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Original Article

Thoracic Sarcoma: A New Glance at the Epidemiological Characteristics of Disease in Iran from 2009 to 2014

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Abstract

Background: The heterogeneity, high rate of mortality and lack of comprehensive diagnostic methods have categorized primary sarcomas of the thorax as a malignancy with dismal outcomes and unknown etiology. Given the fundamental role of epidemiological analysis in establishing management strategies, we designed a study with focus on the epidemiological characteristics of primary thoracic sarcomas in Iran.

Methods: This national population-based cancer study was conducted on patients with histologically confirmed sarcoma of the thorax referred to the Iranian National Cancer Registry between 2009 and 2014. The incidence was calculated as number of cases per 100,000 person-years and was age-adjusted by the direct method using the weight of the 1960 world standard population.

Results: Over a 6-year period, 1477 cases with pathologically confirmed thoracic sarcomas were registered in Iran, of which 896 were male and 581 were female. Khuzestan Province had the highest incidence of thoracic sarcomas as compared to other provinces. Malignant mesothelioma was the most common histological subtype (20.85%). Moreover, the age-standardized incidence rate (ASR) of the disease was 1.94 per 100,000 which was more common in males than females with the highest incidence rate in men aged more than 65 years.

Conclusion: Our study provided valuable epidemiologic data on characteristics of thoracic sarcomas. This data can be used for strategizing preventive measures.

Keywords: Sarcoma; Thorax; Mesothelioma; Incidence; Iran

Introduction

Sarcomas are infrequent and diverse malignant tumors, accounting for less than 1% of all adult malignancies and 12% of pediatric cancers (1-3). They arise from a mesenchymal origin, which includes bone, cartilage, fat, vascular or hematopoietic tissues. The heterogeneous characteristics and diverse tissue distribution, coupled with varied

clinical manifestations puts obstacles in the way of the successful diagnosis and management of thoracic sarcomas, making this disease one of the less known and deadliest malignancies globally (1-3). According to the WHO, "there are more than 50 histologic subtypes of sarcoma" (4).

However, to provide a better perspective of the disease, sarcomas are divided into two groups in terms of origin: bone sarcomas and soft tissue sarcomas (STS) which can occur in any anatomic site as well as the thorax. Although the etiologies of sarcoma is still a hot topic to be debated, the importance of genetic predisposition, gene mutations, chemical carcinogens, and chronic irritation in the initiation and progression of the malignancies should not be underestimated (5). Among different types of sarcomas, primary sarcoma of the thorax, (6), is one of the least-known sarcomas, likely due to difficulties in diagnosis of this malignancy.

Apart from lack of comprehensive diagnostic criteria, the heterogeneity of histologic features of primary thoracic sarcomas together with its high mortality rate have made the clinical outlook of the disease dismal. We designed a study with focus on the epidemiological characteristics of primary thoracic sarcomas in Iran.

Materials and Methods

The study protocol was approved by Ethics Committee of Shahid Beheshti University of Medical Sciences and all patients' data will be remained confidentially.

All patients with histologically confirmed sarcoma of the thorax, who were registered in the Iranian National Cancer Registry (INCR) between 2009 and 2014, were assessed in this study. It is worth mentioning the data in question is national data and was released after 3 years from the last registration. Moreover, the national data of Ministry of Health is currently available until 2014. Epidemiologic, clinical and pathologic data were collected for each patient, including Age at diagnosis, sex, province of residence, year of diagnosis, site of involvement based on clinical and pathologic findings and histologic subtype of sarcoma based on pathology report. Of note, for our study, we used the population and census data based on data from the Statistics Center of Iran in 2006, 2011 and 2016, as well as the data of the Civil Registration Center of Iran.

To study the frequency of thoracic sarcoma in different age groups, we divided patients into 3 categories based on their age at the time of diagnosis. Patients between 0-14 years of age, patients between 15-64 years of age and patients over 65 years old (Table 1). The incidence rates were calculated as number of cases per 100,000 person-years and were age-adjusted by the direct method using the weight of the 1960 world standard population (Segi (world) standard).

Table 1: Incidence of thoracic sarcomas by age, gender, and year, Iran, 2009-2014.

