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Spinal location is prognostic of survival for solitary-fibrous tumor/hemangiopericytoma of the central nervous system

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Abstract

Background: Prior studies have highlighted infratentorial tumor location as a prognostic factor for solitary fibrous tumor (SFT) and hemangiopericytoma (HPC) of the central nervous system (CNS), and spinal location is considered a positive prognostic factor for other tumors of the CNS. While SFT/HPC of the CNS is known to frequently arise from the spinal meninges, there are no case series that report outcomes for spinally located CNS tumors, and their prognosis in relation to intracranial and other CNS-located tumors is unknown.

Objective: To investigate outcomes for patients with SFT/HPC of the spinal meninges.

Methods: The Surveillance, Epidemiology, and End-Results Program was used to identify patients with SFT/HPC within the CNS from 1993 – 2015. We retrospectively analyzed the relationship between tumor location (spinal vs. brain and other CNS) and survival.

Results: We identified 551 cases of CNS SFT/HPC, 64 (11.6%) of which were primary tumors of the spinal meninges. Spinal tumors were more likely than brain and other CNS tumors to be SFT vs. HPC (37.5 vs. 12%, $p < 0.001$), benign (42.2 vs. 20.3%, $p < 0.001$), and less than 5 cm (53.1 vs. 35.7%, $p < 0.001$). The 10-year survival rates for spinal and brain/other CNS tumors

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were 85 and 58%, respectively. Median survival time was significantly longer for spinal tumors (median survival not reached vs. 138 months, $p = 0.03$, $HR = 0.41$ [95% CI 0.18 – 0.94]). On multivariable analysis, spinal tumor location was associated with improved survival over tumors located in the brain and other CNS ($HR = 0.36$ [95% CI 0.15 – 0.89], $p = 0.03$).

Conclusion: Spinal tumor location is associated with improved survival in patients with SFT/HPC of the CNS. Larger institutional studies are necessary to characterize the relationship between tumor location and other relevant factors such as presentation and amenability to gross-total resection and adjuvant radiotherapy. Future studies exploring optimal management of spinally located tumors are also needed.

Keywords

Solitary-Fibrous Tumor; Hemangiopericytoma; Central Nervous System; Meningeal Tumor; Spinal Tumor

Introduction:

Previously separated on the basis of their clinical and pathological behaviors, the 2016 World Health Organization (WHO) classification of tumors of the central nervous system (CNS) combined solitary fibrous tumor (SFT) and hemangiopericytoma (HPC) into one category [1]. SFT and HPC are now known to demonstrate the NAB2-STAT6 fusion, which can be identified by immunohistochemistry [2–6]. Although these tumors are understood to share a biologic basis [7], they remain behaviorally distinct: SFTs occurring in the CNS tend to be classified as benign and are usually treated with resection alone [8, 9], while HPCs are aggressive, known to metastasize systemically, and commonly recur [10–13]. HPCs of the CNS are frequently treated with adjuvant radiotherapy after resection [14, 15].

The annual incidence rate of CNS SFT/HPC in the United States is 3.77 per 10,000,000 people and is rising [14]. The rarity of this disease and accompanying paucity of clinical trials has made optimal management difficult to establish [16]. Since the recent update of WHO CNS classifications, few studies have analyzed the prognostic factors and related outcomes in SFT and HPC combined [14, 17–19]. Those that have are often limited by small sample sizes and a subsequent inability to identify and validate prognostic factors. The capability of assessing rare diseases on a large scale renders the Surveillance, Epidemiology, and End Results (SEER) Program and similar cancer registry databases uniquely positioned to address these limitations [20–23].

Prior studies have stratified this disease on the basis of tumor subtype, grade, and histopathology [14, 19, 24]. We chose to investigate prognostic factors, outcomes, and overall survival associated with tumor location (spinal versus brain and other CNS) while incorporating the latest WHO classification for CNS tumors. Previous literature has highlighted infratentorial tumor location as a major prognostic factor [12, 20–22, 25], along with patient age [20, 22], tumor size [13, 26], and extent of resection [12, 13, 25, 27, 28], among others. CNS SFT/HPC can arise from dura of the brain or from the spinal meninges. Other types of CNS spinal tumors carry varied prognoses: spinal cord gliomas, while rare, have a notoriously poor prognosis [29, 30] comparable to their intracranial counterparts.

