Spinal Hemangioblastoma Incidence and Outcomes: A Surveillance Epidemiology and End Results Study

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Introduction

Intradural spinal hemangioblastoma are typically benign tumours of vascular origin arising either in association with von Hippel-Lindau disease or sporadically. Approximately 1000 cases have been described with varying epidemiological factors and outcomes. This study's objective was to evaluate epidemiological factors related to spinal hemangioblastoma with the surveillance, epidemiology, and end results (SEER) database.

Methods

The SEER database was examined for spinal hemangioblastomas between 2000-2010. Incidence was evaluated for age, gender, and race. Outcome was evaluated in the form of survival using Cox proportionate hazards ratios, isolating age, gender, location, treatment modality, pathology, and number of primaries (p=0.05).

Illustrative Case

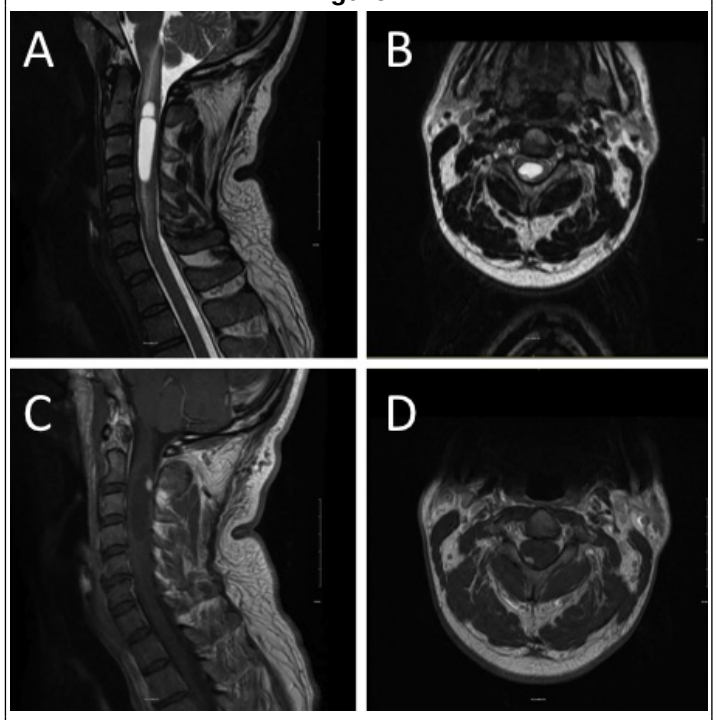
43 year old male with progressive myelopathy and C2 intradural-intramedullary hemangioblastoma with syrinx. Magnetic resonance imaging (figure 1) and intraoperative images (figure 2).

Results

Between 2000-2010, there were 133 cases of spinal hemangioblastoma with an age-adjusted incidence of 0.014 (0.012-0.017) per 100 000, and was the tenth most common intradural spinal tumor type, 2.1% (133/6156) of spinal tumours. There was no difference in incidence between males and females. The average age of patients was 48.0 (45.2 – 50.9), and a lower incidence was noted in patients younger than 15 compared to all other age groups (p<0.05). There was no difference in incidence amongst the different races. Treatment included surgical resection in 106 (79.7%) cases, radiation with surgery in 7 (5.3%) cases, and no treatment was performed in 17 (12.8%) cases. Mortality was noted in 12 (9%) cases, and median survival of 27.5 months (range 1 – 66 months) over the 10-year period. There was no statistically significant different in Cox hazard ratios for mortality for sex, race, treatment modality, pathology, or number of primaries.

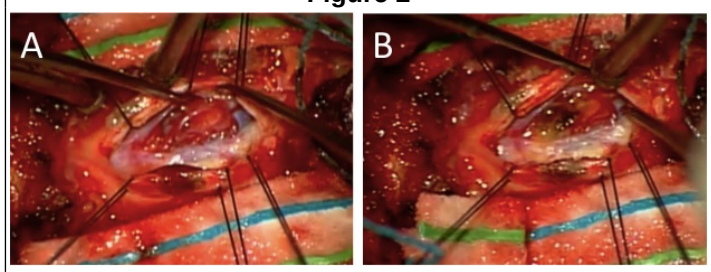
Results are tabulated in Table 1.

Figure 1



Typical MRI findings in illustrative case

Figure 2



Intraoperative findings of typical case of spinal hemangioblastoma A) before and B) after resection

Conclusions

Spinal hemangioblastoma account for a small fraction of primary intradural spinal tumors, with no difference in incidence between genders. Overall prognosis is good, with 9% observed mortality over the 10-year period, with 2% mortality attributable to the malignancy.

Table 1

	Variable of Interest	No Cases (%)	Mortality within subgroup (%)	Cox Hazard Ratio (95% CI)	Cox Hazard Ratio P-value	Log-Rank Kaplan Meier Chi-square (P-value)
Age	0-14	2 (1.5%)				19.4 (0.002)
	15-44	53 (39.8%)				
	45-54	**27 (20.3%)**	**3 (11.1%)**	**0.1 (0.01-0.9)**	**0.04**	
	55-64	28 (21.1%)				
	65-74	**15 (11.3%)**	**2 (13.3%)**	**0.08 (0.008-0.8)**	**.04**	
	75+	8 (6.0%)	4 (50%)	Reference		
Sex	Male	62 (47%)	7 (11.3%)	0.4 (0.07-2.2)	0.3	0.75 (0.4)
	Female	71 (53%)	5 (7%)	Reference		
Race	White	106 (79.7%)	11 (10.4%)	NS	1.0	4.7 (0.2)
	Black	12 (9.0%)				
	American Indian	4 (3.0%)				
	Asian	8 (6.0%)				
	Unknown	3 (2.3%)				
Location	Spinal Meninges	9 (6.8%)				**41.5 (9×10^{-9})**
	Spinal Cord	123 (92.5%)	10 (8.1%)	NS	0.9	
	Cauda Equina	1 (0.8%)				
Treatment	Radiation and Surgery	7 (5.3%)	2 (28.6%)	NS	1.0	3.1 (0.4)
	Surgery Alone	106 (79.7%)	10 (9.4%)	NS	1.0	
	Radiation Alone	1 (0.8%)	0 (0)	NS	1.0	
	No Treatment	17 (12.8%)	0 (0)	Reference		
	Unknown	2 (1.5%)	0 (0)	NA		
Number of Primaries	1	104 (78.2%)	10 (9.6%)	0.906 (0.2-5.0)	0.9	0.09 (0.8)
	2 or more	29 (21.8%)	2 (6.9%)	Reference		
Pathology	9161/0 Acquired tufted hemangioblastoma	4 (3.0%)				2.6 (0.3)
	9161/1 Hemangioblastoma	127 (95.5%)				
	9161/3 Hemangioblastoma, malignant	2 (1.5%)				

Hemangioblastoma survival analysis. Cox hazard ratios and Kaplan-Meier Log-Rank analysis for survival. Data with small group sizes have been blacked-out in accordance with SEER guidelines to prevent identification of individual cases

Learning Objectives

By the conclusion of this session, participants should be able to: 1) Describe incidence of intradural spinal hemangioblastoma. 2) Describe outcomes of intradural spinal hemangioblastoma. 3) Describe treatment modalities of intradural spinal hemangioblastoma