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Meningiomas With Rhabdoid Features Lacking Other Histologic Features of Malignancy: A Study of 44 Cases and Review of the Literature

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Abstract

The behavior of rhabdoid meningiomas otherwise lacking malignant features remains unknown as most of the originally reported aggressive cases showed anaplastic histologic features independently of rhabdoid phenotype. We studied 44 patients with rhabdoid meningiomas lacking anaplastic features. Median age at diagnosis was 48.6 years (range 10–79). Location was supratentorial in 28 (63.6%), skull base in 15 (34.1%), and spinal in 1 (2.3%). Tumor grade was otherwise World Health Organization grade I (n = 22, 50%) or II (n = 22, 50%). Rhabdoid cells represented <20% of the tumor in 12 cases (27.3%), 20% to 50% in 18 (40.9%), and >50% in 14 (31.8%). Median clinical follow-up, available for 38 patients, was 5.0 years (range 0.17–14.2). Recurrence occurred in 9 patients (5-year recurrence-free survival, 73.7%) with a significantly higher risk in subtotaly resected tumors (p = 0.043). Rhabdoid cell percentage was not associated with recurrence. Six patients died (4 of disease, 2 of unclear causes); 5-year

overall survival was 86.7%, a mortality in excess of that expected in grade I–II meningiomas but much lower than originally reported. Review of 50 similar previously reported cases confirmed our findings. We suggest that rhabdoid meningiomas be graded analogously to non-rhabdoid tumors, with caution that some may still behave aggressively and close follow-up is recommended.

Key Words: Anaplastic meningioma, Meningioma, Rhabdoid meningioma, WHO grade.

INTRODUCTION

Rhabdoid meningioma is a rare, aggressive meningioma subtype. Histologically, these tumors are characterized by sheets of loosely cohesive, plump cells with eccentric nuclei and glassy, eosinophilic inclusion-like cytoplasm. The tumor cells resemble rhabdoid cells as described in tumors at other sites, in particular the kidney, and in the atypical teratoid rhabdoid tumors in the brain. Rhabdoid meningioma was initially defined in 1998 in 2 case series by Kepes et al (1) and Perry et al (2), who described a total of 19 patients with rhabdoid meningiomas. In the series of 15 cases by Perry et al, 13 patients had tumor recurrence and 8 died, with a median time to death of 3.1 years after initial appearance of rhabdoid morphology. This led to the classification of meningiomas with rhabdoid features as anaplastic (World Health Organization [WHO] grade III), in the 2000 WHO Tumors of the Central Nervous System (3). However, independent of rhabdoid morphology, the majority of rhabdoid meningiomas described in the initial series could be classified as anaplastic/malignant (grade III), based on mitotic rate or frankly anaplastic histology. As such, the behavior of meningiomas showing rhabdoid features in the absence of other features of malignancy has not been well studied and remains largely unknown (3, 4).

In this study, we describe the clinicopathologic features of 44 patients with meningiomas that showed rhabdoid features and lacked other histologic features of malignancy. Additionally, we analyzed 50 previously reported cases with similar features among 160 previously published rhabdoid meningiomas.

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Supplementary Data can be found at <http://www.jnen.oxfordjournals.org>.

TABLE 1. Patient Demographics (N = 44)

Age ^a (years), Median (range)	48.6 (9.8–79.1)
Gender	
Male	18 (40.9%)
Female	26 (59.1%)
Tumor Site	
Supratentorial	28 (63.6%)
Skull base	15 (34.1%)
Spinal cord	1 (2.3%)
Extent of Resection	
GTR or NTR	26 (68.4%)
STR	12 (31.6%)
Unknown	6
Adjuvant Therapy	
None	23 (67.6%)
Radiation therapy	9 (26.5%)
Chemotherapy (for concurrent neuroendocrine tumor)	1 (2.9%)
Gamma knife	1 (2.9%)
Unknown	10
Appearance of Rhabdoid Features	
Primary resection	41 (93.2%)
Recurrence	3 (6.8%)

GTR, gross total resection; NTR, near total resection; STR, subtotal resection.

^aAt first appearance of rhabdoid features.