	Male							***Female***						
			Incidence rates (1/100,000)							Incidence rates (1/100,000)				
Year	N	%	0-14	15-64	+65	CR	ASR	N	%	0-14	15-64	+65	CR	ASR
2009	147	16.41	0.09	0.36	2.11	0.39	0.39	82	14.11	0.07	0.24	0.74	0.22	0.22
2010	121	13.50	0.03	0.28	2.04	0.32	0.32	68	11.70	0.02	0.20	0.68	0.18	0.17
2011	142	15.85	0.06	0.37	1.66	0.37	0.36	99	17.05	0.09	0.28	0.86	0.27	0.26
2012	161	17.97	0.08	0.43	1.90	0.42	0.42	112	19.28	0.02	0.32	1.28	0.31	0.29
2013	168	18.75	0.06	0.45	1.87	0.44	0.42	125	21.51	0.09	0.35	1.12	0.34	0.32
2014	157	17.52	0.08	0.39	2.06	0.41	0.41	95	16.35	0.04	0.25	1.06	0.26	0.24
Total	896	100	0.4	2.28	11.64	2.36	2.34	581	100	0.33	1.64	5.74	1.56	1.51

Stata 14 software and SPSS Statistics 24 (IBM Corp., Armonk, NY, USA) was used to analyze crud data. For subset analysis, tumors were categorized by the site and tissue of origin in the thorax including: tumors in the heart, mediastinum and pleura, bronchus and lung, connective tissues of thorax, ill-defined sites of thorax, ill-defined parts in respiratory system, thymus, trachea and ribs (Table 2). Histological subtypes were coded according to the third edition of the International Classification of Disease for Oncology (ICD-O-3) and then grouped into major histological subtypes based on the WHO Classification of Tumors of Soft Tissue and Bone (2) as shown in Table 3.

Table 2: Frequency and percentage of thoracic sarcomas by primary site, Iran, 2009-2014

Location	*Frequency (%)*	*Incidence rates (1/100,000)*
Connective tissues of thorax	501 (33.92)	0.668
Heart, mediastinum and pleura	430 (29.11)	0.573
Bronchus and lung	295 (19.97)	0.393
Ill-defined sites of thorax	130 (8.80)	0.173
Rib	116 (7.85)	0.155
Trachea	3 (0.20)	0.004
Ill-defined parts in respiratory system	1 (0.07)	0.001
Thymus	1 (0.07)	0.001
Total	1477 (100)	1.968

Table 3: Histologic subtype by ICD10-O-3 Code, percent and incidence rate, Iran, 2009-2014

Histologic subtype	*ICD10-O-3 Code*	*N*	*%*	*Incidence rate (1/100000)*
Soft tissue tumors/ Sarcomas, NOS			30.8	
Sarcoma, NOS	8800	145	9.82	0.193
Spindle cell sarcoma	8801	199	13.47	0.266
Giant cell sarcoma (except of bone)	8802	25	1.69	0.033
Small cell sarcoma (Round cell sarcoma)	8803	15	1.02	0.020
Epithelioid sarcoma	8804	7	0.47	0.009
Undifferentiated sarcoma	8805	4	0.27	0.005
Desmoplastic small round cell tumor	8806	60	4.06	0.080
Fibromatous neoplasms (Fibrosarcoma)			4.39	
Fibrosarcoma, NOS	8810	38	2.57	0.051
Fibromyxosarcoma	8811	12	0.81	0.016
Infantile fibrosarcoma	8814	2	0.13	0.003
Solitary fibrous tumor, malignant	8815	13	0.88	0.017
Malignant fibrous histiocytoma	8830	76	5.15	0.101
Dermatofibrosarcoma			1.9	
Dermatofibrosarcoma, NOS	8832	27	1.83	0.036
Pigmented dermatofibrosarcoma protuberans	8833	1	0.07	0.001

