Surgical Outcomes in Peripheral Nerve Sheath Tumors: Insights from an Institutional Experience with Literature Review

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Abstract

Background Peripheral nerve sheath tumors (PNSTs) encompass schwannomas, neurofibromas, and the more aggressive malignant PNSTs (MPNSTs). This study aimed to evaluate the clinical presentation and surgical morbidity associated with PNST excision.

Methods This retrospective study aims to assess outcomes over the past 10 years at our institution following surgical excision of PNSTs, focusing on the extent of resection, neurological deficits, the need for nerve grafting, and postoperative requirements for chemotherapy and radiotherapy.

Results A palpable lump was the most common symptom, painful in 57% and associated with numbness in 35.7%. Gross total resection was achieved in 78.57% of cases. Histopathology revealed schwannomas in 78.57%, neurofibromas in 14.28%, and one case (7.2%) of MPNST. Two patients required sural nerve grafts. Postoperative motor weakness occurred in two cases, both improved. The mean follow-up duration in the study was 6.5 years, during which no tumor recurrences were observed.

Conclusion PNSTs are relatively common and should be managed at specialized centers by experienced surgeons to ensure safe resection, minimal postoperative deficits, and excellent outcomes with low recurrence rates.

Keywords
- peripheral nerve sheath tumors
- retrospective study
- gross total resection
- nerve grafts

Introduction

Tumors originating from peripheral nerves are broadly classified into nerve sheath and non-nerve sheath tumors. Nerve sheath tumors include schwannomas, neurofibromas, intraneural perineuriomas, and malignant peripheral nerve sheath tumors (MPNSTs), whereas non-nerve sheath tumors encompass intraneural perineuriomas, ganglion cysts, lipomas, desmoid tumors, ganglioneuromas, and hemangiomas.[1] Schwann cells are believed to be the cells of origin for peripheral nerve sheath tumors (PNSTs).[2] Most PNSTs occur sporadically but may also be associated with genetic syndromes such as neurofibromatosis type 1 (NF-1), NF-2, and schwannomatosis.[3] MPNSTs typically arise through malignant transformation of neurofibromas, most commonly in patients with NF-1.

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Schwannomas are the most frequently encountered peripheral nerve tumors, typically presenting as painless, slow-growing, well-circumscribed, firm, and round soft tissue masses.[4] These tumors grow slowly, leading to stretching and elongation of adjacent nerve fascicles while usually preserving neurological function.[5] Schwannomas are categorized into four main subtypes: conventional schwannoma, cellular schwannoma, plexiform schwannoma, melanocytic schwannoma (often misdiagnosed as melanoma), and MPNST.[6] Histologically, schwannomas exhibit a biphasic architecture composed of Antoni A and Antoni B areas, with features such as nuclear palisading (Verocay bodies), a fibrous capsule, displaced parent nerve fascicles, and various degenerative changes.[7] Radiologically, they are isointense to skeletal muscle on T1-weighted magnetic resonance imaging (MRI), heterogeneously hyperintense on T2-weighted images, and may demonstrate variable contrast enhancement. Typical imaging signs include the target sign, fascicular sign, and split fat sign.[8] High-resolution ultrasound is a widely accepted, noninvasive, and repeatable first-line modality for diagnosing and monitoring neurofibromas and other malignant lesions.[9]

Neurofibromas are traditionally classified as solitary or plexiform, both affecting peripheral nerves. Solitary neurofibromas usually present as fusiform enlargement of the nerve trunk and are often amenable to surgical removal.[10] In contrast, plexiform neurofibromas involve multiple adjacent nerve fascicles or components of a nerve plexus. These are commonly associated with NF-1 and are typically difficult to excise completely.[10] Due to their intraneural growth pattern, neurofibromas more frequently cause pain or neurological deficits than schwannomas.

