

Supratentorial hemispheric ependymomas: an analysis of 109 adults for survival and prognostic factors

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OBJECTIVE Survival rates and prognostic factors for supratentorial hemispheric ependymomas have not been determined. The authors therefore designed a retrospective study to determine progression-free survival (PFS), overall survival (OS), and prognostic factors for hemispheric ependymomas.

METHODS The study population consisted of 8 patients from our institution and 101 patients from the literature with disaggregated survival information (n = 109). Patient age, sex, tumor side, tumor location, extent of resection (EOR), tumor grade, postoperative chemotherapy, radiation, time to recurrence, and survival were recorded. Kaplan-Meier survival analyses and Cox proportional hazard models were completed to determine survival rates and prognostic factors.

RESULTS Anaplastic histology/WHO Grade III tumors were identified in 62% of cases and correlated with older age. Three-, 5-, and 10-year PFS rates were 57%, 51%, and 42%, respectively. Three-, 5-, and 10-year OS rates were 77%, 71%, and 58%, respectively. EOR and tumor grade were identified on both Kaplan-Meier log-rank testing and univariate Cox proportional hazard models as prognostic for PFS and OS. Both EOR and tumor grade remained prognostic on multivariate analysis. Subtotal resection (STR) predicted a worse PFS (hazard ratio [HR] 4.764, p = 0.001) and OS (HR 4.216, p = 0.008). Subgroup survival analysis of patients with STR demonstrated a 5- and 10-year OS of 28% and 0%, respectively. WHO Grade III tumors also had worse PFS (HR 10.2, p = 0.004) and OS (HR 9.1, p = 0.035). Patients with WHO Grade III tumors demonstrated 5- and 10-year OS of 61% and 46%, respectively. Postoperative radiation was not prognostic for PFS or OS.

CONCLUSIONS A high incidence of anaplastic histology was found in hemispheric ependymomas and was associated with older age. EOR and tumor grade were prognostic factors for PFS and OS on multivariate analysis. STR or WHO Grade III pathology, or both, predicted worse overall prognosis in patients with hemispheric ependymoma.

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KEY WORDS brain tumor; ependymoma; glioma; prognosis; survival; oncology

INTRACRANIAL ependymomas are the third most common glioma in adults, behind astrocytomas and oligodendrogliomas. Ependymomas constitute 3% of primary CNS tumors and 6.7% of gliomas diagnosed each year in the US.^{31,32} Safe maximal resection remains the mainstay of treatment. Radiation and chemotherapy may be provided as adjuvant treatments, especially in the setting of subtotal resection (STR). Reported 5-year overall survival (OS) rates for adults with intracranial ependymoma range widely from 35% to 90%.^{9,19,21,47,54} Controversy exists regarding important prognostic factors for progression-free

survival (PFS) and OS.^{1,37,57} Reported prognostic factors include age, Karnofsky performance status, tumor location, tumor grade, extent of resection (EOR), and postoperative radiation.^{8,9,19–22,46–48,54}

Evidence supports the idea that supratentorial ependymomas have a worse prognosis than infratentorial tumors do in adults.^{9,19,20,23,38,45,46} Additional data indicate that patients with hemispheric ependymomas have decreased PFS and OS compared with tumors occurring in the third or lateral ventricles.^{9,45} Although often considered an intraventricular tumor, more than half of supratentorial ep-

ABBREVIATIONS EOR = extent of resection; GTR = gross total resection; OS = overall survival; PFS = progression-free survival; STR = subtotal resection.

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ependymomas occur within the cerebral hemispheres.^{9,22,39,57} Their hemispheric location likens ependymomas to the other primary intraaxial gliomas. Survival rates and prognostic factors for the most common hemispheric gliomas, astrocytoma and oligodendroglioma, have been well investigated in several clinical studies.^{10,16,43,50}

A large clinical investigation of hemispheric ependymomas is lacking, with the current literature consisting of case reports and small case series. Because these tumors potentially carry the worst prognosis of all ependymomas, accurate survival rates and prognostic factors would provide essential clinical information. We reviewed and statistically analyzed our institutional case series in conjunction with cases reported in the literature to more accurately determine the survival rates and prognostic factors of hemispheric ependymomas.

