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Case report

Central neurocytoma: Clinical characteristics, patterns of care, and survival

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ABSTRACT

Purpose: To investigate clinical characteristics and patterns of care among patients with central neurocytomas in a large cohort of patients.
Methods: The National Cancer Database (NCDB) was queried to identify patients with biopsy confirmed neurocytoma from 2004 to 2015. Patterns of care were described and univariable and multivariable models were performed to investigate the impact of prognostic factors on overall survival.
Results: Among 223,404 patients with brain tumors in the NCDB, 868 patients were diagnosed with biopsy-proven neurocytoma and analyzed (0.4% or approximately 75 patients annually). Median age at diagnosis was 31 years and median tumor size was 4–5 cm. Diagnosis was similar between male (49.5%) and female (50.5%). Regarding location, 622 (72%) tumors were intraventricular, 168 (19%) were extra-ventricular, and 78 (9%) overlapping or unspecified. Five-year overall survival among all patients was 89%. On multivariable analysis tumor location, extent of resection, and use of radiation, were not predictive for improved survival (each $p > 0.05$); however, patient age ($p < 0.001$), WHO grade ($p < 0.001$), and medical comorbidity scores ($p = 0.002$) were independently associated with overall survival.
Conclusion: Patients with central neurocytoma often present as young adults with sizable tumor burden and are well managed with surgery alone. Considering their favorable survival, efforts to improve tumor control should be carefully weighed against the long-term risks associated with adjuvant therapy like radiation.

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1. Introduction

Central Neurocytomas (CN) are benign tumors categorized as a grade II tumor by the World Health Organization [1]. These tumors are rare, encompassing about 0.1–0.5% of all brain tumors [2]. CN affect a wide age range of individuals from 8 days old to 67 years old, with median age of presentation of 34 years old [3]. CN are typically located within the ventricles, more commonly in the anterior half of the lateral ventricle [4,5]. Presenting tumors tend to attach to the septum pellucidum near the foramen of Monroe and thus can result in obstructive hydrocephalus [6]. The molecular origins of CN have previously been hypothesized to arise from neural precursors or even possibly from an astrocytic lineage [7]. More recent research has alluded to the tumor being derived from stem cells aberrations, however the definite lineage continues to be debated [8].

Clinically, patients may present with increased intracranial pressure and hydrocephalus due to obstruction of the interventricular foramen [9]. Subsequently, patients may experience headaches, seizures, nausea, vomiting, altered mental status, memory or changes in vision [10,11]. Multiple interventions, including surgery, radiation and chemotherapy, can be utilized with varied outcomes [12–16].

Previous studies have evaluated prognostic factors associated with poorer outcomes in CN. In a multicenter study with seventy-one patients, larger tumor volume, incomplete surgery, and a mitotic count ≥3 per 10 high-power fields were all predictors of a higher risk of recurrence [17]. However, due to the paucity of CNs, further investigation with a large cohort of patients is required to elucidate epidemiologic data and the impact of prognostic factors. Here we investigate the clinical characteristics,

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patterns of care and outcomes of patients with CN in a large cohort of patients treated at medical centers in the United States.

2. Methods and materials

2.1. Data source

Established in 1989, the National Cancer Database (NCBD) is a nationally recognized clinical oncology database and is sponsored by the American College of Surgeons and the American Cancer Society. The NCDB collects data from >1500 facilities accredited by the Commission on Cancer and contains information on treatments and outcomes for patients with malignant disease. The current database gathers >70% of new cancer diagnoses in the US and contains >34 million historical records [18].

Data was obtained from the NCDB for patients diagnosed with brain tumors between 2004 and 2015 (223,404 patients). Patients excluded included those with non-neurocytoma histology or neurocytoma without biopsy (222,536 total excluded, Fig. 1). The remaining 868 patients were then stratified and analyzed based on available database information [19].

2.2. Statistical analyses

The primary outcome measured for this study was overall survival (OS) measured from date of diagnosis. Secondary goals included to describe the baseline clinical characteristics of patients with CN, as well as the patterns of care that they receive. In order to prevent bias based on sites with underreporting of data (favoring the inclusion some centers over others) and improve result validity, all patients with inclusion criteria of neurocytoma with biopsy were evaluated, even if they contained one or more missing data elements. Important prognostic factors, including gender, age, race, median income, distance to hospital, Charlson/Deyo score, WHO tumor grade, tumor size, tumor location, extent of resection, radiation, and chemotherapy were evaluated.