MATERIALS AND METHODS

Forty-four cases of meningioma with rhabdoid features were identified in the surgical and consult archives of Mayo Clinic Rochester (15 cases), University of California San Francisco (10 cases), Washington University St. Louis (16 cases), and Johns Hopkins Hospital (3 cases) (Table 1; Supplementary Data Table 1). The hematoxylin and eosin (H&E) stained slides were centrally reviewed by 2 neuropathologists (CG and RAV) and evaluated for the presence of rhabdoid features, characterized by plump cells with eccentric nuclei and abundant eosinophilic inclusion-like cytoplasm (Fig. 1). In 3 tumors, rhabdoid features were present only at tumor recurrence; the slides included in this study were from the first specimen displaying rhabdoid features. For each case, the percentage of the tumor with rhabdoid morphology was estimated semiquantitatively (<20%, 20%–50%, >50%). When other histological patterns were present, they were recorded.

The tumors were graded according to WHO criteria as grade I or II independent of the presence of rhabdoid features (3). WHO II tumors were defined by a mitotic rate of ≥ 4 mitoses per 10 high-power fields or the presence of 3 or more atypical histologic features (hypercellularity, small cell change, macronucleoli, sheet-like growth, and necrosis). Brain invasion, when present, was also noted. WHO III tumors (displaying frank anaplasia or mitotic rate ≥ 20 per 10 high-power fields) were excluded.

Clinical follow-up was obtained by chart review or correspondence with clinicians. Follow-up data were available in 38 of 44 cases. For the 3 patients whose tumors showed

rhabdoid features at recurrence, follow-up was reported from the first appearance of rhabdoid features.

Electron microscopy studies were performed in 9 cases, 1 from tissue primarily fixed in glutaraldehyde for electron microscopy, 2 initially fixed in formalin and transferred to glutaraldehyde, and 6 from tissue that was primarily fixed in formalin and embedded in paraffin. This tissue was deparaffinized and reprocessed for electron microscopy.

Patient data were summarized with medians and ranges or frequencies and percentages, as appropriate. The percentage of the tumor with rhabdoid morphology was categorized into approximate tertiles. The risk of recurrence and death were each compared by rhabdoid percentage (<20% vs 20%–50% vs >50%), WHO grade (I vs II), tumor site (supratentorial vs skull base), and extent of resection (gross or near total vs subtotal) using log-rank tests. The recurrence-free and overall survival rates were each estimated at 2 and 5 years postdiagnosis, using the Kaplan-Meier method, along with 95% confidence intervals (CIs). The distribution of WHO grade was compared with the rhabdoid percentage categories with a Fisher exact test. P values less than 0.05 were considered significant. Analyses were performed using SAS version 9.3 (SAS Institute, Cary, NC).

We identified 160 previously published cases of rhabdoid meningioma through an OvidMedline search followed by careful review of each paper’s bibliography, (Supplementary Data Table 2). We reviewed the cases and tabulated the histological characteristics of each tumor to the best of the information provided. However, many of the case reports lacked details regarding tumor histology, mitotic rate, or the presence of atypical features. In papers in which the WHO grade was not explicitly reported, we assigned the tumors a WHO grade based on the histologic descriptions provided. In some cases, the authors used descriptions of mitotic rate, such as “scattered” or “not conspicuous,” and we were unable to distinguish them reliably as WHO I or II. Descriptors of mitotic rate such as “frequent” or “many” were considered to be WHO II–III. Several clinical series did not provide patient-level data and were excluded from analysis (5–7). Recurrence-free and overall survival rates were estimated using the same methods as described above.

RESULTS

Histologic Features

Rhabdoid features were identified in 41 initial resection specimens from the 44 cases reviewed; in the remaining 3 cases, rhabdoid morphology was present only at tumor recurrence. The extent of the rhabdoid features present within the tumor specimen was variable (Fig. 1). Rhabdoid features accounted for <20% of the examined area in 12 tumors (27.3%), 20% to 50% in 18 tumors (40.9%), and >50% in 14 tumors (31.8%) (Table 2). In addition to rhabdoid morphology, other histologic patterns were present in 36 cases. Additional growth patterns were predominantly meningothelial or transitional (in 33 cases); fibrous and metaplastic morphology were rare. In 2 tumors, papillary features were identified and comprised approximately 30% of the tumor in

