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Incidences of Primary Soft Tissue Sarcoma Diagnosed on Extremities and Trunk Wall

A Population-Based Study in Taiwan

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Abstract: Most epidemiological studies of soft tissue sarcoma (STS) were performed in the Western countries, and only limited data highlighting that in the Asian population. The aim of this study is to conduct a comprehensive analysis for the incidence rates of STS in Taiwan.

This was a population-based study analyzing the incidence rates and trends of the primary STS over extremities and trunk wall during 2003 to 2011 by using the nationwide Taiwan Cancer Registry. More specific analyses were conducted for subtypes. Incidence rates of overall STS by cities and counties were also investigated.

A total of 3843 cases were diagnosed with STS during the study period, giving an age-standardized rate (ASR) of 1.63 per 100,000 person-years. Liposarcoma was the most frequent subtype, followed by undifferentiated pleomorphic sarcoma and leiomyosarcoma. STS was more frequently diagnosed in males and angiosarcoma was the most prominent sex-specific type. ASR increased with age in most of the STS subtypes and varied by histologic subtype. The incidence of peripheral primitive neuroectodermal tumor was highest in children, whereas rhabdomyosarcoma revealed a bimodal age distribution. Annual percent change (APC) of STS was 2.2%, and significant change in trend was only in males (APC, 3.5%, $P < 0.05$). Geographical variations indicated that New Taipei City had a significantly higher rate compared with the rest areas. Significantly lower rates were observed in 1 major offshore island.

Incidence variations of STS by sexes, ages, histologic subtypes, and geographic regions were observed in Taiwanese population. The emerging factors associated STS incidence rates deserve further studies to verify.

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Abbreviations: APC = annual percent change, ASR = age-standardized incidence rate, CI = confidence intervals, DCO = death certificate only, IARC = International Agency for Research on Cancer, M/F = male-to-female, MV = microscopy-verified, NF1 = neurofibromatosis type 1, NOS = not otherwise specified, pPNET = peripheral primitive neuroectodermal tumor, SEER = Surveillance, Epidemiology, and End Results Program, SRR = standardized incidence rate ratio, STS = soft tissue sarcomas, TCR = Taiwan Cancer Registry, UPS = undifferentiated pleomorphic sarcoma.

INTRODUCTION

Soft tissue sarcomas (STS) are rare and heterogeneous diseases accounting for less than 1% of the diagnosed malignancy[1] and are widely found in the connective tissues throughout the human body. STS spans a broad range of differentiation including adipocytes (liposarcoma), peripheral nerve tissues (malignant peripheral nerve sheath tumor), smooth (leiomyosarcoma) or striated muscle (rhabdomyosarcoma), vascular tissues (angiosarcoma), and the other unknown origins (such as undifferentiated pleomorphic sarcoma).[2] Due to the rarity and complexity of STS, large population-based studies are required to elucidate the incidences and the potential contributing factors.

Most of the previous population-based studies on the STS were conducted in the Western countries and the reported incidences ranged from 1.8 to 5.0 per 100,000 annually.[3–9] In recent years, an increasing incidence of STS has also been revealed in several studies.[3–6] However, specific data on STS in the Asia-Pacific region are rare,[10] and only 2 studies reporting the incidence patterns of sarcomas in Asia are available.[11,12] By using the nationwide Taiwan Cancer Registry (TCR) database, the incidences related to bone cancer in the Taiwanese population have been reported in our previously published results.[13] In this study, the TCR database was used for a comprehensive analysis of incidences and trends of STS over the extremities and trunk wall. Furthermore, geographic variations in the incidence of STS were also analyzed.

