Epidemiological characteristics of 1385 primary sacral tumors in one institution in China

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Abstract

Background: Primary sacral tumors (PST) are a rare group of skeletal tumors that can affect children and adults of all ages. Little is known about the epidemiological characteristics of PST in China.

Methods: 1385 patients with primary sacral tumors, which had the clinical record at our bone tumor center from 2000 to November 2018 were analyzed.

Results: 51.7% (716 cases) were malignant and 48.3 % (669 cases) were benign. Of malignant tumors, chordoma was the most common malignant tumor (316 cases, 22.8% of all tumors), followed by chondrosarcoma, myeloma and other histologic types. The most common histological type of benign tumors was giant cell tumor accounting for 14.8% (205 cases) of all tumors, followed by neurofibroma, schwannoma and other types. The most common age group affected by malignant bone tumors was the 51- to 60-year-old group, followed by the 41- to 50-year-old group. The most commonly affected age group for benign tumors was the 31- to 50-year-old group, followed by the 21- to 30-year old group. Furthermore, the following histologic types had the gender predilection. Chordoma, chondrosarcoma, myeloma and osteosarcoma of PST affected more frequently males than females. Malignant peripheral nerve sheath tumor, lymphoma, giant cell tumor, neurofibroma, tuberculosis, teratoma and epidermoid cyst more frequently affected females than males.

Conclusions: The large cohort of primary sacral tumors in our database may reveal their epidemiologic characteristics of primary sacral malignant and benign tumors in China and epidemiological feature of PST is fairly distinct from the mobile spine and extremities.

Introduction

Primary sacral tumors (PST) are a rare group of skeletal tumors, that can affect children and adults of all ages. A great number of specific histologic subtypes of PST have been delineated, each of them with unique appearance and biological potential[1, 2]. A barrier is presented to etiologic study due to the rarity and heterogeneity of PST and it poses a great challenge in the understanding of incidence patterns[3]. Little is known about the epidemiology of PST in China. China is a relatively big country with a steady population and the number of patients with PST is huge and our hospital is a biggest specialized musculoskeletal tumor cencer in China. This provide us a beneficial condition to study the epidemiological characteristics of primary sacral tumors. The present study is aiming to provide the first epidemiological analysis of PST in the Chinese population between 2000 and 2018, specifically to describe epidemiologic characteristics of PST by gender, age and subtype of PST.

Patients And Methods

We retrospectively reviewed 1385 patients with primary sacral tumor, which had the clinical record at our bone tumor center from 2000 to November 2018. There were 709 male and 676 female patients with a mean age of 43.9 ± 17.0 years (range, 2–86 years). Histologic diagnosis was confirmed by biopsy or operative specimen. The inclusion criteria for the present study were as follows: (1) Patients had the definitive histological diagnosis; (2) Diagnostic time was from the year of 2000 to 2018. The exclusion criteria were as follows: (1) Without confirmed histological diagnosis; (2) Diagnostic time was beyond the range of 2000 to 2018. All patients which were included in the present study were gave written informed consent for their data to be included in this study during the follow-up. All data were obtained from the clinical and radiograph records. This study was approved by the Institutional Review Board/Ethics Committee of the authors' institution. Following data were collected in the present study: age, gender, affected sacrum level, pathological diagnosis.

Statistical analysis

Continuous variables were summarized with means and ranges; categorical variables were summarized with frequency counts and percentages. The Student's t test was used to compare the age in different histologic types of male and female in Table-5&6. The SPSS software (version 19.0; SPSS Inc, Chicago, IL, USA) was used for all statistical analyses. P ≦ 0.05 indicated a statistically significant difference.

Results

Incidence of histological subtypes of PST

The histological types of primary sacral malignant and benign tumors were listed in Table-1&2. 51.7% (716 cases) were malignant and 48.3% (669 cases) were benign (Figure-1). Of malignant tumors, chordoma was the most common malignant type of PST (22.8% of all PST, 316 cases), followed by chondrosarcoma (5.3%, 74 cases), myeloma (3.8%, 53 cases), malignant peripheral nerve sheath tumor (3.4%, 47 cases), Ewing sarcoma (3.4%, 47 cases), osteosarcoma (2.9%, 40 cases), lymphoma (2.5%, 35 cases), spindle cell undifferentiated sarcoma (1.2%, 17 cases), malignant giant cell tumor (1.2%, 17 cases), malignant teratoma (0.8%, 11 cases) and other malignant types (Table-1). The most common histological type of benign tumors was giant cell tumor accounting for 14.8% (205 cases) of all PST, followed by neurofibroma (11.2%, 155 cases), schwannoma (8.6%, 119 cases), tuberculosis (1.7%, 24 cases), teratoma (1.7%, 23 cases), sacral canal cysts (1.6%, 22 cases), epidermoid cyst (1.5%, 21 cases), hemangioma (1.0%, 14 cases), meningeal cysts (0.9%, 13cases), primary aneurysmal bone cyst (0.9%, 12 cases), fibrous dysplasia (0.6%, 9 cases), osteoblastoma (0.6%, 9 cases) and other types shown in Table-2. The top six of primary sacral tumors were summarizd in Figure-2.

Table 1
Histopathological diagnosis of 716 patients with primary sacral malignant tumors in 1385 patients

Primary malignant tumor	No in 716 patients with PMT (%)	No in 1385 patients (51.7%)
Chordoma	316 (44.1%)	316 (22.8%)
Chondrosarcoma	74 (10.3%)	74 (5.3%)
Myeloma	53 (7.4%)	53 (3.8%)
Malignant peripheral nerve sheath tumor	47 (6.6%)	47 (3.4%)
Ewing sarcoma	47 (6.6%)	47 (3.4%)
Osteosarcoma	40 (5.6%)	40 (2.9%)
Lymphoma	35 (4.9%)	35 (2.5%)
Spindle cell undifferentiated sarcoma	17 (2.4%)	17 (1.2%)
Malignant giant cell tumor	17 (2.4%)	17 (1.2%)
Malignant teratoma	11 (1.5%)	11 (0.8%)
Solitary fibrous tumor	11 (1.5%)	11 (0.8%)
Liposarcoma	10 (1.4%)	10 (0.7%)
Hemangiopericytom	9 (1.3%)	9 (0.6%)
Ependymoma	4 (0.6%)	4 (0.3%)
Angiosarcoma	3 (0.4%)	3 (0.2%)
Fibrosarcoma	3 (0.4%)	3 (0.2%)
Hemangioendothelima	3 (0.4%)	3 (0.2%)
Myelocytic sarcoma	3 (0.4%)	3 (0.2%)
Yolk sac tumor	3 (0.4%)	3 (0.2%)
Leiomyosarcoma	2 (0.3%)	2 (0.1%)
Alveolar soft part sarcoma	2 (0.3%)	2 (0.1%)
Leukemia	2 (0.3%)	2 (0.1%)
Epithelioid sarcoma	1 (0.1%)	1 (0.07%)
Granulocyte sarcoma	1 (0.1%)	1 (0.07%)
Myofibroblastic sarcoma	1 (0.1%)	1 (0.07%)
Synovial sarcoma	1 (0.1%)	1 (0.07%)

