ORIGINAL ARTICLE

Incidence, distribution of histological subtypes and primary sites of soft tissue sarcoma in China

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ABSTRACT

Objective: Soft tissue sarcomas (STSs) are rare malignancies deriving from mesenchyme. In this study, we reported the epidemiology of STS in China using population-based cancer registry data.

Methods: In 2017, qualified data from 339 cancer registries were included in the national database. All STS cases were retrieved based on the morphological and topographical codes of International Classification of Diseases for Oncology, and were categorized into different histological subtypes and primary sites accordingly. Nationwide new STS cases were estimated using incidence rate of STS and the national population, and were reported for gastrointestinal stromal tumor (GIST) and STSs other than GIST separately by sex and region. Distribution of histological subtypes and primary sites of STS were calculated, as well as primary sites of GIST.

Results: Approximately 39,900 new STS cases occurred nationwide in China in 2014, accounting for 1.05% of overall cancer incidence. The crude incidence rate was 2.91/100,000 and generally increased with age. An overall female predilection was found. GIST was the most common histological subtype, followed by nerve sheath tumor and malignant peripheral nerve sheath tumor, leiomyosarcoma, liposarcoma, and fibrosarcoma. About 67.5% of GIST occurred in stomach while 1.4% were recorded outside the gastrointestinal tract. Connective, subcutaneous and other soft tissues were the most common primary site, of which extremities were the major subsite.

Conclusions: The burden of STS is not serious in China relatively. However, due to their histological and topographical complexity, STSs should not be unnoticed, and more basic and clinical studies should focus on STSs.

KEYWORDS
Soft tissue sarcoma; incidence; gastrointestinal stromal tumor; epidemiology; China

Introduction

Sarcomas, consisted of soft tissue sarcomas (STs) and bone sarcomas, are a heterogeneous group of mesenchymal malignancies that can develop at any age, comprising approximately 1% of all adult malignancies and 15% of pediatric malignancies. Compared with other common cancer types, STs can develop at almost any anatomical sites, and are more prone to occur at childhood. Although the etiology of STs is still unclear, known risk factors include inherited syndromes, chemical and radiation exposures, viral infections and genetic mutations.

Due to their rarity, STs are always outnumbered by carcinomas in many primary sites, therefore not reported in most researches categorizing cancers with the International Classification of Diseases. In United States, it is estimated that 13,040 new cases and 5,150 deaths were caused by STs in 2018, representing 0.75% of overall cancer incidence and 0.84% of overall cancer mortality. In Europe, nearly 23,600 new ST cases rose annually and the crude incidence rate was 4.7 per 100,000. In UK alone 3,300 new cases were diagnosed each year, with about 90 cases in children under 15 years old. So far, most national and subnational epidemiological researches on the burden of STs were carried out in developed countries.

Of all histological subtypes of STs, gastrointestinal stromal tumor (GIST) was categorized as ST in the 2013 World Health Organization (WHO) Classification. Compared with other common cancer types in gastrointestinal tract, GIST starts in different types of cells and needs different types of treatment. Therefore, GIST was either treated as an entity equal to STs and bone sarcomas, or reported...
separately from other STS subtypes\textsuperscript{18-20} in some researches. The crude incidence rate of GIST was 1.1–1.4 per 100,000 according to French and Spanish researches\textsuperscript{20,21}. Stomach is the most common site for GIST\textsuperscript{21} while there are some extremely rare tumors that arise outside the gastrointestinal tract but show the features of GIST. These tumors were therefore named extra-GIST\textsuperscript{22}. Similar to the condition of overall STSs, few researches on the epidemiology of GIST and extra-GIST were carried out in developing countries.

With world’s largest population, 23.7% of the global new cancer cases were estimated to occur in China in 2018\textsuperscript{23}. However, little is known about the incidence of STS in this country. In order to have a comprehensive understanding on the cancer burden of STS in China, in this study we described the distribution of histological subtypes and primary sites of STS using updated data from 339 population-based cancer registries. Estimation of the incidence of STS in the whole Chinese population was made as well and was reported for GIST and STSs other than GIST (non-GIST STSs) separately. We also described the distribution of GIST and explored the proportion of extra-GIST.