Patients who have undergone a prior biopsy or unsuccessful tumor excision attempt often experience significant pain and neurological impairment in both schwannoma and neurofibroma cases.[11,12]

Solitary benign PNSTs are frequently incidental findings in asymptomatic individuals and are commonly managed with serial imaging and clinical observation. Surgical excision of benign PNSTs is generally reserved for cases exhibiting progressive neurological decline, intractable pain, increasing tumor size, or the need for a definitive tissue diagnosis.

MPNSTs may arise in patients with neurofibromas, occurring both sporadically and in those with NF-1. However, individuals with NF-1 face a significantly elevated risk of MPNST development, necessitating early imaging and regular clinical assessments for early detection.[13] Management of MPNST includes wide local excision with negative margins, followed by radiotherapy and chemotherapy. Nerve grafting is generally avoided due to the detrimental effects of adjuvant radiotherapy on nerve regeneration. Nonetheless, functional outcomes can be improved through tendon transfer procedures, enhancing quality of life in affected patients.[14]

Methods

The study was designed with the aim of assessing outcomes including extent of resection, neurological deficit, need for nerve grafting, and postoperative need for chemotherapy and radiotherapy following surgical excision of PNSTs. It is a retrospective study performed at the All India Institute of Medical Sciences, New Delhi, India. All those patients who had undergone surgery for PNSTs in the past 10 years at the institute were included in the study, given their medical records and follow-up scans and clinical status were available. Data was collected using a self-designed questionnaire and patients were followed at least 1 year to watch for recurrence. A detailed history was obtained followed by thorough physical examination to look for neurological deficits, Tinel sign, stigmata of neurocutaneous syndromes, nerve conduction studies, and it was finally augmented with contrast-enhanced MRI (CE-MRI) of the region involved.

Statistical Analysis

The data was entered in MS-Excel and STATA 15.0 software was used for statistical analysis. The normality of the data was tested by the Shapiro–Wilk test. Descriptive statistics was performed, and categorical variables were presented in frequency and percentage and continuous variables were presented as mean ± standard deviation.

Surgical Management

Intraoperatively, a linear skin incision is made along the course of the affected nerve and overlying tumor, followed by meticulous dissection of the subcutaneous tissues (►**Fig. 1**). Surgical exposure begins with identification of the normal nerve anatomy both proximally (cranially) and distally (caudally), which is then extended toward the region of the tumor. A nerve stimulator is routinely employed to assist in locating an "a-fascicular" entry point for intraneural dissection. In cases of large tumors, internal debulking is often performed first to facilitate the development of an a-fascicular dissection plane, allowing safer and more complete tumor removal. Nerve grafting may be considered if multiple nerve fibers are sacrificed or damaged during excision. However, splayed and thinned fascicles—if anatomically preserved—generally retain normal function. The tumor capsule is typically left in situ, as its removal has not been shown to reduce the risk of recurrence. In patients with plexiform neurofibromas, the primary surgical objective is adequate debulking to alleviate symptoms caused by mass effect.

Results

A total of 14 patients were included in the study, with a mean age of 42.6 years (range: 22–74 years). Of these, 64.28% were male. One of these patients was a known case of NF-1 and had previously undergone surgery for an intradural schwannoma at the L2–L3 level 6 years ago.

The most common presenting symptom in all the cases was a palpable lump, which was painful in 57% of patients and associated with numbness in 35.71%. Preoperatively, motor deficit was observed in only one patient, who had a large tumor. The average duration of symptoms was 2.28 years. ►**Table 1** shows the demographic profile of patients.

