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Incidence and distribution of chordoma: A study analysing data from the "Surveillance, Epidemiology and End Results" program.

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Background:
Only a few studies exist that describe the frequency distribution and incidence of chordoma, a rare tumor originating from remnants of the notochord. Apart from single-institution case series there are two bigger population-based surveys analyzing a number of 400 (National Cancer Institute, 1973-1995) and 409 (California Cancer Registry, 1989-2007) cases. With the use of the most recent dataset from the Surveillance, Epidemiology and End Results (SEER) program of the National Cancer Institute we conducted a retrospective analysis calculating distribution and age-adjusted incidence rates for 706 cases of microscopically confirmed chordoma from 2000 to 2009.
Methods:
The Surveillance, Epidemiology and End Results program combines the information of 18 registries throughout the United States covering approximately 28% of the population. The WHO's "International Classification of Diseases for Oncology, 3rd Edition" morphological Codes for chordoma (9370/3 chordoma NOS, 9371/3 chondroid chordoma, 9372/3 dedifferentiated chordoma) were used to identify and include relevant cases. With the help from the SEER*Stat statistical software, we calculated frequencies and age-adjusted incidence rates and analyzed them by gender, age, race, and primary site of presentation.
Results:
The 706 cases are composed of 654 chordomas not otherwise specified, 46 chondroid chordomas and 6 dedifferentiated chordomas. The overall age adjusted incidence rate for chordoma is 0.09 per 100,000 and concerning gender it is higher in males (0.11/100,000) than in females (0.07/100,000; rate ratio: 0.61). The median age at diagnosis is 57 (range: 0-91) and the incidence rates increase with age. In blacks the incidence rate is with 0.03/100,000 significantly lower than in whites (0.10/100,000). Hispanics have a chordoma incidence rate of 0.08/100,000 in comparison to a rate of 0.09/100,000 in non-Hispanics. The distribution of the primary site of presentation is as follows: Cranial (42%, n=300); spinal (26%, n=182); sacral (30%, n=212); extra-axial, non categorizable and unknown site (2%, n=12).
Conclusion:
With the use of the latest Surveillance, Epidemiology and End Results data (SEER18), which has been released at the end of spring 2012, this study provides substantial and up-to-date information on distribution and incidence patterns of chordomas in the United States.

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