Results of Spinal Fusion After Spinal Nerve Sheath Tumor Resection

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■ INTRODUCTION: **Intradural extramedullary spine tumors, approximately one-half of which are peripheral nerve sheath tumors (PNSTs), comprise two-thirds of primary spinal neoplasms. Given the rarity of PNSTs and the restricted indications for adding fusion to laminectomy for tumor resection, analyses of spinal fusion outcomes are limited.**

■ METHODS: **Demographics, clinical presentation, tumor characteristics, extent of resection, spinal fusion, complications, and clinical follow-up were recorded retrospectively.**

■ RESULTS: **A total of 221 tumors in 199 patients were identified (53 neurofibromas, 163 schwannomas, 5 malignant PNSTs); 78 patients underwent fusion (70 instrumented; 8 noninstrumented). Fusion rates were higher for extradural versus intradural lesions (60% vs. 29%; $P = 0.001$) and for tumors involving the cervicothoracic junction (88% vs. 31%, $P < 0.001$). There was no difference in fusion rates based on pathology. Rates of new or worsening sensory (19% in fusion vs. 13% in nonfused) or motor deficits (8% in fused vs. 4% in nonfused), wound infection (3% in fused vs. 6% in nonfused) and cerebrospinal fluid (CSF) leak or pseudomeningocele (6% in fused vs. 4% in nonfused) were not statistically different. There were 10 fusion-related complications: 6 adjacent segment disease, 3 implant failures, and 1 pseudoarthrosis. Mean time from surgery to last follow-up was 32 months.**

■ CONCLUSIONS: **In this cohort, PNSTs in the cervical spine, spanning the cervicothoracic junction, and extradural tumors were associated with higher rates of spinal fusion. Fusion was not associated with new or worsening motor/sensory deficits, CSF leak, pseudomeningocele, wound infection, or spinal deformity. Overall, spinal fusions were well tolerated and did not increase the risk of postoperative complications.**

INTRODUCTION

Primary spinal nerve sheath tumors represent ~15% of spinal tumors in children and 24% in adults.[1-3] They can be classified by their anatomic location as intramedullary, intradural extramedullary, dumbbell (intradural both intraspinal and extraspinal), and extradural. Intramedullary tumors are primarily glial tumors (ependymomas and astrocytomas), whereas extradural lesions consist primarily of metastases, hematopoietic tumors, and primary bony tumors. Intradural extramedullary spinal cord tumors consist primarily of peripheral nerve sheath tumors (PNSTs) and meningiomas.[4] Less common entities include lipomas, spinal nerve sheath myxomas, paragangliomas, sarcomas, and vascular tumors.[5]

Among patients with neurofibromatosis type 1 (NF1), spinal neurofibromas are found in up to 38%.[6-9] However, they are estimated to cause symptoms or become clinically significant in only ~5% of NF1 patients, though this estimate is likely too low.[9-11] There is an alternate form of neurofibromatosis known as familial spinal neurofibromatosis (FSNF) that is categorized by multiple neurofibromas symmetrically affecting the entire axial spine.[12-14] On histologic examination, neurofibromas are composed of Schwann cells with abnormal nuclei and scant cytoplasm and fibroblasts in a matrix of collagen fibers and

Key words
- Spinal fusion
- Peripheral nerve sheath tumor
- Outcomes
- Complications

Abbreviations and Acronyms
CPT: Current procedural terminology
FSNF: Familial spinal neurofibromatosis
GTR: Gross total resection
MPNST: Malignant peripheral nerve sheath tumor
NF1: Neurofibromatosis 1
NF2: Neurofibromatosis 2
PNST: Peripheral nerve sheath tumor
STR: Subtotal resection

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myxoid material.[15] They grow along the course of nerve fibers and are surrounded by a thick epineurium. Schwannomas are made up of neoplastic Schwann cells forming 2 basic patterns: compact elongated cells with occasional nuclear palisading (Antoni A pattern) or less cellular and loosely textured cells with indistinct processes (Antoni B pattern).[15] Approximately 90% of schwannomas are solitary and sporadic, 4% arise in the setting of neurofibromatosis type 2 (NF2), and another 5% are multiple but unassociated with NF2.[16]

Spinal nerve sheath tumors, particularly neurofibromas and schwannomas are rare entities, and published studies are limited to single-institution case series.[17-24] Unfortunately, there are limited data assessing the outcome of spinal fusions or related surgical complications in patients treated surgically for these lesions. Bony fusion rates are dependent on a number of factors, including age,[25,26] smoking,[25,27] number of segments fused,[28] and use of instrumentation.[29] Contemporary rates of wound infection in adult patients undergoing spine surgery range from 1.4%–4.2% and are highest in cases of spinal deformity, revision surgery, spinal fusion, and the use of implants.[30] Cerebrospinal fluid (CSF) leaks are not well studied in intradural tumors, but rates in the literature range from 0.8% to 3%.[31-34] Rates for spinal nerve sheath tumors, particularly intradural lesions, are likely higher. We sought to characterize our institutional experience in the management of these lesions with a particular focus on spinal fusion and fusion-related complications.

