Original Article

Epidemiologic trend of mobile spine and sacrum chordoma: A National population-based study

ABSTRACT

Introduction: Chordoma is a rare sarcoma of the axial skeleton. The incidence of this tumor is different between races. To understand the epidemiologic characteristic and due to rarity of this pathology, large number of cases should be evaluated through national data registries.

Materials and Methods: All pathologically confirmed cases of chordoma were derived from the Iran National Cancer Registry. Descriptive analysis was performed to extract age-standardized and age-specific incidence rates. Data regarding tumor location and chordoma subtypes were derived and analyzed.

Results: One hundred twenty-two cases of chordoma including 80 male and 42 female were identified. One hundred seven cases of nonotherwise specified chordoma, 14 chondroid chordoma, and one dedifferentiated chordoma were detected. The age-standardized incidence rate (ASIR) of chordoma was 0.28. Chordoma of the sacrum composed 67.2% of cases. The mean survival time was 4.5 years.

Conclusion: Epidemiology characteristic of chordoma in Iran is similar to other studies; however, the total ASIR was lower and the incidence in sacrum is nearly twice respect to the mobile spine. While men affected by sacral chordoma in relatively older age, the female patients had higher mean age in case of mobile spine involvement. The survival rate of chordoma is significantly lower in comparison with other studies.

Keywords: Chordoma, epidemiology study, sacrum, sarcoma, spine

INTRODUCTION

Chordoma is a rare sarcoma of the bone which interest solely the axial skeleton and accounts for 1%–4% of all bone malignancies and <0.2% of all new cancers.[1,2] Chordoma arises from notochordal remnants and occurs along the axial skeleton including skull base, mobile spine, and sacrum.[3] The slow-growing nature of chordoma leads to late diagnosis with a large size tumor and destruction of the bone and extension to surrounding soft tissue which may complicate tumor management process and affect the survival rate.[4]

Due to the relative rarity of this tumor, a large number of cases can be provided solely through national data registries. The Surveillance, Epidemiology, and End Results (SEER) Registry, National Cancer Database (NCDB), National Cancer Intelligence Network, Scandinavian sarcoma group, and similar registries in other countries have been stabilized to provide large number of patients for epidemiological studies of different cancers.[5-8] Moreover, epidemiologic characteristics of each cancer should be evaluated for

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different populations with their own races and ethnicities separately. In light of the need to perform a similar study in the territory of Iran, a comprehensive study aimed to evaluate the epidemiologic characteristics of the mobile spine and sacrum chordoma among the Iranian population according to the Iran National Cancer Registry (INCR).[9,10] The latter is a preliminary registry of cancers in the territory of Iran based on data provided by the Ministry of Health and Medical Education.

MATERIAL AND METHODS

We performed a national population-based study based on data derived from INCR between 2009 and 2015. All microscopically and pathologically confirmed cases of patients with chordoma were detected and registered. Only patients with involvement of the mobile spine including cervical, thoracic, and lumbar spine (C-code 41.2) and sacrum and coccyx (C-code 41.4) were enrolled and chordoma of the skull (C-code 41.0) and any other region were eliminated from this study. According to the third edition of the International classification of diseases for oncology,[11] three morphology types of chordoma were detected and searched including nonotherwise specified (NOS) chordoma (M-code: 9370), chondroid chordoma (M-code: 9371), and dedifferentiated chordoma (M-code: 9372). Due to incomplete histopathology registration of the patients and lack of histology codes on INCR, histology evaluation was excluded completely from this study. To perform data cleansing, cases with the same first name and surname were identified as duplicate suspects, and their records were scrutinized and excluded if they had a same national code.

The descriptive analysis was expressed with number of cases (percentage) and mean standard deviation (SD) for all chordoma patients. The binomial proportion test was used to compare the percentage between groups. The age-standardized incidence rate (ASIR) was analyzed by sex, disease site, and tumor subtype using the new World Health Organization (WHO) standard population.[12] The 95% confidence intervals (C. I) was expressed for each ASIR. Patients were divided in different age groups. The age-specific incidence rate (ASR) was calculated for each age group and sex.[13]

The Kaplan–Meier survival analysis was performed to estimate the survival probability of patients; the mean/median survival time was used to summarize the survival of patients. To analyze the survival rate, patients were communicated through a phone call and by a questionnaire; the data regarding the survival were collected and registered. Informed consent was explained telephonically for each patient and compiled. Ethical approval for this study was obtained under a number: IR. SBMU.CRC.REC.1398.010. Death due to chordoma and the interval between diagnoses to death were registered as treatment failure and survival time, respectively. Data analyzing were performed using the IBM SPSS Statistics for Windows, Version 25.0. (Armonk, NY: IBM Corp.), and $P \leq 0.05$ was set as the significance level.

