, chondrosarcoma, as well as Ewing’s sarcoma\(^12\).

Out of 16,114 cases examined in this study, 586 cases (3.65%) of patients were diagnosed with sarcomas, among which 59 patients (10.06%) suffered from sarcomas of the head and neck, the study cohort. The mean age of the patients was 32.22 ± 8.31 years with an age range of 1.5–83 years, of which 17 patients (28.8%) were <16 years of age, whereas 42 patients (71.2%) were > 16 years. A total of 26 patients (44.05%) were males and 33 (55.9%) were females. More than half of patients (59.3%) lived in Yazd (Table 1). As demonstrated in Table 2, 41 cases (69.5%) of

Materials and Methods

In this descriptive, cross-sectional, and retrospective study, 16114 patients records (case notes), diagnosed as malignant tumors were examined from the archive of the Shahid Sadooghi Dental School and several other hospitals (Shahid Sadoughi, Shahid Rahnemoon, Mojibiyian, Mortaz, ShohadayeKargar, Seyedosohada and SavanehSookhtegi) over a 20-year period (1994–2014). The records were obtained by proposing a research study from the Shahid Sadoughi medical university of Yazd (Ethical code: p.17.1.77710; date: 1393.4.22).

It should be noted that case records that were incomplete or cases in which the patients reported a written dissatisfaction were excluded from the study. In order to glean the study data, a checklist was devised consisting of the following variables: case record number, pathology department identification number, age, sex, occupation, place of residence, type of sarcoma, tumor location, tumor grade, the occurrence of metastasis, recurrence, history of head and neck irradiation. The patients’ medical records as well as their pathology reports available in the mentioned health centers were collected and analyzed utilizing the SPSS software (Ver. 17) through descriptive statistics.

Results

Out of 16,114 cases examined in this study, 586 cases (3.65%) of patients were diagnosed with sarcomas, among which 59 patients (10.06%) suffered from sarcomas of the head and neck, the study cohort. The mean age of the patients was 32.22 ± 8.31 years with an age range of 1.5–83 years, of which 17 patients (28.8%) were <16 years of age, whereas 42 patients (71.2%) were > 16 years. A total of 26 patients (44.05%) were males and 33 (55.9%) were females. More than half of patients (59.3%) lived in Yazd (Table 1). As demonstrated in Table 2, 41 cases (69.5%) of
The most prevalent type of soft tissue sarcomas was rhabdomyosarcoma (27.1%). Among the bone tissue sarcomas, affecting 18 patients (30.5%), osteosarcoma (15.3%) and chondrosarcoma (11.9%) were the most prevalent (Table 2). Rhabdomyosarcoma was reported to be the most common sarcoma (13.5 %) in the both the age groups, as well as in men (10.2%) and women (16.9%). The lowest incidence was found to be Kaposi sarcoma, which was only observed in one man >16 years old (1.7 %) (Table 2).

Table 1. Distribution of the head and neck sarcomas according to the demographic characteristics

<table>
<thead>
<tr>
<th>Variable</th>
<th>Number</th>
<th>Percent</th>
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<tr>
<td>Age</td>
<td></td>
<td></td>
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<tr>
<td>≤16</td>
<td>17</td>
<td>28.8</td>
</tr>
<tr>
<td>&gt;16</td>
<td>42</td>
<td>71.2</td>
</tr>
<tr>
<td>Sex</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td>26</td>
<td>44.05</td>
</tr>
<tr>
<td>Female</td>
<td>33</td>
<td>55.9</td>
</tr>
<tr>
<td>Habitat</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Yazd</td>
<td>35</td>
<td>59.3</td>
</tr>
<tr>
<td>Out of Yazd</td>
<td>24</td>
<td>40.7</td>
</tr>
</tbody>
</table>

Table 2: Distribution of the head and neck sarcomas according to demographic variables and frequency