PMT, primary malignant tumor

Table 2
Histopathological diagnosis of 669 patients with primary sacral benign or tumor-like lesions in 1385 patients

Primary benign tumor	No in 669 patients with PBT (%)	No in 1385 patients (48.3%)
Giant cell tumor	205 (30.6%)	205 (14.8%)
Neurofibroma	155 (23.2%)	155 (11.2%)
Schwannoma	119 (17.8%)	119 (8.6%)
Tuberculosis	24 (3.6%)	24 (1.7%)
Teratoma	23 (3.4%)	23 (1.7%)
Sacral canal cysts	22 (3.3%)	22 (1.6%)
Epidermoid cyst	21 (3.1%)	21 (1.5%)
Hemangioma	14 (2.1%)	14 (1.0%)
Meningeal cysts	13 (1.9%)	13 (0.9%)
Primary aneurysmal bone cyst	12 (1.8%)	12 (0.9%)
Fibrous dysplasia	9 (1.3%)	9 (0.6%)
Osteoblastoma	9 (1.3%)	9 (0.6%)
Eosinophilic granuloma	7 (1.0%)	7 (0.5%)
Simple bone cyst	5 (0.7%)	5 (0.4%)
Spinal meningioma	5 (0.7%)	5 (0.4%)
Gut-tail cyst	4 (0.6%)	4 (0.3%)
Benign fibrous histiocytoma	4 (0.6%)	4 (0.3%)
Fibromatosis	3 (0.4%)	3 (0.2%)
Diffuse giant cell tumor of tendon sheath	3 (0.4%)	3 (0.2%)
Chondroblastoma	3 (0.4%)	3 (0.2%)
Lipoma	2 (0.3%)	2 (0.1%)
Phosphouria stromal tumor	2 (0.3%)	2 (0.1%)
Osteoidosteoma	2 (0.3%)	2 (0.1%)
Liomyoma	1 (0.1%)	1 (0.07%)
Paget disease	1 (0.1%)	1 (0.07%)
Osteochondroma	1 (0.1%)	1 (0.07%)
PBT, primary benign tumor		

Age Distribution of Pst

For a total of 1385 primary sacral tumors, the mean age was 43.9 ± 17.0 years (range, 2–86 years), 51.2% (709 cases) of the tumors occurred in males and 48.8% (676 cases) in females, with a mean age of 44.5 ± 17.2 and 43.1 ± 16.7 years. The mean age of 716 malignant and 669 benign tumors group were respectively 46.9 ± 17.5 years (range, 2–86 years) and 40.6 ± 15.7 years (range, 2–85 years).

The incidences were presented separately by gender (males and females) and age at diagnosis was grouped into nine subgroup (from 0 to 10 years cohort to ≥ 80 years cohort). The most common age group affected by malignant bone tumors was the 51- to 60-year-old group (21.1%, 151cases), followed by the 41- to 50-year-old group (19.4%, 139 cases) (Table-3). The most commonly affected age group for benign tumors were the 31- to 40-year-old and 41- to 50-year-old groups (21.1%, 141 cases and 21.1%, 141 cases), followed by the 21- to 30-year old group (19.6%, 131 cases) (Table-4).

Table 3
Age distribution of primary sacral malignant tumors

Histology	0–10	11–20	21-30	31-40	41-50	51-60	61-70	71-80	81-90	Total
Primary malignant tumor	9(1.3%)	54(7.5%)	88(12.3%)	96(13.4%)	139(19.4%)	151(21.1%)	122(17.0%)	52(7.3%)	5(0.7%)	716
Chordoma	0(0)	1(0.3%)	17(5.4%)	29(9.2%)	58(18.4%)	85(26.9%)	82(25.9%)	39(12.3%)	5(1.6%)	316
Chondrosarcoma	0(0)	5(6.8%)	14(18.9%)	16(21.6%)	19(25.7%)	16(21.6%)	4(5.4%)	0(0)	0(0)	74
Myeloma	0(0)	0(0)	2(3.8%)	7(13.2%)	13(24.5%)	13(24.5%)	12(22.6%)	6(11.3%)	0(0)	53
Malignant peripheral nerve sheath tumor	1(2.1%)	5(10.6%)	10(21.3%)	5(10.6%)	11(23.4%)	9(19.1%)	5(10.6%)	1(2.1%)	0(0)	47
Ewing sarcoma	5(10.6%)	19(40.4%)	9(19.1%)	9(19.1%)	4(8.5%)	1(2.1%)	0(0)	0(0)	0(0)	47
Osteosarcoma	1(2.5%)	15(37.5%)	13(32.5%)	5(12.5%)	3(7.5%)	3(7.5%)	0(0)	0(0)	0(0)	40
Lymphoma	0(0)	2(5.7%)	4(11.4%)	5(14.3%)	9(25.7%)	9(25.7%)	4(11.4%)	2(5.7%)	0(0)	35
Spindle cell undifferentiated sarcoma	0(0)	0(0)	1(5.9%)	2(11.8%)	3(17.6%)	5(29.4%)	4(23.5%)	2(11.8%)	0(0)	17
Malignant giant cell tumor	0(0)	2(11.8%)	6(35.3%)	4(23.5%)	2(11.8%)	2(11.8%)	1(5.9%)	0(0)	0(0)	17
Malignant teratoma	0(0)	0(0)	2(18.2%)	4(36.4%)	1(9.1%)	2(18.2%)	2(18.2%)	0(0)	0(0)	11
Solitary fibrous tumor	0(0)	0(0)	2(18.2%)	2(18.2%)	2(18.2%)	2(18.2%)	3(27.3%)	0(0)	0(0)	11
Liposarcoma	0(0)	0(0)	0(0)	2(20.0%)	4(40.0%)	1(10.0%)	3(30.0%)	0(0)	0(0)	10
Hemangiopericytom	0(0)	0(0)	2(22.2%)	3(33.3%)	2(22.2%)	0(0)	1(11.1%)	1(11.1%)	0(0)	9
Ependymoma	0(0)	0(0)	1(25.0%)	0(0)	2(50.0%)	0(0)	0(0)	1(25.0%)	0(0)	4
Angiosarcoma	0(0)	0(0)	0(0)	0(0)	1(33.3%)	2(67.7%)	0(0)	0(0)	0(0)	3
Fibrosarcoma	0(0)	1(33.3%)	0(0)	0(0)	2(67.7%)	0(0)	0(0)	0(0)	0(0)	3
Hemangioendothelima	1(33.3%)	0(0)	0(0)	1(33.3%)	0(0)	0(0)	1(33.3%)	0(0)	0(0)	3
Myelocytic sarcoma	0(0)	0(0)	1(33.3%)	2(67.7%)	0(0)	0(0)	0(0)	0(0)	0(0)	3
Yolk sac tumor	1(33.3%)	2(67.7%)	0(0)	0(0)	0(0)	0(0)	0(0)	0(0)	0(0)	3
Leiomyosarcoma	0(0)	0(0)	1(50.0%)	0(0)	1(50.0%)	0(0)	0(0)	0(0)	0(0)	2
Alveolar soft part sarcoma	0(0)	0(0)	2(100.0%)	0(0)	0(0)	0(0)	0(0)	0(0)	0(0)	2
Leukemia	0(0)	1(50.0%)	0(0)	0(0)	1(50.0%)	0(0)	0(0)	0(0)	0(0)	2
Epithelioid sarcoma	0(0)	1(100.0%)	0(0)	0(0)	0(0)	0(0)	0(0)	0(0)	0(0)	1
Granulocyte sarcoma	0(0)	0(0)	0(0)	0(0)	1(100.0%)	0(0)	0(0)	0(0)	0(0)	1
Myofibroblastic sarcoma	0(0)	0(0)	0(0)	0(0)	0(0)	1(100.0%)	0(0)	0(0)	0(0)	1
Synovial sarcoma	0(0)	0(0)	1(100.0%)	0(0)	0(0)	0(0)	0(0)	0(0)	0(0)	1