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MATERIAL AND METHODS

Data Qualification and Acquirement

The data of primary soft tissue cancers diagnosed and registered in the TCR database during 2003 and 2011 were acquired.[14] Soft tissue cancers were classified according to the International Classification of Diseases for Oncology, version 3 (ICD-O-3: C47, C49 cancer of connective, subcutaneous, and other soft tissues).[15] The data included the soft tissue cancers over extremities and trunk wall, but those from central nervous system, head and neck regions, breast, intrathoracic and intraabdominal cavities, endocrine system, and skin were excluded. In TCR, the primary soft tissue cancers were further stratified into the subgroups including undifferentiated pleomorphic sarcoma (UPS, including malignant fibrohistiocytoma and fibrosarcoma), liposarcoma, leiomyosarcoma, rhabdomyosarcoma,

TABLE 1. Incidence (Per 100,000 Person-Yr) of Primary Soft Tissue Cancers by Sex and Histologic Subtype, Taiwan (2003–2011)

Histologic Subtype	Sex	*n*	Crude Rate	ASR*	95% CI	SRR	95% CI	MV%
All types	Both sexes	3843	1.86	1.63	1.58–1.68			97.6
	Male	2211	2.12	1.87	1.79–1.95	1.34‡	1.26–1.43	
	Female	1632	1.60	1.39	1.32–1.46			
Undifferentiated pleomorphic sarcoma	Both sexes	728	0.35	0.29	0.27–0.32			99.5
	Male	432	0.41	0.35	0.32–0.38	1.48‡	1.28–1.71	
	Female	296	0.29	0.24	0.21–0.26			
Liposarcoma	Both sexes	885	0.43	0.36	0.33–0.38			98.9
	Male	537	0.51	0.44	0.40–0.48	1.58‡	1.39–1.80	
	Female	348	0.34	0.28	0.25–0.31			
Leiomyosarcoma	Both sexes	292	0.14	0.12	0.10–0.13			99.1
	Male	150	0.14	0.12	0.10–0.14	1.09	0.87–1.37	
	Female	142	0.14	0.11	0.09–0.13			
Rhabdomyosarcoma	Both sexes	201	0.10	0.11	0.10–0.13			97.2
	Male	115	0.11	0.12	0.10–0.14	1.18	0.88–1.58	
	Female	86	0.08	0.10	0.08–0.13			
Angiosarcoma	Both sexes	282	0.14	0.11	0.10–0.13			97.8
	Male	182	0.17	0.15	0.12–0.17	1.79‡	1.42–2.26	
	Female	100	0.10	0.08	0.06–0.10			
Synovial sarcoma	Both sexes	172	0.08	0.08	0.07–0.09			100
	Male	75	0.07	0.07	0.05–0.08	0.74	0.55–1.00	
	Female	97	0.10	0.09	0.07–0.11			
Malignant nerve sheath tumor (including malignant schwannoma)	Both sexes	222	0.11	0.10	0.08–0.11			98.8
	Male	117	0.11	0.10	0.08–0.12	1.05	0.80–1.36	
	Female	105	0.10	0.10	0.08–0.11			
Peripheral primitive neuroectodermal tumor	Both sexes	112	0.05	0.07	0.05–0.08			97.6
	Male	66	0.06	0.08	0.06–0.10	1.31	0.89–1.93	
	Female	46	0.05	0.06	0.04–0.08			
Other specified sarcoma	Both sexes	519	0.25	0.22	0.20–0.23			98.5
	Male	289	0.28	0.24	0.21–0.27	1.29‡	1.08–1.53	
	Female	230	0.23	0.19	0.16–0.21			
Sarcoma NOS	Both sexes	337	0.16	0.14	0.12–0.15			94.8
	Male	200	0.19	0.16	0.14–0.19	1.47‡	1.19–1.83	
	Female	137	0.13	0.11	0.09–0.13			
Other malignancy†	Both sexes	93	0.05	0.04	0.03–0.05			63.9
	Male	48	0.05	0.04	0.03–0.05	1.12	0.74–1.67	
	Female	45	0.04	0.04	0.03–0.05			

ASR = age-standardized incidence rate; CI = confidence interval; MV% = percentage of microscopically verified cases; NOS = not otherwise specified; SRR = male-to-female standardized incidence rate (ASR) ratio.

* ASRs were age-standardized to the 2000 world standard population.