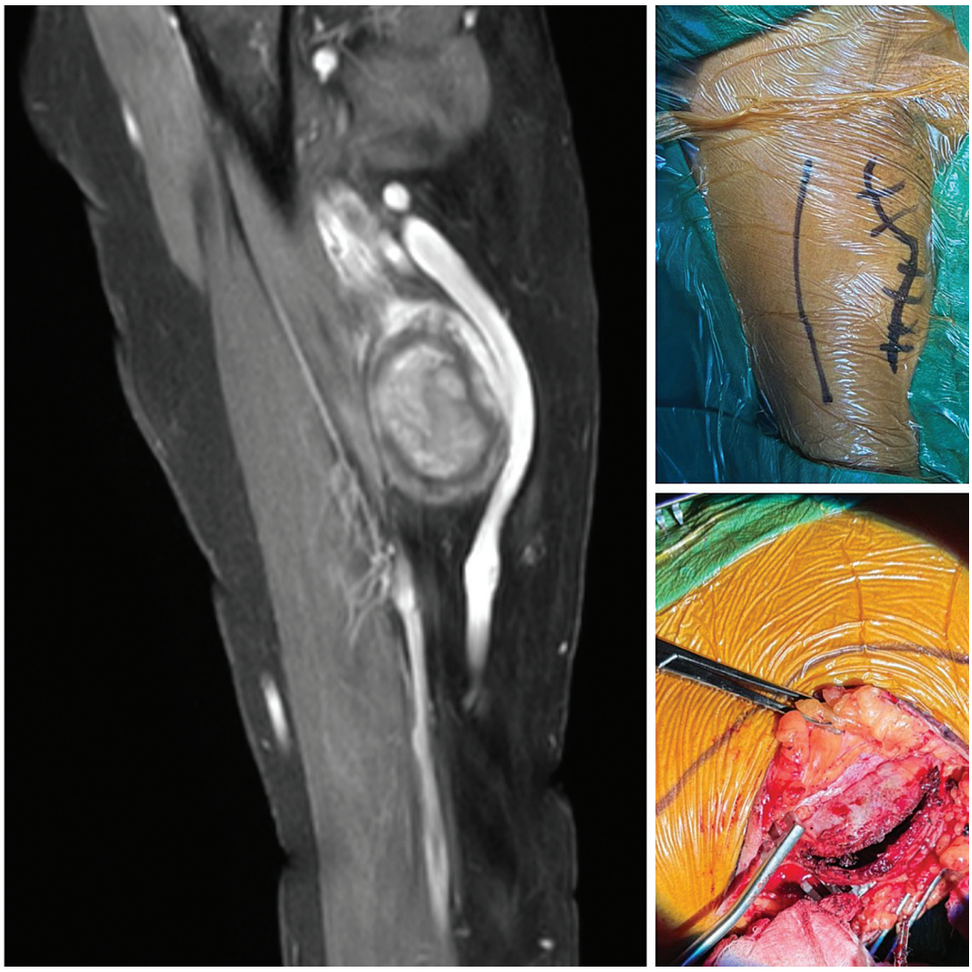

Fig. 1 Contrast-enhanced magnetic resonance imaging (CE-MRI) demonstrates a right median nerve schwannoma (left image). The top-right image shows intraoperative marking for tumor excision, while the bottom-right image depicts the exposed tumor capsule following internal debulking.

Intraoperatively, nerve monitoring was routinely employed in all cases. Gross total resection (GTR) was achieved in 78.57% of patients, while subtotal resection was performed in 21.42% to minimize the risk of postoperative sensorimotor deficits. In two patients, end-to-end sural nerve grafts were required due to intraoperative resection of a significant number of involved fascicles. ►**Table 2** shows postoperative details and ►**Table 3** shows the distribution of nerves involved in the study.

The average duration of surgery was 132 minutes, with a mean estimated blood loss of 92.5 mL. Postoperatively, two patients experienced a reduction in motor power.

Histopathological analysis revealed schwannomas in 78.57% of cases, neurofibromas in 14.28%, and a MPNST in one case (7.2%). ►**Fig. 2** shows the histological distribution of tumors in the study. We routinely perform a CE-MRI 3 months postoperatively to assess for any residual tumor or recurrence.

At the initial 1-year follow-up, both patients with postoperative motor weakness showed meaningful improvement in muscle power. The average duration of follow-up was 6.5 years. No tumor recurrence was observed in any patient. The patient diagnosed with MPNST received adjuvant radiotherapy and remains under regular follow-up.