Conversely, spinal meningiomas are known to be benign and more amenable to surgical resection [31, 32] than intracranial meningiomas. While SFT/HPC tumors of the CNS are known to occur in the spinal meninges, there have been no large case series examining outcomes for these tumors. We used retrospective data from the SEER database to further examine the demographics and potential prognostic factors of patients diagnosed with SFT or HPC.

Methods:

Data Source

The SEER Program, which is the National Cancer Institute's authoritative source for information on cancer incidence and survival [33], is considered the gold standard for cancer data collection internationally [34]. Data is collected from population-based cancer registries around the country, covering approximately 28% of the United States population. The database routinely undergoes quality-control checks and is updated annually for follow-up on patient vital status. Data was collected and coded as previously described [14, 35–37].

Sample Selection and Coding

We queried the SEER database (November, 2017 submission, 1973 – 2015)[38] to identify all cases of SFT (International Classification of Diseases- (ICD-) O-3 code 8815) and HPC (ICD-O-3 9150) within the CNS (ICD-O-3 codes C70.1-C72.9), diagnosed between January 1, 1993 and December 31, 2015. The following variables were collected and coded: age at diagnosis, sex, race, percent of persons below poverty, ICD-0–3 histology, ICD-0–3 behavior, primary site, size, and surgery at primary site. Information on receipt of radiation therapy was not available in the SEER 2017 submission. Cases that did not receive surgery or for which surgical status was unknown (n=40) were excluded from the analysis.

The percent of persons below the poverty line variable in SEER is calculated based on county information from the American Community Survey by the United States Census Bureau. ICD-0–3 behavior coding in SEER is a histological parameter indicating the likely behavior of the tumor regarding its potential to invade surrounding tissue, based on what most pathologists believe is usual for that tumor. ICD-0–3 behavior is determined by the coding pathologist and classified as benign, borderline malignant, or malignant. ICD-O-3 behavior was analyzed in this study as an ordinal variable. WHO grade was not included in the analysis due to obvious inconsistencies between historical and current WHO grading guidelines in the tumors that were diagnosed before 2007 [39]. Tumor location was coded as spinal (ICD-O-3 C70.1 and C70.2, spinal cord and spinal meninges) or brain and other CNS (all other ICD-O-3 codes of the brain and CNS). Tumors of cranial nerves or other parts of the CNS not otherwise specified were included in this category. Tumor size was coded as < 5 cm, 5 cm or greater, or unknown. Extent of resection was coded as subtotal resection (STR), gross-total resection (GTR), or unknown from the surgery at primary site variable. Surgery codes in SEER are submitted by individual state registries based on definitions in the American College of Surgeons Commission on Cancer's Facility Oncology Registry Data System (FORDS) manual [40]. Information on extent of resection, however, is not routinely coded for spinal tumors. Certain variables, including invasiveness and metastasis at

diagnosis [11] were only available for tumors diagnosed after 2004 and were, therefore, not included in the analysis. While patients in this series overlap with those in our previous published work [11, 14] and in other SEER studies [20, 22, 41], we conducted this study using the latest SEER submission and all data extraction, coding, and analysis were conducted independently.

Statistical Analysis

All statistical analyses were conducted with the IBM SPSS Statistics software package (International Business Machines Corporation, Armonk, New York). Median survival times were determined using the Kaplan-Meier method and significance was determined using the log-rank test. Univariable and multivariable analyses of overall survival was conducted using the Cox Proportional Hazards Ratios (HR) model to estimate hazard ratios and 95% confidence intervals (95% CI). Associations between tumor location and other variables were determined using the Pearson's chi-square test and column proportions were compared using Bonferroni correction to adjust for multiple comparisons. Tests with two-tailed P values < .05 were considered statistically significant. Demographical and clinical features that were significantly associated with survival were included in multivariable analyses.

Results:

Patient Characteristics

551 patients were identified that matched search criteria, 487 of which had tumors that were located in the brain and other CNS, and 64 which were in the spinal meninges. Patient and clinical characteristics are presented in Table 1. There was a slight female predominance for all SFT/HPC tumors (51.4%), although subgroup analysis revealed a slight male predominance (57.8%) in spinal tumors. Most patients with spinal tumors were white (82.5%) and patients identifying as black (9.5%) were the largest minority affected. There was no significant difference in the percentage of patients below the poverty line. A larger proportion of spinal vs. brain/other CNS tumors were classified as SFT (38 vs. 13%, $p < 0.001$) although the majority were classified as HPC (62.5%). Similarly, a larger proportion of the spinal tumors were considered benign (42 vs. 23%, $p < 0.001$) and less than 5 cm in size (53 vs. 36%, $p < 0.001$). There was a trend for older age at diagnosis in the spinal cases (median age 56 vs. 53 years old, $p = 0.06$). Many patients with spinal tumors received GTR (35.9%), although data on extent of resection was unavailable the majority of spinally located tumors (60.9%).