Table 4
Age distribution of primary sacral benign or tumor-like lesions

Histology	0–10	11–20	21-30	31-40	41-50	51-60	61-70	71-80	81-90	Total
Primary benign tumor	11(1.6%)	58(8.7%)	131(19.6%)	141(21.1%)	141(21.1%)	108(16.1%)	60(9.0%)	16(2.4%)	3(0.4%)	669
Giant cell tumor	0(0)	24(11.7%)	61(29.8%)	62(30.2%)	40(19.5%)	12(5.9%)	6(2.9%)	0(0)	0(0)	205
Neurofibroma	1(0.6%)	6(3.9%)	18(11.6%)	24(15.5%)	39(25.2%)	41(26.5%)	20(12.9%)	5(3.2%)	1(0.6%)	155
Schwannoma	0(0)	2(1.7%)	16(13.4%)	24(20.2%)	33(27.7%)	24(20.2%)	16(13.4%)	4(3.4%)	0(0)	119
Tuberculosis	1(4.2%)	2(8.3%)	4(16.7%)	5(20.8%)	4(16.7%)	4(16.7%)	1(4.2%)	3(12.5%)	0(0)	24
Teratoma	1(4.3%)	3(13.0%)	8(34.8%)	6(26.1%)	3(13.0%)	1(4.3%)	1(4.3%)	0(0)	0(0)	23
Sacral canal cysts	0(0)	1(4.5%)	3(13.6%)	4(18.2%)	3(13.6%)	4(18.2%)	6(27.3%)	0(0)	1(4.5%)	22
Epidermoid cyst	0(0)	2(9.5%)	3(14.3%)	2(9.5%)	5(23.8%)	4(19.0%)	2(9.5%)	3(14.3%)	0(0)	21
Hemangioma	1(7.1%)	2(14.3%)	1(7.1%)	2(14.3%)	1(7.1%)	4(28.6%)	1(7.1%)	1(7.1%)	1(7.1%)	14
Meningeal cysts	0(0)	1(7.7%)	1(7.7%)	1(7.7%)	1(7.7%)	6(46.2%)	3(23.1%)	0(0)	0(0)	13
Primary aneurysmal bone cyst	2(16.7%)	8(66.7%)	1(8.3%)	0(0)	1(8.3%)	0(0)	0(0)	0(0)	0(0)	12
Fibrous dysplasia	0(0)	1(11.1%)	2(22.2%)	3(33.3%)	2(22.2%)	1(11.1%)	0(0)	0(0)	0(0)	9
Osteoblastoma	3(33.3%)	3(33.3%)	1(11.1%)	0(0)	1(11.1%)	1(11.1%)	0(0)	0(0)	0(0)	9
Eosinophilic granuloma	1(14.3%)	1(14.3%)	1(14.3%)	2(28.6%)	0(0)	2(28.6%)	0(0)	0(0)	0(0)	7
Simple bone cyst	0(0)	0(0)	1(20.0%)	1(20.0%)	1(20.0%)	2(40.0%)	0(0)	0(0)	0(0)	5
Spinal meningioma	0(0)	0(0)	1(20.0%)	2(40.0%)	2(40.0%)	0(0)	0(0)	0(0)	0(0)	5
Gut-tail cyst	0(0)	0(0)	3(75.0%)	0(0)	1(25.0%)	0(0)	0(0)	0(0)	0(0)	4
Benign fibrous histiocytoma	0(0)	0(0)	1(25.0%)	1(25.0%)	1(25.0%)	0(0)	1(25.0%)	0(0)	0(0)	4
Fibromatosis	0(0)	0(0)	0(0)	0(0)	2(66.7%)	1(33.3%)	0(0)	0(0)	0(0)	3
Diffuse giant cell tumor of tendon sheath	0(0)	1(33.3%)	1(33.3%)	1(33.3%)	0(0)	0(0)	0(0)	0(0)	0(0)	3
Chondroblastoma	1(33.3%)	0(0)	2(66.7%)	0(0)	0(0)	0(0)	0(0)	0(0)	0(0)	3
Lipoma	0(0)	0(0)	0(0)	0(0)	0(0)	1(50.0%)	1(50.0%)	0(0)	0(0)	2
Phosphouria stromal tumor	0(0)	0(0)	1(50.0%)	0(0)	0(0)	0(0)	1(50.0%)	0(0)	0(0)	2
Osteoidosteoma	0(0)	1(50.0%)	1(50.0%)	0(0)	0(0)	0(0)	0(0)	0(0)	0(0)	2
Liomyoma	0(0)	0(0)	0(0)	0(0)	1(100.0%)	0(0)	0(0)	0(0)	0(0)	1
Paget disease	0(0)	0(0)	0(0)	0(0)	0(0)	0(0)	1(100.0%)	0(0)	0(0)	1
Osteochondroma	0(0)	0(0)	0(0)	1(100.0%)	0(0)	0(0)	0(0)	0(0)	0(0)	1