Myxomatous neoplasms (Myxosarcoma)	8840	1	0.07	0.001
Lipomatous neoplasms (Liposarcoma)			4.33	
Liposarcoma, NOS	8850	14	0.95	0.019
Liposarcoma, well differentiated	8851	13	0.88	0.017
Myxoid liposarcoma	8852	22	1.49	0.029
Round cell liposarcoma	8853	3	0.20	0.004
Pleomorphic liposarcoma	8854	7	0.47	0.009
Mixed liposarcoma	8855	3	0.20	0.004
Dedifferentiated liposarcoma	8858	2	0.14	0.003
Myomatous neoplasms (Leiomyosarcoma)			3.92	
Leiomyosarcoma, NOS	8890	48	3.25	0.064
Epithelioid leiomyosarcoma	8891	4	0.27	0.005
Angiomyosarcoma	8894	1	0.07	0.001
Myosarcoma	8895	2	0.13	0.003
Myxoid leiomyosarcoma	8896	3	0.20	0.004
Rhabdomyosarcoma			1.56	
Rhabdomyosarcoma, NOS	8900	10	0.68	0.013
Pleomorphic rhabdomyosarcoma, adult type	8901	2	0.13	0.003
Mixed type rhabdomyosarcoma	8902	1	0.07	0.001
Embryonal rhabdomyosarcoma, NOS	8910	7	0.47	0.009
Spindle cell rhabdomyosarcoma	8912	1	0.07	0.001
Alveolar rhabdomyosarcoma	8920	2	0.14	0.003
Complex mixed and stromal neoplasms			0.96	
Mixed tumor, malignant, NOS	8940	4	0.28	0.005
Malignant rhabdoid tumor	8963	3	0.20	0.004
Carcinosarcoma, NOS	8980	6	0.41	0.008
Mesenchymoma, malignant	8990	1	0.07	0.001
Fibroepithelial neoplasms				
Phyllodes tumor, malignant	9020	1	0.07	0.001
Synovial neoplasms (Sarcoma)			4.26	
Synovial sarcoma, NOS	9040	46	3.11	0.061
Synovial sarcoma, spindle cell	9041	9	0.61	0.012
Synovial sarcoma, epithelioid cell	9042	1	0.07	0.001
Synovial sarcoma, biphasic	9043	7	0.47	0.009

Table 3: Continued…

Histologic subtype	***ICD10-O-3 Code***	***N***	***%***	***Incidence rate (1/100000)***
Mesothelial neoplasms			22.07	
Mesothelioma, malignant	9050	308	20.85	0.410
Fibrous mesothelioma, malignant	9051	18	1.22	0.024
Blood vessels tumors			2.5	
Hemangiosarcoma	9120	9	0.61	0.012

Hemangioendothelioma, malignant	9130	1	0.07	0.001
Epithelioid hemangioendothelioma, malignant	9133	3	0.20	0.004
Kaposi sarcoma	9140	21	1.42	0.028
Hemangiopericytoma, malignant	9150	3	0.20	0.004
Osseous & chondromatous neoplasms (Osteosarcoma)			2.65	
Osteosarcoma, NOS	9180	31	2.10	0.041
Chondroblastic osteosarcoma	9181	5	0.34	0.007
Telangiectatic osteosarcoma	9183	1	0.07	0.001
Central osteosarcoma	9186	1	0.07	0.001
Small cell (Round cell) osteosarcoma	9185	1	0.07	0.001
Chondrosarcoma			6.3	
Chondrosarcoma, NOS	9220	79	5.35	0.105
Myxoid chondrosarcoma	9231	9	0.61	0.012
Mesenchymal chondrosarcoma	9240	4	0.27	0.005
Dedifferentiated chondrosarcoma	9243	1	0.07	0.001
Giant cell tumors			3.79	
Giant cell tumor of bone malignant	9250	9	0.61	0.012
Malignant giant cell tumor of soft parts	9251	1	0.07	0.001
Ewing sarcoma	9260	46	3.11	0.061
Miscellaneous tumors			0.14	
Chordoma, NOS	9370	1	0.07	0.001
Chondroid chordoma	9371	1	0.07	0.001
Nerve sheath tumors			4.19	
Malignant peripheral nerve sheath tumor	9540	59	3.99	0.079
Neurilemoma, NOS	9560	2	0.13	0.003
Perineurioma, malignant Perineural MPNST	9571	1	0.07	0.001
Granular cell tumors and alveolar soft part sarcomas			0.61	
Granular cell tumor, malignant	9580	1	0.07	0.001
Alveolar soft part sarcoma	9581	8	0.54	0.01
Hematopoietic sarcoma			0.34	
Myeloid sarcoma	9930	3	0.20	0.004
Mast cell sarcoma	9740	1	0.07	0.001
Follicular dendritic cell sarcma	9758	1	0.07	0.001

Theory: Given the lack of epidemiologic information on the primary thoracic sarcomas in Iran, we aimed to design a study with focus on the epidemiological characteristics of this malignancy in Iran (from 2009 to 2014) to improve the management and prevention strategies of this disease. This study was based on the data provided by the population based Iran National Cancer Registry data (INCR).

