Analysis of Prognostic Factors for Patients with Chordoma with Use of the California Cancer Registry

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Background: Chordoma is the most common primary malignant tumor of the spine. It is extremely rare and has been studied primarily in single-institution case series. Using data from a large, population-based cancer registry, we designed the present study to examine the outcome for patients with chordoma and to determine relevant prognostic factors.

Methods: A retrospective analysis of the California Cancer Registry database was performed to identify patients with a diagnosis of chordoma in the years 1989 to 2007. Comparisons examined differences in demographics, disease characteristics, treatment, and survival. Survival analyses were performed with use of the Kaplan-Meier method with log-rank tests and Cox proportional hazards models.

Results: Four hundred and nine patients with chordoma were identified; 257 (62.8%) were male and 152 (37.2%) were female. With regard to racial or ethnic distribution, 266 patients (65%) were white; ninety-three (22.7%), Hispanic; forty-three (10.5%), Asian or other; and seven (1.7%), black. The site of presentation was the head in 202 patients (49.4%), spine in 106 patients (25.9%), and pelvis and/or sacrum in 101 patients (24.7%). Hispanic race ($p = 0.0002$), younger age (less than forty years; $p < 0.0001$), and female sex ($p = 0.009$) were associated with cranial presentation, whereas older age (forty years or older; $p < 0.0001$) was associated with pelvic presentation. After adjustment for clinically relevant factors, a significantly decreased risk of death for chordoma-specific survival was seen for Hispanic race (hazard ratio = 0.51, 95% confidence interval [95% CI], 0.28 to 0.93; $p = 0.03$), high socioeconomic status (hazard ratio = 0.8, 95% CI, 0.67 to 0.95; $p = 0.01$), and local excision and/or debulking (hazard ratio = 0.38, 95% CI, 0.18 to 0.81; $p = 0.01$). Large tumor size was independently associated with an increased risk of death (hazard ratio = 2.05, 95% CI, 1.01 to 4.20; $p = 0.048$).

Conclusions: In this study, the survival of patients with chordoma was significantly better for those who were Hispanic and had a small tumor, high socioeconomic status, and surgical intervention.

Level of Evidence: Prognostic Level II. See Instructions for Authors for a complete description of levels of evidence.

Arising from notochordal remnant tissue, chordoma is the most common primary malignant tumor affecting the spine¹⁻⁸. Its occurrence is extremely rare and has been described primarily in small, single-institution case series^{1,2,6,8-11}. Although few population-based surveys of chordoma exist, they are limited by small numbers of cases or they describe only the incidence and mortality without a detailed analysis on prognostic factors for survival³⁻⁵. These surveys

include a review of the Leeds Regional Bone Tumour Registry in the United Kingdom, which recorded only twenty-eight cases of chordoma from 1958 to 2001. In addition, 221 cases from 1973 to 1987 and 400 cases from 1973 to 1995 were reviewed in two separate studies with use of the Surveillance, Epidemiology, and End Results (SEER) program¹². Neither study included an analysis of prognostic factors nor contained substantial data on race or ethnicity or socioeconomic status.

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In contrast to other cancer registries, the California Cancer Registry (www.ccrca.org) contains detailed data on patient demographics, tumor staging, and treatment modalities¹³. This makes the California Cancer Registry particularly valuable for the study of rare tumors such as chordoma, and allows us to describe the epidemiological patterns of this uncommon disease. Using the California Cancer Registry, we evaluated the cases of 409 patients with chordoma from 1989 to 2007 to examine the outcome and determine the relevant prognostic factors for survival.

Materials and Methods

Study Population

We performed a retrospective, case-only analysis of patients with chordoma in the California Cancer Registry database. The California Cancer Registry is part of the National Cancer Institute's SEER program and is the largest contiguous-area population-based cancer registry in the world¹³. Standardized data collection and quality control procedures have been in place since 1988¹³⁻¹⁶. Case reporting is estimated at 99% for the entire state of California, with a follow-up completion rate of >95% for most tumors¹⁴. Data were abstracted from medical and laboratory records by trained tumor registrars¹⁵. Tumor site and histological findings were coded according to World Health Organization criteria in the International Classification of Diseases for Oncology, Third Edition (ICD-O-3)¹⁶. Cases of patients were extracted on the basis of the histological types for chordoma not otherwise specified, chondroid chordoma, and dedifferentiated chordoma (histological codes 9370, 9371, and 9372, respectively) according to ICD-O-3. Cases of patients were excluded if they had nonspecific or unknown SEER extent of disease classification or if their diagnosis dates and follow-up dates were inconsistent.

