Epidemiological Evaluation of Head and Neck Sarcomas in Iran (the Study of 105 Cases Over 13 Years)

Batoul Alishahi,¹ Neda Kargahi,² and Solmaz Homayouni¹*

¹Department of Oral and Maxillofacial Pathology, Dental Research Center, School of Dentistry, Isfahan University of Medical Sciences, Isfahan, IR Iran
²Department of Oral and Maxillofacial Pathology, Dental Implant Research Center, School of Dentistry, Isfahan University of Medical Sciences, Isfahan, IR Iran
*Corresponding author: Solmaz Homayouni, Department of Oral and Maxillofacial Pathology, Dental Research Center, School of Dentistry, Isfahan University of Medical Sciences, Isfahan, IR Iran. Fax: +98-3136687080, E-mail: solmazhomayoni@yahoo.com

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Background: Head and neck sarcomas are exceedingly rare and they include 4% - 10% of all sarcomas and less than 1% of all neoplasm of head and neck.

Objectives: The aim of this study is to evaluate the epidemiological characteristics of head and neck sarcomas of patients in Isfahan, Iran.

Patients and Methods: In this retrospective study, from the 16000 patients whose files were evaluated, the total number of 105 head and neck sarcomas were collected. They were evaluated with due attention to age, gender of the patients and the most common location of the lesion.

Results: From the total number of 105 (0.6%) patients with sarcomas, 56 were men (53.33%) and 49 women (46.66%). The most common head and neck sarcomas among this population were Osteosarcoma (32 cases, 30.47%), Chondrosarcoma (14 cases, 13.33%), and Ewing sarcoma (11 cases, 10.47%). The most common soft tissue sarcoma was Rhabdiosarcoma. Mandible was the most common location for these lesions.

Conclusions: In this study, the hard tissue sarcomas were more prevalent than soft tissue ones. Hence, special attention should be paid to the patients when being diagnosed.

Keywords: Epidemiology; Sarcoma; Head and Neck

1. Background

Sarcomas are a wide range of malignant neoplasm derived from mesenchymal tissue. Head and neck sarcomas are exceedingly rare and they make about 4% - 10% of all sarcomas and less than 1% of all neoplasm of head and neck (1-7). These lesions are often similar to common benign lesions and include many histological types that can be seen in any anatomical condition. In fact, many of these tumors occur in sites that no mature tissue can be seen (8, 9). For this reason it is often difficult to diagnose oral sarcomas and they are not usually diagnosed easily. Because of high recurrence rate and poor prognosis of these tumors, it is necessary to conduct an epidemiological study on the prevalence of these lesions, with consideration of age, gender of the patients and location of lesions (10, 11). Therefore, it should be noted that clinician and pathologists are expected to have complete knowledge about these tumors for the precise diagnosis.

So far, numerous studies have been carried out in various countries about the prevalence and distribution of sarcomas. 32 patients with oral sarcoma were studied by Yamaguchi et al. (1) in Japan (2004). The patients average age was reported 42 with a male- female ratio of (3 : 1). In 2007, Chidzonga and Mahomva (12) studied 88 sarcomas of the oral and maxillofacial region in a period of 24 years. Osteosarcoma was reported to be the most common sarcoma that could be seen in the third decade of life mostly in male and in the mandible. Rhabdomyosarcoma was the most common sarcoma of maxilla. Guevara-Canales et al. (13) in Peru studied 155 patients with oral sarcoma. They reported that Osteosarcoma was the most common oral sarcoma. The mean age of patients with this type of sarcoma was 36, and the prevalence of the disease was higher in females in mandible. Razmpa (8) conducted a study on head and neck sarcomas in Tehran in 1999. Among 40 patients 28 were males (70%) and 12 were females (30%). The age range of the patients was 31 - 40. The highest occurrences of sarcomas were osteosarcoma and Rhabdomyosarcoma, respectively.