Results

Studying thoracic sarcomas in Iranian patients between 2009 to 2014: age, gender, and year of diagnosis

Over a 6-year period, from 2009 to 2014, 1477 cases with pathologically confirmed diagnosis of sarcoma in the thorax from 31 provinces of Iran were registered in the INCR. 896 were (60.6%) male and 581 (39.4%) were female. As shown in Fig. 1a, ASR for males and females were 2.34 (1/100000) and 1.51 (1/100000) respectively. The overall crude incidence rate (CR) and age standardized incidence rates (ASR) of thoracic sarcoma

was 1.97 per 100,000. The incidence rate of thoracic sarcomas between 2009 and 2014 rose from 0.31 per 100000 to 0.34 per 100000 with a peak at 0.39 per 100000 in 2013 and was higher in males than females (Fig. 1a). To provide a better perspective for our findings, we classified the incidence rates according to age, gender, and year of diagnosis (Table 1). According to age and among newly diagnosed thoracic sarcomas, 69.97% of the cases (40.56% male and 29.11% female) occurred in the age category 2 (15-64 years old), 25.73% (17.54% male and 8.19% female) in the age category 3 (+65 years) and 4.6% (2.57% male and 2.03% female) in the age category 1 (0-14 years old).

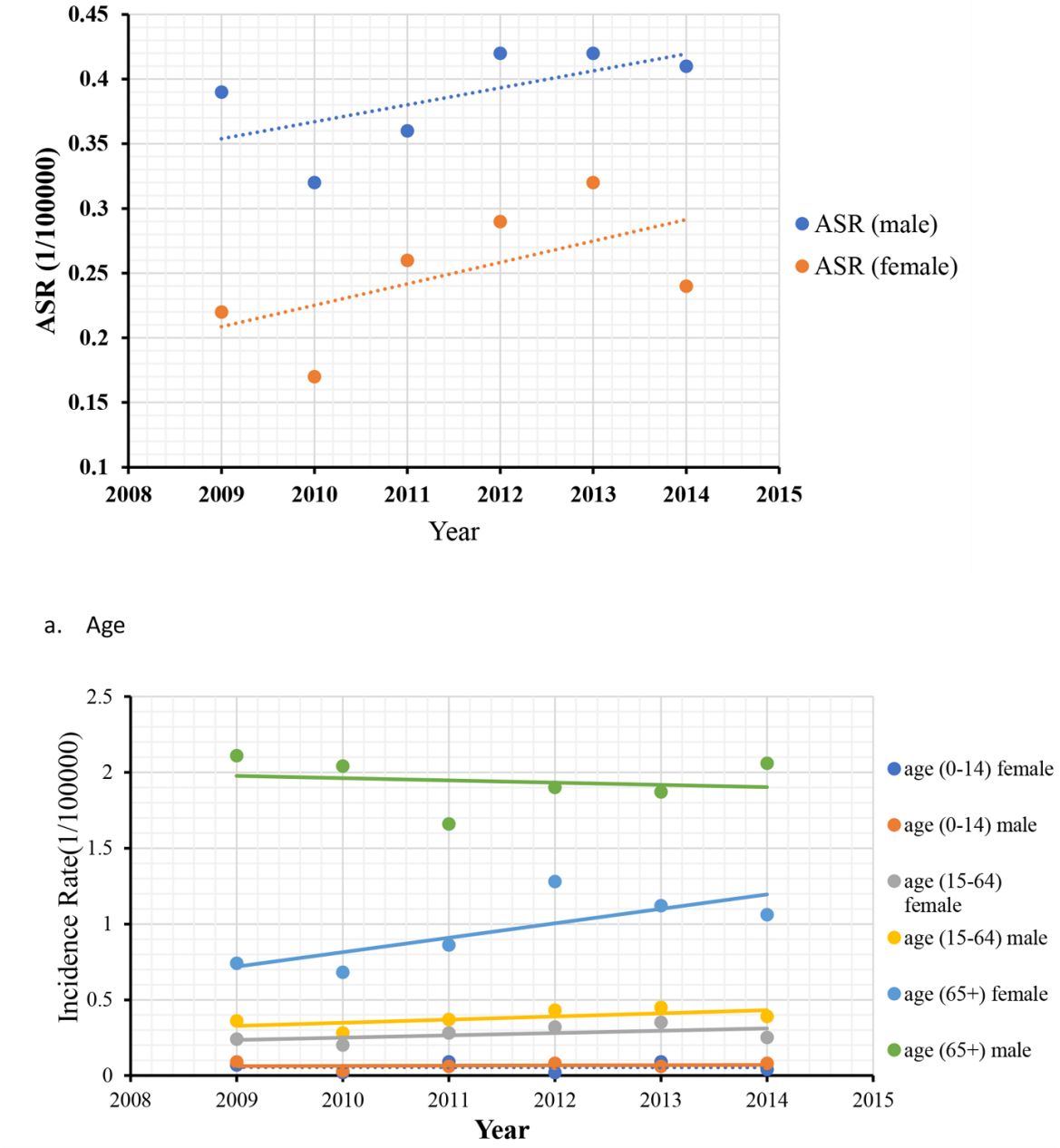

Fig. 1: Trends for age-adjusted incidence of thoracic sarcomas by (a) gender and (b) age between