<table>
<thead>
<tr>
<th>Type of sarcoma</th>
<th>Name of sarcoma</th>
<th>N</th>
<th>%</th>
<th>Total</th>
<th></th>
<th>Sex</th>
<th>Age</th>
</tr>
</thead>
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<tr>
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<td></td>
<td></td>
<td></td>
<td></td>
<td>Female</td>
<td>Male</td>
</tr>
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<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>n</td>
<td>n</td>
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<td></td>
<td>16&lt;</td>
<td>16≥</td>
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<td>n</td>
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<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Soft tissue sarcomas</td>
<td>Rhabdomyosarcoma</td>
<td>16</td>
<td>27.1</td>
<td>16.9</td>
<td>10</td>
<td>10.2</td>
<td>6</td>
</tr>
<tr>
<td></td>
<td>Neurofibrosarcoma</td>
<td>5</td>
<td>8.5</td>
<td>6.8</td>
<td>4</td>
<td>1.7</td>
<td>1</td>
</tr>
<tr>
<td></td>
<td>Fibrosarcoma</td>
<td>5</td>
<td>8.5</td>
<td>6.8</td>
<td>4</td>
<td>1.7</td>
<td>1</td>
</tr>
<tr>
<td></td>
<td>Liposarcoma</td>
<td>3</td>
<td>5.1</td>
<td>1.7</td>
<td>1</td>
<td>3.4</td>
<td>2</td>
</tr>
<tr>
<td></td>
<td>Malignant fibrous</td>
<td>3</td>
<td>5.1</td>
<td>1.7</td>
<td>1</td>
<td>3.4</td>
<td>2</td>
</tr>
<tr>
<td></td>
<td>histiocytoma</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
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</tr>
<tr>
<td></td>
<td>Synovial Sarcoma</td>
<td>3</td>
<td>5.1</td>
<td>1.7</td>
<td>1</td>
<td>3.4</td>
<td>2</td>
</tr>
<tr>
<td></td>
<td>Angiosarcoma</td>
<td>3</td>
<td>5.1</td>
<td>3.4</td>
<td>2</td>
<td>1.7</td>
<td>1</td>
</tr>
<tr>
<td></td>
<td>Leimyosarcoma</td>
<td>2</td>
<td>3.4</td>
<td>0</td>
<td>0</td>
<td>3.4</td>
<td>2</td>
</tr>
<tr>
<td></td>
<td>Kaposi's Sarcoma</td>
<td>1</td>
<td>1.7</td>
<td>0</td>
<td>0</td>
<td>1.7</td>
<td>1</td>
</tr>
<tr>
<td>Hard tissue sarcomas</td>
<td>Osteosarcoma</td>
<td>9</td>
<td>15.3</td>
<td>10.2</td>
<td>6</td>
<td>5.1</td>
<td>3</td>
</tr>
<tr>
<td></td>
<td>Chondrosarcoma</td>
<td>7</td>
<td>11.9 (30.5%)</td>
<td>6.8</td>
<td>4</td>
<td>5.1</td>
<td>3</td>
</tr>
<tr>
<td></td>
<td>Ewing’s sarcoma</td>
<td>2</td>
<td>3.4</td>
<td>0</td>
<td>0</td>
<td>3.4</td>
<td>2</td>
</tr>
</tbody>
</table>
Discussion

Sarcomas are rare and involve 1% of human cancers, among which currently 4%-10% occur in the head and neck.\textsuperscript{(14)} In the present study, 59 cases (10.06%) of 586 sarcomas patients were observed to suffer from the head and neck sarcomas.

In this study, 12 types of sarcoma histopathology were observed, which in turn demonstrates the wide histological variety of these tumors. Pacheco et al.\textsuperscript{(16)} in a study on 36 patients with the head and neck sarcomas, reported 12 types of sarcomas. Lajer et al. observed 15 histopathological types of sarcoma in a study consisting of 36 patients, which are in line with the findings of the present study.

Most of the head and neck sarcomas belong to the soft tissue category, and only 20% are bone sarcomas. In a study on 36 patients with the head and neck sarcomas, 41 (69.5%) soft tissue sarcomas, and 18 cases (30.5%) were hard tissue sarcomas. In a similar study in the Canadian population, Aljabab et al. observed 80% of sarcomas in the hard tissue and 20% of sarcomas in the soft tissue.\textsuperscript{(18)}

In the current study, the most prevalent type of sarcoma, rhabdomyosarcoma, involved 27.1% of the total tumors. Multiple reports\textsuperscript{(19-23)} have noted that approximately half of rhabdomyosarcomas commonly occur in the head and neck areas. This is in concurrence with the findings of the current study. However, the incidence of head and neck rhabdomyosarcoma can vary. Few studies mention the incidence to be in the range of 8% to 16\%\textsuperscript{(24-26)}, or higher(33.3\%)\textsuperscript{(11)}.

This could be because of geographical and racial differences as well as the different sample size of the different studies. Various studies mention that no geographical preference can be taken into consideration for the occurrence of soft tissue sarcomas\textsuperscript{(36, 37)}. The majority of the patients in our study\textsuperscript{(59,35)} lived in Yazd, and the rest were from other cities traveled to Yazd for treatment. However, this might affect follow-up of patients in the study centers.

In this study, more than half of the involved patients with the head and neck sarcomas (56\%) were females, which is in line with the findings by Bree et al.\textsuperscript{(27)}, although in several other studies\textsuperscript{(23, 28-31)} men have been reported to be affected more than the women. Some of these studies have indicated the predilection of males for the disease is nearly twice as much than females\textsuperscript{(29, 31)}. Due to the rarity of head and neck sarcomas, and limited sample size of most studies conducted in Iran and the world, drawing conclusions and comparisons is not very effective, yet; perhaps one of the reasons for the higher prevalence of females in this study was that women were referred to health centers more than men.

