Cancer Epidemiology 54 (2018) 90–94

Contents lists available at ScienceDirect

Cancer Epidemiology

journal homepage: www.elsevier.com/locate/canep

Evaluation of racial disparities in pediatric optic pathway glioma incidence: Results from the Surveillance, Epidemiology, and End Results Program, 2000–2014

Erin C. Peckham-Gregory[a,b], Roberto E. Montenegro[c], David A. Stevenson[d], David H. Viskochil[e], Michael E. Scheurer[a,b,1], Philip J. Lupo[a,b,1], Joshua D. Schiffman[f,*,1]

[a] Department of Pediatrics, Section of Hematology-Oncology, Baylor College of Medicine, One Baylor Plaza, MS: BCM305, Houston, TX 77030, United States
[b] Texas Children's Cancer Center, Texas Children's Hospital, Feigin Center, 1102 Bates St, Houston, TX 77030, United States
[c] Department of Psychiatry and Behavioral Medicine, The University of Washington School of Medicine, 4800 Sand Point Way NE, Seattle, WA 98105, United States
[d] Division of Medical Genetics, Stanford University, 300 Pasteur Drive, Boswell Building A097, Stanford, CA 94304, United States
[e] Division of Medical Genetics, University of Utah School of Medicine, 295 Chipeta Way, Salt Lake City, UT 84108, United States
[f] Department of Pediatrics and Department of Oncological Sciences, Huntsman Cancer Institute, University of Utah School of Medicine, 2000 Circle of Hope, Salt Lake City, UT 84112, United States

ARTICLE INFO

Keywords:
Optic pathway gliomas
Racial disparity
Epidemiology
SEER

ABSTRACT

Background: Racial predilection to pediatric cancer exists; however optic pathway glioma (OPG) risk differences by race/ethnicity are undefined. We estimated differences in OPG incidence across racial/ethnic groups in a multi-state cancer surveillance registry in the United States.

Methods: OPG data were obtained from the Surveillance, Epidemiology, and End Results (SEER-18) Program, 2000–2014. Race/ethnicity was categorized as: White; Black; Asian; Other; and Latino/a ("Spanish-Hispanic-Latino"). Latino/a included all races, while all other categories excluded those identified as Latino/a. Age-adjusted incidence rates and rate ratios (IRR) with 95% confidence intervals (CIs) were generated in SEER-STAT (v8.3.4).

Results: Data on 709 OPG cases ages 0–19 were abstracted from SEER-18. Minority children experienced lower age-adjusted OPG incidence rates compared to White children (IRR_{Black} = 0.38, 95% CI: 0.28–0.50; IRR_{Asian} = 0.41, 95% CI: 0.29–0.58; and $IRR_{Latino/a}$ = 0.39, 95% CI: 0.32–0.48). In subgroup analyses among the highest risk age categories (0–4, 5–9), minority children experienced lower incidence rates compared to White children. Specific patterns for Latinos/as also emerged. Latino/a children ages 0–4 experienced the lowest incidence rates of all racial/ethnic groups compared to Whites (0.24 per 100,000 person-years versus 0.66 per 100,000 person-years, respectively), whereas among those ages 5–9, Black and Asian children experienced the lowest incidence rates (0.08 per 100,000 person-years each).

Conclusions: Incidence of OPGs was highest among White children. This study represents one of the largest to assess differences in OPG susceptibility by race/ethnicity. These findings may inform future studies that seek to evaluate modifying factors for this pediatric tumor including tumorigenesis, treatment, outcome, and long-term late effects.

1. Introduction

Optic pathway gliomas (OPG) represent 2–5% of intracranial brain tumors in children [1], and are generally low-grade pilocytic astrocytomas with growth patterns ranging from indolent to highly proliferative [2]. Tumor development occurs in the structures of the visual pathway, including the optic nerve and chiasm [3]. Over 65% of OPGs are diagnosed in children under the age of 5 years old [4,5], with the majority of remaining cases diagnosed between 5 and 15 years of age [1,6]. Overall incidence is estimated at 3–4 cases per 100,000 [7],

Abbreviations: OPG, optic pathway glioma; NF1, neurofibromatosis type 1; SEER Program, surveillance, epidemiology, and end results; US, United States; IR, incidence rate; IRR, age-adjusted incidence rate ratio; CIs, confidence intervals; OR, odds ratio

* Corresponding author.
E-mail addresses: Erin.Peckham@bcm.edu (E.C. Peckham-Gregory), Joshua.Schiffman@hci.utah.edu (J.D. Schiffman).
[1] These authors contributed equally to this work.

https://doi.org/10.1016/j.canep.2018.04.005
Received 9 December 2017; Received in revised form 6 April 2018; Accepted 7 April 2018

with a 5-year survival rate over 95% [8]. While the mortality rate of OPGs is low, over 75% of patients suffer from severe visual impairment [9,10], which can often lead to life-long blindness [11]. One of the strongest risk factors for developing OPGs includes germline mutations in the *NF1* tumor suppressor gene, as reflected by 50–60% of OPGs occurring in individuals diagnosed with neurofibromatosis type 1 (NF1) [3,12–14].

Aside from NF1, relatively little is known regarding the impact of other inherited factors, such as race/ethnicity, on the epidemiology of OPGs [8,15]. Some reports suggest that NF1-associated OPG incidence rates may be lower in minority groups compared to non-Latino/a Whites; however, these studies have been limited in sample size and largely descriptive in nature [16,17]. Other pediatric tumors, including Ewing Sarcoma [18], demonstrate strong racial predilection impacting predisposition, clinical manifestations, and health outcomes [18]. Therefore, we sought to evaluate racial and ethnic differences in the incidence of OPGs using the most recently available data from the Surveillance, Epidemiology, and End Results (SEER) Program of the National Cancer Institute in the United States (US).