Materials and methods

Cancer registry data source

By 30th August 2017, 449 cancer registries from 31 provinces in China submitted cancer registry data for 2014 to National Central Cancer Registry of China (NCCRC). Local population data were provided along with the cancer data. After quality control based on the criteria of Chinese Guideline for Cancer Registration, data from 339 registries met the criteria and were finally included in this analysis. Among them, 129 were located in urban areas (covering a population of 144,061,915) and 210 were in rural areas (covering a population of 144,181,432). The overall population covered by these 339 cancer registries was 288,243,347 (146,203,891 men and 142,039,456 women), accounting for 21.07% of the national population that year.

All cancer cases were coded according to the International Classification of Diseases for Oncology, 3rd edition (ICD-O-3) and the International Statistical Classification of Diseases and Related Health Problems 10th Revision (ICD-10). Of all STS cases, the proportion of morphology verified cases (MV%) was 84.97%. More information on the national cancer registry program and data quality control procedure could be found elsewhere\textsuperscript{24,25}.

Data extraction, classification and statistical analysis

STS cases in the 339 registries were retrieved and categorized into different histological groups according to their ICD-O-3 morphological codes as shown in Table 1 based on a previous study\textsuperscript{12}. The primary site of a case was defined by the ICD-O-3 topographical code. Cases with topographical codes being C40-C41 were deleted to exclude sarcomas in bones, joints and articular cartilages.

Crude incidence rates were calculated by area (urban/rural), sex (male/female) and age group (0, 1–4, 5 to 80 by 5 years, 85+) based on the retrieved cases. To estimate the number of new STS cases in the whole country, incidence rates stratified by area, sex and age group were multiplied by corresponding nationwide population in each stratum. National population data were obtained from Nation Bureau of Statistics. Chinese population in 2000 and World Segi’s population were used for age-standardization.

The estimated nationwide incidence was reported for overall STSs, GIST, and non-GIST STSs separately. The distribution of histological subtypes was reported by sex and age group (0–19, 20–64, and over 65 years old) in order to reveal the sex and age predilection. Primary sites of all STS cases were classified into 14 groups and reported by sex.

Results

Incidence of soft tissue sarcoma

Approximately 39,900 new STS cases (19,200 men and 20,700 women) occurred nationwide China in 2014, as shown in Table 2. The crude incidence rate was 2.91/100,000 (2.72/100,000 in men and 3.11/100,000 in women), while the age-standardized rate by Chinese population (ASRcn) and world population (ASRwld) were 2.21 and 2.13 per 100,000, respectively.

There were 5,700 GIST cases (3,200 men and 2,500 women) nationwide in 2014, accounting for about 14.3% of the overall STSs. The crude incidence rate was 0.42/100,000 (0.45/100,000 in men and 0.38/100,000 in women), while the age-standardized rate by Chinese population (ASRcn) and world population (ASRwld) were 2.21 and 2.13 per 100,000, respectively.

On the other hand, approximately 34,200 non-GIST cases (16,000 men and 18,200 women) were diagnosed, accounting for about 85.7% of the overall STSs. The crude incidence rate was 2.50/100,000 (2.27/100,000 in men and 2.73/100,000 in women). The incidence of overall STSs and non-GIST STSs were higher in women than in men in urban and rural areas, whereas the incidence of GIST was higher in men than in women in both areas.
As shown in Figure 1, the age-specific incidence rates of overall STSs and non-GIST STSs were the lowest in children aged 5 to 9 years, increased dramatically after 35 years old and peaked at 75–79 years old in women and 80–84 years old in men for overall STSs, and 80-84 years old in both sexes for non-GIST STSs. As for GIST, the age-specific incidence rate was unstable due to the scarcity in individuals under 25 years old. The rate increased dramatically after 45 years old and peaked at 75–79 years old in women and 70–74 years old in men.