Data were obtained on the cases of 409 patients with chordoma during 1989 to 2007 in the California Cancer Registry. The cases of patients were identified through high-quality reporting sources (hospital inpatient-outpatient centers; oncology treatment centers; laboratories; private practitioners; or nursing home, convalescent home, and hospice facilities). Recorded data included demographic information, anatomical site of presentation, presence of metastasis, treatment during the first course of therapy, socioeconomic status, and vital status^{14,15}. Sites of presentation were compiled according to ICD-O-3 topography codes, and were grouped in similar fashion to the review by McMaster et al.⁵. Cranial sites included the oropharynx-nasopharynx and sinuses; bones of the skull and face; connective, subcutaneous, and soft tissues of the head, face, and neck; eye; and any structure in the brain. Spinal sites included the spinal cord; mediastinum; retroperitoneum, connective, subcutaneous, and soft tissue of the trunk and thorax; nervous system not otherwise specified; and the vertebral column exclusive of sacrum and coccyx. Included within sacral and/or pelvic sites were bones of the pelvis and sacrum; connective, subcutaneous, and soft tissue of the pelvis; and pelvis not otherwise specified.

Socioeconomic status is denoted as a single index variable in the California Cancer Registry with use of statewide measures of education, income, and occupation from census data, as described previously by Yost et al.¹⁷. The socioeconomic status index used is based on the principal component analysis of census block-level data in the California Cancer Registry linked to census data assessing the following areas: education level, median household income, proportion below 200% of the poverty level, median house value, median rent, percent employed, and percent with blue-collar employment¹⁷⁻²¹. It is of note that the proportion "below 200% of poverty" was used because of its greater sensitivity; it is defined as all those in poverty and those who have income above poverty but less than two times their poverty threshold²². Essentially, each aspect mentioned serves as a proxy for socioeconomic status and is weighted together to derive a score. Scores were divided into five categories, with the higher scores representing higher socioeconomic status. Quintiles for the socioeconomic status score were analyzed²³.

TABLE I Demographic and Clinicopathologic Data

	No.	%
All patients	409	100
Sex		
Male	257	62.8
Female	152	37.2
Age (yr)		
0-19	38	9.3
20-39	71	17.4
40-49	63	15.4
50-59	73	17.8
60-69	76	18.6
≥70	88	21.5
Race		
White	266	65.0
Hispanic	93	22.7
Black	7	1.7
Asian	38	9.3
Other	5	1.2
Socioeconomic status		
Lowest	49	11.9
Low	80	19.6
Middle	80	19.6
High	102	24.9
Highest	98	24.0
Primary site		
Head	202	49.4
Spine	106	25.9
Pelvis and/or sacrum	101	24.7
Tumor size		
<5cm	92	22.5
≥5cm	106	25.9
Unknown	211	51.6
*Radiation		
Yes	187	46.1
No	219	53.9
*Chemotherapy		
Yes	21	5.2
No	380	94.8
Metastasis		
Yes	35	8.6
No	374	91.4
*Surgery		
Yes	342	84.4
No	63	15.6
Operation		
None	63	15.4
Local	140	34.2
Radical	139	34.0
Not otherwise specified	63	15.4
Unknown	4	1.0
*Treatment		
Surgery	184	45.4
Radiation	28	6.9
Both	158	39.0
Neither	35	8.6

*Information not available for all patients.

Cause of death was recorded according to ICD-O criteria in effect at the time of death. Chordoma-specific survival was defined as death due to musculoskeletal cancer. Hospital registrars contacted chordoma patients annually, and California Cancer Registry staff annually reviewed state death certificates to identify the cases of deceased patients in the registry. The last date of follow-up was either the date of death or the last date of contact. In 2006 to 2007, complete follow-up was available for 394 (96.3%) of the 409 patients.

