Clinical Outcomes of Surgical Management of Primary Brachial Plexus Tumors

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Introduction

Brachial plexus tumors (BPTs) are rare and constitute 5% of all the upper-limb tumors. These include benign or malignant tumors arising from neural or surrounding nonneural tissues. Preoperative evaluation and planning depend on origin, characteristics, extension, and pathology of tumor. The tumor often causes displacement in the already complex neurovascular anatomy. Procedures such as tumor resection or even biopsy are prone to risk of losing limb functions thus making it a clinical and surgical challenge. Successful management of these lesions requires a thorough understanding of anatomy and familiarity with surgical variations that may be encountered.

The surgical management of BPTs has undergone a sea change from en-masse excision of tumor and nerve to enucleation of encapsulated tumors. The first description of tumor excision in 1886 resulted in significant morbidity. Refinements in surgical techniques aim at preservation of limb functions. The improved functional outcomes have been achieved with the help of better preoperative imaging, higher magnification, and intraoperative neural monitoring techniques. There are few recent reports demonstrating the surgical approaches and variations expected. However, most of these are either isolated case reports or series owing to the heterogeneity in clinicopathological presentations and is often limited by a retrospective design. This inevitably leads...

Keywords
- schwannoma
- neurofibroma
- microneurolysis
- paresthesia
- dumbbell tumor
to frequent misdiagnosis and delay in management. With a thorough understanding of varied presentations and surgical challenges involved, the potential for preservation of limb functions can be achieved. Hence, this study was performed to describe the clinical presentation, tumor characteristics, and clinical outcomes of surgically treated primary BPTs.

**Methods**

This was a prospective descriptive analysis of patients with BPTs performed from June 2015 to August 2020. Institute Research Committee approval and informed consent were obtained from all patients. Fourteen patients who underwent surgical management for BPTs were included.

BPT was diagnosed based on clinical presentation, imaging, and tissue diagnosis. Patients presenting with swelling over neck, shoulder or axilla, with or without pain or tingling in upper-limb were suspected to have BPT. Detailed history regarding the progression of symptoms, size of swelling, relevant family history, presence of neurofibromatosis (NF1) and history of attempted excision were noted. Clinical examination of location, size, mobility, and tenderness of swelling and detailed neurological assessment was performed. All patients underwent magnetic resonance imaging (MRI) to define the tumor's location and the extent and its relation to the neural elements. In cases of dumbbell tumors involving roots and intravertebral foramina, computerized-tomography imaging was obtained. Tissue-diagnosis was performed with fine needle aspiration cytology (FNAC) for primary cases and histopathological review of previous biopsy slides if available.

**Surgical Technique**

All patients underwent surgical exploration under magnification. An anterior supraclavicular and infraclavicular approach was used for upper lesions (roots/trunks) and lower lesions (divisions/cords/terminal branches), respectively. The axillary incision extended onto medial arm was used to access the distal end of axillary lesions. The authors were able to gain wide access with this combined approach and the need for clavicle resection was avoided in all the cases. The tumor was identified in relation to plexus and mobilized from adjacent structures. The nerves proximal and distal to the lesion were identified and protected using silicon vascular slings.

The thickened epineurium over the lesion was opened, entering extracapsular plane carefully until mass was separated from adjacent nerve fascicles. Fascicles entering the substance of the tumor were isolated at proximal and distal poles of mass. Intraoperative electrical stimulation (0.5–2mA; 2 Hz) was used to differentiate functioning and nonfunctioning fascicles within the tumor. Microneurolysis was performed under 10X magnification to preserve the functioning nerve fascicles, which were displaced, splayed out, or thinned due to the mass effect of expanded tumor. Smaller tumors could be shelled out as a single mass easily with careful microneural dissection. However, very large tumors often needed debulking over nonneural areas to help in the ease of dissection. Once adequate debulking was achieved, the plane of cleavage between tumor and functioning nerve fascicles was identified and tumor was excised completely. Malignant tumors were excised with 1 cm margin of surrounding tissue to ensure complete oncological clearance. The nerves were included in the excision if required to achieve free margins and secondary functional reconstruction procedures such as distal nerve/tendon transfers were planned.