† Included 9 specified carcinoma (male/female, 7/2) and 4 malignant mesenchymoma (male/female, 3/1).

‡ Indicates statistical significance at the 0.01 level.

angiosarcoma, synovial sarcoma, malignant nerve sheath tumor (including malignant schwannoma), peripheral primitive neuroectodermal tumor (pPNET), other specified sarcoma (including spindle cell sarcoma, giant cell sarcoma, small cell sarcoma, epithelioid sarcoma, and so on), sarcoma not otherwise specified (NOS), and other malignancy (including carcinomas, malignant mesenchymoma, and other nonsarcoma neoplasms). Lymphoma and multiple myeloma were excluded in the current study, except for analyzing the geographical variations. Census and land data were obtained from the Department of Statistics, Ministry of the Interior, Taiwan.[16]

The TCR have collected cancer data since 1979. Regarding the data quality defined by the International Agency for Research on Cancer (IARC), improvements in the percentage of microscopy-verified registration (MV, %) and decreased percentage of death certificate only (DCO, %) were demonstrated in previous reports.[17,18] In Taiwan, the Cancer Control Act effective in 2003 began mandating that all the hospitals implemented with >50 beds need to submit cancers data to the central registry. For all cancers registration, the ratio of MV was raised from 85.6% in 2003 to 91.6% in 2011, and the percentage of DCO was lowered from 2.7% to 0.82% during the same period. In fact, the variations in these indicators were markedly reduced than the years before 2003. These indicators demonstrated that the completeness and validity of TCR have been enhanced by mandated registration since 2003. Therefore, the present study was designed to acquire the data registered in the period between 2003 and 2011.

The dataset used in this study was obtained from TCR, Health Promotion Administration, Ministry of Health and Welfare, Taiwan (http://tcr.cph.ntu.edu.tw/main.php?Page=A1, http://tcr.cph.ntu.edu.tw/main.php?Page=SA3&-KeyID=190388277454d4356b5ece3, and http://www.hpa.-gov.tw/BHPNet/Web/Stat/Statistics.aspx). These data are publicly available and do not contain any identifiable personal information. Based on Taiwan regulations, ethical approval was waived for studies using these data (such as ours).

Data Analysis

This study employed IARC's published methods to calculate the incidence, standard errors, 95% confidence intervals (CI), and standardized incidence rate ratio (SRR).[19,20] Age-specific incidences were stratified into 18 subgroups by 5-year age interval (0–4 to 85+ years). An age-standardized incidence rate (ASR) is a weighted average of the age-specific (crude) rate, where the weight is the proportion of individuals in the corresponding age group of a standard population. Calculating the ASRs of subgroups using the same standard population can correct the potential confounding effect derived from the differences in ages. In this study, the world standard population in 2000 was used to calculate the ASRs and to examine geographic variations. The trends were analyzed by using the joinpoint regression model and permutation tests (Joinpoint Regression Program, version 4.0.4; Statistical Methodology and Applications Branch, Surveillance Research Program, National Cancer Institute, Bethesda, MD) to identify significant changes,[21,22] in which up to 2 joinpoints were produced to express the annual percent change (APC). The APC was considered significant if the 95% CI did not include 0. The relative risks of cancer, ratio of ASRs (SRRs), and 95% CI were calculated to compare the cancer incidences by cities/counties. The SRRs were considered significantly different if the estimated 95% CI did not contain 1. To avoid the effect of comparing heavily populated cities/counties (eg, New Taipei City, with 16.3% of the total population), which is itself affected by their contribution, the rate for each city/county was compared with the rest area of Taiwan (eg, New Taipei City vs. Taiwan-minus-New Taipei City).

