Neural sheath tumors of major nerves

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Over a 22-year period, operations were performed on 263 patients for 288 primary benign tumors of major peripheral nerves. The tumors included 85 schwannomas, 197 neurofibromas, and six plexiform neurofibromas. Total removal was achieved in 83 of the 85 schwannomas, and 76 of these patients were available for follow-up evaluation. Motor function either improved or was unchanged in 87% of these patients and 85% of those with pain in the distribution of the involved nerve had either total or partial resolution of their symptoms. Of the neurofibromas, 123 occurred in 121 patients without von Recklinghausen's disease. All tumors within this group were completely excised using a fascicular approach to the tumor. Of the 99 patients available for follow-up evaluation, 90% had either improved or unchanged motor function and 88% had partial or complete resolution of pain syndromes. Fifty-nine patients with von Recklinghausen's disease had 80 tumors removed: 74 fusiform tumors (58 of which were completely removed) and six plexiform tumors. Forty-eight of the 58 patients with gross total removal of fusiform tumors were available for follow-up evaluation, of whom 83% had improved or unchanged motor function and 74% had partial or complete resolution of pain syndromes. All six patients with plexiform tumors had progression of symptoms postoperatively. One brachial plexus schwannoma recurred and was re-excised without subsequent recurrence at the 5-year follow-up evaluation. Several incompletely excised plexiform neurofibromas have recurred with a symptomatic presentation.

Key Words: schwannoma • neurofibroma • neurofibromatosis • peripheral nerve • brachial plexus • neural sheath

MANAGEMENT of peripheral nerve tumors with neural sheath origin has been a matter of debate for decades. Much of the concern has centered around the question of whether neurofibromas are safely resectable, and arguments have been presented that suggest they are not encapsulated. Some authors have indicated an unacceptably high and therefore prohibitive incidence of neurological injury with attempted resection of these lesions. Our experience suggests otherwise. In the period between October, 1968, and December, 1991, we performed 354 operations for tumors of major peripheral nerves: 263 of these were performed on 288 benign nerve sheath tumors. We present evidence suggesting that benign tumors of neural sheath origin can usually be resected with a small but acceptable risk to the nerve involved, regardless of histology.

Clinical Material and Methods

Patient Population

Between October, 1968, and December, 1991, operations for the treatment of benign nerve sheath tumors were performed on 263 patients at our institution. Of these, 248 operations were carried out for suspected tumor based upon historical, clinical, or radiological data. In the other 15 cases, the tumor was not diagnosed preoperatively, but was encountered at exploration. Tumors of intraspinal or intracranial origin or those in soft tissues other than major nerves were not included in this study, but tumors located at the intra- or extrarapinal spinal nerve or root level were included.

All patients had pre- and postoperative evaluation of symptomatology; evaluation was directed specifically to the presence or absence of spontaneous local or referred pain, weakness, or numbness. Strength and sensation were graded from 0 (absent) to 5 (excellent) using the Louisiana State University Medical Center (LSUMC) grading scale (Table 1). Individual muscle grades were combined to provide an overall motor grade for the nerve or plexus element involved. Electromyography was performed preoperatively in 90% of the cases.

Follow-up evaluation was obtained in person or by mail in 229 cases between 1 and 96 months postoperatively (mean 15.3 months). Patients were reevaluated for strength, and notations were made regarding the disappearance or persistence of preoperative pain and sensation.
Neural sheath tumors of major nerves

**TABLE 1**

<table>
<thead>
<tr>
<th>Grade</th>
<th>Evaluation</th>
<th>Description</th>
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<tbody>
<tr>
<td>0</td>
<td>absent</td>
<td>no contraction</td>
</tr>
<tr>
<td>1</td>
<td>poor</td>
<td>trace contraction</td>
</tr>
<tr>
<td>2</td>
<td>fair</td>
<td>movement against gravity only</td>
</tr>
<tr>
<td>3</td>
<td>moderate</td>
<td>movement against gravity and some (mild) resistance</td>
</tr>
<tr>
<td>4</td>
<td>good</td>
<td>movement against moderate resistance</td>
</tr>
<tr>
<td>5</td>
<td>excellent</td>
<td>movement against maximal resistance</td>
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**TABLE 2**