**Postoperative Protocol**

Patients were immobilized with crepe bandage dressings for up to 3 weeks postoperatively. The patient was started on passive range of motion exercises for 1 week and followed up by active range of motion exercises. In case of motor deficit postoperatively, passive physiotherapy and galvanic current electrical stimulation were applied till M2 power was recovered and was then followed by active range of motion exercises.

**Statistical Analysis**

All the continuous variables were expressed as means and categorical variables were expressed as proportions. Descriptive analysis was performed using SPSS software V.21.

**Results**

**Clinical and Demographic Characteristics**

Fourteen patients underwent surgical management of 15 BPTs. Four male and ten female patients were included with a mean age of 37.8 ± 12.3 years (range: 16–55 years). Majority of patients (66.6%) were index cases except 4 patients who had history of attempted excision at outside centers. None of the patients had history of neurofibromatosis. The symptom duration ranged from 3 months to 1 year. The most common clinical presentations were swelling (100%), pain (86.6%), and paresthesia (76.9%). Sensory and motor deficits were present in 5 and 4 patients, respectively. Tinel’s sign could be elicited at location of tumor in 11/15 cases. The summary of the various clinicopathological features of the patients is shown in **Table 1**.

**Tumor Characteristics**

The tumors were varied in their location, extent, and pathological characteristics. MRI demonstrated ovoid mass along the nerves with homogeneous enhancement in T1 and hyperintense with postcontrast enhancement on T2-weighted images (**Fig. 1**). Malignant lesions were characterized by large size, irregular peripheral enhancement, and intratumoral cystic appearance (**Fig. 1**). The lesions involved various parts of brachial plexus such as roots (5/15), trunk (5/15), division (1/15), and cords (4/15) (**Table 1**). One patient had bilateral tumor (Schwannoma) involving all the roots on right and divisions on left side (**Fig. 2**). One patient had dumbbell-shaped tumor having intraspinal and extraspinal extension onto C5 root (**Fig. 3**). Thirteen patients had benign pathology: 8 schwannomas, 3 neurofibromas, and 2 lipomas. Two patients had malignant neurofibrosarcoma.

**Intraoperative Features**

All tumors were excised as a single mass with preservation of fascicles. Gross total resection (GTR) was achieved...
in 14/15 cases. Schwannomas appeared as firm well-defined masses without much deformation of nerve and could be easily dissected out preserving neural tissues (►Fig. 2). Neurofibroma resulted in firm masses with splaying and enlargement of neural tissue making the dissection tedious (►Fig. 4). Electrical stimulation to identify and preserve functional fascicles was performed meticulously. In the patient with dumbbell tumor, subtotal resection (STR) was performed leaving behind intraspinal part of tumor (►Fig. 3). In one case with a large tumor with cystic degeneration due to central tumor necrosis, debulking of the benign tumor by aspirating the contents was performed that facilitated retrieval of the deflated tumor, thus preserving function. In malignant peripheral nerve sheath tumor (MPNST) patients, GTR with 1 cm margin was achieved in one case with preservation of all neural fascicles, whereas another patient required partial excision of median nerve fascicles and adventitia of axillary vessels (►Fig. 4). A line diagram representing the differences in dissection and excision between the various types of tumors is shown in ►Fig. 5.

### Postoperative Recovery Characteristics

The mean follow-up period was 24 ± 5 months. Postoperatively, all patients reported improvement in pain and paresthesia. None of the patients developed new sensory/motor deficit. All patients had developed initial motor weakness (Grades 2–4) in the muscles supplied by the involved/adjacent nerves; however, full power (Grade 5) was recovered in all those muscles by 3 to 5 months. For example, patient with

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**Table 1** Summary of the various clinicopathological features of the patients in the series