We analyzed the age and gender related epidemiologic characteristics of the top seven primary sacral malignant tumors (Figure-3). Of 316 chordomas, the mean age was 55.4 ± 13.7 (range, 18–86 years) and 210 cases occurred in males and 106 cases in females, with a mean age of 56.1 ± 13.3 and 54.1 ± 14.4 years. Of 74 chondrosarcoma, the mean age was 40.9 ± 13.3 (range, 17–69 years) and 41 cases occurred in males and 33 cases in females, with a mean age of 37.7 ± 14.8 and 44.9 ± 10.0 years. Of 53 myeloma, the mean age was 54.1 ± 13.2 (range, 22–78 years) and 41 cases occurred in males and 33 cases in females, with a mean age of 53.4 ± 14.4 and 55.6 ± 10.6 years. Of 47 malignant peripheral nerve sheath tumor, the mean age was 40.8 ± 16.7 (range, 8–75 years) and 19 cases occurred in males and 28 cases in females, with a mean age of 40.4 ± 18.3 and 41.0 ± 15.8 years. Of 47 Ewing sarcoma, the mean age was 23.3 ± 12.8 (range, 2–56 years) and 24 cases occurred in males and 23 cases in females, with a mean age of 23.8 ± 13.1 and 22.7 ± 12.8 years. Of 40 osteosarcoma, the mean age was 26.0 ± 12.7 (range, 10–58 years) and 23 cases occurred in males and 17 cases in females, with a mean age of 29.0 ± 13.6 and 22.0 ± 10.3 years. Of 35 lymphoma, the mean age was 47.9 ± 15.0 (range, 20–78 years) and 15 cases occurred in males and 20 cases in females, with a mean age of 46.2 ± 16.4 and 49.2 ± 14.3 years.

Moreover, we analyzed the age and gender related epidemiologic characteristics of the top seven primary sacral benign tumors (Figure-3). Of 205 giant cell tumor, the mean age was 34.4 ± 11.6 (range, 11–67 years) and 69 cases occurred in males and 86 cases in females, with a mean age of 32.7 ± 10.3 and 35.8 ± 12.5 years. Of 155 neurofibroma, the mean age was 46.7 ± 14.5 (range, 9–83 years) and 95 cases occurred in males and 110 cases in females, with a mean age of 44.6 ± 13.8 and 48.4 ± 14.9 years. Of 119 schwannoma, the mean age was 46.1 ± 13.5 (range, 13–79 years) and 60 cases occurred in males and 59

cases in females, with a mean age of 44.6 ± 13.1 and 47.7 ± 13.9 years. Of 24 tuberculosis, the mean age was 42.0 ± 18.9 (range, 8–76 years) and 8 cases occurred in males and 16 cases in females, with a mean age of 46.9 ± 21.6 and 39.5 ± 17.7 years. Of 23 teratoma, the mean age was 31.7 ± 12.9 (range, 5–62 years) and 4 cases occurred in males and 19 cases in females, with a mean age of 33.8 ± 17.3 and 31.3 ± 12.4 years. Of 22 sacral canal cysts, the mean age was 48.8 ± 17.1 (range, 19–81 years) and 11 cases occurred in males and 11 cases in females, with a mean age of 46.0 ± 21.2 and 51.5 ± 12.3 years. Of 21 epidermoid cyst, the mean age was 47.0 ± 18.6 (range, 19–78 years) and 4 cases occurred in males and 17 cases in females, with a mean age of 32.8 ± 23.0 and 50.3 ± 16.4 years.

Gender Distribution Of Pst

The distribution of different histological types in males and females was shown as follows (Figure-4): chordoma (316 cases, M: F = 1.98:1), chondrosarcoma (74 cases, M: F = 1.24:1), myeloma (53 cases, M: F = 2.12:1) and osteosarcoma (40 cases, M: F = 1.35:1) affected more frequently males than females. Meanwhile, malignant peripheral nerve sheath tumor (47 cases, M: F = 0.68:1), lymphoma (35 cases, M: F = 0.75:1), giant cell tumor (205 cases, M: F = 0.86:1), neurofibroma (155 cases, M: F = 0.80:1), tuberculosis (24 cases, M: F = 0.50:1), teratoma (23 cases, M: F = 0.21:1) and epidermoid cyst (21 cases, M: F = 0.24:1) affected more frequently females than males (Tanle-5&6).

Table 5
Gender distribution of primary sacral malignant tumors

Histology	No.	Male		Female		M:F	Age range (years)	Mean ± SD (age)		
	No.	No.	%	No.	%			Male	Female	Total P value
Chordoma	316	210	66.5%	106	33.5%	1.98:1	18-86	56.1±13.3	54.1±14.4	55.4±13.7 P=0.236
Chondrosarcoma	74	41	55.4%	33	44.6%	1.24:1	17-69	37.7±14.8	44.9±10.0	40.9±13.3 P=0.015
Myeloma	53	36	67.9%	17	32.1%	2.12:1	22-78	53.4±14.4	55.6±10.6	54.1±13.2 P=0.567
Malignant peripheral nerve sheath tumor	47	19	40.4%	28	59.6%	0.68:1	8-75	40.4±18.3	41.0±15.8	40.8±16.7 P=0.903
Ewing sarcoma	47	24	51.1%	23	48.9%	1.04:1	2-56	23.8±13.1	22.7±12.8	23.3±12.8 P=0.791
Osteosarcoma	40	23	57.5%	17	42.5%	1.35:1	10-58	29.0±13.6	22.0±10.3	26.0±12.7 P=0.085
Lymphoma	35	15	42.9%	20	57.1%	0.75:1	20-78	46.2±16.4	49.2±14.3	47.9±15.0 P=0.574

Table 6
Gender distribution of primary sacral benign or tumor-like lesions

Histology	No.	Male		Female		M:F	Age range (years)	Mean ± SD (age)		
	No.	No.	%	No.	%			Male	Female	Total P value
Giant cell tumor	205	95	46.3%	110	53.7%	0.86:1	11-67	32.7±10.3	35.8±12.5	34.4±11.6 P=0.057
Neurofibroma	155	69	44.5%	86	55.5%	0.80:1	9-83	44.6±13.8	48.4±14.9	46.7±14.5 P=0.108
Schwannoma	119	60	50.4%	59	49.6%	1.02:1	13-79	44.6±13.1	47.7±13.9	46.1±13.5 P=0.212
Tuberculosis	24	8	33.3%	16	66.7%	0.50:1	8-76	46.9±21.6	39.5±17.7	42.0±18.9 P=0.380
Teratoma	23	4	17.4%	19	82.6%	0.21:1	5-62	33.8±17.3	31.3±12.4	31.7±12.9 P=0.741
Sacral canal cysts	22	11	50.0%	11	50.0%	1.00:1	19-81	46.0±21.2	51.5±12.3	48.8±17.1 P=0.461
Epidermoid cyst	21	4	19.0%	17	81.0%	0.24:1	19-78	32.8±23.0	50.3±16.4	47.0±18.6 P=0.089

Discussion

This series of primary sacral tumors treated at our tumor center provide valuable data of epidemiological characteristics contributing to our understanding of the diagnosis and therapy for primary bone tumors at the sacrum in the clinical practice. Available reports on the epidemiologic features of primary spine tumors were mostly among the mobile spine[1, 4, 5]. The present report demonstrated epidemiologic characteristics based on the largest series of primary sacral tumors.

