Journal of Clinical Neuroscience 44 (2017) 169–174

Contents lists available at ScienceDirect

Journal of Clinical Neuroscience

journal homepage: www.elsevier.com/locate/jocn

Clinical commentary

Clinical management and survival of patients with central nervous system hemangiopericytoma in the National Cancer Database

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ARTICLE INFO

Article history:
Received 10 April 2017
Accepted 15 June 2017

Keywords:
Hemangiopericytoma
NCDB
Survival
Radiation
Surgery
Radiosurgery
CNS

ABSTRACT

Purpose/objectives: Hemangiopericytomas are rare central nervous system (CNS) tumors. We sought to investigate existing clinical management strategies and overall survival (OS) among patients with hemangiopericytomas of the CNS.

Methods/materials: All patients diagnosed with CNS hemangiopericytoma from 2004 to 2014 in the National Cancer Database were included. Clinical and treatment-related characteristics were analyzed for an association with OS following diagnosis using univariable and multivariable analyses.

Results: Nine-hundred and eighty-one patients were included (0.22% of all CNS tumors). At diagnosis, 22 patients had spinal tumors (2%), 21 patients had multifocal tumors (2%) 28 had disseminated disease (3%), and the remainder were unifocal intracranial tumors. Patients either underwent surgical resection and radiation (48%), surgery alone (37%), radiation alone (6%), or biopsy alone (9%). Of patients with known extent of resection, 53% underwent gross total resection, and, of patients with known radiation modality, 15% received stereotactic radiosurgery. Among the total cohort, 3 and 10 year OS was 87% and 59%, respectively. On multivariable analysis, factors associated with inferior OS included age (HR = 1.05, $p < 0.001$), WHO grade ($p < 0.001$), multifocal disease (HR = 2.59, $p = 0.04$), disseminated disease (HR = 2.67, $p = 0.01$), and chemotherapy (HR = 2.66, $p = 0.01$). Patients receiving surgery alone or surgery and radiation demonstrated improved OS compared to biopsy alone (HR = 0.45, $p = 0.01$ and HR = 0.47, $p = 0.02$, respectively). However radiation utilization did not impact OS ($p = 0.691$).

Conclusions: The present data provide large-scale prognostic information from a contemporary cohort of patients with hemangiopericytoma and support an initial attempt at surgical extirpation. The benefits of ionizing radiation are likely limited to improved local control and neurologic function.

1. Introduction

Hemangiopericytomas are rare meningeal tumors that are thought to arise from pericytes surrounding capillary walls [1]. They are distinguished from meningioma histopathologically, but also clinically by their propensity for local, regional, and distant recurrence [2]. While they can arise from anywhere in the body [3], they pose unique clinical challenges when located within the central nervous system (CNS).

Owing to their rarity, the management of hemangiopericytomas is not discussed in the National Comprehensive Cancer Network Clinical Practice Guidelines [4], and clinical evidence guiding the treatment of hemangiopericytomas is limited to institutional retrospective series [2,5–7] and Surveillance, Epidemiology and End Results (SEER) analyses [3,8,9]. While SEER database analyses can be informative because of their large sample size, current published SEER series typically contain fewer than 90 patients [8,9]. Likewise, while single-institutional series comprise the majority of available data on the topic, they are limited in their sample size and external validity.

In an effort to provide clinically relevant outcomes data on the topic, our aim was to utilize a large national cancer registry to evaluate the pattern of initial clinical care and survival among patients diagnosed with CNS hemangiopericytoma.

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http://dx.doi.org/10.1016/j.jocn.2017.06.036
0967-5868/

Table 1
Clinical characteristics of the 981 patients with hemangiopericytoma the National Cancer Database 2004–2014.