Statistical Analysis

The clinical characteristics, including age, sex, race or ethnicity, presence of metastasis, tumor size, anatomic tumor site, socioeconomic status quintile, and treatment were analyzed with the Pearson chi-square test or Fisher exact test for categorical and dichotomous variables, and the nonparametric Kruskal-Wallis test was used for comparison of continuous variables (age) for more than two groups. The term *chordoma-specific survival* was used to define death from cancer of the musculoskeletal system. Deaths from all other causes were censored from the analysis of the California Cancer Registry. Other causes of a censoring event in the chordoma-specific survival analysis included patients who were lost to follow-up or were alive at the end of the follow-up period. Life tables and Kaplan-Meier curves were generated for each category, and curves were compared with the log-rank test. Multivariate survival analysis was utilized to calculate chordoma-specific survival with use of Cox proportional hazards ratios. We restricted the analysis to events that occurred in the first six years to adjust for the shorter follow-up times for patients who had received the diagnosis more recently. For the patients with unknown chemotherapy, radiation, surgical treatment, and tumor size, dummy variables were assigned to maintain all cases in the multivariate analysis. Socioeconomic status was viewed as a continuous variable to test for significant trends in our multivariate model.

Ethical Considerations

This study involved analysis of existing data from the California Cancer Registry database with no subject intervention. No identifiers were linked to subjects. The study was approved by the University of California, Irvine institutional review board under the category "exempt status."

Source of Funding

Funding in support of this project was derived from the University of California Department of Medicine and the Chao Family Comprehensive Cancer Center.

Results

Demographic Data

The cases of 409 patients with chordoma were identified in the years 1989 to 2007. The specific histological findings included 395 patients with chordoma that was not otherwise specified, twelve with chondroid chordoma, and two with de-differentiated chordoma. The median age was fifty-five years, and demographic data for all patients are shown in Table I. Due to the relatively small numbers of patients, those who were black, Asian or Pacific Islanders, or other race or ethnicity were combined into one group labeled "other" for the purposes of statistical analyses.

Across the racial and ethnic groups, a significantly smaller proportion of Hispanic patients (8.6%) were in the oldest age group (seventy years or older) compared with others (34% were seventy years or older) and whites (24% were seventy years or older) ($p = 0.0008$). Significant differences in socioeconomic status were also observed across the major ethnic groups. While a large proportion of whites and those categorized as "other" belonged to the highest socioeconomic status category (28.2% and 28.0%, respectively), significantly

TABLE II Six-Year Chordoma-Specific Survival

Variable	6-Year Chordoma-Specific Survival (%)	P Value
Sex		0.84
Male	73.7	
Female	73.4	
Age (yr)		0.02
0-19	59.7	
20-39	78.8	
40-49	84.7	
50-59	76.3	
60-69	71.7	
≥70	63.5	
Race		0.65
White	73.0	
Hispanic	74.2	
Other	76.0	
Socioeconomic status		0.37
Primary site		0.48
Head	71.1	
Spine	78.2	
Pelvis and/or sacrum	73.3	
Tumor size		0.11
<5 cm	77.5	
≥5 cm	66.8	
Radiation		0.50
Yes	73.7	
No	74.1	
Chemotherapy		0.01
Yes	57.2	
No	75.0	
Metastasis		1.00
Yes	75.0	
No	73.5	
Surgery		0.27
Yes	74.6	
No	70.6	
Operation		<0.0001
None	70.6	
Local	83.8	
Wide	69.4	
Not otherwise specified	69.0	
Unknown	37.5	
Treatment		0.03
Surgery	82.7	
Radiation	76.4	
Both	72.3	
Neither	59.9	

fewer Hispanic patients (9.7%) belonged to this socioeconomic status category ($p < 0.0001$). A significantly larger proportion of Hispanic patients belonged to the lowest socioeconomic status category (29%) compared with those in other ethnic groups (5.6% of the white patients and 14% of those categorized as "other"; $p < 0.0001$). Socioeconomic status was not

significantly associated with age ($p = 0.11$), site of presentation ($p = 0.85$), metastatic disease ($p = 0.76$), large tumor size ($p = 0.98$), radiation therapy ($p = 0.29$), chemotherapy ($p = 0.16$), or surgical intervention ($p = 0.83$).