Discussion

Schwannomas are benign, slow-growing lesions that are typically painless. Their shape often varies depending on the size of the involved nerve: lesions associated with smaller nerves tend to be spindle-shaped, whereas those involving larger nerves usually present as spherical masses.[15] On the other hand, neurofibromas are usually painful due to intraneural pattern of growth and tend to be spindle shaped. Anatomically, schwannomas arise from the periphery of the nerve bundles, making them more amenable to surgical removal, whereas neurofibromas are interwoven with multiple nerve fascicles, which makes complete excision more challenging.[16]

In this study, the number of male patients was twice that of female patients, which contrasts with the findings reported by Levi et al and Desai.[17,18] The most common presenting complaint in our study was a painful lump, reported in 57% of cases. This finding aligns with the characteristic sign of nerve sheath tumors—severe, radiating, electric shock-like neurogenic pain elicited by tapping over the swelling. Preoperative biopsy is not routinely performed in cases of PNSTs due to the risk of fascicular damage and hemorrhage, as advised in the

Table 1 Demographic profile of participants in the study

Demographic details	
Age	
Mean	42.6 y
Range	22–74 y
Gender	
Male	9 (64.28%)
Female	5 (35.71%)
Presentation	
Palpable lump	14 (100%)
Pain at site of lesion	8 (57.1%)
Sensory/motor weakness	1 (7.1%)
Duration of symptoms	
Mean	2.28 y
Range	0.5–10 y
Associated syndromes	
NF-1	1
NF-2	0
Schwannomatosis	0
Preoperative biopsy	
Obtained	No

Abbreviation: NF, neurofibromatosis.

Table 2 Postoperative details of participants in the study

Postop details	
Resection	
GTR	11 (78.57%)
NTR	3 (21.43%)
Follow-up (mo)	
Mean	6.5 y
Range	0.5–10 y
Postop motor function	
Improved	1 (7.1%)
Same as preop	11 (78.57%)
Deterioration	2 (14.28%)
Postop pain status	
Improved	8 (100%)
Increased	0
Same as preop	0
Tumor recurrence	
Yes	0
Postop chemo/targeted therapy	
Yes	1 (7.1%)
Postop radiation	
Yes	1 (7.1%)

Abbreviations: GTR, gross total resection; NTR, near total resection.

Table 3 Distribution of nerve fibers involved with tumor in the study

Nerves involved	No. of cases
Axillary	1
Radial	0
Median	1
Ulnar	4
Sciatic	2
Common peroneal	1
Tibial	5
Femoral	0
Grand total	14

literature. However, in cases where a MPNST is suspected, a biopsy is warranted.[19] Levine et al concluded that the presence of a target sign on MRI is highly indicative of a benign PNST (BPNST), whereas gallium uptake on ^67Ga MRI scintigraphy suggests malignancy. In a separate study, Reynolds et al reported that infiltrative margins observed on ultrasonography are also indicative of malignant transformation.[20,21] Use of intraoperative neurophysiological monitoring significantly reduced the incidence of postop deficits in the study. In studies conducted by Wilson et al and Desai, GTR was achieved in 85 and 81.2% of cases, respectively, while our study demonstrated a GTR rate of 78.57%.[18,22] In certain cases, GTR was intentionally avoided in our cohort to prevent the risk of postoperative sensory and motor deficits. The incidence of postoperative numbness and weakness in patients with neurofibromas was comparable to that observed in patients with schwannomas, suggesting that these complications are more closely related to intraoperative nerve dissection and manipulation than to the tumor type itself. Although schwannomas and neurofibromas are histologically benign, they have the potential for local recurrence. Reported recurrence rates for BPNSTs in the literature range from 1.3 to 35.9%.[23] In this study, the incidence of schwannomas was much higher than neurofibromas, which is contrary to a higher incidence of neurofibromas in previously reported series.[24,25] A similar study conducted at our institute by Bharadwaj et al reported a significantly higher incidence of brachial plexus schwannomas—a subset of PNSTs—compared with neurofibromas.[26] Levi et al observed that surgery had the greatest impact on functional outcomes in cases of MPNSTs and recommended the use of chemotherapy, aligning with findings from the Italian Sarcoma Group.[17,27] Chemotherapy was found to aid in forming a more defined pseudocapsule, thereby facilitating surgical excision.[17] Therefore, we recommend referral to a tertiary care center with CE-MRI of the lesion without prior biopsy whenever a PNST is suspected for better results.

