



Tumor Pathology and Location are Associated with Clinical Outcomes in 221 Spinal Nerve Sheath Tumors

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Introduction

Intradural extramedullary spine tumors represent two-thirds of all primary spinal neoplasms. Approximately half of these are peripheral nerve sheath tumors, mainly neurofibromas and schwannomas. Given the rarity of this disease, analyses of clinical outcomes are limited.

Methods

Patients were identified through a search of our institutional neuropathology database and a separate review of current procedural terminology (CPT) codes. Age, gender, clinical presentation, presence of neurofibromatosis (NF), tumor type, location, extent of resection characterized as gross total resection (GTR) or subtotal resection (STR), and clinical follow-up were recorded.

Learning Objectives

By the conclusion of this session, participants should be able to 1) understand the anatomic and pathologic distribution of spinal nerve sheath tumors, 2) symptoms associated with spinal nerve sheath tumors, 3) factors associated with extent of resection

Results

221 tumors in 199 patients were identified with a mean age of 45 years. 53 were neurofibromas, 163 were schwannomas, and 5 were malignant peripheral nerve sheath tumors. Spinal pain was the most common presenting symptom (76%), followed by weakness (36%) and sensory symptoms (34%). Mean symptom duration was 16 months.

Conclusions

Surgical resection is an effective treatment for spinal nerve sheath tumors. Neurofibromas occur more commonly in the cervical spine and are associated with higher rates of recurrence and lower rates of GTR, particularly in patients with NF1 or NF2.

Results

Neurofibromas were more common in the cervical spine (74% vs. 27%, p<0.001), while schwannomas were more common in the thoracic and lumbosacral spine (73% vs. 26%, p<0.001). Rates of GTR were lower among neurofibromas compared to schwannomas (51% vs. 83%, p<0.001), regardless of location. Tumors in the cervical spine had lower rates of GTR (54%) compared to thoracic (90%) and lumbosacral (86%) lesions (p<0.001). Additionally, NF was associated with lower rates of GTR among all tumors (43% vs. 86%, p<0.001). Mean follow-up was 32 months. Recurrence/progression was more common in neurofibromas compared to schwannomas (17% vs. 7%, p=0.03), although the mean time to recurrence/progression did not differ between tumor pathology (45 vs. 53 months, p=0.63). As expected, GTR was associated with lower recurrence rates (4% vs. 22%, p<0.001). In a multivariate model, cervical location (OR=0.239, 95% CI: 0.110-0.520) and presence of NF (OR=0.166, 95% CI: 0.054-0.507) were associated with lower rates of GTR. In a separate model, only GTR (OR=0.141, 95% CI: 0.046-0.429) was associated with tumor recurrence.

Conclusions

Our multivariate analysis found that both cervical location and presence of NF were associated with lower rates of GTR. In a second multivariate model, the only variable associated with tumor recurrence was extent of resection. Maximal safe resection remains ideal for these lesions, however patients with cervical tumors or NF should be counseled about their increased risk of recurrence.

Table 1. Patient Demographics	
Characteristic	n=221
Age (years)	
Mean	45
Median	46
Range	1-87
Gender	
Male	123 (56%)
Female	98 (44%)
Neurofibromatosis	
Type 1	42 (19%)
Type 2	11 (5%)
None	168 (76%)
Symptom duration (months)	
Mean	16
Median	6
Range	0-120
Clinical presentation	
Spinal pain	168 (76%)
Weakness	79 (36%)
Numbness/paresthesias	76 (34%)
Gait disturbance	12 (5%)
Bowel/bladder incontinence	12 (5%)
Incidental	4 (2%)
Tumor type	
Neurofibroma	53 (24%)
Schwannoma	163 (74%)
MPNST	5 (2%)
Tumor level	
Cervical	85 (38%)
Thoracic	62 (28%)
Lumbosacral	74 (34%)
Location	
Intradural	160 (72%)
Extradural	47 (21%)
Paraspinal	14 (6%)
Extent of resection	
GTR	166 (75%)
STR	54 (24%)
Biopsy	1 (0.5%)
Postoperative radiotherapy	14 (6%)
Recurrence	20 (9%)
Time to recurrence (months)	
Mean	49
Median	39
Range	4.5-119
Follow-up duration (months)	
Mean	32
Median	16
Range	0-162

Table 2. Comparison of Tumor Types			
Characteristic	Neurofibroma (n=53)	Schwannoma (n=163)	Significance (p value)
Age (years)			
Mean	36	48	<0.001
Median	35	49	
Range	1-76	14-88	
Gender			
Male	28 (53%)	92 (56%)	0.646
Female	25 (47%)	71 (44%)	
Neurofibromatosis			
Type 1	39 (74%)	2 (1%)	<0.001
Type 2	1 (1%)	10 (6%)	
None	13 (25%)	151 (93%)	<0.001
Symptom duration (months)			
Mean	19	15	0.560
Median	6	6	
Range	0.5-120	0-120	
Clinical presentation			
Spinal pain	31 (58%)	132 (81%)	0.003
Weakness	23 (43%)	55 (34%)	
Numbness/paresthesias	14 (26%)	62 (38%)	0.168
Gait disturbance	2 (4%)	10 (6%)	
Bowel/bladder incontinence	5 (9%)	7 (4%)	0.146
Incidental	0 (0%)	4 (2%)	
Location			
Cervical	39 (74%)	44 (27%)	<0.001
Thoracic	7 (13%)	55 (34%)	
Lumbosacral	7 (13%)	64 (39%)	<0.001
Location			
Intradural	35 (66%)	123 (75%)	0.179
Extradural	13 (25%)	31 (19%)	
Paraspinal	5 (9%)	9 (6%)	0.315
Extent of resection			
GTR	27 (51%)	135 (83%)	<0.001
STR	26 (49%)	27 (17%)	
Biopsy	0 (0%)	1 (0.1%)	0.568
Postoperative radiotherapy	4 (8%)	6 (4%)	0.245
Time to recurrence (months)			
Mean	45	53	0.640
Median	37	53	
Range	10-118	5-119	
Follow-up duration (months)			
Mean	38	31	0.300
Median	17	16	
Range	0-163	0-157	

Table 3. Multivariate analysis of factors associated with GTR			
Variable	Odds Ratio	95% CI	p value
Cervical location	0.231	0.109-0.488	<0.001
NF mutation	0.199	0.073-0.541	0.002
Tumor pathology	0.880	0.310-2.500	0.811
Intradural location	1.259	0.572-2.768	0.567

Table 4. Multivariate analysis of factors associated with tumor recurrence			
Variable	Odds Ratio	95% CI	p value
GTR	0.160	0.051-0.503	0.002
Tumor pathology	0.716	0.149-4.387	0.677
Cervical location	1.646	0.517-5.241	0.399
NF mutation	0.890	0.181-4.387	0.886
Radiotherapy	0.326	0.037-2.910	0.316