<table>
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<tr>
<th>Operative Site</th>
<th>Schwannoma</th>
<th>NF</th>
<th>NF + VRD</th>
<th>Totals</th>
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<tr>
<td>supraclavicular</td>
<td>24 (8%)</td>
<td>37 (13%)</td>
<td>14 (5%)</td>
<td>75 (26%)</td>
</tr>
<tr>
<td>brachial plexus</td>
<td>13 (5%)</td>
<td>21 (7%)</td>
<td>12 (4%)</td>
<td>46 (16%)</td>
</tr>
<tr>
<td>infracavicular</td>
<td>13 (5%)</td>
<td>21 (7%)</td>
<td>12 (4%)</td>
<td>46 (16%)</td>
</tr>
<tr>
<td>brachial plexus</td>
<td>14 (5%)</td>
<td>13 (5%)</td>
<td>8 (3%)</td>
<td>35 (12%)</td>
</tr>
<tr>
<td>median nerve</td>
<td>4 (1%)</td>
<td>16 (6%)</td>
<td>14 (5%)</td>
<td>34 (12%)</td>
</tr>
<tr>
<td>ulnar nerve</td>
<td>6 (2%)</td>
<td>5 (2%)</td>
<td>7 (2%)</td>
<td>18 (6%)</td>
</tr>
<tr>
<td>radial nerve</td>
<td>4 (1%)</td>
<td>7 (2%)</td>
<td>5 (2%)</td>
<td>16 (6%)</td>
</tr>
<tr>
<td>lumbarplexus</td>
<td>4 (1%)</td>
<td>7 (2%)</td>
<td>3 (1%)</td>
<td>14 (5%)</td>
</tr>
<tr>
<td>femoral complex</td>
<td>16 (6%)</td>
<td>17 (6%)</td>
<td>17 (6%)</td>
<td>50 (17%)</td>
</tr>
<tr>
<td>sciatic complex</td>
<td>10 (4%)</td>
<td>13 (5%)</td>
<td>10 (4%)</td>
<td>37 (13%)</td>
</tr>
<tr>
<td>totals</td>
<td>85 (30%)</td>
<td>123 (43%)</td>
<td>80 (27%)</td>
<td>288</td>
</tr>
</tbody>
</table>

* NF = neurofibroma; VRD = von Recklinghausen's disease; LS = lumbosacral. Percentages are of the total number of tumors.

**Histological Evaluation**

Most of the histological determinations were made by Dr. William Mitchell, a pathologist at the Ochsner Hospital, and by Dr. Carlos Garcia, neuropathologist at the LSUMC. Tumors that were difficult to categorize were studied histologically using the following stains: alcian blue, a stain for mucopolysaccharide; Gridley stain for reticulum; a Bodian stain to show axons; and the commonly used hematoxylin and eosin stain. Neurofibromas do not have the compact array of cells characteristic of schwannomas, but have a myxomatous matrix with a more prominent mucopolysaccharide staining and reticulum. Neurofibromas are also more likely to show axons on a Bodian stain than are schwannomas. Some of these differential histological features are depicted in Fig. 1.

**Operative Technique**

In each of the 263 patients, complete excision of the lesion was attempted. The technique used has been described previously for brachial plexus tumors. At exploration, the tumor and the nerve fascicles proximal and distal to the tumor were dissected. The usual finding was of a tumor growing within the substance of the nerve with uninvolved fascicles splayed circumferentially around the center of the mass (Fig. 2). With a microsurgical technique, these fascicles were gently dissected free of the tumor in the extracapsular plane. As the tumor was thus gradually exposed and the proximal and distal poles were approached, care was taken to isolate fascicles entering either the substance of the tumor or its capsule. A common finding in the case of either neurofibromas or schwannomas was of one or more fascicles running between the leaves of the capsule of the tumor but not entering the tumor proper. In many cases, these fascicles were not evident until dissection of the tumor poles was undertaken. Occasionally, when it was considered helpful to debulk large tumors in order to simplify subsequent dissection, a longitudinal incision was made in the capsule between fascicles for this purpose.