Studying thoracic sarcomas in Iranian patients from 2009 to 2014: gender and province

Incidence of thoracic sarcomas was also classified according to gender and province (Fig. 2). While Khuzestan Province has the highest incidence rate of thoracic sarcoma, the occurrence of this malignancy in Kermanshah Province was lowest for both genders. Among men, the highest incidence rates occurred in Khuzestan, Lorestan, Isfahan, Kohgiluyeh and Boyer-Ahmad, and Tehran with age-standardized rates (ASRs) of 4.75, 4.34, 3.49, 3.21, and 3.05 per 100000, respectively. For women, West Azerbaijan, Khuzestan, and Isfahan had the highest incidence rates with ASRs of 3.24, 3.06, and 2.15 per 100000, respectively.

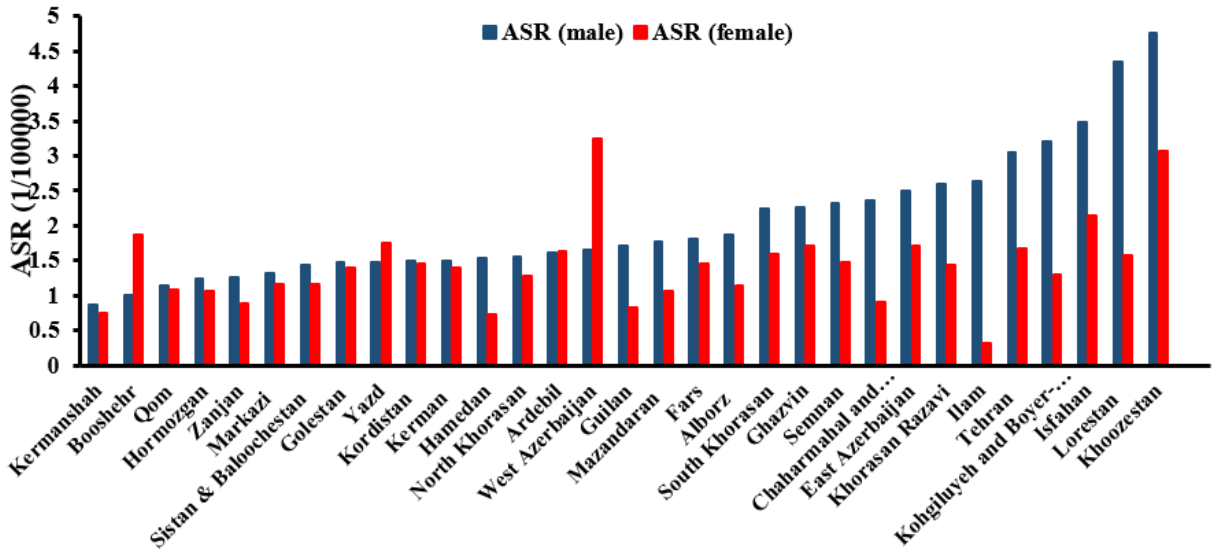

Fig. 2: Incidence of thoracic sarcoma in Iranian patients during 2009-2014 according to gender and province

Studying thoracic sarcoma in Iranian patients from 2009 to 2014: tissue origin

Another important criterion for the classification of thoracic sarcomas is its tissue of origin. In this study, 1378 of 1477 cases (93.30%) of thoracic sarcomas originated from soft tissue and the origin of the remaining cases was the bone. Table 4 describes the incidence of bone or soft tissue thoracic sarcomas according to gender and age between 2009 and 2014.

Table 4: Incidence of thoracic sarcomas by age, gender, and bone or soft tissue, Iran, 2009-2014.