METHODS

Data Collection

Patients were identified through a search of our institutional neuropathology database and a separate review of current procedural terminology (CPT) codes by authors N.M.B. and C.P.A. All research activities were approved by the Committee on Human Research of our Institutional Review Board (CHR 10–04026). Pathology records were reviewed to include the following PNSTs: neurofibromas, schwannomas, and malignant peripheral nerve sheath tumors (MPNSTs). Medical records were reviewed to include patient age, sex, presence of NF1 or NF2 according to clinical criteria, presenting symptoms and duration, use of radiotherapy, tumor location, extent of resection (defined by postoperative magnetic resonance imaging [MRI] or operative report), time to last follow-up, and date of recurrence. Tumor location was classified by the level of the involved nerve root (cervical, thoracic, or lumbosacral) and relationship with adjacent dura as follows: intradural extramedullary, extradural intraspinal (tumor within the spinal canal), extradural paraspinal (tumor at the neural foramen or extending outward into adjacent tissues). For the purposes of this manuscript they will be described as intradural, extradural, and paraspinal. Extent of resection was defined as gross total resection (GTR) if there was no evidence of residual disease on postoperative MRI and as subtotal resection (STR) in cases where residual tumor was present. Use of radiotherapy was limited to a small number of patients in this study. All of the patients who received radiation were treated with stereotactic radiosurgery, but in some cases this was done at an outside institution with treatment regimens unavailable for review. Patients with MPNSTs were almost exclusively treated at outside facilities with the use of fractionated radiotherapy. In 1 case, a patient received intensity-modulated radiation therapy in addition to proton-beam therapy.

Statistical Analysis

Univariate analysis of continuous variables was performed using the Student t test and categoric variables compared by means of the chi-square test or Fisher exact test when cell counts were <5. Multivariate analysis was performed with the use of binary logistic regression. Variables were included in the multivariate model if they were found to demonstrate a statistically significant relationship on univariate analysis, defined as $P < 0.05$. To assess recurrence-free survival (the likelihood of a recurrence event over time), we generated Kaplan-Meier plots and used the log-rank test. All analyses were performed with the use of SPSS version 22 (IBM).

RESULTS

Patient Demographics

A total of 221 tumors in 199 patients were identified. Patient demographics are summarized in **Table 1**. The mean age was 45 years with a range of 1–88 years. There were 11 tumors in pediatric patients, defined as those $\leq$18 years of age, with the remaining 210 tumors in adults. Tumors were slightly more common in males with 98 females (44%) and 123 men (56%). Neurofibromatosis was present in 53 patients: NF1 in 42 and NF2 in 11. Mean symptom duration was 16 months. The most common symptom at presentation was pain (76%), followed by weakness (36%), numbness and paresthesia (34%), gait disturbance (5%), and bowel or bladder incontinence (5%). Pathology was classified as neurofibroma in 53 cases, schwannoma in 163 cases, and MPNST in 5 cases. The mean age among patients with neurofibromas was 36 years compared with 48 years in those with schwannomas ($P < 0.001$). NF was strongly associated with neurofibromas: The syndrome was present in 40 of 53 cases compared with 12 of 163 cases of schwannomas ($P < 0.001$). The remaining case of NF was found in a patient with a MPNST. Among the neurofibromas, 39 cases involved NF1 and 1 case NF2; among the schwannomas, 10 were NF2 and 2 NF1.

Tumors were localized to cervical nerve roots in 85 cases (38%), thoracic nerve roots in 62 cases (28%), and lumbosacral nerve roots in 74 cases (24%). Tumors were intradural in 160 cases (72%), extradural but within the spinal canal in 47 cases (21%), and paraspinal in 14 cases (6%). The majority of patients underwent GTR (166 cases, 75%), 54 patients underwent STR (24%), and 1 patient underwent biopsy followed by radiation. Mean duration of follow-up was 32 months.

Fusion Characteristics

Among the 221 tumors, 78 underwent spinal fusions. There was no significant difference in age, sex, or incidence of NF1 or NF2. Spinal fusion patient demographics are presented in **Table 2**. Symptom duration was slightly longer in patients who underwent fusions (20 mo vs. 13 mo), but the difference was not significant. Tumor location was associated with spinal fusion; patients who underwent fusions had a significantly higher proportion of