**Distribution of histological subtypes**

As shown in Table 3, altogether 8,475 new STS cases (4,033 men and 4,442 women) were reported from the 339 cancer registries. The most common histological subtype was GIST (14.38%), followed by nerve sheath tumor and malignant peripheral nerve sheath tumor (MPNST) (12.48%), leiomyosarcoma (5.97%), liposarcoma (5.75%), and fibrosarcoma (5.73%).

Overall male/female ratio was 0.9, indicating a female predilection. Highest ratios were found in malignant myoepithelioma, clear cell sarcoma, malignant fibrous histiocytoma and primitive neuroectodermal tumor NOS, whereas lowest ratios were found in endometrial stromal sarcoma, granular cell tumors and alveolar soft part sarcoma, leiomyosarcoma, carcinosarcoma NOS, and myxosarcoma.
Children and adolescents under 20 years old accounted for 3.12% of all STS cases, while adults aged 20-64 years and over 65 years accounted for 64.64% and 32.24%, respectively. In major histological subtypes, children and adolescents represented 44.63% of all rhabdomyosarcoma cases, whereas endometrial stromal sarcoma (84.13%) and nerve sheath tumor and MPNST (80.72%) occurred mainly in adults aged 20 to 64 years, and proportions of malignant fibrous histiocytoma (47.93%) and GIST (43.07%) were relatively high in elders above 65 years old.

### Distribution of primary sites

As shown in Table 4, more than 22% of STSs were located in the connective, subcutaneous and other soft tissues, of which 574 were in lower limb and hip (lower extremities), 234 were in upper limb and shoulder (upper extremities). About one-fifth of STSs occurred in digestive organs, of which 952 were in stomach, 294 were in small intestine, and 153 were in liver. Retroperitoneum and peritoneum and eye, brain and other parts of the central nervous system both accounted for nearly 14% of all STSs.

### Table 2 Estimated number of nationwide new cases of soft tissue sarcomas in China, 2014

| Areas          | Gender | Number of new cases | Incidence rate (1/100,000) | ASRcn (1/100,000) | ASRwld (1/100,000) |
|---------------|--------|---------------------|-----------------------------|-------------------|-------------------|
| Overall STS   | All areas | Total               | 39,900                      | 2.91              | 2.21              | 2.13              |
|               | Male    | 19,200              | 2.72                        | 2.10              | 2.05              |
|               | Female  | 20,700              | 3.11                        | 2.32              | 2.23              |
| Urban areas   | Total   | 26,600              | 3.55                        | 2.55              | 2.47              |
|               | Male    | 12,900              | 3.38                        | 2.47              | 2.40              |
|               | Female  | 13,700              | 3.71                        | 2.65              | 2.54              |
| Rural areas   | Total   | 13,300              | 2.14                        | 1.74              | 1.69              |
|               | Male    | 6,300               | 1.94                        | 1.61              | 1.58              |
|               | Female  | 7,000               | 2.36                        | 1.89              | 1.81              |
| GIST          | All areas | Total               | 5,700                       | 0.42              | 0.29              | 0.29              |
|               | Male    | 3,200               | 0.45                        | 0.32              | 0.32              |
|               | Female  | 2,500               | 0.38                        | 0.26              | 0.25              |
| Urban areas   | Total   | 3,900               | 0.52                        | 0.34              | 0.34              |
|               | Male    | 2,200               | 0.57                        | 0.38              | 0.38              |
|               | Female  | 1,700               | 0.47                        | 0.31              | 0.30              |
| Rural areas   | Total   | 1,800               | 0.29                        | 0.22              | 0.21              |
|               | Male    | 1,000               | 0.30                        | 0.24              | 0.23              |
|               | Female  | 800                 | 0.28                        | 0.20              | 0.19              |
| Non-GIST STS  | All areas | Total               | 34,200                      | 2.50              | 1.92              | 1.85              |
|               | Male    | 16,000              | 2.27                        | 1.78              | 1.73              |
|               | Female  | 18,200              | 2.73                        | 2.06              | 1.97              |
| Urban areas   | Total   | 22,700              | 3.03                        | 2.21              | 2.13              |
|               | Male    | 10,700              | 2.81                        | 2.09              | 2.02              |
|               | Female  | 12,000              | 3.25                        | 2.34              | 2.25              |
| Rural areas   | Total   | 11,500              | 1.85                        | 1.52              | 1.48              |
|               | Male    | 5,300               | 1.64                        | 1.37              | 1.35              |
|               | Female  | 6,200               | 2.09                        | 1.69              | 1.61              |