RESULTS

A total of 3843 subjects were diagnosed as primary soft tissue cancer during the 9-year study period with a crude rate of 1.86 and an ASR of 1.63 in 100,000 (Table 1). The cumulative risk developing soft tissue cancer from birth to age 74 years (or age 84 yr) was 0.15% (0.22%). Soft tissue cancer comprised 0.6% of the diagnosed cancers in 2003, and 0.51% in 2011. The median age of the registered soft tissue cancers was 51 years in 2003, and 56 years in 2011. This result implied the growing trend of aging population in Taiwan. Liposarcoma was the most common subtype, accounting for 23% of soft tissue cancers, followed by UPS (18.9%) and leiomyosarcoma (7.6%). The 3 subtypes comprised approximately half of all primary soft tissue cancers diagnosed in Taiwanese population. Soft tissue cancers were more frequently diagnosed in males than females. The male-to-female (M/F) SRR was 1.34 ($P < 0.01$). In addition, males were significantly more susceptible to the cancer subtypes including UPS, liposarcoma, angiosarcoma, other specified sarcoma, and sarcoma NOS, with the SRRs ranging between 1.29 and 1.79. The most prominent sex difference was found in angiosarcoma (M/F SRR = 1.79, 95% CI, 1.42–2.26, $P < 0.01$). By contrast, the lowest M/F SRR was found for synovial sarcoma (M/F SRR = 0.74), although not statistically

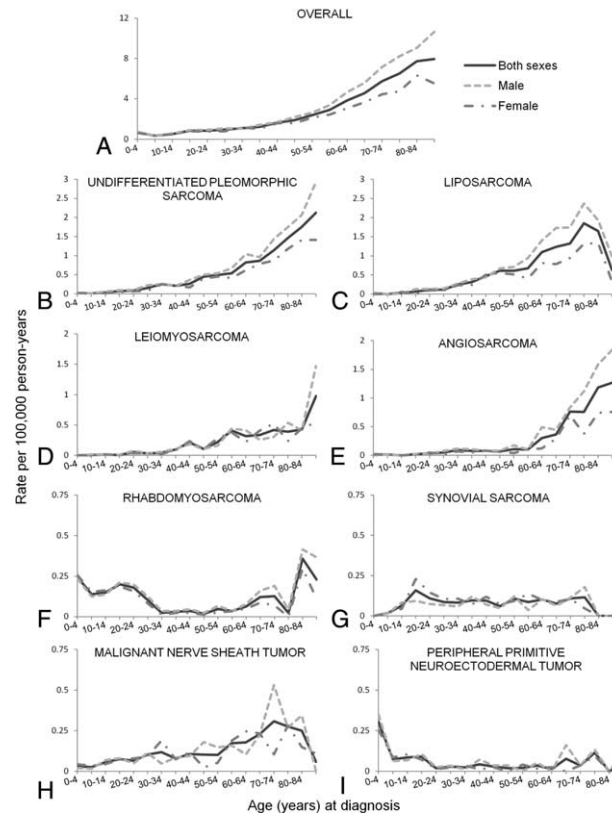

FIGURE 1. Incidence rates of primary soft tissue cancers in 5-year age groups with histologic subtypes and by sexes in Taiwan during 2003 to 2011.

significant. The percentage of MV was variable between 94.8% and 100% for all acquired subtypes of soft tissue cancers data but that of the subtype ''other malignancy'' showed a lower MV rate, 63.9% (Table 1).

Age-Specific Incidences

The age-specific incidences of soft tissue cancer increased with age and peaked for the elderly population (Figure 1A). The incidences of UPS (Figure 1B), liposarcoma (Figure 1C), leiomyosarcoma (Figure 1D), and angiosarcoma (Figure 1E) showed a similar trend among the different age groups. It was slowly increased for the population aged 30 to 50 years, a rapid increase was found in the age group of 60 to 70 years, and the highest incidence was shown in the population aged 85 and over. It is noteworthy that an exceptionally decreased incidence of liposarcoma in the population older than those

TABLE 2. Annual Percent Changes in Incidence Rates of Primary Soft Tissue Cancers by Sex and Histologic Subtype, Taiwan (2003–2011)