	Male							***Female***						
			Incidence rates (1/100,000)											
Tissue	N	%	0-14	15-64	+65	CR	ASR	N	%	0-14	15-64	+65	CR	ASR
Bone	60	6.69	0.02	0.19	0.45	0.16	0.15	39	6.71	0.03	0.13	0.09	0.11	0.09
Soft tissue	836	93.31	0.39	2.08	11.24	2.21	2.19	542	93.29	0.30	1.51	5.68	1.46	1.42
Total	896	100	0.41	2.27	11.64	2.36	2.34	581	100	0.33	1.64	5.77	1.56	1.51

Studying thoracic sarcoma in Iranian patients between 2009 and2014: site of tumor

33.92% of sarcomas were located in the connective tissues of thorax (Table 2). Sarcomas of the thymus and trachea had the lowest incidence with only 3 and 1 cases, respectively.

Studying thoracic sarcoma in Iranian patients between 2009 and2014: age, gender and histologic subtype

Seventy one different histological subtypes of sarcoma were identified in the thorax (Table 3). The highest incidence rates belonged to age category 3 (Age 65+) in both sexes except for Ewing sarcoma with highest incidence rate in age category 2 (Age 15-64) in men and age category 1 (Age 0-14) in women. All histologic sub types had higher incidence rates in male except for leiomyosarcoma, Granular cell tumors and alveolar soft part sarcomas, myxosarcoma (1 case), which had higher incidence rates in females. The most common histologic sub types in age category 1 were rhabdosarcoma and Ewing sarcoma, malignant mesothelioma and chondrosarcoma in age category 2 and malignant mesothelioma and malignant fibrous histiocytoma in age category 3. Table 5 summarizes the incidence of thoracic sarcomas by age, gender, and histologic type, Iran, 2009-2014.

Table 5: Incidence of thoracic sarcomas by age, gender, and histologic type, Iran, 2009-2014.

Histologic subtype	***Male*** Incidence rates (1/100,000)							***Female***						
	N	%	0-14	15-64	+65	CR	ASR	N	%	0-14	15-64	+65	CR	ASR
Sarcoma, NOS	260	29.02	0.18	0.648	3.13	0.695	0.633	195	33.62	0.11	0.544	1.88	0.523	0.464
Fibrosarcoma	41	4.58	0.04	0.11	0.45	0.113	0.103	24	4.13	0.01	0.07	0.24	0.071	0.049
Malignant fibrous histiocytoma	55	6.14	0	0.11	1.12	0.15	0.14	21	3.61	0.02	0.05	0.33	0.06	0.06
Dermatofibrosarcoma	18	2.01	0	0.054	0.13	0.043	0.042	10	1.72	0	0.03	0.05	0.03	0.02
Myxosarcoma	0	0	0	0	0	0	0	1	0.17	0	0.004	0	0.003	0.002
Liposarcoma	44	4.9	0.02	0.13	0.45	0.116	0.093	20	3.43	0	0.068	0.15	0.061	0.04
Leiomyosarcoma	20	2.23	0	0.05	0.36	0.055	0.046	38	6.53	0	0.106	0.53	0.101	0.097
Rhabdomyosarcoma	17	1.89	0.04	0.046	0.04	0.043	0.04	6	1.03	0.04	0.008	0	0.016	0.016
Complex mixed and stromal neoplasms	11	1.22	0	0.03	0.17	0.029	0.026	3	0.51	0	0.01	0.05	0.008	0.009
phyllodes tumor malignant	1	0.11	0	0	0.04	0.003	0.002	0	0	0	0	0	0	0
Synovial sarcoma	37	4.16	0.03	0.104	0.31	0.093	0.082	26	4.47	0	0.08	0.24	0.065	0.056
Mesothelial neoplasms	207	23.1	0.02	0.45	3.9	0.54	0.55	119	20.48	0.01	0.32	1.63	0.31	0.3
Blood vessel sarcoma	32	3.57	0	0.084	0.4	0.087	0.064	5	0.86	0.01	0.012	0.05	0.014	0.012

Osteosarcoma	25	2.79	0.01	0.088	0.09	0.066	0.057	14	2.41	0.02	0.028	0.09	0.036	0.024
Chondrosarcoma	57	6.37	0	0.174	0.58	0.143	0.141	36	6.2	0	0.12	0.19	0.09	0.08
Giant cell tumors	7	0.78	0	0.012	0.18	0.023	0.012	3	0.51	0	0.014	0	0.008	0.008
Ewing sarcoma	24	2.68	0.04	0.06	0.04	0.06	0.05	22	3.79	0.09	0.06	0	0.06	0.06
Miscellaneous tumors	2	0.22	0	0.006	0	0.006	0.003	0	0	0	0	0	0	0
Nerve sheath tumors	32	3.57	0.02	0.1	0.13	0.08	0.07	30	5.16	0.02	0.094	0.19	0.078	0.068
Granular cell tumors and alveolar soft part sarcomas	3	0.33	0	0.01	0	0.008	0.006	6	1.03	0	0.024	0.05	0.013	0.012
Hematopoietic Sarcoma	3	0.33	0	0.004	0.08	0.008	0.007	2	0.34	0	0	0.1	0.006	0.006
Total	896	100	0.41	2.27	11.64	2.36	2.34	581	100	0.33	1.64	5.77	1.56	1.51