Methods

Patient Population and Article Selection

Our patient population consisted of 2 groups. The first group was all patients 18 years of age or older diagnosed with supratentorial hemispheric ependymomas at the University of Michigan from 1981 to 2014. Approval from the University of Michigan Institutional Review Board was obtained before medical records were accessed. A total of 11 patients were identified. One patient was excluded because tissue diagnosis demonstrated ependymoblastoma, one patient was excluded due to myxopapillary histology, and the third was excluded due to lack of survival data.

The second patient population consisted of all adults (≥ 18 years of age) with supratentorial hemispheric ependymoma reported in the literature with disaggregated clinical information. This was defined as any patient reported in the literature with individually listed patient characteristics (e.g., age, sex, tumor side and location, treatment) and survival times (PFS and OS). Statistical data or calculated values from the literature were not included for analysis. The English-language literature was reviewed for clinical articles pertaining to supratentorial hemispheric ependymomas. PubMed searches for the terms “hemispheric ependymoma,” “ectopic ependymoma,” “cortical ependymoma,” “extraventricular ependymoma,” and “lobar ependymoma,” combined with a “human species” filter, was performed. We chose to use the term “hemispheric” to emphasize that these tumors are intraaxial parenchymal tumors located within the cerebral hemispheres. After review of study abstracts resulting from the search, a total of 53 studies performed between 1985 and 2014 were deemed appropriate for article review. Each article was individually reviewed for disaggregated patient data. Studies with only aggregated patient data were excluded. Patients were excluded if they were less than 18 years of age, had no overall survival data available, or had an intraventricular tumor location. Ultimately, clinical data were obtained from 27 of 53 articles, and 101 patients from the literature were identified.^{3,4-7,11,13-15,17,18,24-26,27-29,34,35,40-42,44,49,52,53,58} Article authors were not contacted for additional data. A total of 109 patients from both study populations were included in our investigation, and 77 patients had complete clinical information as described below.

Data Collection

The following patient data were collected either from chart review or extracted from case reports and case series obtained in the literature search outlined above: age at diagnosis; sex; tumor side and location; extent of tumor resection (dichotomized to subtotal resection [STR] vs gross-total resection [GTR]); WHO tumor grade, histology, or both; postoperative radiation; postoperative chemotherapy; total follow-up time; time to recurrence; PFS; and OS. All patients underwent craniotomy for tumor resection. No patient had a biopsy only. Gross-total resection was defined as no evidence of enhancement on T1-weighted postgadolinium MRI for our institutional series. Subjects with absent or unknown data for any of the above clinical items were excluded from statistical analyses for that item to avoid inaccuracy or estimation. The number of patients included for each statistical analysis is indicated in the appropriate tables.

Statistical Analysis

Age, PFS, and OS were analyzed as continuous variables. Age > 40 or ≤ 40 years, sex, tumor side, tumor location, tumor grade, EOR, postoperative radiation, and chemotherapy were analyzed as categorical variables. Two survival outcomes were assessed: PFS and OS. PFS was defined as time from resection to radiological evidence of recurrence for our institutional series or PFS time interval reported in the article. OS was defined as the time from tissue diagnosis to death or OS time interval reported in the article. Eight patients in the series did not have data regarding recurrence or PFS. These patients were excluded from the statistical analysis of PFS. Kaplan-Meier survival function curves were constructed both for PFS and OS, using each prognostic factor (age, sex, tumor side, tumor location, EOR, tumor grade, postoperative chemotherapy, and postoperative radiation). The log-rank test was used to calculate p values for statistical significance between survival curves. Univariate Cox proportional hazard analysis was used to evaluate variables as prognostic factors for PFS and OS. Multivariate analysis was completed and included age, sex, tumor side, tumor location, EOR, tumor grade, postoperative chemotherapy, and postoperative radiation. Statistical significance was determined by ANOVA for continuous variables. The Pearson chi-square test or Fisher exact test (expected count < 5) was used for categorical variables. Statistical significance was defined as a p value < 0.05 . All p values were calculated using a 2-tailed test. Variance about the mean was reported as 95% confidence interval (CI) or standard deviation. All statistical analyses were completed using SPSS software (version 22, IBM Corp.).