Univariable and multivariable analyses (Cox proportional hazards models) were performed to investigate factors associated with OS. Potentially prognostic variables in the multivariable models were chosen through purposeful selection and univariable analyses to investigate significance. Factors associated with a $p < 0.10$ on univariate analysis were included in the multivariable Cox regression models. All statistical analyses were performed using the SPSS program (SPSS, version 24.0; SPSS Inc., Chicago, IL) and $p < 0.05$ on multivariable analysis were considered statistically significant.

3. Results

3.1. Clinical characteristics

Patient characteristics are shown in Table 1. Among 223,404 patients with brain tumors in the NCDB, 868 patients were diagnosed with biopsy-proven neurocytoma, representing 0.4% of brain tumors. Over the years 2004–2015, 62–82 patients were diagnosed annually across all centers. Median age at diagnosis was 31 years. Diagnosis was similar between male (49.5%) and female (50.5%) gender. Regarding tumor location, 622 (71.7%) were intraventricular, 168 (19.3%) were extra-ventricular, and 78 (9.0%) overlapping or unspecified. The median tumor size was 4–5 cm.

3.2. Treatment characteristics

All patients included underwent biopsy or resection (Table 1). Extent of resection was unknown in 491 (57%) patients, 146 (16.8%) underwent gross total resection, and 122 (14.1%) underwent subtotal resection. The majority (83.6%) did not receive radiotherapy. Chemotherapy was utilized in 18 (2.1%) patients, and withheld in 841 (96.9%) patients.

3.3. Overall survival

Five-year overall survival among all patients was 89%. Among those who underwent gross total resection, five-year survival

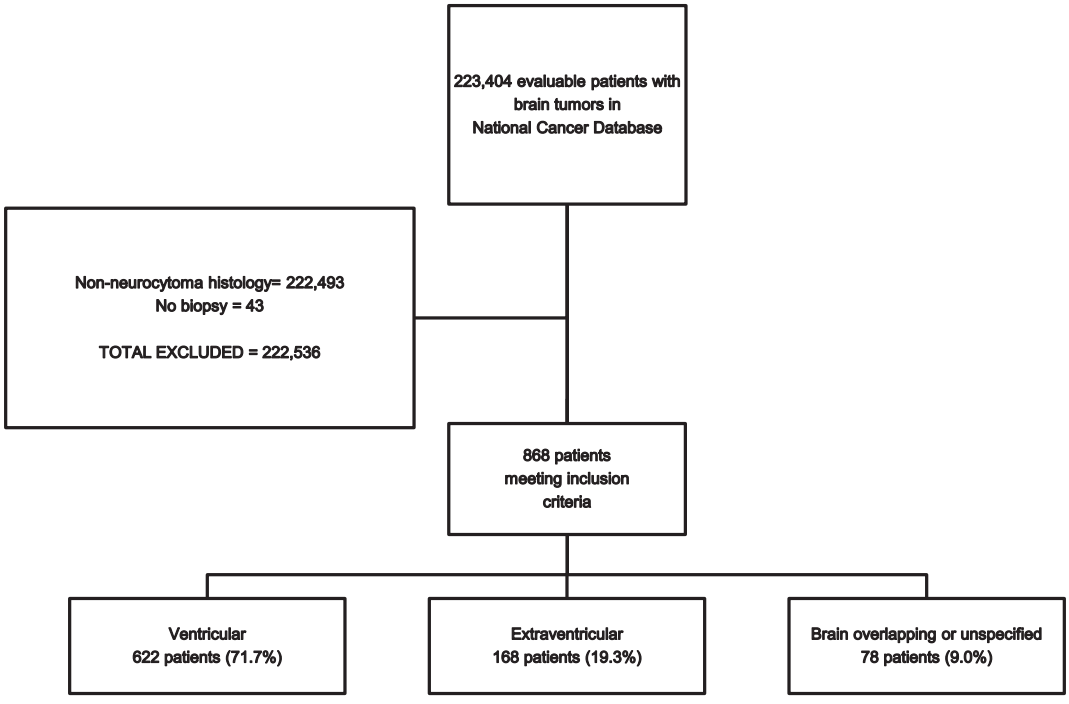

Fig. 1. National Cancer Database cohort selection diagram.

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