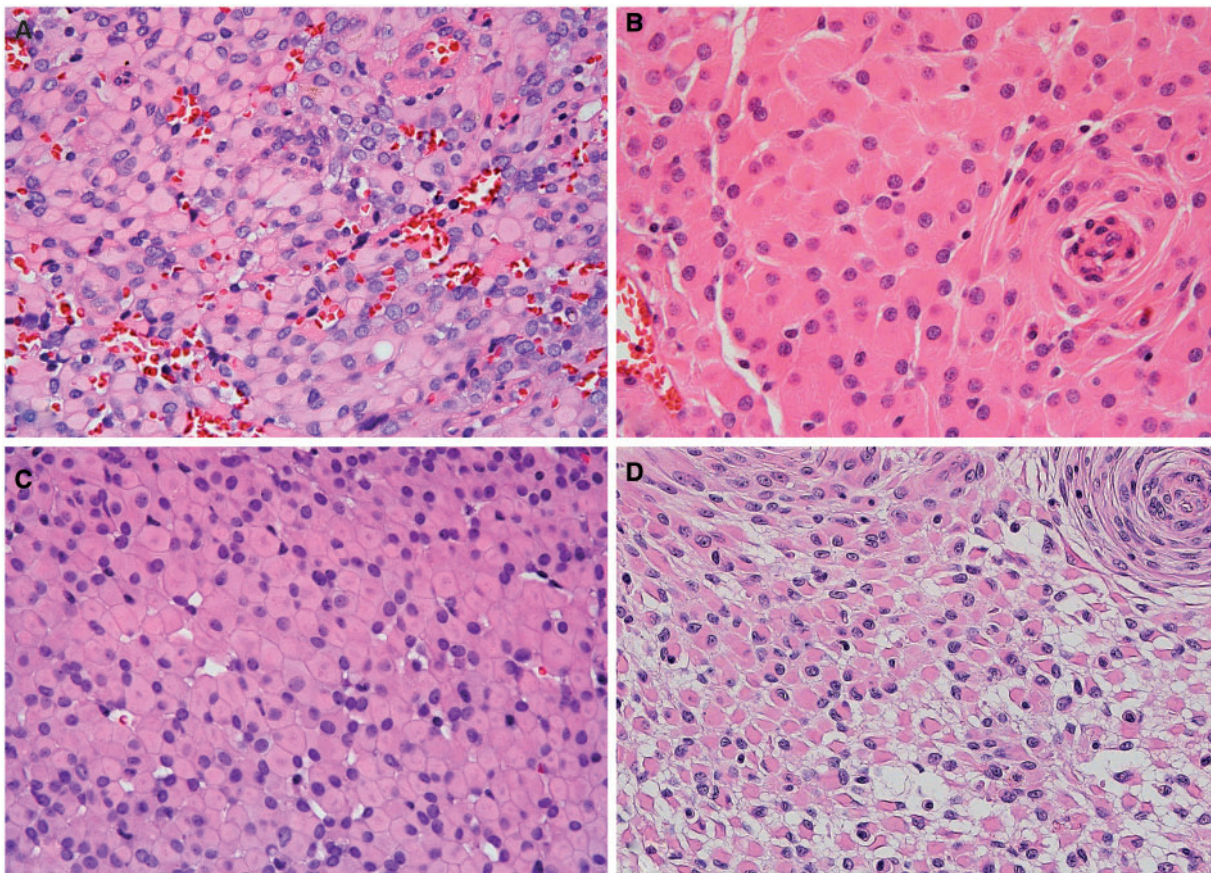


FIGURE 1. (A–D) Histologic features of rhabdoid meningioma, characterized by plump cells with eccentric nuclei and abundant eosinophilic cytoplasm. **(A, case 5), (B, case 24), and (C, case 7),** were predominantly rhabdoid. **(D)** Case 14 also had a prominent meningothelial growth pattern (hematoxylin and eosin [H&E], 400x).

TABLE 2. Patient Outcomes by Percentage of Rhabdoid Component

Percent Rhabdoid	All Tumors (N = 44)	<20% (N = 12, 27.3%)	20%–50% (N = 18, 40.9%)	>50% (N = 14, 31.8%)
WHO Grade				
WHO I	22 (50%)	7 (58.3%)	6 (33.3%)	9 (64.3%)
WHO II	22 (50%)	5 (41.7%)	12 (66.7%)	5 (35.7%)
Tumor Recurrence				
None	27	8	10	9
1	7	2	3	2
≥ 2	2	0	1	1
Unknown or no follow-up	8	2	4	2
Death				
DOD	4	1	2	1
Other/unknown cause	2	0	1	1
Follow-up Available, N				
	38	10	15	13
Follow-up, Median (range)				
	5.0 years (61 days–14.2 years)	5.1 years (182 days–14.2 years)	5.5 years (219 days–13.8 years)	4.0 years (61 days–6.0 years)

DOD, died of disease; WHO, World Health Organization.