Histologic Subtype	Sex	Year	APC*	95% CI
All types	Both sexes	2003–2011	2.2†	0.3–4.1
	Male	2003–2011	3.5†	1.1–6.0
	Female	2003–2011	0.5	−2.5–3.6
Undifferentiated pleomorphic sarcoma	Both sexes	2003–2011	−0.7	−4.0–2.8
	Male	2003–2011	2.0	−2.9–7.1
	Female	2003–2011	−4.2†	−8.1 to −0.2
Liposarcoma	Both sexes	2003–2011	4.2†	0.5–8.1
	Male	2003–2011	5.3†	2.4–8.2
	Female	2003–2011	2.8	−3.2–9.0
Leiomyosarcoma	Both sexes	2003–2011	0.7	−3.5–5.1
	Male	2003–2011	−1.4	−4.4–1.7
	Female	2003–2011	2.5	−4.5–10.0
Rhabdomyosarcoma	Both sexes	2003–2011	4.4	−8.6–0.1
	Male	2003–2011	−5.0	−12.9–3.6
	Female	2003–2011	−3.2	−12.5–7.0
Angiosarcoma	Both sexes	2003–2011	6.4	−1.3–14.7
	Male	2003–2011	6.8	−5.4–20.5
	Female	2003–2011	6.2	−2.0–15.2
Synovial sarcoma	Both sexes	2003–2011	3.2	−1.8–8.4
	Male	2003–2011	6.5	−0.6–14.0
	Female	2003–2011	0.8	−6.8–9.1
Malignant nerve sheath tumor (including malignant schwannoma)	Both sexes	2003–2011	2.0	−8.0–13.1
	Male	2003–2011	8.7	−3.3–22.2
	Female	2003–2011	−4.4	−13.9–6.1
Peripheral primitive neuroectodermal tumor	Both sexes	2003–2011	4.3	−4.4–13.8
	Male	2003–2011	8.4	−2.6–20.6
	Female	2003–2011	−2.3	−13.1–10.0
Other specified sarcoma	Both sexes	2003–2011	4.4	−0.7–9.7
	Male	2003–2011	3.7†	1.3–6.1
	Female	2003–2011	5.6	−4.4–16.5
Sarcoma NOS	Both sexes	2003–2011	2.0	−8.0–13.2
	Male	2003–2011	4.0	−5.8–14.8
	Female	2003–2011	−0.4	−11.7–12.3
Other malignancy	Both sexes	2003–2011	0.4	−10.3–12.4
	Male	2003–2011	−3.2	−12.4–7.1
	Female	2003–2011	4.2	−8.9–19.0

APC = annual percent change; CI = confidence interval; NOS = not otherwise specified.
* The APC was calculated via weighted least-squares regression.
† Indicates statistical significance at the 0.05 level.

aged 75 to 79. The age-specific incidence of rhabdomyosarcoma (Figure 1F) revealed a bimodal distribution, showing a peak incidence in children and young adults and another peak amongst the elderly. The age-specific incidence of synovial sarcoma (Figure 1G) was increased in the adolescent population and remained stable throughout the ages from 15 to 79. The incidence of malignant nerve sheath tumor (Figure 1H) steadily increased since early childhood, and peaked in the elderly. In contrast to most subtypes of soft tissue cancers with incidences peaking in the elderly, pPNET (Figure 1I) was the most frequently diagnosed cancer in early childhood aged 0 to 4 years. The incidence was lowered thereafter and remained stable up to age 10 to 20 years. It became rarely found in those aged over 20 years but a slight increase in incidence was found in the elderly.

Temporal Trends

The annual changes in incidence of soft tissue cancers by histologic subtypes between 2003 and 2011 are demonstrated in Table 2 and Figure 2. There was a statistically significant change in the trend for all soft tissue cancers, with an average APC of 2.2% (95% CI, 0.3–4.1%, $P < 0.05$, Table 2). Specifically, the significant increasing trend was only found in males (APC, 3.5%; 95% CI, 1.1–6.0%, $P < 0.05$, Table 2 and Figure 2A). In terms of histologic subtypes, significant decreasing trend in incidence rates was only found in females diagnosed with UPS (APC, −4.2%, Figure 2B), whereas annual changes in incidences of liposarcoma (APC = 5.3%, Figure 2C) and other specified sarcoma (APC = 3.7%, Figure 2D) revealed significantly upward trends in males.