In our series, chordoma was the most common primary malignant tumor, accounting for 44.1% (316/716), followed by chondrosarcoma (10.3%, 74/716), myeloma (7.4%, 53/716), malignant peripheral nerve sheath tumor (6.6%, 47/716), Ewing sarcoma (6.6%, 47/716), osteosarcoma (5.6%, 40/716) and lymphoma (4.9%, 35/716).

Chordoma is a relatively rare neoplasm and accounts for 1%-4% of all primary malignant bone tumors which arise from embryonic remnants of notochord[6]. It has been reported that chordomas occur most commonly within the sacrum (50–60%), followed by the spheno-occipital vertebrae (25–30%), cervical region (10%) and thoracolumbar vertebrae (5%) and chordoma affects males more commonly than female[7]. Our results also showed the male predominance was most pronounced among 306 chordomas, accounting for 68.6% and age of 51–70 yrs was the most common of age interval, which was concordant with the previous studies. The peak age in our cohort was the range of 51 to 60 and it was distinctly uncommon in patients younger than 30 years, only accounting for 5.9% (18/306).

Chondrosarcomas (CS) rank the second in the incidence of all primary malignant bone tumors, with reporting about 25% incidence and following after osteosarcoma. Regarding age, it is more common in adults between 40–80 years old, and is slightly more common in men[8]. CS constituted over 20.4% of the malignant tumors in Mayo Clinic series and more than two-thirds of chondrosarcomas were in the trunk and the upper ends of the femur and humerus. CS was a relatively rare and the incidence at the sacrum was 1.9% (24/1293) among all chondrosarcomas in Mayo Clinic series[9]. However in our cohort, the analysis of epidemiological feature of CS showed an obvious predilection of male (M: F = 1.24:1) and the peak age ranged from 41 to 50 years. Moreover, chondrosarcoma was the second common histological type at the region of the sacrum for the primary sacral malignant tumors and the incidence of CS among the malignant tumors was 10.3% (74/716).

Myeloma, a tumor of hematopoietic derivation, is the most common primary neoplasm of bone. There were more than 5000 patients with myeloma documented in the Mayo Clinic files. They reported 67.7% were males and the largest concentration of age was in the sixth and seventh decades of life. The well-known rarity of myeloma in patients who were younger than 40 years was shown in the Mayo Clinic series[9]. Likewise, myeloma at the sacrum had the gender predilection (M: F = 2.12:1) and 67.9% of all myelomas were males in our series. The median age of myeloma was 54.1 ± 13.2 yrs and the peak age ranged from 41 to 50 years. Only two patients in our cohort were younger than 30 years and no patient was in the first and second decades of life. Generally, the epidemiological characteristics of sacral myeloma were similar to other boney myeloma.

Malignant peripheral nerve sheath tumors (MPNST) include malignant schwannoma (malignant peripheral schwannoma) and neurofibrosarcoma. MPNST represents a relatively common subtype of soft tissue sarcoma and is particularly likely to occur in individuals with type 1 neurofibromatosis (NF1)[10–13]. A SEER database analysis showed 64 MPNST in the spinal location. Their mean age at diagnosis was 50.9 years with more patients in the higher age group and fifty-six percent of patients were male. However, it had no description of the incidence at the sacrum[14]. In our cohort, the analysis of epidemiological

feature of MPNST showed an obvious predilection of female (M: F = 0.68:1) and the peak age ranged from 41 to 50 years. Our result showed the epidemiologic characteristic of age predilection was in concordance with the previous study in SEER database, but gender predilection of MPNST at the sacrum was not similar to the result of SEER database [14].

Ewing sarcoma (ES) is a distinctive, small, round cell sarcoma that is considered one of the most lethal of all bone tumors. In Mayo Clinic database, ES comprised 8.6% of the total malignant tumors and had a distinct predilection for males (62%). Furthermore, appromimately 75% were in the first two decades of life and the incidence of ES at the sacrum accounted for 5.9% (36/611) of all patients with ES[9]. The sacrum is not the frequent site for Ewing sarcoma. In our PKUPH database, Ewing sarcoma located at the sixth top of all primary sacral malignant tumors. The peak incidence was in the second decade (40.4% of all 47 Ewing sarcomas), followed by the third and fourth decades (38.2%) and it had no predilection of gender, which was not concordant with the Mayo Clinic experience.

Osteosarcoma is the most common malignant bone tumor. Mayo Clinic series files recorded 1952 osteosarcomas, accounting for 27.5% of all malignant tumors and 19.2% of all bone tumors and approximately 58% of patients with OS were male. In their database, the incidence of OS at the sacrum accounted for 2.0% of all patients with OS and the anatomic site around the knee was the most common site. The second decade was the most common age distribution and among 1952 osteosarcomas, 192 patients were older than 60 years who had the preexisting condition such as Paget disease, previous radiation, infarct, chronic osteomyelitis and cyst of degenerative joint disease as the second peak[9]. According to our results, osteosarcoma at the sacrum was not frequent as the site around the knee and placed as the sixth top of all primary sacral malignant tumors. Although only one patient with the sacral OS was in the first decade of life, the peak incidence was in the second decade (37.5% of all 40 osteosarcomas), followed by the third decade (32.5%). We noticed no one patient aged more than 60 years. This result demonstrated that the age distribution of sacral osteosarcoma had the peak age of 11–20 and didn't show another age peak of more than 60 years. Moreover, osteosarcoma was not common at the region of sacrum.

Bone lymphoma is a rare disease. It is estimated that bone lymphoma is accounting for about 5% of extranodal lymphomas and 3–7% of all malignant bone tumors[15]. Parker et al. firstly described the malignant lymphoma of bone and separated it from Ewing sarcoma[16]. The Mayo Clinic database showed the 905 cases of malignant lymphoma comprised as 12.7% of the malignant bone tumors in their series and males predominated at a ratio of 4 to 3 in their cohort. In our cohort, approximately 51.4% of lymphoma occurred in patients between 41–60 years, with a peak incidence in the fifth and sixth decade of life. Only 5.7% of the patients with lymphoma were younger than 20 years and no one was younger than 10 years old. However, our series revealed lymphoma at the sacrum had the female predilection (M: F = 0.75:1), which was not concordant with the Mayo Clinic experience.