2. Materials and methods

2.1. Study population

OPG incidence and case listing data were obtained from the newest SEER database: SEER 18 (April 2017 release). Incidence data from the time period 2000–2014 were used since SEER 18 only has data available from 2000 onwards for all eighteen registry sites included. SEER registries represent the following states: Alaska, Arizona, Connecticut, California (Greater California, Los Angeles, San Francisco-Oakland, and San Jose-Monterey), Georgia (Atlanta and Rural Georgia), Hawaii, Iowa, Louisiana, Kentucky, Michigan (Detroit), New Jersey, New Mexico, Utah, and Washington (Seattle-Puget Sound). Case data were obtained by these SEER registries from medical records [19]. The SEER Program provides population-based cancer incidence and survival data, representing approximately 30% of the US population and a large proportion (38%) of the US Latino/a population, based on 2013 data [19]. Of note, the terms “Hispanic,” “Hispanic/Latino/a,” and “Latino/a” are used interchangeably to describe individuals of Spanish speaking descent. Hispanic can refer to those from Spanish speaking countries including Spain, while Latino is now recognized to reflect those individuals originally from Latin (Central) America. The SEER Program currently uses the all-inclusive term “Spanish-Hispanic-Latino,” but similar to other publications [20], we will use the term *Latino/a* to reflect the geographical origins of a large proportion of this ethnic group.

OPG cases were abstracted from SEER 18 if they had malignant behavior; a primary site originating in the optic nerve (C72.3); and histologic types 9380 (Glioma), 9382 (Mixed Glioma), or 9400–9421 (Astrocytoma) as defined by the International Classification of Diseases for Oncology 3rd edition (ICD-O-3). SEER has defined the term "malignant" to mean cases had a behavior code of 3 (“malignant”) in both ICD-O-2 and ICD-O-3 databases (https://seer.cancer.gov/behavrecode/). As OPGs occur primarily in those patients less than 19 years of age, we restricted our analysis to those individuals with tumors between 0–19 years of age [3]. Age at and year of diagnosis, sex, race/ethnicity, and number of primary tumors were abstracted for these analyses.

2.2. Statistical analyses

SEER 18 incidence data were used to estimate incidence rates (IR), age-adjusted incidence rate-ratios (IRR), and 95% confidence intervals (CIs) to assess the association between race/ethnicity and incidence of OPGs during 2000–2014. IRs, IRRs, and 95% CIs were estimated using SEER*Stat software version 8.3.4 (seer.cancer.gov/seerstat). Incidence rates were age-adjusted to the 2000 US standard population using single ages up to 19 years of age for OPGs, and the Tiwari method was used for confidence interval calculations [21]. We estimated incidence rates for all cases ages 0–19. To identify any differences in OPG incidence by race/ethnicity among those in the highest risk age categories (0–4 and 5–9), we conducted subgroup analyses. Demographic characteristics were tabulated in STATA v13.1.

Using the SEER*Stat variables, we created a merged variable “race/ethnicity” by combining the SEER variables “race recode” and “origin recode NHIA.” This variable allowed us to obtain more accurate incidence rates by race/ethnicity for the following racial/ethnic groups: non-Latino/a White (White); non-Latino/a Black (Black); non-Latino/a Asian/Pacific Islander (Asian); non-Latino/a Other (Other); and Latino/a (regardless of race). American Indians and those of multiple races were merged with the “Other” category due to limited sample size. Due to sparse data concerns in specific racial/ethnic groups (e.g. non-Latino/a Other), we present results for White, Black, Asian, and Latino/a racial/ethnic groups.

Table 1
Select Demographic and Clinical Characteristics of Pediatric Optic Pathway Gliomas (Ages 0–19), SEER 18 2000–2014.

	Optic Pathway Gliomas
Characteristic	Ages 0–19 (n = 709)
Age at Diagnosis, n (%)	
0–4 years	394 (55.57)
5–9 years	193 (27.22)
10–14 years	91 (12.83)
15–19 years	31 (4.37)
Sex, n (%)	
Male	342 (48.24)
Female	367 (51.76)
Racial/Ethnic Group [a], n (%)	
White	478 (67.42)
Black	52 (7.33)
Asian	36 (5.08)
Other	23 (3.24)
Latino/a	120 (16.93)
Number of Primary Tumors, n (%)	
1	631 (89.00)
≥2	78 (11.00)
Time Period of Diagnosis, n (%)	
2000–2006	292 (41.18)
2007–2014	417 (58.85)
Diagnostic Confirmation, n (%)	
Positive Microscopic Confirmation	171 (24.1)
No Microscopic Confirmation	22 (3.1)
Radiography and Other Imaging Techniques, No Microscopic Confirmation	504 (71.1)
Unknown if Microscopically Confirmed, Death Certificate Only	12 (1.7)

[a] The Latino/a category includes those of all races, while all other race categories exclude those identified as Latino/a.

3. Results

A total of 709 incident OPGs in children ages 0–19 were abstracted from SEER 18 for the period 2000–2014 (Table 1). Over 80% of OPG cases occurred prior to 10 years of age, with 56% percent of cases occurring in those ages 0–4. Nearly 70% of cases occurred in White children; no difference in OPG incidence was detected by sex. For the majority of children (89%), this OPG diagnosis was the first primary tumor experienced. Slightly under half of the children (41%) were diagnosed in the period 2000–2006.

Overall, children from racial minority backgrounds ages 0–19 experienced lower rates and age-adjusted incidence rate ratios of OPGs compared to White children (Table 2). In White children, the rate of OPGs (IR_{White} = 0.29 per 100,000 person-years) was more than double