<table>
<thead>
<tr>
<th>No.</th>
<th>Age/ gender</th>
<th>Clinical symptoms</th>
<th>Tinel’s sign</th>
<th>Preoperative neural deficit</th>
<th>Location</th>
<th>Excision</th>
<th>Postoperative histopathology</th>
<th>Postoperative neural deficit</th>
</tr>
</thead>
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<tr>
<td>1</td>
<td>45/F</td>
<td>Swelling, pain, paresthesia</td>
<td>+</td>
<td>Sensory, motor</td>
<td>C5 root</td>
<td>GTR</td>
<td>Schwannoma</td>
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<td>+</td>
<td>C5, 6 root</td>
<td>GTR</td>
<td>Schwannoma</td>
<td></td>
<td></td>
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<tr>
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<td>C5 root</td>
<td>GTR</td>
<td>Schwannoma</td>
<td></td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>16/F</td>
<td>Swelling, paresthesia</td>
<td>–</td>
<td>Upper trunk</td>
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<td>Schwannoma</td>
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<tr>
<td>5</td>
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<td>+</td>
<td>Motor</td>
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<td>Neurofibroma</td>
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<tr>
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<td>Lipoma</td>
<td></td>
<td></td>
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<tr>
<td>7</td>
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<td>Lower trunk</td>
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<td>Neurofibroma</td>
<td></td>
<td></td>
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<tr>
<td>9</td>
<td>30/F&lt;sup&gt;a&lt;/sup&gt;</td>
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<td>+</td>
<td>Sensory</td>
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<td>Sensory</td>
<td>Medial cord</td>
<td>GTR</td>
<td>Lipoma</td>
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<td>Schwannoma</td>
<td></td>
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<td></td>
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<tr>
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<td></td>
<td>Motor</td>
<td>Medial cord</td>
<td>GTR</td>
<td>Neurofibrosarcoma</td>
<td>Index finger and thumb flexion weakness (Grade 3)</td>
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</table>

**Abbreviations**: GTR, gross total resection, STR, subtotal resection.
<sup>a</sup>Patient with bilateral tumor.
<sup>b</sup>The extraspinal tumor was resected completely; the intraspinal component was left behind.

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**Fig. 1** Characteristic magnetic resonance imaging (MRI) findings of brachial plexus tumors: (A) Benign brachial plexus tumor—ovoid lesion along the axis of the nerve, hyperintense with postcontrast enhancement showing the target sign in T2-weighted images (black arrow). (B) Malignant tumor—MRI shows characteristic irregular peripheral enhancement and intratumoral cystic appearance (black arrow).
tumor arising from C5 root had postoperative weakness of shoulder abduction; however, it improved to full power by 3 to 5 months. The patient with bilateral tumor had residual Horner’s syndrome postoperatively on one side. Patient with dumbbell tumor who underwent STR reported improvement in sensory and motor deficits as well as myelopathy symptoms. One patient with malignancy succumbed to disease due to systemic metastasis at 9 months follow-up. Second patient had persistent motor deficit in the form of weakness (Grade 3) of index finger and thumb flexion. None of the patients developed recurrence.

Discussion

The primary BPTs constitute an unfamiliar and surgically challenging territory. With a better understanding of anatomy and surgical variations, the goal of curative resection with preservation of limb function is feasible. The first report of excision of brachial plexus schwannoma resulted in significant deltoid paralysis. Further studies reported significant postoperative neurological deficit or STRs. There has been constant modifications in the surgical technique with the increasing use of intraoperative neural monitoring and magnification aimed at improving outcomes and minimizing morbidity. There are few large series demonstrating the variations and surgical challenges. Huang et al reported a series of 20 cases with good outcomes. Recent reports from China and India also demonstrated the importance of appropriate surgical management to protect limb functions. However, majority of these reports were limited by sample size and retrospective design.
Current understanding of the approach to BPTs remains incomplete. This is reflected in a large number of misdiagnosed cases despite characteristic clinical signs. Jia et al. reported >50% patients who were wrongly diagnosed and treated as cervical spondylosis thus leading to delay in presentation to a specialist. Desai reported 12% cases misdiagnosed as either nonneural tumor or lymph nodes and underwent incomplete excision and abandonment of procedure. Similarly, in the present study 28% cases were diagnosed following previous attempted excision and biopsy for mistaken neck swellings. Although a rare diagnosis, this underscores the potential need to create awareness and have a high index of suspicion.

The mean age at diagnosis was 45 to 48 years in the previous reports; however, a relatively lower age was reported in the present study as well as in previous reports from India. Among benign BPTs, neurofibroma was the most common followed by schwannoma in reports from Western countries. The recent series of 143 cases from China reported 83% schwannoma and 8% neurofibromas. Desai also reported 60% schwannomas and 40% neurofibromas. Similar results were noted in the present study with 50% cases being schwannoma. This geographical variation in epidemiology and tumor type is surprising and may be attributed to genetic or environmental factors.