Results

Patient Characteristics

A summary of baseline patient characteristics for all patients is presented in Table 1. Mean follow-up time was 5.29 ± 5.87 years with a range of 0.8 to 28.0 years. Mean age was 40.0 ± 16.3 years with a range of 18 to 74 years. Males and females were approximately equally affected. The 2 most common tumor locations were the frontal (33%) and parietal lobes (22%). All patients underwent

TABLE 1. Patient characteristics

Characteristic	No. of Patients	%
Age at diagnosis (yrs)	86	
<40	47	55
≥40	39	45
Sex	82	
Male	40	49
Female	42	51
Side	103	
Lt	63	61
Rt	40	39
Location	106	
Frontal	35	33
Parietal	24	22
Temporal	8	8
Other*	39	37
EOR	85	
GTR	71	84
STR	14	16
WHO grade	109	
III	68	62
II	41	38
Postop radiation	103	
Yes	68	66
No	35	34
Postop chemotherapy	102	
Yes	20	20
No	82	80

* Other tumor locations included frontoparietal, frontotemporal, parietooccipital, occipital.

surgical resection, with GTR achieved in 84% of cases. WHO Grade III tumors constituted the majority of hemispheric ependymomas (62%). Radiation type and dose was available for 30/68 patients (focal only, n = 21; focal with whole brain, n = 5; focal with craniospinal radiation, n = 1; whole brain only, n = 3). All patients received a radiation dose of 50 Gy or more.

There was a statistically significant correlation between age and higher tumor grade. Although WHO Grade III tumors were the most common type of tumor to occur in both younger and older age groups, patients 40 years and older were more likely to be diagnosed with a WHO Grade III tumor than younger patients were (OR 2.41, 95% CI 1.07–8.12; p = 0.046). EOR was not associated with tumor grade (p = 0.416). STR was not associated with postoperative chemotherapy (p = 0.241) or postoperative radiation (p = 0.632).

All patient information from our institutional case series is presented in Table 2. Common presenting symptoms were focal neurological deficit (5/8), headache (4/8), and seizure (2/8). Tumors were located in the frontal (4/8) and parietal (4/8) lobes. GTR was achieved in 4/8 patients.

TABLE 2. Summary of patient characteristics from institutional case series of hemispheric ependymomas

Age at Diagnosis (yrs), Sex	Symptoms	WHO Grade	Side	Location	EOR	Radiation	Dose (Gy)	Chemotherapy	Recurrence	Time (yrs)	Alive/Deceased	Survival (yrs)
51, F	Rt hand clumsy	III	Lt	Parietal	GTR	Focal	60	No	Local	3	Deceased	17
74, F	Behavior changes; it hand weakness	III	Rt	Frontal	STR	No	NA	No	No	NA	Alive	1.6
28, F	Headache	II	Rt	Frontal	GTR	WBRT, focal	45, 9	No	No	NA	Alive	28
48, M	Headache; word-finding	III	Lt	Parietal	STR	Focal	56	Temodar (7 cycles) adjuvant; oral VP-16 after third resection	Local	1.5	Deceased	3
26, F	Seizures	III	Lt	Frontal	GTR	Focal	Unknown	No	No	NA	Alive	10
44, M	Headache	III	Rt	Parietal	STR	Focal	56	No	Local	1.5	Deceased	2
61, M	Headache; right-sided weakness; simple partial seizures	III	Lt	Frontal	STR	Focal, craniospinal	59, 36	Temodar and lapatinib; then bevacizumab & carboplatin	Local, leptomeningeal dissemination	NA	Alive	3
22, M	Aphasia; word-finding	II	Lt	Parietal	GTR	Focal	54	No	No	1	Alive	1

NA = not applicable; VP-16 = etoposide; WBRT = whole-brain radiation therapy.