Our PKUPH database showed that giant cell tumor (30.6%, 205/669) was the most frequent benign tumors, followed by neurofibroma (23.2%, 155/669), schwannoma (17.8%, 119/669), tuberculosis (3.6%, 24/669), teratoma (3.4%, 23/669), sacral canal cysts (3.3%, 22/669) and epidermoid cyst (3.1%, 21/669). The epidemiological characteristics of primary sacral benign tumors were distinct from the one of spinal benign tumors. It has been reported that hemangioma was the most frequent benign primary spine tumor in one hospital of China, which was different from the region of sacrum and our database showed giant cell tumor was the most common histological type at the sacrum [1]. Hemangioma at the sacrum accounted for 2.1% (14/669) of all sacral benign tumor and 1.0% (14/1385) of all PST. It illustrated that hemangioma was not a frequent tumor at the sacrum.

Giant cell tumor (GCT) is an invasive benign bone tumor consisting of proliferative mononuclear cells and osteoclast-like multinucleated giant cells. In the present study, GCT was the most common benign tumor at the sacrum and the incidences of GCT among all PST and primary sacral benign tumors were respectively 14.8% (205/1385) and 30.6% (205/669). This result was concordant with the study of Zhou et al about the epidemiological feature of all spinal tumors[1]. The Mayo Clinic series showed the female predominated in bone GCT, with 376 females and 295 males[9]. However, Niu et al. reported 621 patients with GCT in the extremity and male predominance (1.4:1) was observed in their series[17]. Their epidemiological feature of extremities was different from the characteristics of our sacral GCT and this illustrated that predominate gender may depend on the anatomical site. Our cohort also had 110 cases of female, accounting for 53.7% of all GCTs in our series. Approximately 79.5% of GCT occurred in patients between 21–50 years, with a peak incidence in the third and fourth decade of life. Only 8.8% of the patients with GCT were older than 50 years and only 11.7% of GCT occurred in patients before 20 years old. Furthermore, no one was younger than 10 years old.

Benign peripheral neurogenic tumors include neurofibroma and peripheral schwannoma. Neurogenic tumors arising from the sacrum are rare, with only about 7% of intraspinal neurogenic tumors involving the sacrum. Neurofibromas can occur in any site, both deep soft sissue and superficial cutaneous lesions. They are usually painless and they are often excised for cosmetic purpose. Individuals with NF1 may have multiple neurofibromas that need excision for functional purpose as well as to exclude the possibility of malignant transformation. The analysis of epidemiological feature of neurofibroma in our series showed a little predilection of female (M: F = 0.80:1) and the peak age ranged from 41 to 60 years. Meanwhile, schwannomas are relatively common benign lesions of the peripheral nerves, which are thought to derive from Schwann cell. They affect all age groups and are usually solitary sporadic tumors. Schwannomas can also develop along the spine adjacent to the neural foramina and frequently show a "target sign" on imaging that can be very suggestive of this specific diagnosis. They are usually slow growing and often discovered incidentally. It has been reported that sacral schwannomas typically occur in middle-aged patients without any predilection for gender. Pennington et al. performed a systematic review and reported presacral schwannoma can reasonably be treated with either en bloc or piecemeal GTR and recurrence was infrequent. In their review, the patients showed no gender predilection[18]. Our database analysis revealed epidemiological feature of schwannoma also had no gender predilection and the peak age ranged from 31 to 60 years. This result was concordant with the result of previous study in the literature.

Over the past decades, the incidence of boney tuberculosis has continued to increase due to population growth, acceleration of mobility. Little is known about the incidence of spinal tuberculosis. In our cohort, we evaluated the incidence of sacral tuberculosis, which was the fourth top of sacral benign tumors (3.6%, 24/669). Moreover, it had the female predilection and it occurred in a patient of any age.

Sacrococcygeal teratoma is the most common extragonadal germ cell tumor in neonates and infants[19–21]. However, this lesion sometimes occurs in the adults. This lesion consists of a solid and/or cystic component and develops either from the tip of the sacrum, protruding outward from the buttocks, or within the pelvic cavity. In our series, the analysis of epidemiological feature of sacrococcygeal teratoma also showed obvious gender predilection and females occurred in 82.6% of all patients with teratoma. Moreover, the peak age ranged from 21 to 40 years and this feature of age distribution may have some bias due to having more adult cases not neonates and infants in our tumor center.

Epidermoid cysts of spinal lesions are very rare and they are often found in the lumbosacral region, which are commonly intradural-extramedullary tumors[22–24]. They commonly occur in children and the cause of epidermoid cysts is believed to be the introduction of ectoderm cells arising in early fetal life between 3 and 5 weeks. The formation of epidermoid cysts may be associated with defective closure of the dural tube[25]. Epidermoid cysts in our cohort had 17 cases of female, accounting for 81% of all epidermoid cysts in our series. Approximately 42.9% of epidermoid cysts occurred in patients between 41–60 years. It also had significant gender predilection and females occurred in 81% of all patients with epidermoid cysts, like the epidemiologic feature of sacrococcygeal teratoma.

In conclusion, the large cohort of primary sacral tumors in our database may reveal their epidemiologic characteristics of primary sacral malignant and benign tumors in China and epidemiological feature of PST is fairly distinct from the mobile spine and extremities.

Declarations

Conflict of interest statement and ethical approval

The authors declare that they have no conflict of interest. This article does not contain any studies with human participants or animals performed by any of the authors and informed consent was obtained from all individual participants included in the study.

Availability of data and materials

All data generated or analyzed during this study was included in this published article.

Ethics approval and consent to participate

This study was approved by the Institutional Review Board/Ethics Committee of the authors' institution and the study was performed according to the Helsinki Declaration guidelines. Informed consent was obtained from the patients prior to enrolling in the study.

Consent for publication

We obtained consent for publication from the patients.

Competing interests

The authors declare that they have no competing interests.

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Authors' contributions

WJ carried out the data analysis and drafted the manuscript. LDS collected data and performed the statistical analysis YRL, TXD and YTQ participated in its design. GW designed the study and reviewed the article. All authors have been actively involved in the drafting and critical revision of the manuscript, and each provided final approval of the version to be published.