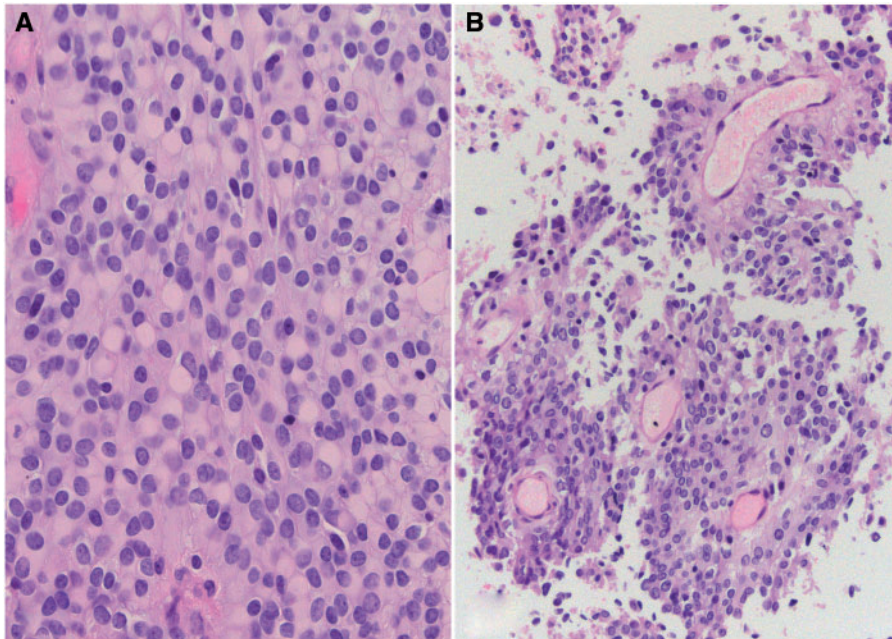


FIGURE 2. Composite rhabdoid-papillary meningioma. Both rhabdoid (**A**) and papillary features (**B**) were present in case 35. The tumor was otherwise a WHO grade II tumor, based on mitotic rate.

both (Fig. 2). Based on mitotic rate and/or the presence of atypical histology features, 22 tumors (50%) were graded as WHO I, and 22 tumors (50%) were graded as WHO II (Table 2). Brain invasion was identified in a single WHO II tumor. The extent of rhabdoid morphology did not correlate with WHO grade ($p = 0.24$).

To confirm the presence of a true rhabdoid phenotype, electron microscopy was performed in a subset of 9 tumors. Seven cases demonstrated the presence of intracytoplasmic whorls of intermediate filaments, which define a true rhabdoid phenotype (Fig. 3). Electron microscopy from paraffin-embedded tissue was unsuccessful in 2 cases due to inadequate tissue preservation.

Clinical/Therapeutic Data

The overall clinicopathologic features of the 44 cases are summarized in Table 1; individual case details are in Supplementary Data Table 1. There were 26 (59.1%) female patients and 18 (40.9%) male patients; the median age at diagnosis was 48.6 years (range 9.8–79.1). The majority of the tumors involved the supratentorial compartment (63.6%) and skull base (34.1%), with a single tumor occurring in the spinal cord (2.3%). The extent of resection was known in 38 cases; 26 (68.4%) patients underwent gross total or near total resection; subtotal resection was achieved in 12 (31.6%) cases. Nine patients (26.5%) received adjuvant radiotherapy, and 1 received chemotherapy for a concurrent neuroendocrine tumor in addition to radiotherapy.

Patient Outcome

Clinical follow-up information was available for 38 patients, with a median follow-up length of 5.0 years (range 61 days–14.2 years) after the first identification of rhabdoid features (Table 2). Overall, there were 9 tumor recurrences corresponding to a 5-year recurrence-free survival of 73.7% (95% CI: 57.8%, 89.6%). Of the 9 recurrences, 4 tumors were WHO I tumors and 5 were WHO II. Recurrences occurred in tumors that were <20% rhabdoid ($n = 2$), 20%–50% rhabdoid ($n = 4$), and >50% rhabdoid ($n = 3$). Recurrence-free survival was not significantly associated with the extent of rhabdoid morphology ($p = 0.77$) (Table 3). Similarly, recurrence-free survival was not significantly different between WHO I and WHO II tumors (5-year recurrence-free survival was 75.0% for WHO I and 71.6% for WHO II, $p = 0.73$). Only the extent of tumor resection correlated with recurrence-free survival, with subtotally resected tumors significantly more likely to recur (5-year recurrence-free survival 53.6% vs 84.6%, $p = 0.043$). Skull base tumors showed a trend toward lower recurrence-free survival when compared to supratentorial tumors but this was not significant ($p = 0.08$). Adjuvant radiation therapy was not associated with recurrence-free survival ($p = 0.87$).