Clinicopathologic Characteristics

Clinicopathologic data for all patients are summarized in Table I. With use of chi-square tests, large tumors were not found to be associated with low socioeconomic status ($p = 0.98$), metastatic disease ($p = 0.4$), chemotherapy ($p = 0.22$), radiation therapy ($p = 0.85$), or surgical intervention ($p = 0.11$). Large tumors occurred more frequently in sacral and/or pelvic sites and small tumors, in cranial sites. Furthermore, 86.6% of sacral and/or pelvic tumors were large, while only 48.5% of spinal and 32.7% of cranial tumors were large ($p < 0.0001$). There was a significant correlation between age and location of disease, with 70.6% of younger patients (less than forty years old) having cranial involvement and only 41.7% of older patients (forty years or older) having cranial involvement ($p < 0.0001$). Patients forty years or older more commonly presented with sacral and/or pelvic involvement (30.7% of older patients compared with 8.3% of younger patients; $p < 0.0001$). Location of disease was also related to sex, as 59.2% of female patients had cranial involvement compared with only 43.6% in male patients ($p = 0.009$). Across the racial and ethnic groups, the proportion of patients with cranial involvement was significantly larger in the Hispanic group (67.7%) than in the group of white patients (42.5%) and the group categorized as “other” (52.0%) ($p = 0.0002$). The location of disease was not related to the presence of metastasis, radiation therapy, or chemotherapy.

The treatment profile for all patients is also included in Table I. Our analysis showed that metastatic disease was associated with a higher rate of chemotherapy ($p < 0.0001$). Also, the rate of chemotherapy was higher for patients in the youngest age group (zero to nineteen years) than for those in all other age groups (28.9% and 2.7%, respectively). Among the patients who did not have surgery, forty (63.5%) had not had surgery recommended, six (9.5%) had contraindications to surgery because of comorbidities, and five (7.9%) had declined surgery; the reason was unclear for twelve patients (19.0%). For the forty patients who had not had surgery recommended, thirty-two (80%) were fifty years or older. The six patients who had contraindications to surgery because of comorbidities were fifty years of age or older ($p < 0.0001$) and white ($p = 0.0039$). Overall, the proportion of patients who did not have surgery was significantly greater in the oldest age group (more than seventy years) than in all other age groups (38.8% and 9.4%, respectively; $p < 0.0001$). A difference in surgical intervention was also seen across the racial and ethnic groups, with surgical treatment in only 70.0% of the patients categorized as “other” compared with 85.1% of white patients and 90.3% of Hispanics ($p = 0.005$). Surgical intervention was also seen more frequently with cranial disease (90.1%) than with spinal (84.0%) or pelvic (73.5%) disease ($p = 0.001$).

Treatment groups for the 405 patients with available information included radiation therapy only (twenty-eight

TABLE III Multivariate Analysis (Cox Models) of Chordoma-Specific Survival

	Hazard Ratio (95% Confidence Interval)	P Value
Age	1.01 (0.99-1.02)	0.23
Sex		
Male	1.00*	–
Female	0.94 (0.59-1.52)	0.81
Race or ethnicity		
White	1.00*	–
Hispanic	0.51 (0.28-0.93)	0.03
Other	0.58 (0.27-1.25)	0.16
Socioeconomic status	0.8 (0.67-0.95)	0.01
Stage		
Local disease	1.00*	–
Metastasis	0.79 (0.37-1.69)	0.54
Tumor size		
<5 cm	1.00*	–
≥5 cm	2.05 (1.01-4.20)	0.048
Anatomic site		
Spine	1.00*	–
Head	1.74 (0.97-3.15)	0.07
Pelvis and/or sacrum	0.78 (0.39-1.57)	0.49
Radiation		
No	1.00*	–
Yes	0.81 (0.52-1.27)	0.35
Chemotherapy		
No	1.00*	–
Yes	2.85 (1.26-6.45)	0.01
Surgery		
None	1.00*	–
Local excision and/or debulking	0.38 (0.18-0.81)	0.01
Wide excision	0.76 (0.39-1.50)	0.43
Not otherwise specified	0.83 (0.39-1.78)	0.63

*Reference group.

patients; 6.9%), surgery only (184 patients; 45.4%), both surgery and radiation therapy (158 patients; 39%), and neither surgery nor radiation (thirty-five patients; 8.6%).