	Biopsy only		Radiation only		Surgery only		Surgery & radiation	
	n	%	*n*	%	*n*	%	*n*	%
N	86	9%	61	6%	365	37%	469	48%
Patient age*	52		54		53		50	
Race								
White	63	73%	43	71%	268	73%	366	78%
African American	10	12%	6	10%	30	8%	35	8%
American Indian	0	0%	0	0%	3	1%	0	0%
Asian/Pacific Islander	1	1%	4	7%	16	4%	22	5%
Unknown	1	1%	2	3%	14	4%	11	2%
Hispanic	11	13%	6	10%	34	9%	35	8%
Facility Type								
Community Cancer Program	5	8%	2	4%	12	4%	8	2%
Comprehensive Community Cancer program	29	48%	19	38%	86	29%	101	30%
Academic/Research Program	22	36%	23	46%	164	55%	204	60%
Integrated Network Cancer Program	5	8%	6	12%	34	12%	29	9%
Distance to Hospital								
<25 miles	68	79%	36	59%	201	5%	296	63%
25–100 miles	14	16%	18	30%	116	32%	115	25%
>100 miles	4	5%	6	10%	46	13%	52	11%
Unknown	0	0%	1	2%	2	1%	6	1%
Charlson/Deyo Score								
0	63	73%	48	79%	273	75%	378	81%
1	13	15%	11	18%	63	17%	63	13%
2	10	12%	2	3%	29	8%	28	6%
Size								
0.1–19 mm	10	1%	9	15%	21	6%	18	4%
20–39 mm	18	21%	15	25%	56	15%	94	20%
40–59 mm	21	24%	17	28%	116	32%	148	32%
60–79 mm	13	15%	5	8%	58	16%	76	16%
80–99 mm	2	2%	0	0%	10	3%	19	4%
≥100 mm	1	1%	0	0%	5	1%	6	1%
Unknown	21	24%	15	25%	99	27%	108	23%
WHO Grade								
I	2	2%	0	0%	10	3%	7	2%
II	23	27%	15	25%	98	27%	111	24%
III	9	11%	12	20%	60	16%	152	32%
IV	1	1%	1	2%	4	1%	10	2%
Unknown	51	59%	33	54%	193	53%	189	40%
Laterality								
Right	26	30%	19	31%	85	23%	123	26%
Left	22	26%	12	20%	86	24%	123	26%
Midline/Bilateral	38	44%	30	49%	194	53%	223	48%
Focality								
Unifocal	52	61%	43	71%	124	34%	196	42%
Multifocal	7	8%	3	5%	7	2%	4	1%
Unknown	27	31%	15	25%	234	64%	269	57%
Extent of Resection								
Biopsy alone	86	100%	43	71%	0	0%	0	0%
Subtotal	0	0%	0	0%	34	9%	73	16%
Gross total	0	0%	0	0%	95	26%	129	28%
Unknown	0	0%	18	30%	236	65%	267	57%
Radiotherapy Technique								
None	86	100%	0	0%	365	100%	0	0%
Fractionated External Beam	0	0%	44	72%	0	0%	389	83%
Radiosurgery	0	0%	12	20%	0	0%	63	13%
Technique not specified	0	0%	5	8%	0	0%	17	4%

Abbreviations: WHO, World Health Organization.
* Median.

2. Methods

2.1. Data source and cohort selection

The National Cancer Database (NCDB) is a nationwide facility-based comprehensive clinical surveillance resource that is managed jointly by the American College of Surgeons and the American Cancer Society and comprises 70% of new cancer diagnoses in the United States annually [10]. It contains patient-level data informing several clinical and pathologic characteristics of their initial treatment course as well as overall survival (OS) following diagnosis [11].

Patients in the NCDB, treated between 2004 and 2014, with tumors arising from the CNS (448,453 patients) were evaluated and excluded if they were not pathologically diagnosed with hemangiopericytoma (447,472 patients excluded). Clinical characteristics including year of diagnosis, patient age, gender, race, facility type, distance from home to treatment facility, and Charlson/Deyo comorbidity index were evaluated along with various tumor and treatment-related characteristics (Table 1).