There were 6 patient deaths over the follow-up interval, corresponding to a 5-year overall survival of 86.7% (95% CI: 74.2%, 99.1%) (Table 2). Of these, 4 male patients, aged 18, 43, 53, and 56 years, died of disease. Additionally, 2 patients died of unknown causes. One patient was a 59-year-old man with no clinical follow-up; the other was a 78-year-old woman with a history of both renal cell carcinoma and insular thyroid carcinoma. There were too few deaths for formal statistical analysis; however, the deaths occurred in tumors

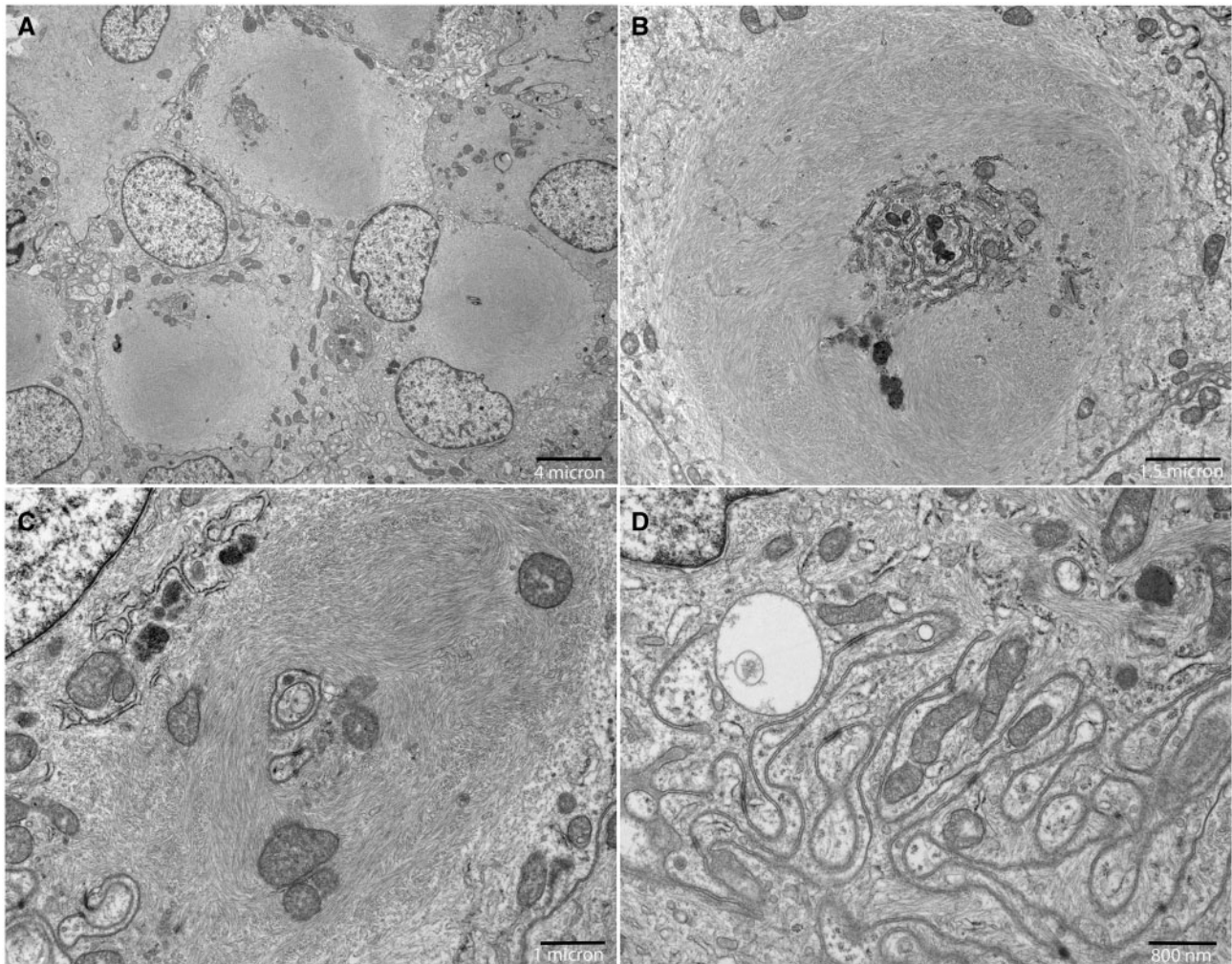


FIGURE 3. Rhabdoid meningioma ultrastructure. **(A–C)** Electron microscopy was performed on glutaraldehyde-fixed tissue in case 5; the corresponding H&E section is shown in Figure 1A. Rhabdoid morphology is defined by intracytoplasmic whorls of intermediate filaments **(A)**, which entrap cellular organelles, including endoplasmic reticulum **(B)** and mitochondria **(C)**. **(D)** The characteristic features of meningotheial cells, including intercellular junctions formed by interdigitating cell processes, were also present.

across the range of focal to predominantly rhabdoid tumors. Disease-related deaths occurred in 3 patients with WHO II tumors and 1 with a WHO I tumor. Of note, 2 of the disease-related deaths occurred in the patients (aged 18 and 43 years) whose tumors also had papillary features.

When including only the patients in which the rhabdoid phenotype was present at first diagnosis (excluding the 3 patients whose tumors showed rhabdoid features only at recurrence), there were 35 patients with clinical follow-up. Five-year overall survival in this group was 88.5% (95% CI: 76.0%, 100.0%), and 5-year recurrence-free survival was 81.7% (95% CI: 67.0%, 96.5%). Each of the 3 patients with tumors that were rhabdoid at recurrence had additional tumor recurrences and 1 patient died.

Literature Review

Since the initial case series reports (1, 2), we identified 52 publications in the English language literature detailing

