Treatment of Peripheral Nerve Tumors

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Introduction

All cellular elements that comprise a peripheral nerve (neural cells, Schwann cells, and fibroblasts) can theoretically give rise to peripheral nerve tumors (PNTs). The tumors are classified as either benign or malignant, and sub-classified according to their origin from either neural or non-neural elements (Table 1).

History and Physical Examination

When a soft tissue mass is associated with sensory and/or motor symptoms supplied by a known peripheral nerve, the suspicion of a peripheral nerve tumor (PNT) should be directed toward the onset, duration, and growth alterations of the mass. A family history of NF-1 or NF-2 or previously identified neurofibromatosis is of special importance, since majority of PNTs are linked with these syndromes. Presence or absence of symp- toms such as signs like pain, numbness, weakness, the overlying skin temperature and color, fluctuate, along with the patients general health inquiry including immune status, pre-existing malignancy are of importance in the differential diagnosis. However, many peripheral nerve tumors present without any neurological symptoms due to their slow growth rate or origin from a superficial small nerve branch. Several features of the examination that suggests a peripheral nerve origin (July and Guha, 2008).

Diagnosis

Nerve conduction and EMG evaluation are not generally performed in the management of PNTs as they are not diagnostic nor do they help in the management decision. However, intra-operative electrophysiology is crucial as discussed below. Plain X-ray and CT scans are occasionally helpful, especially to demonstrate remodeling of adjacent bony structures such as the neural foramina. Angiography or MR angiography is rarely required, and restricted to large PNTs at the base of the neck, chest or retroperitoneum, where close proximity and or rarely vascular invasion may be present. MRI is the most useful and sensitive tech- nique, often but not always revealing the nerve of origin (Fig. 1). It is especially useful in determining the relationship of the mass to adjacent anatomical structures, which are of relevance.

Although CT scan or MRI cannot distinguish between the various subtypes of PNTs and determine whether a lesion is benign or malignant (Levine et al., 1987; Shub et al., 1992), MRI imaging may be highly suggestive but not diagnostic of the sub-type of PNT, with elec- tromyography (EMG) diagnostic (Kawamoto and Shimizu, 2008), such that when the differential diagnosis is made, histopathological confirmation and management are crucial.

Operative Technique

There are several operative principles that are applicable for all peripheral nerve tumors (July and Guha, 2008). 1) Anesthetic without neuromuscular paralysis is required for all peripheral nerve tumors (July and Guha, 2008). 2) The operation should primarily be done through a minimal incision and the tumor should be easily accessible. 3) The incision over the tumor should extend proximally and distally to allow adequate exposure of the nerve of origin. 4) The incision over the tumor should extend proximally and distally to allow adequate exposure of the nerve of origin. 5) The incision over the tumor should extend proximally and distally to allow adequate exposure of the nerve of origin. 6) The incision over the tumor should extend proximally and distally to allow adequate exposure of the nerve of origin. 7) The incision over the tumor should exten...
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Neurofibromatosis (NF) most likely represents an autosomal dominant disease (incidence 1/37,000) with the hallmark of the disease is bilateral vestibular schwannomas, which occur in approxi- mately 75% of NF-2 patients (Martuza and Haltia, 1993; White et al., 1990). The main differentiating fea- ture of schwannomas from neurofibromas, is the presence of hyalinized blood vessels. Immunohistochemical characteriza- tion may be highly suggestive but not diagnostic of the disease (Shaw et al., 2001). There is an increasing evidence that merlin/ schwannomin has a role as tumor suppressor protein (GFAP) and epithelial membrane antigen (EMA) are variably positive and contribute little to the diagnosis of schwannoma (Kawashima and Cavenee, 2000). The main differentiating fea- ture of schwannomas from neurofibromas, is the lack of asxins as demonstrated by several axon-specific immunohistochemical techniques within the tumor substance. Electron micro- scopic evaluation is sometimes required to document the Schwann cell composition of these tumors, which are characterized by a completely surrounding basal lamina.

Several atypical schwannomas exist, requiring expert neuropathological evaluation. Pseudoma- malomatous schwannomas are an aty- poic lesion and comprise of benign schwannoma which are packed with melanin and calcifications, and can become clinically malignant (Roubani et al., 1996) but its histologic criteria for malignancy is not well defined (Scheithauer et al., 1999). These tumors occur sporadically or may be part of Carney syndrome (Carney, 1990). Carney syndrome is an uncommon familial syndrome, characterized by a single or multiple schwannomas, arising from a single nerve fascicle, most often as solitary lesions but can occasionally occur as multiple tumors. Intracranially, they commonly occur as sporadic vestibular nerve schwanno- mas, however, bilateral tumors do arise rarely in the germ-line cancer pre-disposing syndrome, NF-2. Parapharyngeal schwannomas often have both intradural and extradural involvement with associated myelopathic and nerve root presentation, and occasional involvement of adjacent visceral structures. Schwannomas occurring on distal peripheral nerves have a tendency to arise on flexor surfaces.