The epidemiological characteristics of disease vary in geographical areas, and numerous studies have demonstrated geographical variation affects the incidence of different types of sarcomas, ages and genders prevalence. Therefore, conducting such studies is undeniable in com-
2. Objectives

Head and neck sarcomas are exceedingly rare and they include 4% - 10% of all sarcomas and less than 1% of all neoplasms of head and neck. Therefore, the aim of this study is to evaluate the epidemiological characteristics of head and neck sarcomas of patients in Isfahan, Iran.

3. Patients and Methods

In the present retrospective study, all the medical records of the patients in the archives of Departments of Pathology in the largest academic medical centers of University of Isfahan from January 2001 - July 2014 were reviewed. The files with different types of sarcomas were extracted, then the samples were evaluated according to age, gender and location of the lesions. The data were analyzed by SPSS (version 13), T-test, Chi-square and Fisher test. The P Value < 0.05 was statically significant.

4. Results

Totally, according to reports obtained from patients' files in the archives of Departments of Pathology in Isfahan, the following results have been revealed: From the total number of 16000 files studied from the year 2001 - 2014, the number of 105 cases were with head and neck sarcomas. In other words, about 6% of all samples were head and neck sarcomas. From the total number of diagnosed sarcomas 56 males (53.33%) and 49 females (46.66%) suffered from these malignancies. Fisher's test did not indicate statistically significant difference between males and females. The most common head and neck sarcomas in this population were osteosarcoma (32 cases, 30.47%), Chondrosarcoma (14 cases, 13.33%), Ewingsarcoma (11 cases, 10.47%) respectively. The sarcomas with the lowest frequency among head and neck sarcomas in this study were Kaposi sarcoma seen in 2 cases, and Spindle cell sarcomas and liposarcomas each in 3 cases. The most common location for these sarcomas were mandible, maxilla, ears and neck respectively. The patients with head and neck sarcomas were from 1 - 85 years old with the mean age of 41. The frequencies of different types of head and neck sarcomas according to the prevalence of each sarcoma, gender, mean age and the most common location are shown in Table 1.

Table 1. Frequencies of Head and Neck Sarcomas

| Type of Disease       | Total No. (%) | Man | Woman | Mean Age | Common Location \(^{a}\) |
|----------------------|---------------|-----|-------|----------|--------------------------|
| Osteosarcoma         | 32 (30.47)    | 19  | 13    | 34       | 1- mandible; 2- maxilla  |
| Chondrosarcoma       | 14 (13.33)    | 7   | 7     | 30       | 1- mandible; 2- maxilla  |
| Ewing’s sarcoma      | 11 (10.47)    | 5   | 6     | 14       | 1- mandible; 2- maxilla  |
| Rhabdomyosarcoma     | 8 (7.61)      | 3   | 5     | 9        | Ear                      |
| Neurogenic sarcoma   | 8 (7.61)      | 3   | 5     | 48       | Cranial nerves           |
| Synovial sarcoma     | 6 (5.71)      | 3   | 3     | 29       | Neck                     |
| Fibro sarcoma        | 6 (5.71)      | 3   | 3     | 36       | Mandible                 |
| Fibro histocytoma    | 4 (3.80)      | -   | 4     | 40       | Ear                      |
| Hemangio sarcoma     | 4 (3.80)      | 3   | 1     | 72       | Ear                      |
| Metastatic sarcoma   | 4 (3.80)      | 3   | 1     | 35       | 1- lymph node; 2- palate; 3- nasopharynx |
| Liposarcoma          | 3 (2.85)      | 2   | 1     | 76       | 1- thyroid; 2- trachea; 3- ear |
| Spindel cell sarcoma | 3 (2.85)      | 3   | -     | 48       | Maxilla                  |
| Kaposi sarcoma       | 2 (1.90)      | 2   | -     | 67       | 1- ear; 2- mandible      |
| Total                | 105           | 56  | 49    | 41       |

\(^{a}\) The most prevalent location of sarcomas in order of their frequency.
5. Discussion

Sarcomas are a group of malignancies that derived from mesenchymal tissue. Head and neck sarcomas are rare. Epidemiological evaluations in different countries are limited and they present different results about prevalence of various sarcomas. In 2007, Lung et al. (14) studied 1072 patients with head and neck malignancy. It was concluded that 93% of malignancies were carcinomas and 4% were sarcoma. These sarcomas were 66% more common in males. In the present study the total number of 105 cases (0.6%) with head and neck sarcomas were reported during this period. In other studies the prevalence of head and neck sarcomas included less than 1% of all neoplasm of the body (1, 2, 4, 5).