160 cases of rhabdoid meningioma (1, 2, 4–54) (Table 4; Supplementary Data Table 2). In 29 of these publications, describing a total of 50 patients, the authors either assigned the tumors a WHO grade independent of rhabdoid phenotype or reported histologic information that we considered sufficient to grade the tumors as WHO I or II. The majority of these patients ($n = 35$) had WHO II tumors; there were fewer WHO I lesions ($n = 9$). Six additional tumors were definitely in the WHO I–II range, but sufficiently detailed data were not available to distinguish between grades I or II (Table 5). Only a small number of cases contained information regarding the extent of rhabdoid features, precluding analysis of this variable. Intriguingly, a high proportion of the WHO grade I and II tumors ($n = 11$, 22%) was also reported to have papillary features in addition to rhabdoid morphology.

The overall patient characteristics in these 50 published cases of rhabdoid meningioma lacking anaplastic features were similar to our series, although the median age of 36 years

TABLE 3. Five-Year Recurrence-Free Survival

	Number of Cases	Number of Recurrences	Five-Year Recurrence-Free Survival (95% Confidence Interval)	P Value ^a
Overall	44	9	73.7% (57.8%, 89.6%)	
Extent of Resection				
Gross or Near Total	26	3	84.6% (68.5%, 100%)	0.043
Subtotal	12	6	53.6% (23%, 84.2%)	
Tumor Site^b				
Supratentorial	28	3	86.6% (68.4%, 100.0%)	0.08
Skull base	15	5	61.5% (35.1%, 88.0%)	
Adjuvant Radiation				
Radiation	25	5	86.7% (72.7%, 100.0%)	0.87
None	9	2	87.5% (64.6%, 100.0%)	
% Rhabdoid				
< 20%	12	2	78.8% (52.5%, 100%)	0.77
20%–50%	18	4	75.2% (50.6%, 99.8%)	
> 50%	14	3	70.0% (41.6%, 98.4%)	
WHO Grade				
WHO I	22	4	75.0% (53.7%, 96.4%)	0.73
WHO II	22	5	71.6% (47.3%, 95.8%)	

WHO, World Health Organization.

^aLog-rank test.

^bThe single spinal cord tumor was excluded from statistical analysis.

TABLE 4. Patient Demographics of 50 Reported Cases of WHO Grade I–II Rhabdoid Meningiomas

Age^a (years), Median (range)	36.0 (3–84)
Gender	
Male	20 (40.0%)
Female	30 (60.0%)
Tumor Site	
Supratentorial	31 (62.0%)
Skull base, posterior fossa, or infratentorial	16 (32.0%)
Spinal cord	3 (6.0%)
Extent of Resection	
GTR or NTR	32 (68.1%)
STR	15 (31.9%)
Unknown	3
Adjuvant Therapy	
None	32 (65.3%)
Radiation therapy	12 (24.5%)
Chemotherapy	1 (2.0%)
Radiation and chemotherapy	4 (8.2%)
Unknown	1
Appearance of Rhabdoid Features	
Primary	46 (92.0%)
Recurrence	4 (8.0%)
Papillary Morphology	11 (22.0%)

GTR, gross total resection; NTR, near total resection; STR, subtotal resection.