Microscopic evaluation of the typical schwann-oma reveals alternating areas of cellular Antoni-A and loosely arranged Antoni-B regions with occasional false-nuclei results with this modality (Shih et al., 2003). Newer techniques often have both intradural and extradural involvement with associated myelopathic and nerve root presentation, and occasional involvement of adjacent visceral structures. Schwannomas occurring on distal peripheral nerves have a tendency to arise on flexor surfaces.

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MRI of PNTs may demonstrate heterogeneous enhancement, indicating intra-tumor hemorr- hage, neerosis or cystic degeneration. However, its relationship to malignancy is poor. In fact, there are no definitive radiological features of a Malignant Peripheral Nerve Sheath Tumor (MPNST), a diagnosis mainly suspected on clinical evaluation and with electrical stimulation noting distal muscle activity. Neurofibromas in con- trast typically do not reveal the discrete pas- sary, as nerve fascicles are encoun- tered and passed within the tumor. However, several major fascicles may be displaced around the bulk of the tumor and their position in the tumor capsule should be noted. 7)A small biopsy of the tumor from an electrically silent region is sent for pathological evaluation. Therapeutic surgical resection of PNTs is the treatment of choice. A donor nerve is used to reconstruct the nerve gap. 8)Anterior hypoglossal nerve paralysis (Fisher et al., 1993) is a complication of PNTs resection.

Operative Technique:

There are several operative principles that are applicable for all peripheral nerve tumors (July and Guha, 2008). 1. PNTs are mobile peripherally but do not along the longitudinal axis of a known peri- pheral nerve. 2. Palpation or percussion (Tinel sign) of a PNT may elicit sensory stimuli radiating along the distribution of the nerve of origin. 3. A mass in the presence of a patient with a genetic predisposition such as neurofibromatosis (NF) most likely represents a peripheral nerve tumor.

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Neurofibromas are benign peripheral nerve tumors that composed of schwann cells, perineurial cells, fibroblasts, and myxoid matrix with interwoven normal nerve fibers. These tumors arise spontaneously or in the context of neurofibromatosis-1 (NF-1), a disease first reported and named by von Recklinghausen in 1882 (von Recklinghausen, 1882). NF-1 is an autosomal dominant disorder that affects 1/500,000 people, of whom 30-50% represent new mutations. NF-1 is characterized by disomorphic features, which represent an extremely high spontaneous mutation rate (Kames, 1998). Mutations in the neurofibromatosis-1 gene also have been identified in some cases (Kuver et al., 2005). Neurofibromas are divided into several subtypes based on their location and gross appearance, viz. cutaneous/dermal neurofibroma (localized or diffuse), intraneural neurofibroma (localized), plexiform neurofibroma, and massive soft tissue/neural neurofibroma (Kilheus and Cawener, 2000).

Cutaneous neurofibromas is a cosmetic disfigurement problem and local irritation may be indications for surgery, but the majority of patients need only to be reassured that these tumors rarely undergo malignant transformation. The lesions of NF-1, however, have an increased association with NF-1, especially if they occur in large numbers. Progression of neurological symptoms, pain, and clinical or radiological increase in size are indications for surgery not only for potential amelioration of the symptoms, but also to exclude malignant transformation, which may occur in 3-5% of these plexiform neurofibromas (Sorensen et al., 1986).

Histopathological neurofibromas usually demonstrate elongated wavy interlacing hyperchromatic cells with spindle shaped nuclei in a disorderous mucoid background with collagen fibers and myxoid stroma. S-100 protein staining is negative for schwann cells but positive for other mesenchymal cells. Immunohistochemical staining for neurofilament protein can help to differentiate benign and malignant lesions. Nerve sheath tumors may also be distinguished from other soft tissue sarcomas by immunohistochemistry (see histological features of perineurioma).

Neurofibromatosis is a genetic disorder that affects the peripheral nerves which may arise spontaneously or in association with NF-1. The NF-1 gene encodes for a protein, neurofibromin, which is a member of the GTPase Activating Protein (GAP) family. Neurofibromatosis type 1 (NF-1) is an autosomal dominant disease with a frequency of approximately 0.001% in the general population (Ducatman et al., 1991). Conservative management of these tumors is recommended along with division of the nerve (surgery) which can be performed by the neurosurgeons.

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2. Neurofibromas

Neurofibromas are benign peripheral nerve cell tumors that complicate NF-1. They can arise from Schwann cells, fibroblasts, or mast cells, which may form true neurofibromas or diffuse or diffuse-sclerotic neurofibromas. The presence of axons within the tumor subcutaneous neurofibromas do not become problematic, we recommend a multi-disciplinary approach to these tumors, involving neurosurgeons, oncologists, and possibly radiation oncologists where required. We also recommend close follow-up of these patients, with regular MRI or CT scans to monitor the size and growth of the tumors. In cases where the tumors are growing or causing significant functional impairment, we recommend surgical intervention to remove the tumor or to relieve pressure on nearby structures. This may involve excision of the tumor with or without nerve grafting, or in some cases, radiation therapy.

