BRIEF REPORTS •

Pediatric Brain Tumors in Puerto Rico

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Objective: To determine the frequency of pediatric brain tumors treated at the University Pediatric Hospital of Puerto Rico over a 6-year period and examine the demographic data and tumor histology of patients in different age groups.

Methods: A retrospective study was undertaken at the University Pediatric Hospital. We included patients with brain tumors that had been newly diagnosed during the period covering from January 2002 to December 2007. All cases were analyzed by age, gender, histologic diagnosis, and affected area.

Results: One hundred thirty-six patients were included in the study. Overall, males were more frequently affected than were females. Children in the 1 to 4 years old age group had the highest number of newly diagnosed brain tumors. Regarding anatomic location, supratentorial tumors were more frequent than were infratentorial tumors. The most common single tumor was the pilocytic astrocytoma (WHO grade I), representing 31% of the total pediatric brain tumors.

Conclusion: Our results provide an objective platform for further epidemiological studies and for the development of local health strategies for the timely diagnosis and treatment of the most common pediatric tumors in Puerto Rico. [P R Health Sci J 2011;30:195-197]

Key words: Pediatric, Brain Tumors, Hispanic

ediatric brain tumors represent the most common solid malignancy affecting children, second only to leukemia in overall incidence (1). The biological behavior and management of any given childhood tumor depends not only in its histological character but also its location within the central nervous system (CNS) (2). Brain tumors in children differ significantly from adult lesions in terms of their sites of origin, histological features, and clinical presentation (3, 4).

During the past two decades, an increase in the incidence of pediatric brain tumors has been documented, precipitating a debate about their etiology (5). Possible explanations are that there are better diagnostic imaging techniques, that cases are better reported, and that there has been an actual increase in the number of brain tumors being suffered by children (5, 6). Past studies have demonstrated racial disparities in both tumor development and patient survival rate (7, 8, 9).

Although important advances have been made in understanding the molecular mechanisms of tumor behavior and in potential treatment strategies, the prognosis of pediatric brain tumors remains poor (10). Brain tumors are the leading cause of death from solid tumors (1). Mortality is about 0.8 per 100,000 children per year, double that of leukemia, which is 0.4 (1). Because of either the effects of the tumor or the treatment required (or both), survivors of childhood brain tumors often have severe neurologic, cognitive, and psychosocial sequelae (11, 12, 13).

According to the Central Brain Tumor Registry of the United States (CBTRUS), the incidence of pediatric brain tumors in 2005 and 2006 was 4.3 per 100.000 children (4). These data were obtained from 18 state cancer registries and included cases of non-malignant and malignant primary brain and central nervous system tumors. Data from Puerto Rico were not included.

In 2004, the Puerto Rico Cancer Registry reported that the incidence of central nervous system cancer was 20.1 per 100,000 (14). They did not specify location or type of tumor. A complete analysis of pediatric brain tumors with regard to location, tumor typing, and age distribution in the Puerto Rican population has not been made.

In view of the epidemiological importance of this pathology, a retrospective study was performed at the University Pediatric Hospital (UPH) of Puerto Rico in order to investigate the distribution of pediatric brain tumors in a cohort of children living in Puerto Rico.

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Methods

A retrospective cohort study was performed by doing a record review of patients who had been treated at the UPH in San Juan, Puerto Rico, between January 2002 and December 2007 (inclusive). The study focused on patients who were younger than 18 years old, had been newly diagnosed with a brain tumor, and who underwent surgical resection versus biopsy (resulting in histologic confirmation). We excluded patients without such confirmation, with recurrent brain tumors, or with spinal or skull tumors. The collected data were analyzed by age, gender, tumor location, and tumor histology. Descriptive statistics were performed.

Results

We reviewed 157 medical records, culling from them 136 from patients who met the inclusion criteria. Males (55%) were more commonly affected than were females (45%). Children in the 1 to 4 years old age group were the most commonly affected, representing 29% of the cases (Table 1). Regarding anatomic location, supratentorial tumors (57%) were more common than were infratentorial tumors (43%).

Table 1. Demographic features of pediatric patients with brain tumors (n=136)

Variable		N (%)
Sex	Male Female	71 (55) 57 (45)
Age	<1 year old 1 - 4 5 - 9 10 - 14 15 - 17	6 (4) 40 (29) 34 (25) 30 (22) 26 (19)

The most common tumor was the pilocytic astrocytoma (WHO grade I), representing 31% of all cases, and it was also the most frequent tumor in all age groups, except in children less than 1 year old. An infratentorial location was the most common location, representing 71% of all tumors. There was no gender predominance (males: 47%; females: 52%).

Other common pathologies were medulloblastoma (8%), ependymoma (8%), astrocytoma (WHO grade II) (8%), craniopharyngioma (8%), and high grade astrocytoma (WHO grades III and IV) (4%). Other less common tumors were below 3% in terms of the frequency of their occurrence and are shown in Table 2.