Survival

Median follow-up time for all tumors was 52 months. Over this time period, 134 deaths occurred and 417 cases were censored. Median survival time for all tumors was 139 months. Median survival time was significantly longer for spinal tumors than tumors in the brain and other CNS (median survival not reached vs. 138 months, $p = 0.03$, HR = 0.41 [95% CI 0.18 – 0.94]). The corresponding 10-year survival rates for spinal and brain/other CNS tumors were 85% and 58%, respectively (Figure 1, Table 2). Other prognostic factors of survival included older age (HR = 1.04 [95% CI 1.03 – 1.05], $p < 0.001$), race ($p = 0.03$), regional poverty levels (HR = 1.05 [95% CI 1.02 – 1.08] per percentage point, $p = 0.001$), and

increased malignant histological behavior (HR = 1.41 [95% CI 1.18 – 1.8], $p = 0.005$). Asian/Pacific Islander race was significantly associated with increased survival when compared with white race (HR = 0.28 [95% CI 0.13 – 0.65], $p = 0.003$). The relationship between overall survival and tumor subtype, tumor size, and extent of resection did not reach statistical significance in this analysis. It should be noted that GTR vs. STR had reached statistical significance in our previous SEER analysis [11], but this was only after accounting for combined modality treatment with radiation. Information on extent of resection was not available for the majority of spinal tumors and was, therefore, not a major focus of this analysis.

Age, race, regional poverty level, tumor location, and histological behavior were included in the multivariable analysis. After accounting for these other variables, spinal location versus brain and other CNS was still significantly associated with improved survival (HR = 0.36 [95% CI 0.15 – 0.89], $p = 0.03$). Older age (HR = 1.04 [95% CI 1.03 – 1.05], $p < 0.001$), regional poverty level (HR = 1.04 [95% CI 1.01 – 1.07], $p = 0.01$), and aggressive histological behavior (HR = 1.40 [95% CI 1.09 – 1.80], $p = 0.009$) were associated with decreased survival, while Asian/Pacific Islander versus white race (HR = 0.34 [95% CI 0.15 – 0.78], $p = 0.01$) remained associated with increased survival.

Discussion:

In this study we show that a previously unreported tumor feature, spinal location, is a predictor of increased survival in SFTs/HPCs of the CNS. To our knowledge, the current analysis of 551 tumors, including 64 spinal tumors, is the largest combined series of SFT/HPC to date. Although CNS SFT/HPCs may arise from the meninges of the spine, there have been no series that specifically examine outcomes in this group.

Few studies have presented a combined analysis of SFT/HPC since the WHO guidelines were updated in 2016 [1], in part due to the rarity of these tumors [14, 42, 43]. Those that have report conflicting results [7, 9, 14, 18, 19] with regards to classification, optimal management, and outcome. Prior literature has stratified SFT/HPC tumors by location [11, 14, 18, 19] and demonstrated a correlation between infratentorial or posterior fossa location and outcome. However, no correlation between spinal location and clinical behavior has been previously recognized. Our finding may be due in part to common characteristics of spinal tumors such as benign histology and smaller size, both of which have been associated with an improved prognosis [42]. However, after accounting for other factors that were significantly associated with survival, spinal location remained an independent prognostic factor in this study. This paper's novel finding of tumor location as a prognostic feature suggests that it should be considered during clinical management of HPCs/SFTs.

It is important to distinguish SFT/HPC of the spinal meninges versus SFT/HPC of the osseous spine. The former is classified as a CNS tumor by WHO, whereas the latter is categorized as a soft tissue and is commonly referred to as osseous HPC. When CNS SFT/HPCs manifest in the spinal meninges, they are most often intradural and extramedullary [44, 45]. They also often occur in the cervical and thoracic segments of spine, as opposed to osseous spinal tumors, which most often affect the lumbosacral spine [45]. One combined

study of osseous SFT/HPC demonstrated that updated WHO grade and total resection were significantly associated with recurrence and overall survival [46]. However, most studies were conducted when HPC and SFT were treated as separate entities. Several studies have documented local recurrence of both osseous and meningeal spinal tumors [45, 47], and outcomes for both types were found to be similar [45].