According to the results of this study head and neck sarcomas can occur in an age range (from 1 - 85 years old with the mean age of 41). The results of these findings are similar to those of several studies (1, 12, 15, 16). But in the studies by Guevara-Canales et al. (13), Eeles et al. (17) and Pandey et al. (18), the mean age of the patients was slightly less than the present results (13, 17, 18).

In this study, the number of the males with sarcomas were slightly more than females, but this difference between the two genders was not statistically significant (P ≥ 0.05). In Yamaguchi (1), Razmpa (8), Chidzonga and Mahomva (12) and Chidzonga (15), Lung et al. (14) and Lajer et al. (19) studies the prevalence of head and neck sarcomas in males were higher than it was in females, but in the study that was done by Guevara-Canales et al. (13) and Singh et al. (16) the prevalence of sarcomas were higher in females.

Osteosarcoma was the most common sarcoma in the present study with a mean age of 34 years and was more common in males. It was mainly seen in mandible. The results were similar to the findings from other studies (1, 6, 8, 12, 14, 15, 20) in spite of the Guevara-Canales et al. (13) study that osteosarcoma was found mainly in females. An investigation in Shiraz revealed the location of these lesions was more prevalent in maxilla. The data of this study indicated that Chondrosarcoma was the second most common head and neck sarcoma with equal gender distribution and predilection for mandible. But in the study conducted by Guevara-Canales et al. (13), it was more common in females. In studies that was done by Chidzonga and Mahomva (12) and Chidzonga (15) this lesion was the most common head and neck sarcoma that was ranked in the third place after Osteosarcoma and Rhabdomyosarcoma, respectively.

According to current result, Ewing’s Sarcoma was the third most common one. But there were no gender differences in their prevalence and it was in mandible predominantly. The results of the study done by Guevara-Canales et al. (13) showed this sarcoma was more common in males and in maxilla. In most studies, Rhabdomyosarcoma after osteosarcoma was the second most common sarcoma in head and neck, but in this study it is placed after Osteosarcoma, Chondrosarcoma and Ewing’s sarcoma. (1-4, 8, 12, 13, 21, 22). In other words, the result of this study demonstrated that the hard tissue sarcomas were more prevalent than the soft tissue ones. This can particularly be considered for Rhabdomyosarcoma.

Similarly, Rhabdomyosarcoma occurred in early ages commonly in girls with the mean age of 9 years (1, 8, 13). Unlike the results from study by Chidzonga et al. (12) in which the disease was prominent in boys and was mainly seen in maxilla. Other studies by Kraus, Andrassy and Patel on the prevalence of soft tissue sarcomas shed light on the fact that this type of sarcoma can be seen in infants and children (22-24). Liposarcoma and Kaposi sarcoma had the least prevalence among all types of sarcomas. Because of fewer occurrences of these malignancies in head and neck, the findings were not unexpected. Other studies also confirm these results (1, 13, 23, 24).

According to searching methods, racial and geographical differences as well as age and gender differences, all may lead to the various distributions of tumors among patients. Due to the patients’ incomplete files and lack of attainment for the patients, following up the information of the patients for evaluating the size of tumor, signs and symptoms, prognosis of the disease and surgical procedures was not possible. According to the results of this study head and neck sarcomas are rare and the hard tissue sarcomas are more prevalent than the soft tissue ones. Osteosarcoma is the most common head and neck sarcoma and Rhabdomyosarcoma is the most common soft tissue sarcoma. The results of this study confirm other studies done in different countries. Because of the poor prognosis and misdiagnosis of these lesions, these studies could be suggested for better evaluation of the patients.

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Authors’ Contributions
None Declared.

Conflict of Interest
None Declared.

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