RESULTS

The search through INCR from March 2009 to March 2015 identified 122 pathologically confirmed cases of chordoma. Demographically 80 cases (65.57%) were male and 42 cases (34.43%) were female. The mean age of diagnosis was 53.4 (SD ± 17.40) years. The ASIR of chordoma was 0.28 per million person-years. The ASIR among males was 0.37 per million person-years which was nearly twice in compare with ASIR among females (0.19 per million person-year) [Table 1].

Totally, 107 cases (87.7%) were NOS chordoma, 14 cases (11.5%) were chondroid chordoma, and one case (0.8%) was dedifferentiated chordoma. Through 107 cases of NOS chordoma, 70 (65.4%) were male and 37 (34.6%) were female ($P < 0.01$). The mean age of NOS chordoma was 57.1 years. Male and female NOS patients showed the mean age of 54.70 and 52.86 years, respectively, which was not significantly different from each other ($P > 0.05$). In addition, in patients with NOS chordoma, the total frequency was significantly higher among males ($P < 0.05$). Among 14 cases of chondroid chordoma, 9 cases were male and 5 cases were female. The results showed that the mean age of chondroid chordoma was 47.43 years. Furthermore, considering gender,

Table 1: The number of new cases (percentage), mean age (standard deviation), and age-standardized incidence rate for patents with chordoma based on the morphology types during 2008-2015 in Iran

M-code	Morphology type	Number of new cases (%)			Mean age (SD)			ASIR (95% CI)		
		Total	Male	Female	Total	Male	Female	Total	Male	Female
9370	NOS**	107 (100.00)	70 (65.44)	37 (34.60)	57.16 (16.10)	54.70 (18.80)	52.86 (13.79)	0.25 (0.20-0.30)	0.33 (0.25-0.41)	0.17 (0.12-0.23)
9371	Chondroid	14 (100.00)	9 (64.30)	5 (35.7)	47.43 (18.48)	42.89 (18.54)	54.60 (15.47)	0.03 (0.01-0.05)	0.04 (0.01-0.07)	0.02 (0.00-0.04)
9372	Dedifferentiated	1 (100.0)	1 (100.0)	-	77.00 (-)	77.00 (-)	-	<0.01	<0.01	-
Total**		122 (100.00)	80 (65.57)	42 (34.43)	53.45 (17.40)	53.65 (19.09)	53.07 (13.80)	0.28 (0.23-0.34)	0.37 (0.29-0.46)	0.19 (0.13-0.25)

**Binomial proportion test for comparing frequency of the cancer between males and females with P<0.01. SD – Standard deviation

the mean age in male and female was 42.89 and 54.60 years, respectively. The chondroid chordoma new cases and mean age were not affected by sex [Table 1].

According to the results, among patients affected by NOS chordoma, the overall ASIR was 0.25 per million person-years (95% C. I: 0.20–0.30), while male and female had the ASIR of 0.33 (95% C. I: 0.25–0.41) and 0.17 (95% C. I: 0.12–0.23). Furthermore, the results showed that the ASIR of chondroid chordoma was approximately 0.03 (95% C. I: 0.01–0.05) per million person-years [Table 1].

The number of cases in the mobile spine was 40 cases (32.8%) including 24 males and 16 females, while 82 cases (67.2%) arose from the sacral region (male: 56 and female: 26). The mean age of chordoma in mobile spine was 52.3 years and was not affected by gender ($P > 0.05$) (male: 49.79 and female: 56.06) [Table 2]. The results showed that the chordoma of the sacrum had a mean age of 54.01 years and had a higher mean age in males (55.30 years) in comparison to females (51.23 years). The mean age of mobile spine chordoma was 52.3 years and in contrast to the sacral chordoma, females had higher mean age of diagnosis (56.06) in compare with males (49.79). ASIR (95% C. I) of the mobile spine and sacrum was 0.09 (0.06–0.12) and 0.19 (0.15–0.23) per million person-years, respectively [Table 2].