Postoperative radiation or chemotherapy, or both, were given to 7 and 2 patients, respectively.

Kaplan-Meier Survival Analysis and Survival Rates for Study Population

Kaplan-Meier survival curves were created for PFS (Fig. 1, left) and OS (Fig. 1, right). Three-, 5-, and 10-year PFS and OS data are presented in Table 3. Median PFS and OS were not achieved (recurrence 42/101; deaths 32/109); estimated median survival rates can be found in Table 3. The majority of patients had tumor recurrence within the first 3 years following tumor resection (43%). Only 15% of tumors recurred between 3 and 10 years.

Subgroup Kaplan-Meier Survival Analysis and Survival Rates Based on Prognostic Factors

Log-rank testing for Kaplan-Meier survival analysis identified the EOR ($p < 0.001$) and tumor grade ($p < 0.001$) as significant predictors of PFS (Fig. 2A and C). Patients with STR and WHO Grade III ependymomas had worse PFS. EOR ($p < 0.001$) and tumor grade ($p = 0.003$) were also significant predictors of OS. STR and WHO Grade III ependymomas predicted worse OS, with STR predicting the worst survival outcome (Fig. 2B and D). Age, sex, tumor side, postoperative chemotherapy, and postoperative radiation were not prognostic for PFS and OS.

EOR and tumor grade subgroup PFS and OS rates are presented in the upper-right corner of the corresponding Kaplan-Meier curves (Fig. 2). Patients with STR or those with WHO Grade III tumors, or both, had significantly worse 3-, 5-, and 10-year PFS and OS. At 10 years, all patients with STR had tumor progression and were deceased.

In our institutional case series, 4 of 8 patients had tumor recurrence with a median PFS of 1.25 years. Three patients had local recurrence and 1 had leptomeningeal seeding. Three patients died, with all deaths resulting from tumor recurrence.

Univariate and Multivariate Hazard Ratio Analysis of Prognostic Factors

Hazard ratios (HRs) for prognostic factors were calculated using both univariate and multivariate Cox propor-

TABLE 3. Rates for PFS and OS

Survival (yrs)	% PFS (n = 101)	% OS (n = 109)
3	57	77
5	51	71
10	42	58
Estimated median*	6.2 yrs	14.8 yrs

* Median rates were not reached (total recurrences = 43/101, total deaths = 32/109).

tional hazard models. Summary of univariate and multivariate results are presented in Tables 4 and 5, respectively. Univariate analysis identified STR and WHO Grade III tumors as prognostic factors for worse PFS, with hazard ratios of 4.156 and 5.407, respectively. Univariate analysis for OS also identified EOR and tumor grade as prognostic factors. STR and WHO Grade III were predictive of worse OS with a hazard ratio of 4.964 and 3.771, respectively. Neither postoperative radiation nor chemotherapy was prognostic for improved PFS or OS.

Multivariate analysis for prognostic factors was completed and included age, sex, tumor side, EOR, tumor grade, postoperative chemotherapy, and postoperative radiation. Only patients with complete clinical information were included in the analysis. A total of 77 patients with PFS and 79 patients with OS were included in the analysis. Both EOR and tumor grade had statistically significant HRs on multivariate analysis. Multivariate HRs are presented in Table 5.