The gross distinguishing factor between neurofibromas and schwannomas was the tendency of neurofibromas to have more fascicles enter and exit the substance of the tumor at its poles or to have one relatively large contributing and exiting fascicle. Stimulation of proximal fascicle(s) usually yielded no motor response, and nerve action potential (NAP) recordings performed across the tumor usually resulted in a flat trace, indicating either failure to develop or loss of function of these fascicle(s). Schwannomas were noted to have fascicles that ran within the capsule, but were unlikely to have intratumoral fascicles of significance, although these did occur in the superficial layers of several very large tumors. The NAP recordings from capsular fascicles usually gave a positive response. When possible, these fascicles were spared.

Intraoperative NAP studies were performed, usually both prior to and following excision of the lesion. Exceptions to this rule occurred in the cases of large pelvic or presacral tumors in which proximal or distal nerve trunks could not be identified prior to excision of the mass. When the status of any fascicle or group of fascicles referable to the tumor was in doubt, they were tested by NAP studies. When necessary, nerve grafting was performed, usually for repair of nerve injuries that had occurred during previous operations (often biopsy procedures).
FIG. 1. Photomicrographs showing features typical of a schwannoma (left) and a neurofibroma (right). Polaroids kindly provided by Dr. Carlos Garcia. A and D: The schwannoma has a more compact array of cells than the neurofibroma, and Antoni A as well as Antoni B changes. H & E, × 130. B and E: The neurofibroma usually tends to have more stainable reticulum. Gridley, × 130. C and F: Many Schwann cells and some fibroblasts can be seen in both types of tumor, but there are fewer axons in the schwannoma than the neurofibroma. Bodian, × 130.
Neural sheath tumors of major nerves

Fig. 2. Left: Operative photograph showing a schwannoma of the sciatic nerve. Right: After partial dissection of fascicles surrounding tumor, the tumor proper becomes evident. Note the natural plane between the tumor and nerve.

Results

Schwannomas

Eighty-five patients underwent operation for removal of 85 schwannomas (Table 2). There were 42 male and 43 female patients with an average age at presentation of 40.2 years. Seventy-six patients were available for follow-up evaluation 3 to 96 months postoperatively (mean 16.7 months). One patient in this group had a giant cell median nerve tumor and another patient, with von Recklinghausen's disease, had an ulnar nerve tumor with the gross and microscopic appearance of a schwannoma. All tumors except two large pelvic schwannomas were completely excised. A nerve graft repair was required in two patients who had sustained damage from prior operations.

Of the 85 patients with schwannomas, 82 (96%) presented with a palpable mass. All but one patient experienced referred dysesthesia (Tinel's sign) when tapping or percussing was performed over the mass.

Table 3 shows the pre- and postoperative motor grades of the 76 patients with excised schwannomas who had follow-up evaluation. Of 51 patients with preoperative weakness, 17 (55%) improved, 10 (32%) were unchanged, and four (13%) worsened. Among the 45 patients who had intact strength preoperatively, 41 (91%) maintained full strength and four (9%) decreased to a grade of 4/5. Twenty-four (32%) of the 76 patients presented with pain syndromes. Of these, 20 patients had pain that was purely radicular in nature and four had pain that, while radicular, had a component of spontaneous pain localized to the site of the tumor. Of the 20 with pain syndromes, 15 (75%) had complete resolution of their pain syndromes at last follow-up evaluation, two (10%) had partial resolution, one (5%) had no change in the severity of his pain, and two (10%) had increased pain (Table 4). All four patients with pain localized to the site of the tumor had