Geographical Variations

The incidences of primary soft tissue cancers calculated by 22 cities/counties during the 9-year study period are shown in Table 3. The dataset used for analyzing geographical variations in overall soft tissue cancers did not contain information for specific subtypes. Therefore, lymphoma and multiple myeloma were both included, giving a total case numbers of 4002 instead of 3843. The SRRs calculated for each city/county were also demonstrated in Table 3. Significance difference in the incidence was only found in 2 cities/counties. New Taipei City had a significantly higher rate (SRR = 1.11, 95% CI, 1.02–1.21, $P < 0.05$). By contrast, Penghu county (an offshore island) showed significantly lower rate (SRR, 0.60).

DISCUSSION

In this study, a total of 3843 subjects were diagnosed with soft tissue cancer during the study period in the TCR database, giving an ASR of 1.63 per 100,000 person-years. ASR of our patient population was lower than those reported by previous studies from Western countries (including all sites),[3–9] and slightly higher than that reported in Korea (extremity only).[12] The differences could be attributed to different tumor sites included and analyzed. Liposarcoma was the subtype with the highest incidence of STS in this study, but ranked third in most reports of Western countries.[3,8,9] Males had a significantly higher risk of angiosarcoma than females in Taiwanese population (M/F SRR, 1.79, $P < 0.01$). Similar results with a sex-specific difference in risk of angiosarcoma (ASR = 2.0) were shown in The Surveillance, Epidemiology, and End Results Program (SEER) of the National Cancer Institute of USA that investigated cutaneous STS.[23] Recently, several hot spots of genomic aberrations were found in angiosarcoma cells.[24–26] The association between the genetic changes and the susceptibility to angiosarcoma found in males deserves further investigation to elucidate.

For most of STS, the age-specific incidence increased with age, except a higher frequency of pPNET in children. The

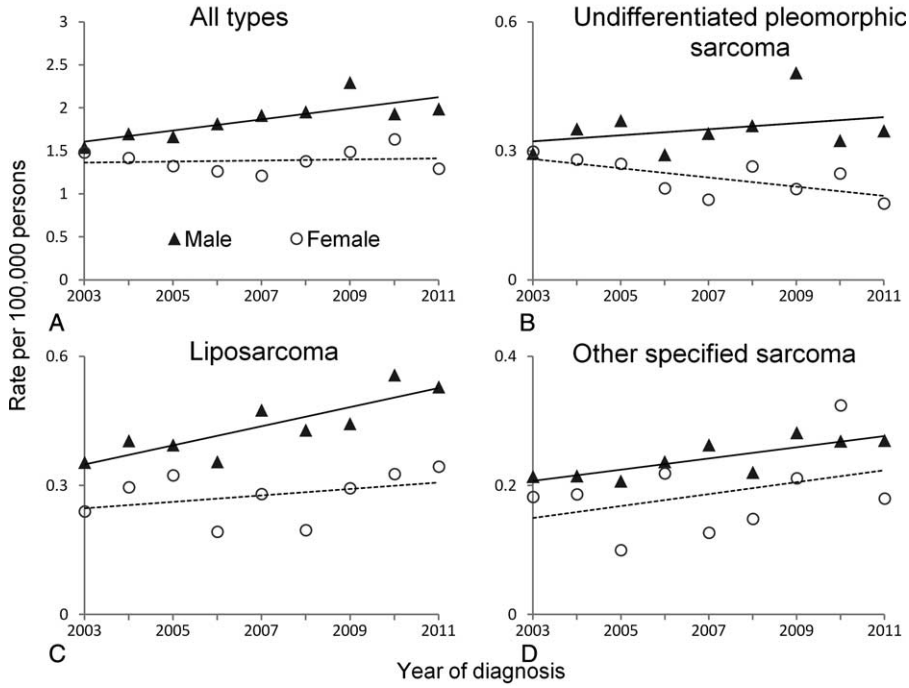

FIGURE 2. Temporal trends in the incidence rates of primary soft tissue cancers by sexes and histologic subtypes in Taiwan during 2003 to 2011.