SFTs are generally benign (WHO grade I) and have a better overall prognosis, while HPCs (WHO grade II and III) have a higher propensity for recurrence and metastases [8]. One exception is anaplastic SFT, which is considered WHO grade III. In our study, histological behavior (benign vs. borderline malignant or malignant) was strongly associated with survival, while histological tumor type (SFT vs. HPC) failed to show a statically significant correlation. These results emphasize that SFT/HPC of the CNS should be considered as a spectrum of the same disease, and are better prognosticated when analyzed that way. Several other studies have also failed to find a significant correlation between tumor type and survival [18, 19, 24], whereas histological aggressiveness, as measured by WHO grade, has been more commonly associated with poorer prognosis [10, 44]. It should be noted that WHO grade was inconsistently annotated for cases in this study and that histological behavior, as annotated by the coding pathologist, was used as a substitute for histological grade. Future population-based studies that incorporate WHO grade would further corroborate our findings. However, we are unaware of any sizeable databases that can reliably provide this information.

The finding of tumor location as a significant prognostic factor may be linked to the achievable extent of total resection, which has been previously established to be a powerful prognostic factor for recurrence and overall survival in HPCs/SFTs [20, 28, 48–50]. We have similarly found that extent of resection is a significant prognostic indicator when accounting for receipt of radiation [14], although this variable was not available in the most recent SEER dataset. While there was a trend towards improved survival with GTR vs STR ($p = 0.08$), statistical significance was not reached. This is likely due to insufficient data, as the extent of resection was unknown for the majority of spinal tumors studied here and methodological limitations prevented us from exploring this association. With regards to optimal management of spinally-located meningeal SFT/HPC, most of the literature supports complete excision [43, 48]. Tumor size may also be related to clinical management, with the expectation that smaller tumors are more amenable to adequate exposure, GTR, and radiation therapy. Other factors influencing GTR and intraoperative complications include vascularity of the tumor [18, 19], with higher grade tumors demonstrating increased vascularity and less amenability to GTR. It is important to note that one study did not find any benefit from GTR with regards to survival or recurrence [49]. Similarly, while radiotherapy has been found to improve local control and overall survival in CNS HPC [11, 14, 22, 48], its role in spinal SFT/HPC is yet to be elucidated.

Tumor size has been previously associated with recurrence and by association, prognosis [13, 26]. In this series, we found that spinally-located tumors were more likely to be < 5 cm, although size was not associated with overall survival. It is possible that spinal structures, such as the spinal cord and nerve roots, are more sensitive to mass effect induced by an enlarging tumor and subsequently present with symptoms earlier in the clinical course.

Unfortunately, the SEER database does not record symptom-related data, so we were unable to explore this aspect further. Larger combined and potentially prospective studies are necessary to more accurately characterize the relationship between tumor size, location, and overall survival.

The data presented here confirm prior findings of age [20, 22], and malignant behavior [10, 17, 27] as significant prognostic factors. We also found that regional poverty level, a previously unreported factor, significantly influenced overall survival when controlling for other prognostic indicators, such as race. Unfortunately, there is a paucity of literature regarding the interaction of socioeconomic status and SFT/HPC, although this interplay has been explored extensively in a variety of more common cancers, including lung, breast, and prostate [51–53]. In meningiomas and gliomas, one study found no correlation between income and tumor development [54], while others have noted an increased incidence of low grade glioma, meningioma, and acoustic neuroma in patients with higher socioeconomic status [55]. The latter finding likely represents diagnostic bias, a result of disparities in access to screening and care [56]. Furthermore, disparities in the receipt of radiation therapy across several cancers and organ systems is well documented [57, 58]. While information regarding radiation therapy was not readily available for this study, understanding the interaction of socioeconomic status, race and access to treatment has significant implications for outcomes research in this field.

Population-based databases are useful for studying rare tumors in the general population [11, 14, 22, 59, 60], but are limited by inherent biases related to missing data, as well as the retrospective nature of data collection [61]. Examples that may be relevant in the study of CNS SFT/HPC include WHO grade and central pathology review, radiation therapy, and patient co-morbidities, amongst others. Nevertheless, all current literature on SFT/HPC continues to be derived from retrospective databases. Given these limitations, we could not perform an analysis on optimal management for spinally located tumors in the current dataset, but future studies are welcome. Furthermore, randomized and prospective clinical trials are sorely needed, even if they are difficult to perform given the rarity of this disease.