^aAt first appearance of rhabdoid features.

(range 3–84 years) was slightly younger (Table 4). Patients received similar treatments, with a comparable proportion of gross-total resection and adjuvant therapies. Available follow-up was shorter than observed for our patients (47 patients with follow-up, median 2 years, range 2 days–17 years).

Of these 47 reported cases with follow-up, 22 patients had tumor recurrence, corresponding to a 2-year recurrence-free survival of 55.2% (95% CI: 37.4%, 73.0%) and a 5-year recurrence-free survival of 34.2% (95% CI: 14.4%, 54%). For this group, 2-year survival estimates are likely more stable as limited follow-up was available at 5 years. However, both 2- and 5-year estimates are reported for comparison to our series. Recurrence-free survival showed a trend towards association with WHO grade ($p = 0.06$) but no clear associations with extent of resection ($p = 0.37$), tumor site ($p = 0.28$), or presence of papillary features ($p = 0.28$). There were 11 total deaths in this cohort, with a 2-year overall survival of 86.8% (95% CI: 75.7%, 98.0%) and 5-year overall survival of 77.6% (95% CI: 62%, 93.3%). Nine patients died from disease, and 2 patients died of unknown or other causes (Table 5).

When we combined all reports of cases of rhabdoid meningioma that described individual patient outcomes (96 cases, including WHO III tumors), outcome was strongly correlated with WHO grade (Table 6). There were 17 patients with WHO III tumors defined by mitotic rate. Of these, 10 patients died, corresponding to a 2-year survival of 60.2% (95% CI: 34.8%, 85.5%) and 5-year survival of 34.4% (95% CI: 8.0%, 60.8%). There were 7 cases in which the histologic grade could not be reliably defined as WHO II or WHO III and 22 cases with insufficient information to reliably assign

TABLE 5. Patient Outcomes of Reported Cases of WHO Grade I–II Rhabdoid Meningiomas

	Overall (N = 50)	WHO I (N = 9)	WHO I-II ^a (N = 6)	WHO II (N = 35)
Tumor Recurrence				
None	26	7	3	16
1	11	1	1	9
≥2	11	1	2	8
Unknown	3	0	0	3
Death				
DOD	9	2	1	6
Other/unknown cause	2	0	0	2
Follow-up Available, N	47	9	6	32
Follow-up, Median (range)	2 years (2 days–17 years)	3.8 years (2 days–10.9 years)	1.7 years (5 days–5.4 years)	1.8 years (1 month–17 years)

WHO, World Health Organization; DOD, died of disease.

^aMitotic rate and/or tumor description were insufficient to differentiate WHO I vs WHO II tumors.

TABLE 6. Five-Year Survival of All Published Cases

WHO Grade	Number of Cases	Number of Deaths	Two-Year Survival (95% CI)	Five-Year Survival (95% CI)
Our series	44	6	91.0% (81.3%, 100.0%)	86.7% (74.2%, 99.1%)
I–II	50	11	86.8% (75.7%, 98.0%)	77.6% (62.0%, 93.3%)
II–III ^a	7	5	80.0% (44.9%, 100.0%)	53.3% (4.7%, 100.0%)
III	17	10	60.2% (34.8%, 85.5%)	34.4% (8.0%, 60.8%)
Unknown ^b	22	9	53.0% (30.4%, 75.7%)	53.0% (30.4%, 75.7%)

CI, confidence interval; WHO, World Health Organization.

^aMitotic rate and/or tumor description were insufficient to differentiate WHO II vs WHO III tumors.

^bCases contained insufficient histologic information to reliably assign any WHO grade.

any histologic grade. These groups, respectively, had intermediate 2-year survivals of 60.3% and 53.0% and 5-year survivals of 53.3% and 53.0% (Table 6).