Discussion

We present the largest clinical investigation of hemispheric ependymomas completed to date. Previous studies have been limited to case reports and case series. Because of the low incidence of these tumors in adults, pooling of institutional and reported data are a useful means to overcome small sample size. A total of 109 patients were included and analyzed for patient characteristics, survival, and prognostic factors. With approximately 350 adult supratentorial ependymomas diagnosed each year in the US,

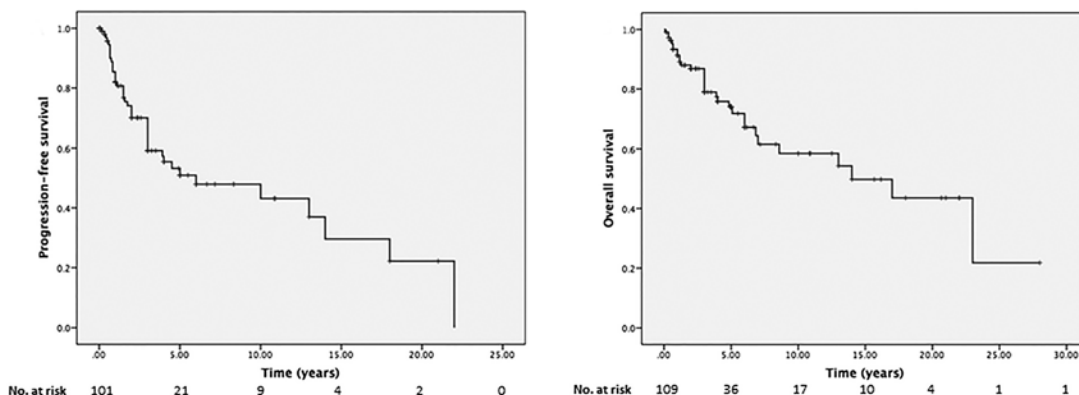


FIG. 1. Kaplan-Meier survival analysis for PFS (left) and OS (right) completed for the study population.