<table>
<thead>
<tr>
<th>TABLE 3</th>
<th>Pre- and postoperative motor grade in 76 patients with schwannoma*</th>
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<tbody>
<tr>
<td>Postop Grade</td>
<td>5</td>
</tr>
<tr>
<td>Totals</td>
<td>41 (54%)</td>
</tr>
<tr>
<td>64</td>
<td>4 (5%)</td>
</tr>
<tr>
<td>35</td>
<td>2 (3%)</td>
</tr>
<tr>
<td>2</td>
<td>2 (3%)</td>
</tr>
<tr>
<td>1</td>
<td>1 (1%)</td>
</tr>
<tr>
<td>0</td>
<td>0 (0%)</td>
</tr>
</tbody>
</table>

* Motor grades of involved muscle groups in patients with resected schwannomas (see Table 1). Overall, 17 patients improved and eight worsened. Of the 51 with no change, 41 had normal strength at presentation. Percentages are of the total schwannoma cases (76) with follow-up evaluation.

<table>
<thead>
<tr>
<th>TABLE 4</th>
<th>Status of pain after operation on benign nerve sheath tumors</th>
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</thead>
<tbody>
<tr>
<td>Tumor Type</td>
<td>Patients Presenting With Pain Symptoms</td>
</tr>
<tr>
<td></td>
<td>No. of Cases</td>
</tr>
<tr>
<td>schwannoma</td>
<td>20</td>
</tr>
<tr>
<td>non-von Recklinghausen's neurofibroma</td>
<td>46</td>
</tr>
<tr>
<td>von Recklinghausen's neurofibroma</td>
<td>23</td>
</tr>
</tbody>
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J. Neurosurg. / Volume 81 / September, 1994

365
TABLE 5
Pre- and postoperative motor grade in 99 patients with neurofibroma*

<table>
<thead>
<tr>
<th>Postop Grade</th>
<th>Preop Grade</th>
</tr>
</thead>
<tbody>
<tr>
<td>5</td>
<td>4</td>
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<td>4</td>
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<td>1</td>
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<td>1</td>
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</tbody>
</table>

* Motor grades of involved muscle groups in patients with resected neurofibromas (see Table 1). Overall, 38 patients improved, 15 worsened, and 46 had no change in grade. Percentages are of the total number of neurofibroma cases with follow-up evaluation (99).

relief of this problem as well as relief of radicular pain. Four (7%) of 56 patients who had no pain syndrome preoperatively suffered mild paresthesias postoperatively.

Of the 85 schwannoma patients, 37 had undergone previous operations; however, only three procedures attempted complete resection of the tumor. These 37 included seven patients who experienced decline in motor function and four who had the onset of new pain postoperatively. Also, the only patients in this group who required graft repair of nerves had undergone biopsy procedures during which fascicles had been divided (seven patients).

One brachial plexus tumor was thought to have been totally excised but in fact recurred. This tumor was re-exercised and has not subsequently recurred during 5 years of follow-up evaluation.

Neurofibromas Without von Recklinghausen's Disease

There were 121 patients not believed to meet the criteria for von Recklinghausen's disease who had removal of 123 solitary neurofibromas. This group included 66 male and 55 female patients, with an average age at presentation of 39.1 years. Of these, 99 patients were available for follow-up evaluation 4 to 72 months postoperatively (mean 15.8 months). All tumors in this group were excised completely. Six patients (6%) required graft repair of nerves. Ninety-two patients had a palpable mass with a Tinel's sign on percussion over the mass.

Of the patients who had neurofibromas without von Recklinghausen's disease, 58 presented with a motor deficit of some degree (Table 5). Among this subgroup, 38 (66%) experienced improved long-term function, 14 (24%) did not change, and six (10%) had some further decrease in function noted at last follow-up evaluation. Forty-one (41%) of the 99 patients had normal function at presentation; of these, 32 (78%) maintained normal function while nine (22%) had postoperative weakness of some degree.