Discussion

Among adult malignancies, sarcomas are rare and diverse tumors with mesenchymal origin that can arises in any anatomic site. Sarcomas account for less than 1% of all adult malignancies (1-3). According to the American college of surgeons, the anatomic distribution of soft tissue sarcomas in 4550 adults was Thigh, buttock, and groin (46 percent) upper extremities (13 percent), trunk (18 percent) retro peritoneum (13%) head and neck (9%) (7). Primary thoracic sarcomas are extremely rare and therefore epidemiologic and etiologic studies to increase knowledge about this disease is difficult (8).

Many epidemiologic studies are available on incidence and mortality of sarcomas worldwide, however, to the best of our knowledge, this is the first study on epidemiologic distribution patterns of sarcomas in Iran. Although small studies on the incidence of cardiac sarcoma, chest wall sarcoma and primary pleuropulmonary sarcomas have been reported in the literature, there are no large scale population based epidemiologic studies available (8-11).

We found that the ASR of thoracic sarcoma was 1.94 per 100,000 and that it was more common in males than females (M/F ratio: 1.54: 1) with the highest incidence rate in men aged 65 years or older. In line with our results, the incidence of soft tissue sarcoma in the extremities and trunk in Taiwan has been reported as 1.63 per 100000 (12). However, the incidence of soft tissue sarcoma is reported to be higher in some other countries, such as those reported by Austrian National Cancer Registry (ASR: 2.4 per 100000) (13), or the population-based study in Shanghai (ASR: 3.4 per 100,000) (14), RARECARE project in Europe (total ASR: 4.2 per 100000) (15) and SEER program in the USA (total 5 with US 2000 standard population) (16). This noticeable difference in incidence could be attributed to the diversity of the studied population and tumor sites.

It is well-established that there is a noticeable connection between incidence of human cancers and age. The incidence of cancers increases with aging and thoracic sarcoma is not an exception. The importance of age in the incidence rate of the disease is to the extent that national cancer intelligence team in the UK conducted a comprehensive research between 1996 and 2010 and reported that the age-standardized incidence of bone sarcoma and soft tissue sarcoma was approximately 7.9 and 45 per million, respectively (17). Although age could be a promising criterion for evaluating the prognosis of cancer, its combination with gender

can act as a beacon to shed more light on the clinical status of a malignancy. Incorporation of both criteria for thoracic sarcoma in the present study provided a piece of valuable information suggesting that while there was a steady upward trend in the incidence rate for all age groups over a 6-year period (Fig.1b), the disease occurrence in males over 65 years old (11.24 per 100000) was significantly higher with the male to female ratio (M/F ratio) of 1.98:1. In agreement with our results, soft tissue sarcomas could be highly detected in males over 85 years old with an M/F ratio of 1.9:1 (17). According to tissue origin, there are some differences in the incidence of sarcoma and age group. Although the results of the previous studies showed that bone sarcoma has the highest peak in teenagers, adolescents, and the elderly, we found that the lowest age specific incidence rates belongs to individuals aged 0-14 entirely.

Among the different solid tumors, thoracic sarcoma has the most diversity in the organs that are infiltrated by the malignant cells. According to histological characteristics, except for the connective tissues of the thorax (33.92%), the most common site of thorax inflicted by sarcoma was the heart, mediastinum and pleura (29.11%), followed by bronchus and lungs (19.97%). After sarcoma NOS (28.56%), the most common histological subtype in this study was malignant mesothelioma that accounted for 20.85% of all sarcoma.

The tissue diversity of thoracic sarcoma has been discussed in many studies, each of them reported a distinct tissue dispersion pattern for the malignancy. Francis et al. (17) reported that in the UK Leiomyosarcoma (including GISTs) with 22% and liposarcoma with 12% were the most common specific types of soft tissue sarcoma. In Europe, "leiomyosarcoma was the most common histologic type" (14). The most common histological subtype excepting of sarcoma NOS, was gastrointestinal stromal sarcoma (GISS) with 14.8 % followed by fibrosarcoma (7.2%), lipoblastoma (6.7%), leiomyosarcomas (6.5%), and osteosarcoma (5.3%) (14).