Among cases of mobile spine chordoma, 34 (85.0%) were NOS chordoma and 6 (15.0%) were chondroid chordoma. In sacrum, 73 cases (89.10%) were NOS chordoma, 8 cases (9.8%) were chondroid chordoma, and one case (1.2%) was dedifferentiated chordoma. The ASIR of mobile spine chordoma was 0.08 and 0.01 per million person-years for NOS and chondroid chordoma, respectively. The ASIR of sacral chordoma showed 0.17 and 0.02 per million person-years for NOS and chondroid chordoma, respectively, and lower than 0.01 per million person-years for dedifferentiated chordoma.

The frequency distribution of chordoma ASR was calculated by morphology types. According to the results, about 80% of cases occurred in patients older than 40 years with the peak age between 50 and 80 years including 62.3% of all patients. Only five cases of chordoma (4%) were detected among patients younger than 25 years. The binomial proportion test showed that only the number of new cases was significantly different among genders in the age group of 55–59 years [Table 3].

Survival data for a total of 24 available chordoma patients were collected and analyzed, in whom 17 cases experienced the death due to the disease from 2009 to 2015. The mean survival time was 54.98 months (95% C. I: 34.48–75.47). The cumulative survival time was 0.29. The 1-year and 3-year survival time was 0.75 and 0.67, respectively. Survival analysis from Kaplan–Meier graphs revealed that before the survival curve arrive at a plateau at about 0.3, this curved experienced a downward trend until 45 months from the beginning [Table 4 and Figure 1].

DISCUSSION

Chordoma is a rare tumor of axial skeleton; however, it remains the most frequent primary cancer of the sacrum and mobile

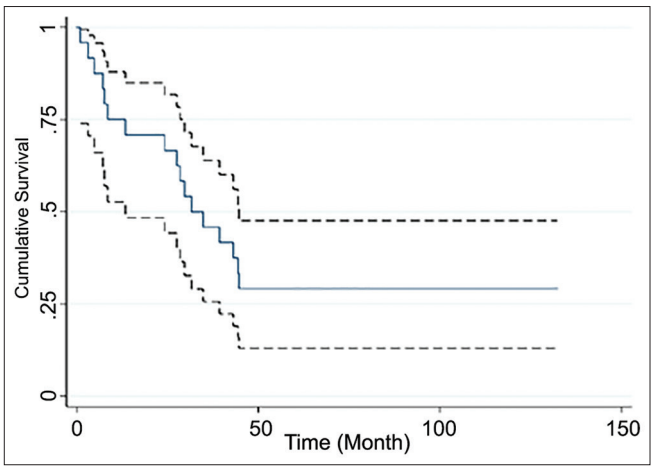

Figure 1: The Kaplan–Meier survival plot for patients with chordoma

Table 2: The number of new cases (percentage), mean age (standard deviation), and age-standardized incidence rate for patents with chordoma based on the topography types during 2008-2015 in Iran

C-Code	Topography type	Number of new cases (%)			Mean age (SD)			ASIR (95% CI)		
		Total	Male	Female	Total	Male	Female	Total	Male	Female
41.2	Malignant neoplasm of vertebral column	40 (100.00)	24 (60.00)	16 (40.00)	52.30 (15.00)	49.79 (15.90)	56.06 (13.12)	0.09 (0.06-0.12)	0.11 (0.06-0.15)	0.08 (0.04-0.11)
41.4	Malignant neoplasm of pelvic bones, sacrum and coccyx**	82 (100.00)	56 (70.70)	26 (29.30)	54.01 (18.50)	55.30 (20.22)	51.23 (14.14)	0.19 (0.15-0.23)	0.27 (0.20-0.34)	0.12 (0.07-0.16)
Total**		122 (100.00)	80 (65.57)	42 (34.43)	53.45 (17.40)	53.65 (19.09)	53.07 (13.80)	0.28 (0.23-0.34)	0.37 (0.29-0.46)	0.19 (0.13-0.25)

**Binomial proportion test for comparing frequency of the cancer between males and females with $P<0.01$. SD - Standard deviation; CI - Confidence interval

Table 3: The number of new cases (age-specific incidence rate/per million person-years) for Chordoma by Morphology Code records during 2008-2014