Conclusion

PNSTs are relatively common and should be managed at specialized centers by experienced surgeons to ensure safe

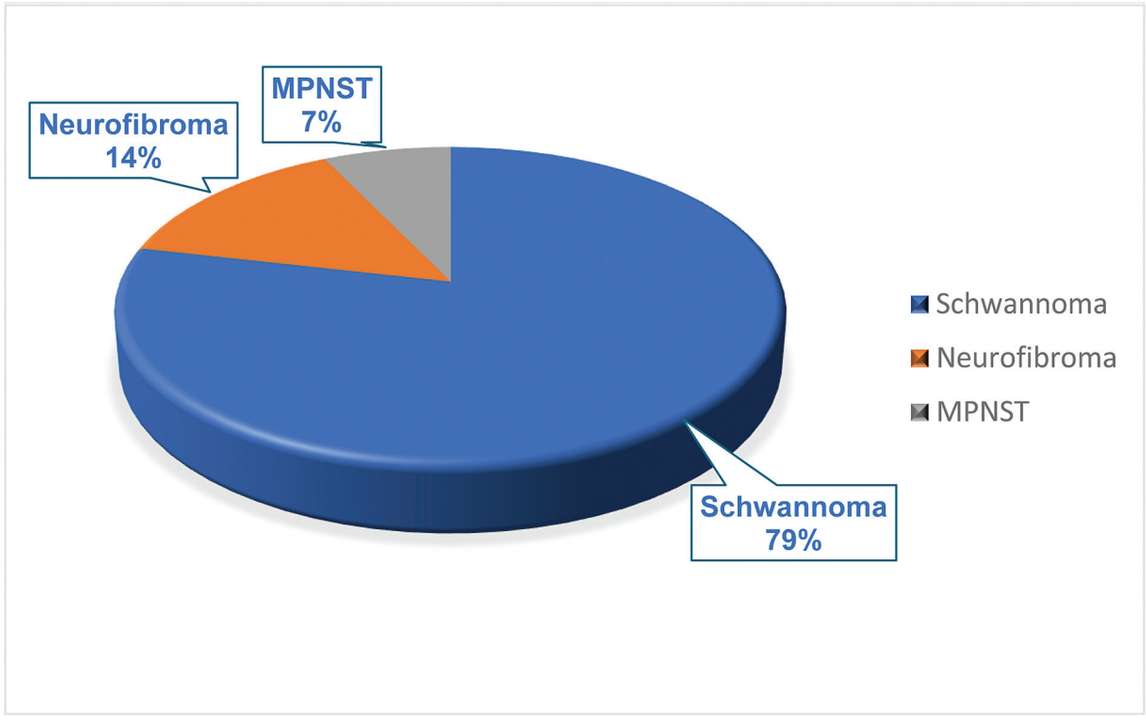

Fig. 2 Pie chart shows histological diagnosis of tumors resected in the study.

resection, minimal postoperative deficits, and excellent outcomes with low recurrence rates.

Declaration of Patient Consent

The authors confirm that they have obtained all necessary patient consent forms. In these forms, the patients have granted permission for their images and other clinical information to be published in the journal. They understand that their name and initials would not be disclosed and that efforts will be made to protect their identity, although complete anonymity cannot be guaranteed.

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Conflict of Interest

None declared.

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