DISCUSSION

Since the initial definition of rhabdoid meningioma in 1998, the behavior of rhabdoid meningiomas lacking the histologic features of malignancy has remained an open question. According to WHO 2007 (4), “A minority of meningiomas with rhabdoid features shows this only focally and lacks other histologic features of malignancy; the behavior of these tumors remains to be determined.” A major challenge to answering this question is that rhabdoid meningioma is a rare entity. Across 4 large academic institutions, we were able to identify 44 cases of rhabdoid meningioma that lacked other features of malignancy treated over a period of 29 years. We also identified 50 additional reported cases of rhabdoid meningioma, lacking other features of malignancy published since 1998. The overall patient age in our study (median of 48.6 years) and in the published cases (median of 36.0 years) was somewhat younger than a typical meningioma case series (Tables 1 and 5). Similarly, there was a slightly higher proportion of male patients than expected in our series (40.9%) and in the published

case series (40.0%). The overall patient presentations and treatment were otherwise typical of meningioma case series (Tables 1 and 5).

Histologically, the tumors in our series demonstrated a range of morphologies from tumors with focal (<20%) to predominant (>50%) rhabdoid features (Fig. 1). However, this percentage did not correlate with recurrence-free survival (p = 0.77, Table 3), suggesting that the extent of rhabdoid features is not an independent risk factor for tumor recurrence. Similarly, the extent of rhabdoid features was not associated with WHO grade (Table 2). In our series, only extent of tumor resection had a significant effect on recurrence-free survival (p = 0.043, Table 3).

Within our series of 44 patients, 9 patients had tumor recurrence, corresponding to a 5-year recurrence-free survival of 73.7% (95% CI: 57.8%, 89.6%). In contrast, within the 50 published WHO I–II cases (Table 5), there were 22 tumor recurrences, corresponding to a 5-year recurrence-free survival of 34.2% (95% CI: 14.4%, 54%). This lower recurrence-free survival may in part reflect a bias toward publication of more aggressive cases. Consistently, the previously published cohort of patients also contained a higher proportion of tumors that were WHO II (70% compared to 50% in our series), as well as a high proportion (22%) of tumors with papillary

features, another aggressive variant classified as WHO III (3). However, the presence of papillary morphology in these cases was not associated with lower recurrence-free survival ($p=0.28$). Within our series of 44 cases, there were 6 patient deaths (Table 2), corresponding to a 5-year overall survival of 86.7% (95% CI: 74.2%, 99.1%). This was comparable to, although slightly higher than, what was observed in our analysis of the published WHO I–II tumors with similar features. In the latter group there were 11 patient deaths (Table 5), corresponding to a 5-year overall survival of 77.6% (95% CI: 62%, 93.3%).

Among the original 15 cases reported by Perry et al (2), 13 patients had tumor recurrence and 8 died, with a median time to death of 3.1 years (range 10 months–10.7 years) after appearance of rhabdoid morphology. Of the 15 tumors, 8 were overtly malignant (WHO III), independent of rhabdoid morphology, 4 were WHO II, and 3 were WHO I at the initial identification of rhabdoid features. Irrespective of rhabdoid morphology, it is likely that the presence of high mitotic rate and other features of malignancy had a significant impact on tumor outcome in this initial series. In our review of the published literature of rhabdoid meningiomas, we identified 17 tumors that were WHO III based on mitotic rate and independent of rhabdoid morphology (Table 6). These tumors behaved extremely aggressively, with 5-year survival of 34.4%. An additional group of 22 similar tumors of unclear histologic grade had a 5-year survival of 53%, comparable to the original series (2).

Our case series and review of the literature suggest that most meningiomas that have rhabdoid features and lack other features of malignancy (WHO I or II) are not as aggressive as rhabdoid meningiomas with independent histological features consistent with WHO grade III. Given the small number of patient deaths in our series, we were unable to perform a formal statistical analysis. However, the 6 deaths in our case series and 11 deaths in the reviewed literature are more than expected in a meningioma series of comparable grade (55). Although the published literature may be biased toward publication of recurrent/more aggressive cases, even in our series, the overall mortality data are higher than expected for a similar cohort of patients with WHO I–II tumors of typical meningothelial morphologies. This suggests that a subset of these tumors may still behave aggressively, even when lacking other features of malignancy. Although the median follow-up in our series (5.0 years) is longer than the median time to death in the initial series (1, 2), it is also possible that a longer follow-up period might have revealed additional tumors with aggressive behavior.

At present, there are no histologic features that can identify the subset of tumors with an aggressive clinical course. Therefore, we suggest that rhabdoid meningiomas be graded similarly to nonrhabdoid meningiomas but with an additional diagnosis comment that a subset of the otherwise grade I and II tumors will behave aggressively. We therefore recommend close clinical follow-up. While it may be the most exhaustive study to date, our case series still contains a relatively small number of cases that were examined retrospectively. Additional studies will be necessary to clarify the significance of rhabdoid morphology.

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