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Not applicable

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Figures

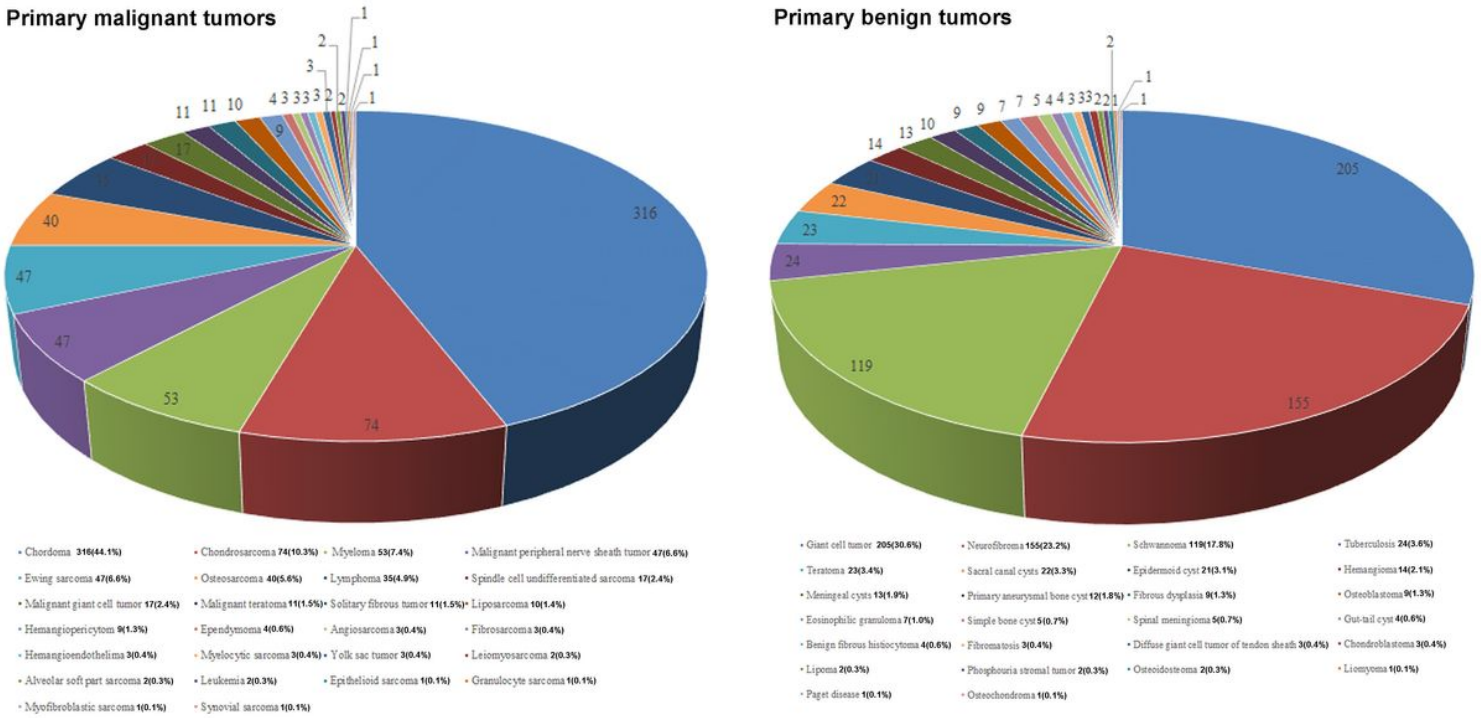

Figure 1

Histological types of primary sacral malignant and benign tumors

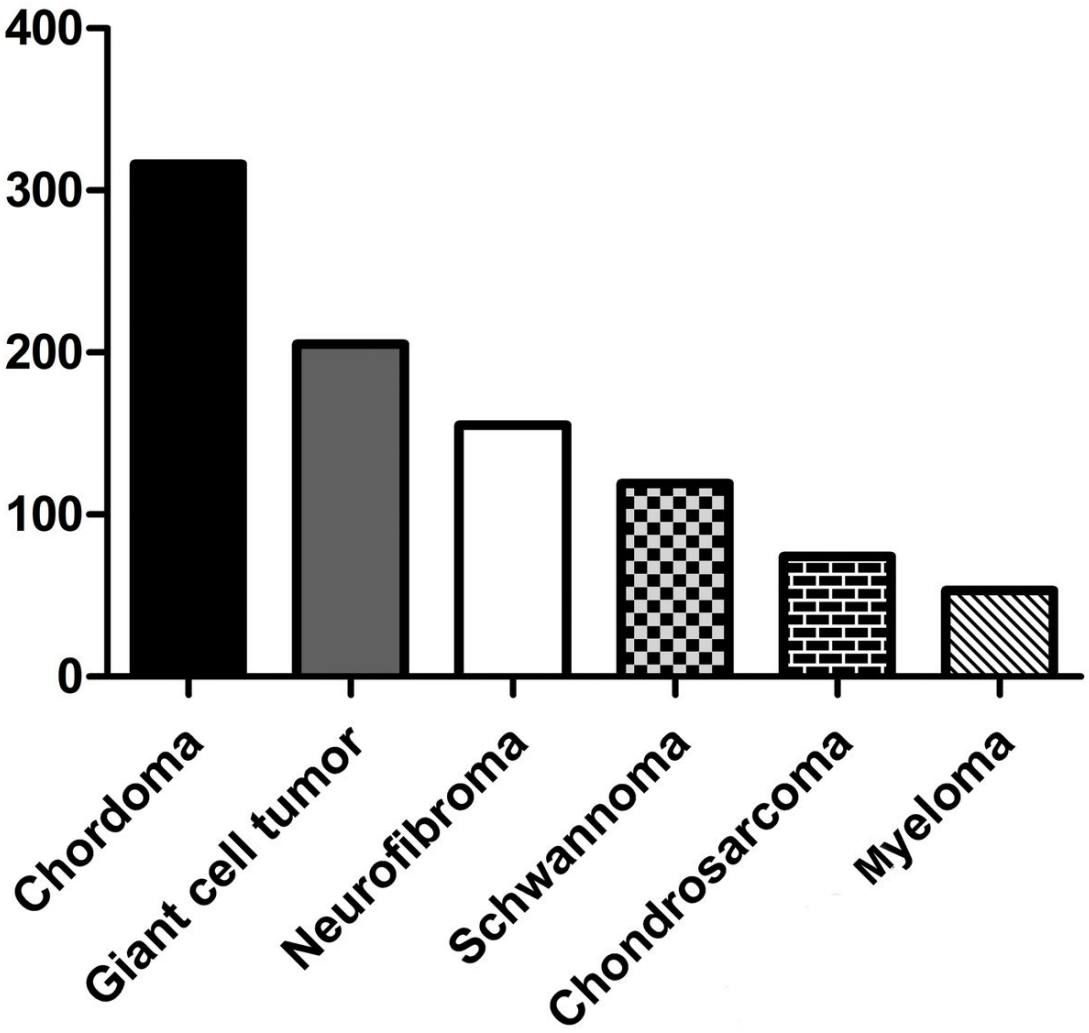

Figure 2

The top six histological types of primary sacral tumors

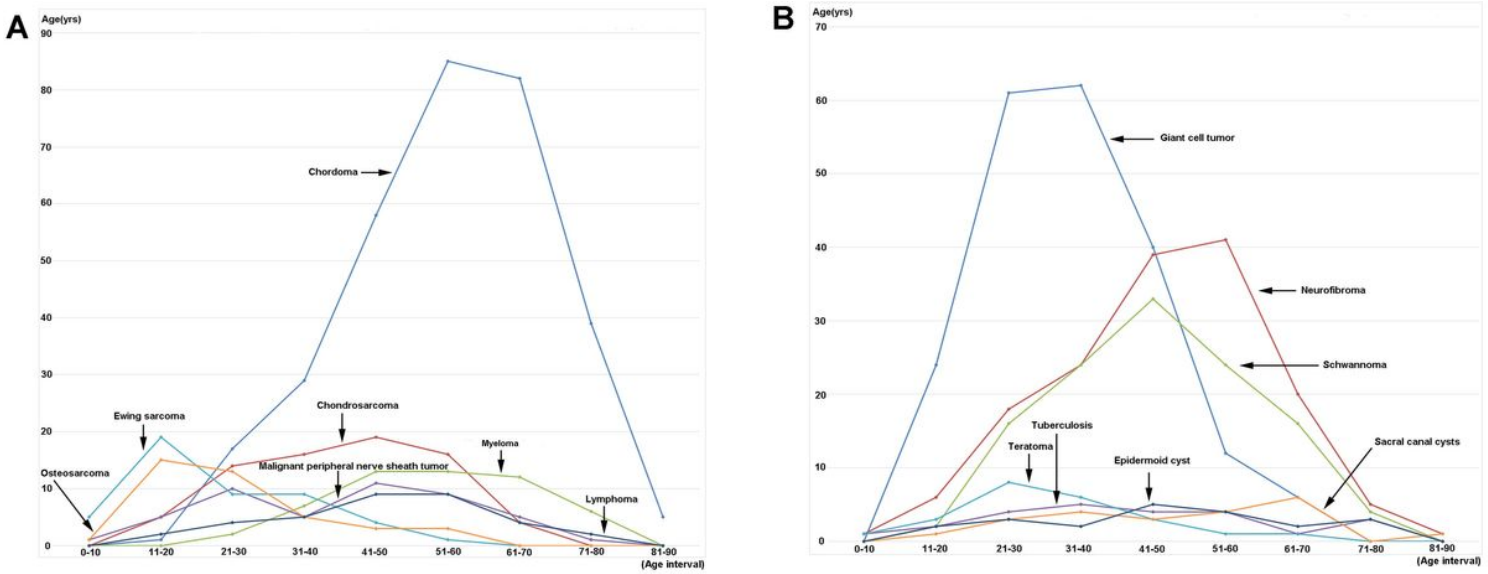

Figure 3