TABLE 3. Incidence Rate (Per 100,000 Person-Yr) of Soft Tissue Cancers According to Cities and Counties, Taiwan (2003–2011)

City/County (n = 22)	*n*	%	Crude Rate	ASR*	95% CI	SRR†	95% CI
Northern Region							
Taipei City	526	13.14	2.23	1.79	1.63 –1.95	1.07	0.97 –1.18
New Taipei City	679	16.97	1.98	1.85‡	1.70–1.99	1.11	1.02–1.21
Taoyuan City	279	6.97	1.61	1.54	1.36–1.73	0.91	0.81–1.02
Keelung City	83	2.07	2.37	2.19	1.69–2.69	1.30	1.00–1.69
Hsinchu City	64	1.60	1.77	1.67	1.25–2.08	0.99	0.77–1.27
Hsinchu County	81	2.02	1.83	1.68	1.31–2.05	0.99	0.79–1.24
Yilan County	97	2.42	2.34	1.87	1.49–2.25	1.11	0.89–1.38
Miaoli County	110	2.75	2.18	1.77	1.43–2.11	1.05	0.86–1.28
Central Region							
Taichung City	407	10.17	1.74	1.65	1.48–1.81	0.97	0.88–1.08
Changhua County	228	5.70	1.93	1.69	1.46–1.91	1.00	0.87–1.14
Nantou County	87	2.17	1.81	1.47	1.14–1.80	0.87	0.70–1.07
Yunlin County	123	3.07	1.88	1.49	1.21–1.77	0.88	0.74–1.05
Southern Region							
Chiayi City	52	1.30	2.12	1.83	1.33–2.33	1.08	0.81–1.44
Chiayi County	97	2.42	1.96	1.56	1.23–1.89	0.92	0.75–1.13
Tainan City	335	8.37	1.99	1.67	1.48–1.85	0.99	0.88–1.11
Kaohsiung City	472	11.79	1.90	1.65	1.50–1.80	0.97	0.88–1.07
Pingtung County	155	3.87	1.94	1.60	1.34–1.85	0.94	0.80–1.10
Eastern Region							
Hualien County	62	1.55	2.00	1.62	1.20–2.04	0.96	0.74–1.24
Taitung County	43	1.07	2.03	1.56	1.08–2.03	0.92	0.69–1.24
Offshore Islands							
Penghu County	13	0.32	1.54	1.02§	0.44–1.60	0.60	0.39–0.94
Kinmen County	7	0.17	‖	‖		‖	
Lienchiang County	2	0.05	‖	‖		‖	
Total	4002	100	1.94	1.69	1.64–1.74		

The dataset used for geographical analysis included overall soft tissue cancers without exclusion of lymphoma/multiple myeloma.
ASR = age-standardized incidence rate; CI = confidence interval; SRR = standardized incidence rate (ASR) ratio.
* ASRs were age adjusted to the 2000 world standard population.
† SRR: ASR of the city/county vs. ASR of the rest area of Taiwan.
‡ Significantly higher than for comparison, $P < 0.05$.
§ Significantly lower than for comparison, $P < 0.05$.
‖ Indicates that the number of cases was less than 10 that did not permit evaluation of the rate.

incidence patterns of rhabdomyosarcoma revealed a bimodal age distribution. Increased rates comprised 2 clusters, 1 for embryonal and alveolar rhabdomyosarcoma in childhood and adolescence, and 1 for pleomorphic rhabdomyosarcoma in elderly population. These findings were comparable with the previous study from SEER.[8] In addition, the incidence of liposarcoma showed a downward trend for the population older than 75 to 79 years. Similar observations from the SEER report have indicated a decreased incidence in dedifferentiated liposarcoma may contribute to a lower frequency of liposarcoma in the population aged older than 75 to 80 years.[8]