As was expected, sarcomas of the head and neck can occur in any age. In fact, studies have demonstrated that soft tissue sarcomas affects 80%-90% of adults and 10%-20% of children.\textsuperscript{(18, 32)} The mean age of the patients involved in this study was 32.22 ± 8.31 years which supports the findings of the studies conducted by Pacheco et al.\textsuperscript{(16)} and Dudhat et al.\textsuperscript{(30)} who noted a similar average mean age of 39.7 ± 25.1. However, Epstein and Gorsky\textsuperscript{(33)} reported the mean age of 40.4 years for this disease. Moreover, Mendenhall et al.\textsuperscript{(34)} reviewed the literature published between 1972 and 2000, and indicated the mean age of 50-55 years for the head and neck sarcomas, though findings of some studies propose that generally sarcomas involving the head and neck affect younger people including children and teenagers compared to squamous cell carcinoma.\textsuperscript{(35)}

In the present study, the anatomical distribution of most of the head and neck sarcomas included the jaw bone as well as the soft tissue of head and neck. Kraus et al.\textsuperscript{(32)} also indicated these two areas as the most prevalent involved locations with the head and neck sarcomas. Tajudeen et al.\textsuperscript{(9)} found the nasal cavity and sinuses as the most commonly involved locations in 22\% of his patients. Penel et al.\textsuperscript{(30)} reported a 39.3\% involvement in the neck tissue that is consistent with the findings of the current study.

In the present study, 5 patients (8.5\%) were found to have metastasis from other parts of the body to the head and neck. Breast is the most common site of tumor metastasis to the bone of the jaw, while the lungs involves the most frequent source for metastasis to the soft tissue of the mouth and teeth. In 30\% of cases, metastasis in the mouth has been found to demonstrate the first sign of an undiscovered cancer in another part of the body.\textsuperscript{(58)} Compared to other head and neck types of sarcoma in a study consisting of 36 pa...
neck neoplasms (e.g., squamous cell carcinoma), soft tissue sarcomas have a lower rate of regional metastasis.\(^{(39)}\) In this study, only 2 cases (3.4%) had metastasis to other parts of the body, whereas in a study conducted by Tajudeen et al.\(^{(9)}\) metastasis to lymph nodes was 6.5% and neural invasion was observed in 6.5% of the cases. Salcedo-Hernández et al.\(^{(5)}\) reported 50% of soft tissue sarcomas of the head and neck involving metastasis. Singh et al.\(^{(31)}\) reported local recurrence in 42% of patients and 42% of metastatic disease development in the lungs. Probably in this study, one reason for the low figures of head and neck sarcoma metastasis to other parts of the body is the lack of follow-up of individual patients specifically from other provinces, who included a large portion of the study sample. Recurrences of the sarcomas were observed in 27.1% of patients in this study. Because the patients were not actively followed up in order to evaluate recurrence, in reality, the recurrence rate in this study might have been more than what is being reported here. It has been demonstrated that local recurrence in head and neck sarcomas is more than that of other organs\(^{(28, 40-43)}\), which is probably due to the fact that reaching negative margins of tumor is more complex during sarcoma surgery of the head and neck.\(^{(44)}\)

Regardless of the location and size of the tumor, one of the main factors for prognosis with sarcomas is the tumor histologic grade.\(^{(45, 46)}\)

In this study, the majority (40.7%) of sarcomas were low-grade tumors, while 30.5% and 28.8% belonged to average and high grades respectively. However, Tajudeen et al.\(^{(9)}\) reported 35% of their cases as high-grade sarcomas.

**Conclusion**

The head and neck sarcomas are rare tumors that demonstrate a high variability in histology. In the current study, soft tissue sarcomas were generally much more prevalent than hard tissue sarcomas, among which rhabdomyosarcoma was the most common soft tissue sarcoma and osteosarcoma was the most common hard tissue sarcoma. Moreover, the age and gender prevalence, as well as the involved anatomic location in the studied population was similar to those of most other studies. However, our results, for the first time, provide an insight into the prevalence of head and neck sarcomas in Yazd, Iran.

The weak points of this study were the incomplete medical and pathological evidence of the patient and the patients were referred to other treatment centers and IHC (Immunohistochemistry) results were unavailable in some cases.

**Acknowledgement**

The authors thank the Vice-Chancellor of the Research Department of Shiraz University of Medical Sciences for supporting this research project. This article is based on the thesis submitted by Bahareh Yaghoobi.

**References**

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