STS, soft tissue sarcoma; GIST, gastrointestinal stromal tumor; ASRcn, Age–standardized incidence rate by China Population; ASRwld, Age–standardized incidence rate by Segi’s Population

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As shown in Table 5, of all GIST cases, 98.6% arose in digestive organs. Stomach was the most common site (67.51%), followed by small intestine (20.92%), other digestive organs (4.59%) and rectum (3.12%). Extra-GIST was found in retroperitoneum and peritoneum, bronchus and lung, soft tissues, thyroid gland, and other unknown sites, representing 1.4% of all GIST cases.

**Discussion**

In this study, using population-based cancer registry data, we found that nearly 39,900 new STS cases (5,700 GIST and 34,200 non-GIST) were diagnosed nationwide in China in 2014, representing 1.05% (0.90% in male, 1.23% in female) of all cancer incidence\(^{24}\), larger than the proportion (0.75%)...
Crude incidence rates of STSs and GIST were 2.91 and 0.42 per 100,000 respectively, lower than the corresponding rates in western countries (4.7 and 0.78 per 100,000).

Incidence of STS in China generally increased with age. The rate in Austria showed similar trend but peaked and dropped at younger ages. Unlike carcinomas, STSs comprise a large proportion of pediatric malignancies and are an important cause of death in adolescents and young adults. In our study, 3.1% of all STSs occurred in individuals aged 0 to 19 years, whereas in United States and Japan, corresponding proportions were 5.6% and 6.7%, respectively. Rhabdomyosarcoma was the only entity with a median age < 20 years in United States. It was also the most common subtype in people under 20 years old in these three studies.