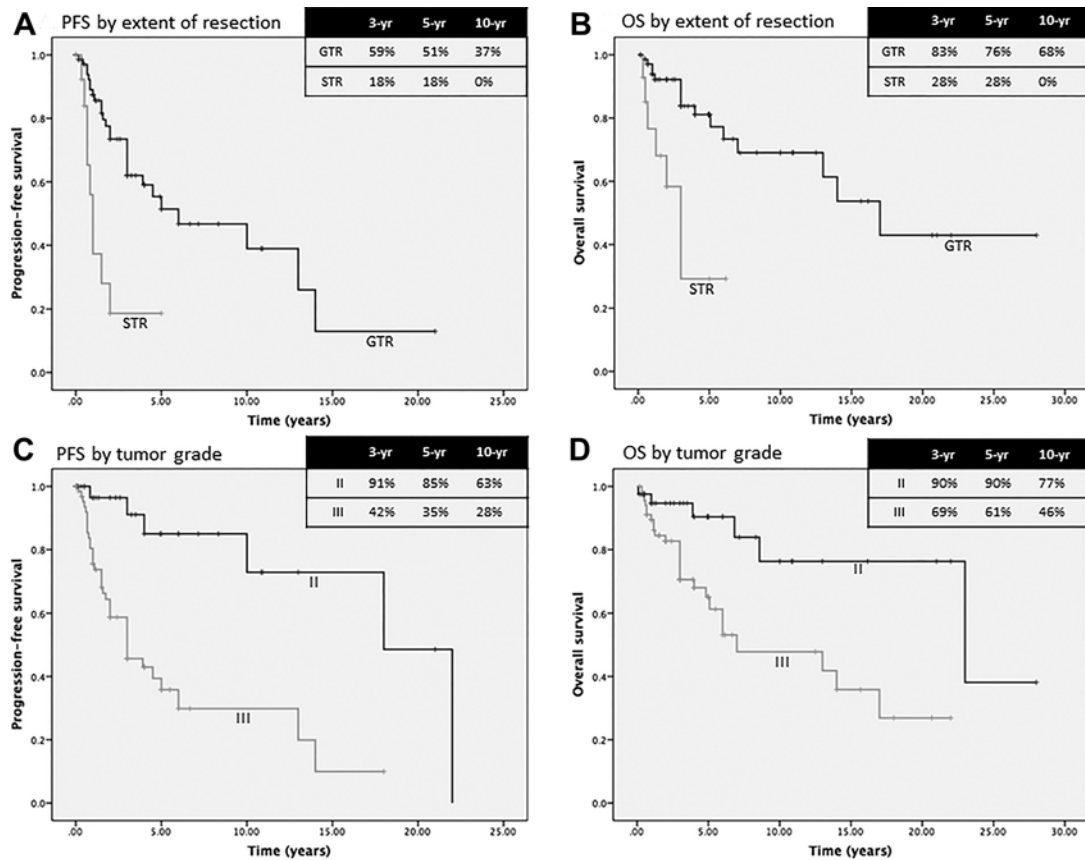


FIG. 2. Subgroup Kaplan-Meier survival analysis and survival rates were determined based on EOR and tumor grade prognostic factors. Kaplan-Meier curves are presented with corresponding survival rates in the upper right corner. **A:** PFS Kaplan-Meier curves for STR and GTR subgroups; there was a statistically significant difference for STR, which had a worse prognosis ($p < 0.001$). **B:** OS Kaplan-Meier curves for STR and GTR subgroups; there was a statistically significant difference for STR, which had a worse prognosis ($p < 0.001$). **C:** PFS Kaplan-Meier curves for WHO Grade II and III tumors; a statistically significant difference was noted for Grade III tumors, which resulted in a worse PFS ($p < 0.001$). **D:** OS Kaplan-Meier curves for WHO Grade II and III tumors; a statistically significant difference was noted for Grade III tumors, which resulted in a worse PFS ($p = 0.003$).

our study captured a representative population of patients with hemispheric ependymomas.⁵⁶

Adult males and females were equally affected. No age group was at increased risk for tumor occurrence. Hemispheric ependymomas were most common in the frontal lobes, but did not show a strong predilection for any location. GTR was achieved in the majority of cases (85%). High rates of GTR may be attributed to the tendency of ependymomas to displace and compress, rather than infiltrate, surrounding brain parenchyma.^{1,3,37} The tumor-brain interface is often well-demarcated both radiographically and intraoperatively. This is a unique feature of ependymomas compared with other gliomas. Surgical management of astrocytoma or oligodendroglioma results in STR in the majority of cases.^{10,30} The more infiltrative nature of other gliomas, such as diffuse astrocytoma or malignant glioma, makes GTR more challenging. As our findings indicate that EOR is an important prognostic factor for OS and PFS, higher rates of GTR may contribute to longer survival times compared with low-grade and malignant gliomas.³²

Anaplastic histology was found in 62% of hemispheric

ependymomas, a larger percentage than previously reported for intracranial ependymomas (23%–54%).^{2,14,21,38,46,47,53} Several series have also found a higher percentage of WHO Grade III tumors in hemispheric ependymomas.^{9,22,47,55} Guyotat and colleagues found that all 10 hemispheric ependymomas in their case series of 34 intracranial ependymomas had WHO Grade III pathology.⁹ Third and lateral ventricle tumors were split evenly between WHO Grade II and Grade III pathology. Another report found that 73% of hemispheric ependymomas (16/22 tumors) were WHO Grade III ($p < 0.05$).²² Six (75%) of the 8 patients in our institutional case series had WHO Grade III tumors. Our results provide evidence that hemispheric ependymomas have a greater likelihood of being WHO Grade III than infratentorial or spinal ependymomas in adults. While the explanation for this is unclear, it may be related to our finding that age was correlated with higher tumor grade. This is a feature common to astrocytomas and oligodendrogliomas, with incidence peaking at 35 to 44 years of age for low-grade tumors and 75 to 84 years of age for high-grade tumors.³¹ In our series, anaplastic histology