A-B Epidemiologic features of gender for top seven of primary sacral malignant tumors; C-D Epidemiologic features of gender for top seven of primary sacral benign tumors

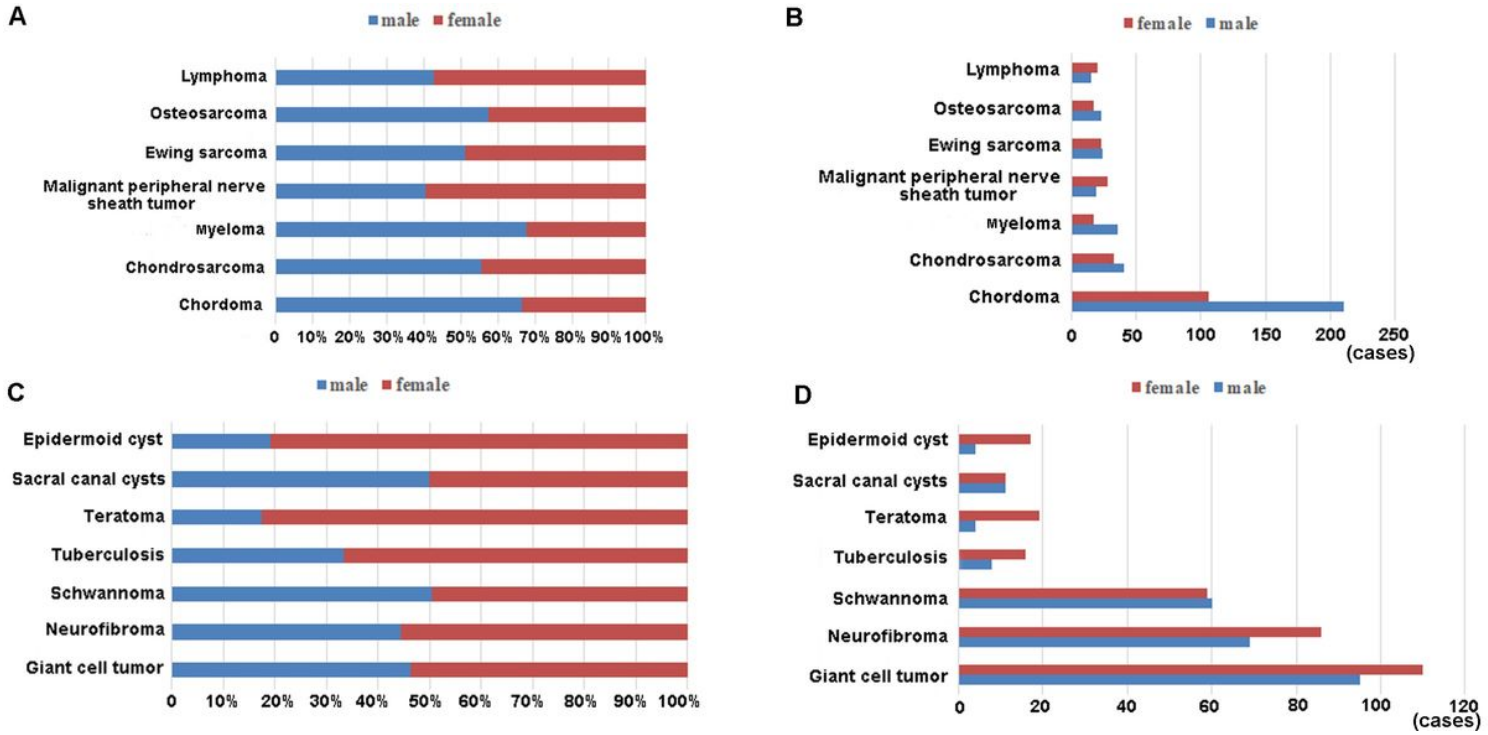

Figure 4

A. Age distribution for top seven of primary sacral malignant tumors. N=numbers of patients; B. Age distribution for top seven of primary sacral benign tumors. N=numbers of patients