A palpable mass was the most common clinical presentation (70–90%) followed by pain (50–60%) in majority of the reports and in the present study as well. The motor and sensory loss was a relatively rare presentation. Among patients with neurological deficits, diagnosis was predominantly neurofibroma than schwannoma. However, Huang et al. reported pain and sensory loss as the major symptom (60%) as opposed to palpable mass (30%). Tinel’s sign can differentiate these tumors from other neck swellings. In the present study, a positive Tinel’s sign was elicited in 73% cases. Similar reports were shown by Desai and Jia et al. MRI has now been widely accepted as imaging of choice. Although it is difficult to differentiate pathological types of benign tumors, differentiation of benign and malignant lesions is possible on MRI. In the present study, the authors have utilized a combination of MRI and FNAC. The core-needle biopsy was avoided as the risk of worsening of neurological symptoms with the needle trauma is larger. Although there is no definite role preoperatively, the importance of intraoperative electrodagnostic studies to help in identification of functioning fascicles has been stressed in literature.

Conservative management with serial MRI monitoring can be used for small tumors without any neurologic or cosmetic issues. However, surgical resection is potentially curative. Relative contraindications to surgery include pregnancy and a poor surgical candidate. Palpable tumors, intractable pain, neurological deficits, and suspicion of malignancy warrant early surgical management. Schwannomas arise from...
Schwann cells and is eccentrically located within the nerve. They are soft to firm in consistency and may contain cystic or hemorrhagic areas. Schwannomas characteristically spare neural fascicles except few which enter and exit the tumors. They have a definite capsule formed of perineurium and fibrous epineurium and hence, intrafascicular dissection and isolation of tumor are relatively easy.8,10,12 Neurofibromas are firm, arise from perineural cells, and lack a definite capsule.8,11,13 They involve larger number of neural fascicles intimately and have a higher risk of loss of neural functions. The older literature distinguished these two entities based on the need for nerve-resection and grafting in neurofibroma.8,6 However, with improvised techniques and intraoperative electrical stimulation, it is possible to excise both with careful preservation of functional neurons.

Huang et al reported improvement in pain without any sensory or neural deficit in 82% neurofibroma and 89% schwannomas.15 Ganju et al reported preservation of functions with total resection in 76% neurofibromas; however, in most of NF1 associated neurofibromas, nerve function could be preserved only at the cost of STR.8 Desai demonstrated total excision was possible in 65% tumors, with STR in 35% cases that were neurofibromas.11 In the present study, GTR could be achieved in 14/15 cases. Although combined supine and prone approach for complete excision of dumbbell tumors has been described, in the present series, STR was performed for dumbbell tumor. The patients did not require further excision of intraspinal part as the patient improved symptomatically. Previous reports demonstrated staged excision of dumbbell tumors and reported that addressing the extraspinal component was important to prevent recurrence.11 Jia et al reported complete recovery of neural functions in 93% patients with total or STR; however, 6% reported persistent neurological deficits.13 Desai demonstrated 95% improvement in pain and paresthesia within weeks; however, 5/115 patients had persistent sensorimotor deficits.11 Similar results were reported with resolution of pain and paresthesia in early postoperative period in all patients in this study. However, all the patients had initial motor weakness owing to neurapraxia due to the intrafascicular dissection and handling of the nerve. Siqueira and Go et al also reported similar transient paralysis following total resection of benign tumors.10,12 In one patient with MPNST who required partial excision of median nerve fascicles, primary nerve reconstruction was not performed as adjuvant radiotherapy was planned. Although nerve resection and reconstruction is feasible, it was deferred keeping in mind the fact that reoperating on a postradiotherapy bed is technically difficult and is likely to have suboptimal outcomes. The study is not without limitations. The follow-up was performed by clinical examination for signs of recurrence and a postoperative MRI could not be performed. However, no recurrence of benign tumors has been reported in up to 5 years follow-up.11,13

**Conclusion**

Total resection of BPTs can be achieved by appropriate microsurgical dissection and electrophysiologic monitoring and is potentially curative with excellent results.

**Conflict of Interests**

None.

**References**