| Histological subtypes                                      | Proportion (%) | Gender | Male/female ratio | 0-19 years old (%) | 20-64 years old (%) | 65+ years old (%) |
|------------------------------------------------------------|----------------|--------|-------------------|--------------------|--------------------|-------------------|
| Sarcoma NOS                                                | 28.65          | 2,428  | 1.0               | 3.50               | 54.08              | 42.42             |
| Gastrointestinal stromal tumor                            | 14.38          | 1,219  | 1.2               | 0.41               | 56.52              | 43.07             |
| Leiomyosarcoma                                             | 5.97           | 506    | 0.2               | 0.40               | 79.05              | 20.55             |
| Endometrial stromal sarcoma                               | 4.46           | 378    | 0.1               | 0.53               | 84.13              | 15.34             |
| Liposarcoma                                                | 5.75           | 487    | 1.4               | 2.26               | 66.74              | 31.00             |
| Malignant fibrous histiocytoma                             | 5.12           | 434    | 1.6               | 0.92               | 51.15              | 47.93             |
| Angiosarcoma                                               | 4.59           | 389    | 1.2               | 3.34               | 59.64              | 37.02             |
| Rhabdomyosarcoma                                           | 1.43           | 121    | 1.1               | 44.63              | 47.93              | 7.44              |
| Fibrosarcoma                                               | 5.73           | 486    | 1.0               | 2.67               | 72.22              | 25.10             |
| Nerve sheath tumor and malignant peripheral nerve sheath tumor | 12.48          | 1,058  | 1.0               | 2.74               | 80.72              | 16.54             |
| Dermatofibrosarcoma                                        | 2.28           | 193    | 1.4               | 6.74               | 78.24              | 15.03             |
| Other specified soft tissue sarcoma                        |                |        |                   |                    |                    |                   |
| Carcinosarcoma, NOS                                        | 1.58           | 134    | 0.4               | 0.00               | 61.19              | 38.81             |
| Synovial sarcoma                                            | 1.18           | 100    | 1.0               | 4.00               | 84.00              | 12.00             |
| Mixed tumor, malignant NOS                                 | 0.86           | 73     | 0.7               | 4.11               | 72.60              | 23.29             |
| Primitive neuroectodermal tumor, NOS                       | 0.55           | 47     | 1.6               | 14.89              | 76.60              | 8.51              |
| Granular cell tumors and alveolar soft part sarcoma        | 1.16           | 98     | 0.2               | 6.12               | 84.69              | 9.18              |
| Paragangliomas and glomus tumors                           | 1.13           | 96     | 0.9               | 3.13               | 79.17              | 17.71             |
| Malignant mesenchymoma                                     | 0.88           | 75     | 1.0               | 2.67               | 68.00              | 29.33             |
| Malignant myoepithelioma                                   | 0.17           | 14     | 2.5               | 0.00               | 64.29              | 35.71             |
| Clear cell sarcoma                                         | 0.34           | 29     | 1.6               | 6.90               | 72.41              | 20.69             |
| Kaposi sarcoma                                             | 0.59           | 50     | 1.4               | 4.00               | 58.00              | 38.00             |
| Rhabdoid tumor                                             | 0.08           | 7      | 1.3               | 28.57              | 57.14              | 14.29             |
| Extraskeletal osteosarcoma and chondrosarcoma              | 0.26           | 22     | 1.4               | 0.00               | 90.91              | 9.09              |
| Myxosarcoma                                                | 0.22           | 19     | 0.6               | 0.00               | 57.89              | 42.11             |
| Malignant giant cell tumors                                | 0.14           | 12     | 1.0               | 16.67              | 58.33              | 25.00             |
| Total                                                      | 100.00         | 8,475  | 0.9               | 3.12               | 64.65              | 32.24             |

NOS, not otherwise specified

in United States. Crude incidence rates of STSs and GIST were 2.91 and 0.42 per 100,000 respectively, lower than the corresponding rates in western countries (4.7 and 0.78 per 100,000).

Incidence of STS in China generally increased with age. The rate in Austria showed similar trend but peaked and dropped at younger ages. Unlike carcinomas, STSs comprise a large proportion of pediatric malignancies and are an important cause of death in adolescents and young adults. In our study, 3.1% of all STSs occurred in individuals aged 0 to 19 years, whereas in United States and Japan, corresponding proportions were 5.6% and 6.7%, respectively. Rhabdomyosarcoma was the only entity with a median age < 20 years in United States. It was also the most common subtype in people under 20 years old in these three studies.
Unlike most carcinomas, to which men are prone to develop, contradictions remained as for the major gender of STS patients. In Europe, there were more female patients, while in Korea and Taiwan China, results showed the opposite. Similar to these Asian results, men outnumbered women in most primary sites and histological subtypes in our study. However, there were more female patients due to the numerous cases in breast and female genital organs, and subtypes like leiomyosarcoma, which occurs mainly in genital organs, bladder and uterus.

GIST was the most common histological subtype in Chinese population, as was found in Europe. In France, 16.7% of the identified sarcoma cases were GIST. French distribution of GIST was similar to our study, with 61% and 27% arose in stomach and small intestine, respectively. The proportion of extra-GIST nevertheless was greater because...
5% of GIST arose in peritoneum. This extravagant percentage can be partly explained by the small sample size. In contrast to our and American results, GIST showed a female predilection in France. In United States, there were relatively less GIST in stomach (55.3%) and more in small intestine (28.7%), while the proportion of extra-GIST was 3.0%, also larger than our result.