Our results on the histologic subtype of sarcoma was different from all previous studies which could be due to the fact that this study was limited to the assessment of thoracic sarcoma alone. In a 6-year period, we reported that the ASR of soft tissue sarcoma in thorax was 1.84 per 100,000 and ASR of the bone sarcoma in the thorax was 0.13 per 100,000 which was lower than reports from other cancer registries.

Conclusion

Our observational study provided valuable data about thoracic sarcomas, which can lead to finding etiologic clues and risk factors of the disease. Apart from some limitations, including the restrictive data of the Iran National Cancer Registry to sarcoma of the thorax, the lack of data on mortality rate, the lack of advanced diagnostic technology such as molecular profile and cytogenetics and the possibility of misdiagnoses of the histology by the pathologist, our findings of this study can have clinical value and may help find the etiology and risk factors.

Journalism Ethics considerations

Ethical issues (Including plagiarism, informed consent, misconduct, data fabrication and/or falsification, double publication and/or submission, redundancy, etc.) have been completely observed by the authors.

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Conflict of interest

The authors declare no conflict of interest.

References

1. Miller R, J Young, Novakovic B (1995). Childhood cancer. *Cancer*, 75: 395-405.
2. Jo VY, CD Fletcher (2014). WHO classification of soft tissue tumours: an update based on the 2013 4th ed. *Pathology*, 46(2): 95-104.
3. Siegel RL, KD Miller, et al (2016). Cancer statistics. *Cancer J Clin*, 66(1): 7-30.
4. Fletcher, C (2002). World Health Organization classification of Tumours. Pathology and genetics of Tumours of soft tissue and bone. Vol 4. Irac.`
5. Zahm SH, J Fraumeni (1997). The epidemiology of soft tissue sarcoma. *Semin Oncol*, 24: 504-14.
6. Gladish GW, Sabloff BM, Munden RF, et al (2002). Primary thoracic sarcomas. *Radiographics*, 22(3): 621-37.
7. Lawrence Jr W, Donegan WL, Natarajan N, et al (1987). Adult soft tissue sarcomas. A pattern of care survey of the American College of Surgeons. *Ann Surg*, 205(4): 349-59.
8. Amano J, Nakayama J, Yoshimura Y, et al (2013). Clinical classification of cardiovascular tumors and tumor-like lesions, and its incidences. *Gen Thorac Cardiovasc Surg*, 61(8): 435-47.
9. Essary LR, SO Vargas, CD Fletcher (2002). Primary pleuropulmonary synovial sarcoma: reappraisal of a recently described anatomic subset. *Cancer*, 94(2): 459-69.
10. Etienne-Mastroianni B, Falchero L, Chalabreysse L, et al (2002). Primary sarcomas of the lung: a clinicopathologic study of 12 cases. *Lung Cancer*, 38(3): 283-9.
11. A Orlandi, A Ferlosio, M Roselli, et al (2010). Cardiac sarcomas: an update. *J Thorac Oncol*, 5(9): 1483-9.
12. GY Hung, CC Yen, JL Horng, et al (2015). Incidences of primary soft tissue sarcoma diagnosed on extremities and trunk wall: A population-based study in Taiwan. *Medicine (Baltimore)*, 94(41):e1696.
13. Wibmer C, Leithner A, Zielonke N, et al (2010). Increasing incidence rates of soft tissue sarcomas? A population-based epidemiologic study and literature review. *Ann Oncol*, 21(5):1106-11.
14. P Bao, Y Zhou, W Lu, et al (2019). Incidence and mortality of sarcomas in Shanghai, China, during 2002-2014. *Front Oncol*, 9: 662.
15. Stiller CA, Trama A, Serraino D, et al (2013). Descriptive epidemiology of sarcomas in Europe: report from the RARECARE project. *Eur J cancer*, 49(3): 684-95.
16. JR Toro, LB Travis, HJ Wu, et al (2006). Incidence patterns of soft tissue sarcomas, regardless of primary site, in the surveillance, epidemiology and end results program, 1978–2001: an analysis of 26,758 cases. *Int J Cancer*, 119(12): 2922-30.
17. Francis M, Dennis N, Charman j, et al (2013). Bone and soft tissue sarcomas UK incidence and survival: National Cancer Intelligence Network 1996 to 2010, pp 7-12.