Age	9370			9371			Total		
	Total	Male	Female	Total	Male	Female	Total	Male	Female
0-4	1 (0.02)	1 (0.04)	0 (0.00)	0 (0.00)	0 (0.00)	0 (0.00)	1 (0.02)	1 (0.04)	0 (0.00)
5-9	0 (0.00)	0 (0.00)	0 (0.00)	0 (0.00)	0 (0.00)	0 (0.00)	0 (0.00)	0 (0.00)	0 (0.00)
10-14	1 (0.02)	1 (0.05)	0 (0.00)	1 (0.02)	1 (0.05)	0 (0.00)	2 (0.05)	2 (0.10)	0 (0.00)
15-19	1 (0.02)	1 (0.04)	0 (0.00)	1 (0.02)	1 (0.04)	0 (0.00)	2 (0.04)	2 (0.08)	0 (0.00)
20-24	2 (0.04)	2 (0.07)	0 (0.00)	0 (0.00)	0 (0.00)	0 (0.00)	2 (0.04)	2 (0.07)	0 (0.00)
25-29	5 (0.09)	5 (0.17)	0 (0.00)	0 (0.00)	0 (0.00)	0 (0.00)	5 (0.09)	5 (0.17)	0 (0.00)
30-34	5 (0.10)	2 (0.08)	3 (0.12)	1 (0.02)	1 (0.04)	0 (0.00)	6 (0.12)	3 (0.12)	3 (0.12)
35-39	8 (0.20)	3 (0.15)	5 (0.26)	0 (0.00)	0 (0.00)	0 (0.00)	8 (0.20)	3 (0.15)	5 (0.26)
40-44	5 (0.15)	2 (0.12)	3 (0.18)	3 (0.09)	1 (0.06)	2 (0.12)	8 (0.24)	3 (0.17)	5 (0.30)
45-49	9 (0.32)	4 (0.28)	5 (0.35)	0 (0.00)	0 (0.00)	0 (0.00)	9 (0.32)	4 (0.28)	5 (0.35)
50-54	11 (0.46)	6 (0.50)	5 (0.41)	3 (0.12)	2 (0.17)	1 (0.08)	14 (0.58)	8 (0.66)	6 (0.50)
55-59	16 (0.87)*	13 (1.42)	3 (0.32)	1 (0.05)	1 (0.11)	0 (0.00)	17 (0.92)*	14 (1.53)	3 (0.32)
60-64	9 (0.68)	6 (0.94)	3 (0.43)	2 (0.15)	2 (0.31)	0 (0.00)	11 (0.83)	8 (1.25)	3 (0.43)
65-69	13 (1.35)	8 (1.71)	5 (1.01)	0 (0.00)	0 (0.00)	0 (0.00)	13 (1.35)	8 (1.71)	5 (1.01)
70-74	9 (1.14)	6 (1.51)	3 (0.76)	2 (0.25)	0 (0.00)	2 (0.51)	11 (1.39)	6 (1.51)	5 (1.27)
75-79	9 (1.48)	7 (2.20)	2 (0.69)	0 (0.00)	0 (0.00)	0 (0.00)	10 (1.64)	8 (2.52)	2 (0.69)
80-85	3 (0.76)	3 (1.50)	0 (0.00)	0 (0.00)	0 (0.00)	0 (0.00)	3 (0.76)	3 (1.50)	0 (0.00)
+85	0 (0.00)	0 (0.00)	0 (0.00)	0 (0.00)	0 (0.00)	0 (0.00)	0 (0.00)	0 (0.00)	0 (0.00)
Total	107 (0.20)*	70 (0.26)	37 (0.14)	14 (0.03)	9 (0.03)	5 (0.02)	122 (0.23)*	80 (0.30)	42 (0.16)

*Binomial proportion test for comparing frequency of the cancer between males and females with $P<0.01$

Table 4: The survival analysis of patients with chordomas

Number of cases (number of events)	Cumulative survival time (95% CI)	1-year survival time	3-year survival time	Mean survival time (months) (95% CI)	Median survival time (months) (95% CI)
24 (17)	0.29 (0.13-0.48)	0.75 (0.60-0.94)	0.67 (0.50-0.89)	54.98 (34.48-75.47)	31.60 (18.60-44.60)

CI – Confidence interval

spine.[14] Due to relative rarity of this tumor, epidemiological studies such as estimation of tumor incidence rate, mean age, sex predilection, tumor location, and survival can be performed by studies based on series collected over a long period of time with highly variable treatment methods or through a large number of cases derived from national or regional registries. To our best knowledge, this is the first epidemiologic study of chordoma according to a National Database Registry in the Eastern Mediterranean Region as described by the WHO.[15]