TABLE 4. Univariate analysis of prognostic factors for PFS and OS

Prognostic Factor*	PFS†	OS†
Age at diagnosis (n)	82	84
HR	1.009	1.010
95% CI	0.987–1.031	0.983–1.039
p Value	0.426	0.472
Sex (n)	78	80
HR	1.040	0.971
95% CI	0.529–2.045	0.428–2.203
p Value	0.909	0.943
Tumor side (n)	90	103
HR	0.912	0.863
95% CI	0.475–1.750	0.412–1.808
p Value	0.782	0.696
EOR* (n)	81	83
HR	4.156	4.964
95% CI	1.886–9.156	1.918–12.848
p Value	0.000	0.001
WHO grade* (n)	96	109
HR	5.407	3.771
95% CI	2.087–14.005	1.439–9.881
p Value	0.001	0.007
Postoperative radiation (n)	93	101
HR	1.006	0.992
95% CI	0.514–1.968	0.446–2.206
p Value	0.986	0.984
Postoperative chemotherapy (n)	92	100
HR	1.543	1.020
95% CI	0.730–3.262	0.409–2.546
p Value	0.256	0.966

* HRs were calculated such that values > 1 reflect worse prognosis for STR and WHO Grade III tumors.

† Boldface values are statistically significant ($p < 0.05$), as determined by Cox proportional hazards model with 2-tailed tests for each prognostic factor.

was found in 80% of patients older than 40 years of age. The combination of hemispheric ependymomas having a greater incidence in adults and high-grade tumors correlating with age may account for our findings.

PFS and OS rates for hemispheric ependymomas did not differ substantially from previously reported rates for supratentorial ependymomas (Table 6). Five- and 10-year OS rates range from 35% to 79% and from 20% to 77%, respectively. The wide range of reported values for PFS and OS in adults with supratentorial ependymomas likely results from small sample sizes, with most studies including less than 25 patients. Vera-Bolanos and colleagues examined the clinical course and PFS of adults with intracranial and spinal ependymomas.⁵⁵ A total of 53 patients with supratentorial ependymomas were included for analysis. Kaplan-Meier survival analysis found similar PFS as that in the current study, with a median PFS of 4 years. Consistent with previous studies, supratentorial location

TABLE 5. Multivariate analysis of prognostic factors for PFS and OS*

Prognostic Factor	PFS	OS
EOR (n)	77	79
HR	4.764	4.216
95% CI	1.952–11.626	1.460–12.169
p Value	0.001	0.008
WHO grade (n)	77	79
HR	10.164	9.117
95% CI	2.216–46.612	1.170–71.076
p Value	0.003	0.035

* Boldface values are statistically significant ($p < 0.05$), as determined by Cox proportional hazards model with 2-tailed tests for each prognostic factor. Age, sex, tumor side, extent of resection, tumor grade, postoperative radiation, and chemotherapy were included in the multivariate analysis. HRs were calculated such that values > 1 reflect worse prognosis for STR and WHO Grade III tumors.

was a statistically significant prognostic factor for PFS. The majority of case series have found that supratentorial ependymomas carry a worse prognosis than infratentorial tumors do in adults.^{9,19,20,23,38,45,46,54}

Our data support EOR and tumor grade as the most important prognostic factors for adult hemispheric ependymomas. An unanticipated finding was that sex, age, and postoperative radiation were not prognostic for PFS or OS. Female sex,²⁰ younger age,^{21,38} and postoperative radiation^{20,46,51,54} have all been reported as prognostic for improved survival in intracranial ependymomas. Postoperative radiation has become commonplace in the management of anaplastic intracranial ependymomas.^{20,46,48,54} However, Vera-Bolanos et al. and our study, the 2 largest series evaluating prognostic factors, did not find a benefit to postoperative radiation. Further clinical investigation will be needed to determine the efficacy of radiation treatment in hemispheric ependymomas.