Cause of Death Analysis

One hundred and ninety-nine (48.7%) of the 409 chordoma patients died. One hundred and sixteen (58.3%) of the deaths were due to musculoskeletal cancer. The cause of death was unknown for twenty-seven patients. There was no significant difference in cause of death among the different racial or ethnic groups ($p = 0.14$), socioeconomic status groups ($p = 0.06$), or by sex ($p = 0.38$).

Univariate Survival Analysis

For all patients, chordoma-specific survival was 78.6% at five years and 62.2% at ten years. Five-year chordoma-specific survival rates over different time periods were similar, with a

Fig. 1

Chordoma-specific survival by treatment group, with 0 (purple) indicating neither surgery nor radiation; 1 (red), surgery only; 2 (green), radiation only; and 3 (brown), both surgery and radiation; the difference was significant ($p = 0.03$).

rate of 77.5% for 1989 to 1994, 78.0% for 1995 to 2000, and 83.1% for 2001 to 2007 ($p = 0.75$). Patients in the youngest (zero to nineteen years) and oldest (more than seventy years) age groups had the poorest chordoma-specific survival rate ($p = 0.02$). Chemotherapy usage was associated with poor chordoma-specific survival ($p = 0.01$). Survival among the treatment groups (surgery without radiation, surgery with radiation, surgery without surgery, and neither) were compared, and patients receiving neither surgery nor radiation had the poorest chordoma-specific survival ($p = 0.03$) (Fig. 1). When treatment groups were compared with each other, no significant difference was seen ($p = 0.57$). Results from univariate survival analyses are summarized in Table II.

Figure 2 illustrates chordoma-specific survival curves for the different surgical treatments for all patients. The different operative approaches were examined separately among cranial, spinal, and pelvic and/or sacral disease presentations and for local versus metastatic disease. Among patients with cranial disease (see Appendix), surgical resection was significantly associated with better chordoma-specific survival ($p < 0.0001$). This was also true for patients with pelvic and/or sacral disease (see Appendix; $p = 0.02$). However, better chordoma-specific survival was seen only for patients who had local resection and not for those who underwent wide resection. For patients with

spinal disease, survival differences were not significant (see Appendix; $p = 0.89$). Furthermore, for all patients with non-metastatic disease at presentation, surgical resection led to significantly better chordoma-specific survival ($p < 0.0001$). Only thirty-five patients had presented with metastatic disease, and no significant survival differences were found between the surgical treatments in this group ($p = 0.13$).

Multivariate Survival Analysis

The variables of age, sex, race or ethnicity, location of disease, tumor stage, socioeconomic status, chemotherapy, radiation therapy, tumor size, and surgery status were included in a multivariate Cox regression model for chordoma-specific survival (Table III). After adjustment for each of the above variables, Hispanic patients were found to have a significantly decreased risk of chordoma-specific death (hazard ratio = 0.51; 95% confidence interval [95% CI], 0.28 to 0.93; $p = 0.03$). Age at diagnosis did not remain an independent risk factor for poor survival in the adjusted analysis. Tumor size of ≥ 5 cm was associated with an increased risk of death (hazard ratio = 2.05; 95% CI, 1.01 to 4.20; $p = 0.048$). Compared with patients who did not undergo surgery, those who received local excision had a significantly decreased risk of death (hazard ratio = 0.38, 95% CI, 0.18 to 0.81; $p = 0.01$). In terms of socioeconomic status, a

Fig. 2

Chordoma-specific survival by surgical intervention, with 1 (red) indicating local resection; 2 (blue), no surgery; 3 (green), wide resection; 4 (brown), surgery not specified; and 5 (purple), unknown; the difference was significant ($p < 0.0001$).

significant trend was found, with patients who had a higher socioeconomic status having a decreased risk of death ($p = 0.01$).

Discussion

Using data from a large, population-based cancer registry, we described the epidemiology of chordoma and identified new prognostic indicators. In our series of 409 patients, chordoma was most commonly found in white males, with a median age of fifty-five years. After adjusting for most demographic and clinicopathologic factors, we found low socioeconomic status and large tumor size to be independent risk factors for poor survival. In contrast, Hispanic race and surgical excision were independently associated with improved survival.

