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Pediatric sinonasal tumors in the United States: incidence and outcomes

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ABSTRACT

Background: Sinonasal tumors in the pediatric population are exceedingly rare.
Materials and methods: Surveillance, Epidemiology, and End Results database was used to identify 250 cases of sinonasal malignancy in patients aged <20 y (1973–2010). Malignant histology codes were based on the International Classification of Disease for Oncology, third edition coding scheme. Incidence rates were adjusted to the 2000 U.S. population. Survival outcomes were plotted using the Kaplan–Meier method and compared with the log-rank test. All other analyses were performed using standard statistical methods.
Results: Overall incidence was 0.052 per 100,000. Rhabdomyosarcoma had the highest incidence among histologic groups. Regional stage was the most common at diagnosis (59%). Overall survival at 5-y follow-up was 62.5%. Patients in age groups 1–4 and 15–19 y had the worst survival rates, as median survival was 205 and 104 mo, respectively. Distant metastases at the time of diagnosis signified a poor prognosis. These were associated with a 39-mo median survival. Survival improved during the study period, $P = 0.003$. Gender, race, site of lesion, or histology did not appear to affect mortality.
Conclusions: Sinonasal tumors are rare in children and adolescents. Long-term survival is dependent on age and clinical stage at the time of diagnosis. Cancer-related surgery confers a survival advantage. Gender, race, and histologic type are not associated with mortality.

1. Introduction

Sinonasal malignancies are a rare and heterogenous collection of malignancies. They comprise <3% of all head and neck tumors [1]. Pediatric head and neck malignancies are less common than their adult counterparts. Within this group, pediatric sinonasal tumors are even less common. Rarity of these lesions has made statistical analysis of their epidemiologic and survival characteristics difficult to ascertain. Surveillance Epidemiology and End Results (SEER) database is a large, multicenter, population-based record that has been collecting data on malignancies diagnosed in the United States since 1973. In this study, we performed an analysis of pediatric sinonasal malignancies. Our goal is to better define the incidence, clinical and histopathologic characteristics, treatment options, and predictors of survival for this subset of patients.

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2. Materials and methods

The SEER database is the single largest cancer database in the United States. Since its inception, the National Cancer Institute (NCI) has collected data on patients diagnosed with malignancies from 18 registries. For this study, the April 2013 release was used to identify all incident cases of malignancy occurring in the nasal cavity, maxillary sinus, ethmoid sinus, sphenoid sinus, and frontal sinus for the pediatric population diagnosed for the period 1973–2010. Tumor location and histology were based on topography and morphology codes according to the International Classification of Disease for Oncology, third edition. International Classification of Disease for Oncology, third edition is the standard coding scheme for malignant histologies used by SEER. Patients >20 y of age were excluded from the analysis. Staging was based on the SEER historic summary stage, derived from the collaborative staging scheme set forth by the NCI.

SEER*Stat software (version 8.0.4; NCI, Bethesda, MD) was used to analyze incidence, trends, and survival. Annual percent change was calculated using the weighted least-squares method. All incidence data were age-adjusted and normalized to the 2000 US standard population. A P value of <0.05 was considered to be statistically significant. Statistical analysis was performed with SPSS version 22 (SPSS Inc, Chicago, IL). Correlations between categorical variables were made using the chi-squared (χ^2) test. Kaplan–Meier method with the Ederer II modification was used to calculate 5, 15, and 30-y survival. Survival was calculated from time of initial diagnosis to the date of last contact. We used the log-rank test to analyze the Kaplan–Meier curves for significant differences in survival outcomes with regards to demographics, clinical presentation, histology, and treatment.

The Institutional Review Board at the University of Miami Leonard M Miller School of Medicine (Miami, FL) deemed this study to be exempt from full review.

Table 1 – Incidence rates of pediatric sinonasal malignancies.

Overall	0.0515
Gender	
Female	0.0543
Male	0.0488
Race	
White	0.0487
Black	0.0676
Other	0.0410
Age (y)	
<1	0.0147
1–4	0.0452
5–9	0.0422
10–14	0.0395
15–19	0.0849
Type	
Rhabdomyosarcomas	0.0214
Sarcomas	0.0052
Germ cell tumors	
Neuroectoderm tumors/Neuroblastomas	0.0099
Lymphomas	0.0085
Other	0.0062
Stage	
In situ	0.0000
Localized	0.0063
Regional	0.0217
Distant	0.0063

3. Results

A total of 250 pediatric patients with sinonasal malignancies were identified during the 37-y study period. Overall incidence was 0.052 per 100,000 per annum (Table 1). Incidence was highest for rhabdomyosarcomas (0.021/100,000 per year). Overall annual percent change was not significant during the study period. Age-adjusted population incidence was highest for females, African Americans and adolescents aged 15–19 y.

Table 2 – Demographics and clinical characteristics.

Category	*n* (% of total)
Gender	
Female	116 (46)
Male	134 (54)
Race	
White	182 (74)
Black	45 (18)
Other	19 (8)
Age (y)	
<1	6 (2)
1–4	42 (17)
5–9	42 (17)
10–14	57 (23)
15–19	103 (41)
Type	
Rhabdomyosarcoma	104 (47)
Sarcomas	23 (10)
Neuroectoderm tumors/neuroblastomas	44 (20)
Lymphoma	40 (18)
Other	12 (5)
Site	
Nasal cavity	95 (38)
Maxillary sinus	76 (30)
Ethmoid sinus	40 (16)
Sphenoid sinus	10 (4)
Overlapping lesion of accessory sinuses	10 (4)
Accessory Sinus, numbers	19 (8)
Surgery	
Surgery	122 (49)
No surgery	127 (51)
Radiation	
Radiation	157 (63)
No radiation	91 (37)
Staging	
In situ	0 (0)
Localized	31 (18)
Regional	102 (59)
Distant	39 (23)
Diagnosis (y)	
1973–1978	9 (4)
1979–1984	14 (6)
1985–1991	25 (10)
1992–1997	33 (13)
1998–2003	66 (26)
2004–2010	103 (41)