Kaposi sarcoma, accounting for less than 0.6% of all STSs, was only a minor subtype in China and even less common (0.2%) in Japan, whereas it had very high incidence in African countries. Kaposi sarcoma is also the most common subtype (31.7%) in United States. This could be explained by the relatively low prevalence of HIV in East Asia because Kaposi sarcoma is strongly related with immune suppression.

Other than Kaposi sarcoma, common subtypes in United States were fibrohistiocytic tumors (30.4%) and leiomyosarcoma (27.4%). Even after the inclusion of GIST into the leiomyosarcoma category, corresponding proportions in our study, which were 5.12% and 20.35% respectively, were still smaller than these results. In Japan, leiomyosarcoma (28.1%) was the most prevalent subtype, followed by liposarcoma (9.6%), and malignant fibrous histiocytoma (9.0%). In contrast, nerve sheath tumor and MPNST is far more prevalent in China, representing more than 12% of all STSs in our study, compared with 3.5% in Japan and 4.6% in United States.

In terms of primary sites, according to American Cancer Society, STSs can develop in soft tissues, internal organs as well as retroperitoneum. However, the WHO classification did not include internal organs as primary sites for STSs and listed only the extremities, trunk wall, and retroperitoneum. Therefore, STSs were recognized as an entity with a variety of primary sites in some studies, while in others, STSs were more confined and sarcomas in internal organs were categorized as visceral sarcoma separately.

In this study, we used the classification of American Cancer Society, and found that connective, subcutaneous and other soft tissues was the most common site. Delving into more specific topographic categories, lower extremity was the most common subsite, and the number of cases in lower extremity was 2–3 times of that in upper extremity. Results in Korea, Japan, and Europe further proved ours. Digestive organs, which was recognized as visceral sarcomas in some studies, were the second most common site for STSs, of which the majority was GIST. We found 17 extremely rare extra-GIST cases based on this large population-based study. This will provide valuable epidemiological evidence for further researches.

STSs have more than 70 histological subtypes. Together with the various primary sites, they make up a large number of combinations that vary tremendously in terms of survival.

### Table 5 Distribution of primary sites of gastrointestinal stromal tumor by sex in China, 2014

| ICD-O-3 code | Primary sites                                      | Gender | Proportion (%) |
|-------------|---------------------------------------------------|--------|----------------|
|             |                                                   | Male, n | Female, n | Total, n |
| C15         | Esophagus                                         | 6      | 3     | 9     | 0.74  |
| C16         | Stomach                                           | 444    | 379   | 823   | 67.51 |
| C17         | Small intestine                                   | 141    | 114   | 255   | 20.92 |
| C18         | Colon                                             | 10     | 9     | 19    | 1.56  |
| C20         | Rectum                                            | 26     | 12    | 38    | 3.12  |
| C21         | Anus and anal canal                               | 1      | 0     | 1     | 0.08  |
| C25         | Pancreas                                          | 1      | 0     | 1     | 0.08  |
| C26         | Other digestive organs                            | 29     | 27    | 56    | 4.59  |
| C34         | Bronchus and lung                                 | 0      | 1     | 1     | 0.08  |
| C48         | Retroperitoneum and peritoneum                    | 5      | 1     | 6     | 0.49  |
| C49         | Connective, subcutaneous and other soft tissues   | 2      | 0     | 2     | 0.16  |
| C73         | Thyroid gland                                     | 1      | 0     | 1     | 0.08  |
| C76         | Other and ill-defined sites                       | 3      | 3     | 6     | 0.49  |
| C80         | Unknown primary site                              | 0      | 1     | 1     | 0.08  |
| Total       |                                                   | 669    | 550   | 1,219 | 100.00|
Conflict of interest statement

No potential conflicts of interest are disclosed.

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Cite this article as: Yang Z, Zheng R, Zhang S, Zeng H, Li H, Chen W. Incidence, distribution of histological subtypes and primary sites of soft tissue sarcoma in China. Cancer Biol Med. 2019; 16: 565-74. doi: 10.20892/j.issn.2095-3941.2019.0041