Although chordoma has historically been addressed surgically, treatment algorithms for the disease are not clearly defined. Our study supports past series, showing surgical excision to be related to significantly better survival^{1-7,9-11,24,25}. This was especially true in our subset analysis of surgical intervention by treatment group as well as by location of disease. For patients with cranial and those with pelvic or sacral presentations, the improved survival with surgical resection was significant. The same was not found with disease in other regions of the spine axis, however, which may reflect a greater difficulty in achieving clear surgical margins with spinal disease. In the

study by Bergh et al.¹¹, patients with a large tumor size and unclear surgical margins had the poorest survival. Radiation therapy in a small proportion of their patients was of limited value. In contrast, the studies by Boriani et al.¹ and Fuller and Bloom²⁶ found that patients treated with radiation therapy in addition to wide excision had improved survival. However, only thirty-seven and twenty-seven patients, respectively, were studied, and no direct comparison was made between patients who had wide excision and those who had wide excision with radiation therapy. In the review of the cases of twenty-three patients with lumbosacral chordoma by Cheng et al.², patients who had marginal tumor excision received radiation therapy. Survival was improved, but a high rate of recurrence remained for all marginally excised tumors. In our review, radiation therapy was not independently related to better survival. In a direct comparison of patients who received surgery alone and those who had surgery with radiation, no difference in survival was found. It may be that chordoma is resistant to radiation therapy, or that proper radiation therapy protocols have yet to be developed. In addition, radiation therapy has been traditionally reserved for patients with complex or inoperable tumors, unclear resection margins, and palliative care. This may create a substantial selection bias and undermine any positive effects of radiation therapy.

The diversity of the California population afforded us the unique opportunity of investigating chordoma as it relates to race or ethnicity. McMaster et al.⁵ reviewed the SEER database and found that the age-adjusted incidence rate of chordoma was 0.08 per 100,000 and that the disease was rare among blacks. California census data in 1990 showed that blacks made up 10.8% of the population, yet only 1.7% of all patients in our series were black²⁷. Interestingly, chordoma-specific survival was significantly and independently improved in Hispanic patients compared with other racial or ethnic groups. This survival difference has not been described before, and the reasons for it are unclear. Hispanic race was found to be significantly associated with a younger age, cranial disease, and a higher rate of surgical intervention, which were all associated with better survival. However, Hispanic race was also significantly associated with low socioeconomic status, which in our multivariate model was related to an increased risk of death. The survival differences may represent competing risks, as other causes of death affect certain races or ethnicities more. We examined the cause of death across the racial and ethnic groups, and no significant association was detected ($p = 0.14$). This finding highlights the possibility of inherent differences in the disease process and warrants further investigation.

The socioeconomic status descriptor used in the present study has been associated with the survival rate of patients with a variety of solid tumor types: breast, pancreas, colorectal, melanoma, and lung cancer^{17,18,20,21}. The socioeconomic status descriptor used in the California Cancer Registry is not related to age, sex, tumor location, large tumor size, metastasis, chemotherapy, radiation therapy, or surgical intervention. Yet, the socioeconomic status descriptor was an independent risk factor for poor survival in patients with chordoma. The socioeconomic status index may reflect many factors, such as access to care and even quality of care. Our data demonstrate that significant health-care disparities exist.

In respect to our study limitations, it is important to identify data that are not included in the California Cancer Reg-

istry. In addition to patient comorbidities, data on specific radiation techniques, doses, and durations; chemotherapy regimens; and the status of surgical resection margins are not included in the California Cancer Registry. Patient medical history, including medications, tobacco or alcohol use, employment, environmental exposures, and family history, was also not recorded in the California Cancer Registry. Other relevant details that may have helped to clarify our analysis but were not in the California Cancer Registry include whether the patient was treated at a cancer center and whether the treating surgeon was specifically trained in oncology. Lastly, although our study is one of the largest population-based reviews of chordoma, the number of patients is small for an adequate study of the epidemiology of the disease.

Appendix

Figures showing chordoma-specific survival for patients with cranial, pelvic and/or sacral, and spinal disease are available with the online version of this article as a data supplement at jbjs.org. ■

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