Only few studies have been conducted to investigate the incidence patterns for synovial sarcoma and malignant nerve sheath tumor. Earlier studies have shown that synovial sarcoma was rarely seen in children.[27] Patient age at initial diagnosis distributed over a wide range (6–82 yr),[28] with the majority of patients presented between 15 and 40 years of age.[29] In this study, the incidence of synovial sarcoma increased in adolescents and remained stable throughout the ages of 15 to 79 years. This is consistent with previous single institution reports such as those from Memorial Sloan Kettering Cancer Center at USA (cases age ranged from 16 to 80)[30] or Rizzoli Orthopedic Institute at Bologna, Italy (more than 80% of cases diagnosed within 18–65 age range).[31] On the other hand, the published studies have shown that the peak incidence of malignant nerve sheath tumor was found in those aged 30 to 50 years.[32] However, patients suffering from malignant nerve sheath tumor with neurofibromatosis type 1 (NF1) were younger than those without NF1,[32,33] and about one third of those non-NF1 patients aged older than 60 years at diagnosis.[33] The present study demonstrated the evidences that an increased incidence of malignant nerve sheath tumor was found during the years of childhood and it was raised dramatically in the elderly. The results of our study were in line with previous observations, suggesting the high incidence in the elderly was most likely contributed by those patients without NF1. In combination, the multiple perspectives in the incidences of STS subtypes were associated with the complex nature of these diseases.

Nationwide trends of the overall STS incidence were inconsistent across countries. A SEER study from the US during 1978 to 2001 revealed that the ASRs significantly increased by 1.2% and 0.8% for Caucasian males and females but declined

slightly in the black population.[8] However, an Austrian population-based study for the period from 1984 to 2004 failed to demonstrate similar variations in the incidences of STS.[9] In this study, the incidence rates increased significantly from 2003 to 2011 in Taiwan, with an APC of 2.2%. Specifically, the trend was only found in males. In addition, sex-specific variations in APC were also found in different histologic subtypes of STS. In Taiwan, a significant downward trend in APC was only found in females with UPS, whereas the annual changes in the incidence of liposarcoma and other specified sarcoma significantly increased in males. Further investigations are required to elucidate the mechanisms underlying the racial and sex-specific differences and the association with histologic subtypes of STS.

Analysis of the geographic differences revealed a significantly higher incidence of STS in New Taipei City, the city with the highest population compared with the rest area of Taiwan. The relationships between the risk factors, such as pollution and life styles, and the increased incidence of STS also deserve further investigation. Despite that Penghu county had significantly lower rate in this study, the small population size and limited number of registered STS might influence the statistical significance of analysis.

There were several limitations in this study. First, the data acquired from the TCR were restricted to the STS of extremities and trunk wall. Second, due to the restriction of data application from TCR, further classifying of rhabdomyosarcoma and liposarcoma into different subtypes, which clearly have distinct clinical behaviors and cancer biology and might display variable incidence patterns, was not done in the current analysis. Third, the data of NF1-associated malignant nerve sheath tumor was not available. Finally, the different coding systems associated with changes in the allocation of registered data might contribute to effects on the results of incidence trends.

In addition, the acquired data stratified as ''other malignancy'' may not exclude the nonsarcoma subjects. However, the effect on the conclusion of this study was limited, because they only comprised less than 2.5% of the total number of STS.

CONCLUSIONS

To our best knowledge, this is the first report analyzing the population-based incidence of STS in Taiwan. Based on the data acquired from the nationwide TCR database, the incidence and temporal trends in Taiwanese population of all ages were elucidated. Age, sex, and histologic subtype-specific variations in the incidence rates of STS suggested complexity and diversity of this disease. The novel findings from this analysis include the upward trend of overall STS incidence in male, angiosarcoma has the most prominent male predilection, and a significant geographic variation with the higher incidence in New Taipei City. All of the mentioned above provide important information for additional research plans to elucidate the underlying risk factors associated with STS in Taiwan.

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