Conclusion:

We used population-level statistics to show that tumor location (spinal meninges vs. brain and other CNS including cranial nerves) is associated with increased overall survival for SFT/HPC of the CNS. This report represents the largest currently published data set of prognostic factors and outcomes in patients with combined SFT/HPC of the CNS, and the first study to compare outcomes between intracranial and spinal tumors. While population-based registries are a useful resource to study rare tumors such as these, we also recognize the limited availability of surgical and adjuvant treatment data in the SEER database. The findings presented here may further inform the workup and management of patients with HPC/SFT of the CNS.

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Number at Risk

Spinal	64	31	6	1
Other	487	251	113	27

Figure 1:

Kaplan-Meier analysis of OS by tumor location. Other = Brain and other CNS (including cranial nerves).

TABLE 1.

Demographic Factors Associated with Tumor Location

	All		Spinal (A)		Brain and Other CNS (B)		P-Value
	Count	%	Count	%	Count	%	
Age*	54 (median)		56 (median)		53 (median)		0.06
Sex	Female	51.4%	27	42.2%	256	52.6%	0.12
	Male	48.6%	37	57.8%	231	47.4%	
Race	American Indian/ Alaska Native	0.4%	0	0%	2	0.4%	0.50
	Asian or Pacific Islander	11.3%	4	6.3%	56	11.9%	
	Black	8.1%	6	9.5%	37	7.9%	
	Unknown	0.6%	1	1.6%	2	0.4%	
	White	79.7%	52	82.5%	373	79.4%	
Histology	SFT	15.6%	24 ^B	37.5%	62	12.7%	< 0.001
	HPC	84.4%	40	62.5%	425 ^A	87.3%	
% Below Poverty Line*	Below median	50.3%	34	53.1%	243	49.9%	0.25
	Above median	49.7%	30	46.9%	244	50.1%	
Histological Behavior	Benign	22.9%	27 ^B	42.2%	99	20.3%	< 0.001
	Borderline malignancy	35.9%	21	32.8%	177	36.3%	
	Malignant	41.2%	16	25%	211 ^A	43.3%	
Tumor Size	< 5 cm	37.7%	34 ^B	53.1%	174	35.7%	< 0.001
	5 cm or greater	37.6%	7	10.9%	200 ^A	41.1%	
	Unknown	24.7%	23 ^B	35.9%	113	23.2%	
Extent of Resection	STR	37.7%	2	3.1%	126 ^B	25.9%	< 0.001
	GTR	37.6%	23	35.9%	231	47.4%	
	Extent Unknown	24.7%	39 ^A	60.9%	130	26.7%	

Alphabetical superscripts represent statistically significant differences in comparisons between groups.

* Associations with age were determined based on the number of patients above and below the median.

* The median percentage of persons below the poverty line was calculated to be 14.16%.

Bold indicates statistically significant.

TABLE 2.

Univariable and Multivariable Analysis of Overall Survival

Variable	Reference	Univariable				Multivariable			
		P-Value	HR	Lower	Upper	P-Value	HR	Lower	Upper
Age		< 0.001	1.04	1.03	1.05	< 0.001	1.04	1.03	1.05
Sex (Male)	Female	0.97	1.00	0.85	1.17	-	-	-	-
Race	White	0.03				0.03			
American Indian/ Alaska Native		0.26	2.25	0.55	9.19	0.11	3.26	0.78	13.57
Asian or Pacific Islander		0.003	0.28	0.13	0.65	0.01	0.34	0.15	0.78
Black		0.92	1.03	0.58	1.83	0.34	1.33	0.74	2.37
Unknown		0.96	0.00	0.00	2.31E197	0.97	0.00	0.00	1.03E210
% Below Poverty Line		0.001	1.05	1.02	1.08	0.01	1.04	1.01	1.07
Tumor Type (HPC)	SFT	0.25	1.39	0.80	2.41	-	-	-	-
Tumor Location (Other)	Spinal	0.03	0.41	0.18	0.94	0.03	0.36	0.15	0.89
Histological Behavior (Benign/ Borderline/ Malignant)		0.005	1.41	1.11	1.80	0.009	1.40	1.09	1.80
Tumor Size	< 5cm	0.16						-	-
5cm or Greater		0.97	1.01	0.67	1.53	-	-	-	-
Unknown		0.10	1.39	0.94	2.06	-	-	-	-
Treatment Modality	STR	0.16				-	-	-	-
GTR		0.08	0.70	0.47	1.05	-	-	-	-
Unknown Extent		0.09	0.68	0.44	1.06	-	-	-	-

Bold indicates statistically significant.