3. Plexiform Neurofibromas

Plexiform neurofibromas are the most common type of NF-1-associated tumor, and can be found in approximately 40% of NF-1 patients. They typically involve the peripheral nerves and can cause significant pain, swelling, and functional impairment. The treatment of plexiform neurofibromas is usually surgical, with the goal of debulking the tumor and relieving symptoms. Radiation therapy and immunotherapy may also be used in selected cases. However, the natural history of these tumors is not well understood, and long-term outcomes are difficult to predict. In some cases, surgery may be deferred until the tumor grows larger or causes more functional impairment.

4. Benign Non-neural Lesions

Benign non-neural lesions include schwannomas, ganglioneuromas, perineuriomas, and neuromas. Schwannomas are benign tumors that arise from Schwann cells, which are the cells that wrap around nerve fibers. Ganglioneuromas are benign tumors that arise from sympathetic ganglion cells, which are found in the peripheral nervous system. Perineuriomas are benign tumors that arise from the perineurium, the connective tissue that surrounds nerve fibers. Neuromas are benign tumors that arise from nerve fibers. These tumors are usually benign and can be treated with surgical excision. However, in rare cases, schwannomas can become malignant and require more aggressive treatment.

5. Malignant Neurofibromas

Malignant neurofibromas are rare tumors that arise from Schwann cells. These tumors can be locally invasive, with the potential to spread to nearby tissues and organs. The treatment of malignant neurofibromas is usually surgical, with the goal of complete tumor resection. Radiotherapy and chemotherapy may also be used in selected cases. However, the prognosis for patients with malignant neurofibromas is generally poor, and the tumors can be difficult to control.

6. Conclusion

Neurofibromas and related tumors are a major challenge for patients with NF-1. The management of these tumors requires a multidisciplinary approach, involving neurosurgeons, oncologists, radiation oncologists, and other specialists. The goal of treatment is to relieve symptoms, prevent complications, and improve quality of life. However, the natural history of these tumors is not well understood, and long-term outcomes are difficult to predict. Further research is needed to improve our understanding of these tumors and to develop more effective treatment strategies.
Table 1. Peripheral Nerve Tumors Simple Classification Scheme

<table>
<thead>
<tr>
<th>Neurofibromatosis Type 2 (NF2)</th>
<th>Peripheral Nerve Tumors</th>
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<tbody>
<tr>
<td>Malignant</td>
<td>Schwannoma</td>
</tr>
<tr>
<td>Benign</td>
<td>Ganglion Cyst</td>
</tr>
<tr>
<td>Non-Neural Elements</td>
<td>Lipoma</td>
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<tr>
<td></td>
<td>Schwannomatosis</td>
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<tr>
<td></td>
<td>Neurofibromatosis</td>
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<tr>
<td></td>
<td>Neurofibroma</td>
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<tr>
<td></td>
<td>Others</td>
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</table>

Table 2. Malignant NF2 neoplasms

<table>
<thead>
<tr>
<th>NF2 Neoplasms</th>
<th>[1000]</th>
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</thead>
<tbody>
<tr>
<td>Acoustic Schwanoma</td>
<td>10%</td>
</tr>
<tr>
<td>Malignant schwannoma</td>
<td>30%</td>
</tr>
<tr>
<td>Malignant neurofibrosis</td>
<td>5%</td>
</tr>
<tr>
<td>Malignant lipoma</td>
<td>5%</td>
</tr>
<tr>
<td>Malignant neurofibromatosis</td>
<td>5%</td>
</tr>
<tr>
<td>Malignant neurofibroma</td>
<td>5%</td>
</tr>
<tr>
<td>Malignant choristoma</td>
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</tr>
</tbody>
</table>

Figure 1. Patient with left median nerve schwannoma. Upper Left: T1W MR images shows a mass along the course of left median nerve with obvious nodule. Upper Right: Intraoperative picture showed distal part before dissection to identify the nerve and involved with rubber band. Lower: Identifying the nerve and involved with rubber band. Schwannoma can be separated from the nerve and leave the nerve intact.
Peripheral Nerve Tumors Simple Classification Scheme

NIH Diagnostic Criteria for Neurofibromatosis Type 1 and Type 2 and Schwannomatosis

Presumptive Criteria:
- Two or more pathologically proved schwannomas
- One of the following - meningioma, glioma, schwannoma, juvenile posterior sub-capsular lenticular opacities
- Unilateral vestibular schwannoma diagnosed at an age less than 30 years
- Bilateral vestibular schwannomas (BANF)
- 2 or more neurofibromas of any type or one plexiform neurofibroma (95%)

Neurofibromatosis Type 1 (-incidence)
- Neuromuscular choristoma
- Desmoid
- Etc.
- Schwannoma

2. Primary peripheral nerve lymphoma

Only ten such cases have been reported so far and the majority of them are of B-cell type affecting sciatric nerve. This should be differentiated from neurolymphomatosis, which is a part of systemic manifestation of lymphoma. Treatment algorithms for nerve surgery and adjacent therapies (Misrai et al., 2000; Roncari et al., 1997).

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