Presumably, the incidence rate of chordoma is different between races which demonstrate the need to perform regional epidemiological studies among patients with nearly similar genetic characteristics. Moreover, the incidence rate of chordoma varied between countries and reported differently from 0.18 to 0.84 per million persons per year.[16-18] While McMaster reported 0.8 per million persons-year as the incidence rate of chordoma in the United States, other series in England, Sweden, and Taiwan showed lower rates as 0.4, 0.5, and 0.4 respectively.[8,19,20] According to our results, the ASIR of chordoma in Iran was 0.28 per million person-years which showed lower incidence rate respect to other studies. The incidence rate among males was nearly twice in comparison with females (0.37 vs. 0.19).

The majority of large series show male predominance for chordoma;[8,16,17] however, in few studies, female predilection has been reported.[21] Our results showed that 65.5% of cases were male which was in concordance with other studies. This predilection was also observed among chordoma of the mobile spine and sacrum and among NOS and chondroid chordoma.

Chordoma of the mobile spine and sacrum affects middle age population in their late fifties and is rare (<5%) among children and adolescents.[4,22] In a study by Stiller *et al*.[23] among European population under RARECARE (Surveillance of Rare Cancers in Europe) project, the crude incidence rate was 1.0 per million persons per year for persons older than 65 years. According to SEER and California Cancer Registry, the median age of chordoma has been reported as 57 and 55 years.[17,24] The mean age of diagnosis in our study was 53.4 years, which showed slightly younger age respect to other series. The current study showed that while sacral

chordoma affects males in older ages, the mean age of mobile spine chordoma is higher among females.

Chordoma is generally restricted to the axial skeleton in three major sites including skull base, mobile spine, and sacrum. While earlier series revealed an equal distribution of chordoma among these anatomical sites,[18] in a more recent study by Lee *et al*.,[17] the majority of cases were found in cranial region and nearly similar distribution between the mobile spine (27.2%) and sacrum (26.9%) was observed. In another study based on California Cancer Registry,[24] by considering only mobile spine and sacrum, 51.2% of cases were mobile spine and 48.2% were sacral chordoma. Ardekani *et al*.[25] reported 39 cases of chordoma in Iran including 21 (53.8%) clivus chordoma, 13 (33.3%) cases in the sacrum, and 5 (12.8%) in the mobile spine. According to our results, the number of chordoma cases in the sacrum (67.2%) was almost more than twice compared to the mobile spine (32.8%). The current study also showed that chondroid chordoma occurs among younger patients in comparison to NOS chordoma.

Survival of chordoma is attributed to different factors such as tumor size, diagnostic and treatment method, surgical technique, and local recurrence.[21,26] According to SEER and NCDB, the 5-year survival of chordoma is 64%–73%.[6,17,27] Higher 5-year survival (84%) also has been reported in smaller series.[21] In the current study, the mean survival time was 4.5 years which is lower than larger series like SEER (6.2 years).[4] The cumulative survival time was 0.29 which indicates the low survival probability for patients with chordoma in Iran with existing therapeutic strategies.

Limitations

This study represents a preliminary report of epidemiologic characteristic of chordoma in Iran according to a national registry (INCR). While INCR has many advantages, it is not without limitations. In this registry, due to incomplete histopathology registration of the patients and lack of histology codes, histology evaluation was not possible. The communication data for the majority of patients were not available or have been changed, and limited patients were available to compile the questionnaire for survival analysis. Due to the low number of cases enrolled for survival analysis, it did not provide reliable data for this valuable issue. Lack of surgical approach and treatment type was also a weak point for this study. Methodological limitations of the study including sparse data and sparse-data bias should also be noted.

Epidemiology characteristic of chordoma in Iran is nearly similar to other studies; however, lower ASIR of chordoma was observed in Iran (0.28 per million person-years). The incidence of sacral chordoma is nearly twice respect to the mobile spine. While men affected by sacral chordoma in relatively older age, the female patients had higher mean age in case of mobile spine chordoma. The survival rate of chordoma is significantly lower in comparison with other studies.

Main points

Cancer epidemiology should be performed for each country separately to understand characteristic of each cancer among patients with nearly similar races. The incidence rate of chordoma in Iran is 0.28 with male predilection and affects more frequently the sacral region.

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Conflicts of interest

There are no conflicts of interest.

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