STR was the strongest predictor of poor PFS and OS in our series. Ependymomas have long been considered a surgical disease best managed with safe maximal resection. Professional guidelines from the National Comprehensive Cancer Network emphasize the importance of complete resection (http://www.nccn.org/professionals/physician_gls/f_guidelines.asp#site). Re-resection may be appropriate in the setting of initial STR with radiographic evidence of residual tumor. For patients with residual tumor, only 28% survived 5 years and no patient survived 10 years. EOR has emerged as an essential prognostic factor for hemispheric low- and high-grade gliomas.^{10,16,43,50} Our study supports the view that, similar to other hemispheric gliomas, patients with hemispheric ependymomas benefit from safe maximal resection. Hemispheric ependymomas are intraaxial parenchymal tumors that warrant the same surgical management strategies as astrocytomas and oligodendrogliomas. Advanced surgical techniques, such as functional pathway mapping and intraoperative MRI, should be used to maximize tumor resection and prolong patient survival.¹²

TABLE 6. Previously reported PFS, OS, and prognostic factors for supratentorial ependymomas

Authors & Year	No. of Patients	Location	5-Yr PFS %	10-Yr PFS %	5-Yr OS %	10-Yr OS %	Prognostic Factors for Worse OS
Marks & Adler, 1982	19	Supratentorial	16		35		Supratentorial
Vanuytsel et al., 1992	40	Supratentorial	37	34	48	40	WHO Grade III, STR, male, no radiation
McLaughlin et al., 1998	10	Supratentorial				20	Supratentorial
Schild et al., 1998	13	Supratentorial			62		Supratentorial, WHO Grade III
Schwartz et al., 1999	23	Supratentorial	42	27	77	77	WHO Grade III
Guyotat et al., 2002	23	Supratentorial	28	23	43	25	Supratentorial, WHO Grade III
Reni et al., 2004	37	Supratentorial			70	31	Supratentorial, age
Kawabata et al., 2005	21	Supratentorial	46	39	63	46	WHO Grade III, STR
Metellus et al., 2007	46	Supratentorial	37	28	77	62	STR, WHO Grade III, age, KPS score
Metellus et al., 2010	22	Supratentorial	53	35	79	74	Supratentorial, STR, KPS score
Current study, 2015	111	Hemispheric	51	42	71	58	WHO Grade III, STR

KPS = Karnofsky Performance Scale.

Previous reports conflict on the prognostic value of tumor grade in adult ependymomas.^{9,14,33,36} Our study provides evidence that WHO Grade III hemispheric ependymomas have a worse PFS and OS compared with WHO Grade II tumors in adults. With only 46% of patients with WHO Grade III tumors surviving to 10 years, compared with 77% of patients with WHO Grade II tumors, tumors with anaplastic histology are likely to demonstrate a more aggressive clinical course. The higher incidence of WHO Grade III pathology and associated poorer prognosis potentially contribute to the worse overall prognosis for hemispheric ependymomas compared with infratentorial ependymomas.

The limitations of our study result from the difficulty of investigating the clinical features and outcomes of a rare brain tumor. The inclusion of patients reported in the literature introduces selection bias and patient heterogeneity into the study population. To standardize the study population and avoid systematic error as much as is feasible, we included common categorical variables that were largely observer independent. Performance status and tumor dimensions were excluded to avoid measurement error. Patient characteristics, PFS, and OS were comparable to that reported in other clinical studies, reflective of a representative patient sample. The efficacy of specific postoperative radiation and chemotherapy regimens was not statistically analyzed due to sample size. Evaluation of each regimen with sufficient sample sizes would be needed to make definitive claims on treatment efficacy.

Conclusions

In summary, we analyzed 109 adults with supratentorial hemispheric ependymomas for survival and prognostic factors. WHO Grade III tumors constituted 62% hemispheric ependymomas, a higher percentage than either infratentorial or spinal ependymomas. Five- and 10-year OS rates were 71% and 58%, respectively. EOR and tumor grade were identified as prognostic factors for both PFS and OS on multivariate analysis. Postoperative radiation

was not found to be prognostic for PFS or OS. Our findings provide hitherto unknown clinical information that we hope will aid in the optimal evaluation and management of patients with hemispheric ependymomas.

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Disclosures

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Conception and design: Orringer, Junck. Acquisition of data: all authors. Analysis and interpretation of data: all authors. Drafting the article: Hollon, Lewis. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Orringer. Statistical analysis: Nguyen. Study supervision: Orringer, Junck.

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