The most common histologies by age are as follows: in the less than 1 year old age group, tumors arising in the choroid plexus (33%) were more frequent. In the 1 to 4 years old age group, pilocytic astrocytoma (WHO grade I) (43%), medulloblastoma (13%), and craniopharyngioma (10%) dominated. In the 5 to

9 years old age group, pilocytic astrocytoma (WHO grade I) (29%), craniopharyngioma (12%), and ependymoma (12%) were most found. In the 10 to 14 years old age group, we saw pilocytic astrocytoma (WHO grade I) (20%), medulloblastoma (17%), and germinoma (10%). Finally, in the 15 to 17 years old age group, pilocytic astrocytoma (WHO grade I) (35%), astrocytoma (WHO grade II) (12%), and supratentorial PNET (7%) topped the list.

Table 2. Tumor histology of pediatric patients with brain tumors (n=136)

Tumor	N
Pilocytic astrocytoma (WHO grade I)	42
Medulloblastoma	12
Ependymoma (WHO grades I , II, III)	11
Astrocytoma (WHO grade II)	11
Craniopharyngioma	11
Astrocytoma (WHO (grades III & IV)	6
Choroid plexus papilloma (WHO grade I)	5
Germinoma	5
PNET	5
Dermoid cyst	4
SEGA (WHO grade I)	4
Meningioma	3
Ganglioglioma (WHO grade I)	2
Metastatic	2
Oligodendroglioma (WHO grade II)	3
Choroid plexus carcinoma (WHO grade III)	1
Epidermoid cyst	1
Hemangiopericytoma	1
Pilomyxoid astrocytoma	1
Pineocytoma (WHO grade II)	1
Pleomorphic xanthoastrocytoma	2
Rathke's cleft cyst	1
Schwannoma (WHO grade I)	1
Pituitary adenoma	1

Table 3. Histology of the most common tumors by anatomic location

Supratentorial = 77 (57%)	Infratentorial = 59 (43%)
Pilocytic astrocytoma: 16%	Pilocytic astrocytoma: 50%
Astrocytoma (WHO grade II): 14%	Medulloblastoma: 20%
Craniopharyngioma:14%	Ependymoma: 12%
Other: 56%	Other: 18%

Discussion

Our study of 136 patients shows in detail the histologic predominance and anatomical location of a variety of tumors for different pediatric age groups. This is the first study that analyzes in depth a cohort of pediatric patients with brain tumors in Puerto Rico.

In our study, male patients (55%) outnumbered female patients (45%). The age group with the highest number of

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cases was the 1 to 4 years old age group (29%). Our data are comparable with those of the Central Brain Tumor Registry of the United States (4).

It has traditionally been taught that most pediatric brain tumors (60%) are infratentorial and that these are equally divided among brainstem gliomas, cerebellar astrocytomas, and medulloblastomas (15,16,17,18,19,20,21). In our study we found a predominance of supratentorial tumors (57%), the frequency and classification of which are as follows: pilocytic astrocytoma (16%), astrocytoma (WHO grade II) (14%), and craniopharyngiomas (14%). In terms of infratentorial tumors, the pilocytic astrocytoma was the most frequent, representing 50% of all pediatric brain tumors, followed by medulloblastoma (20%), and ependymoma (12%).

Data analyzed by age group showed that infratentorial tumors were the most common in children from 1 to 9 years of age. Supratentorial tumors were the most common in children in the 10 to 14 and 15 to 17 years old age groups.

UPH is the main referral center for pediatric brain neoplasms in Puerto Rico. However, the accurate calculation of the incidence of pediatric brain tumors in Puerto Rico was limited in our study by the exclusion of patients without histologic confirmation and by the unavailability of data from other community hospitals.

In summary, this is the first descriptive study that analyzes pediatric brain tumors in depth in Puerto Rico. Our results provide an objective platform for further epidemiological studies and for the development of local health strategies for the timely diagnosis and treatment of the most common pediatric tumors in Puerto Rico.

Resumen

Objetivo: Determinar la frecuencia de tumores cerebrales en pacientes pediátricos tratados en el Hospital Universitario Pediátrico de Puerto Rico durante un periodo de 6 años y examinar la información demográfica e histológica en los distintos grupos de edades. Métodos: Se realizó un estudio retrospectivo en el Hospital Universitario Pediátrico de Puerto Rico. Se incluyeron pacientes diagnosticados con tumores cerebrales de novo desde enero del 2002 a diciembre del 2007. Todos los casos fueron analizados por edad, género, diagnóstico histológico y área anatómica afectada. Resultados: Se incluyeron 136 pacientes en el estudio. Los varones fueron más afectados que las féminas. Los niños de 1 a 4 años fueron los más frecuentemente afectados. En relación a la localización anatómica, los tumores supratentoriales fueron más frecuentes. El tumor más común fue el astrocitoma pilocítico (WHO I) (31%). Conclusión: Nuestros resultados proveen una plataforma objetiva para nuevos estudios epidemiológicos y para el desarrollo de estrategias de salud local para el diagnóstico y tratamiento oportuno de los tumores cerebrales más comunes en la población pediátrica.

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