Within this group, 46 patients presented with spontaneous pain syndromes. The pain was radicular in all 46 cases; only two patients complained of additional pain at the site of the tumor. Postoperatively, pain resolved completely in 29 patients (63%), partially in 11 (24%), not at all in three (7%), and was made worse in three (7%) (Table 4). New pain syndromes were encountered in seven (13%) of the remaining 53 patients, although all were mild.

Fifty-three neurofibroma patients had undergone prior operations, including 11 attempts at complete removal of the tumor. It is important to note that seven of the nine patients who experienced new postoperative paresis, 12 of 15 patients whose paresis worsened, and each of the four who had new pain syndromes had all undergone previous operations.

von Recklinghausen's Neurofibromas

There were 57 patients who had neurofibromatosis (NF) type 1 in this series, comprising 25 males and 32 female patients. In addition, two patients, one male and one female, met the criteria for segmental NF (NF-S). The average age at presentation was 27.7 years. A total of 80 tumors were removed from these patients, with a maximum of seven in one patient. Six of these were plexiform tumors (see below). Many patients were noted to have multiple discrete tumors along the length of a single nerve; in such cases, the smaller tumors (<1 cm) were left alone. Of the tumors that were resected, 58 were completely removed and 16 were subtotally removed. Tumor left behind usually consisted of fragments adherent or intrinsic to functional fascicles. The incompletely removed tumors included a 30-cm pelvic tumor and a 25-cm tumor arising from the proximal brachial plexus that extended deeply into the mediastinum. About 10% of each of these tumors was left behind.

Six patients (11%) required graft repair of nerves following removal of tumor. Three patients required repeat surgery 12 to 21 months postoperatively at the site of the original tumor; however, each of these three patients had had small tumors documented near the site of the initially removed tumor. It was therefore impossible to ascertain whether these later tumors represented recurrences or simply progression of the smaller lesions. One patient had a subsequent occurrence of a malignant tumor at the site of a resected neurofibroma that had been histologically benign. Of the 57 patients in this group, 48 were available for follow-up evaluation (Table 6). Thirty-six patients presented with weakness. Of these, 18 (50%) had improved by the time of their last follow-up evaluation, 12 (33%) had no change from preoperative strength, and six (17%) worsened. Among the 12 patients who had normal strength at presentation, two (17%) had some significant deficit postoperatively. Twenty-three (48%) of the 48 patients had presented with radicular pain syndromes. Postoperatively, 10 patients (43%) had complete resolution of pain, seven (30%) had partial resolution, and four (17%) had no change after surgery (Table 4). In two patients (9%), the pain was worse after operation. Of the 25 patients who presented without
TABLE 6

<table>
<thead>
<tr>
<th>Preop Grade</th>
<th>Postop Grade</th>
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<tbody>
<tr>
<td>5</td>
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<tr>
<td>4</td>
<td>6</td>
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Neural sheath tumors of major nerves

Plexiform Neurofibromas

Six patients with plexiform neurofibromas were encountered. This represents 11% of the 57 patients with von Recklinghausen’s disease and 7.5% of that type of tumor in our study. Only two of these patients had a palpable mass. The tumors were treated as follows: partial excision in two patients; internal neurolysis with gross total excision in one; internal neurolysis with subtotal excision in one; and division of the involved nerve proximal and distal to the tumor with lengthy graft repair in two. The two patients on whom the latter procedure was performed had only trace strength in the associated muscles preoperatively. Neither of these patients had recovered function at their last follow-up evaluation. The tumor recurred in each of the other four patients. The time to symptomatic recurrence, as judged by progressive loss of strength, ranged from 4 to 19 months after the initial procedure. All patients in this group had motor scores at last follow-up evaluation lower than those recorded preoperatively.

Two of these patients had pain syndromes. Pain had lessened in severity with the passage of time after operation, but this was thought to be largely an effect of tumor regression rather than technical success.

Discussion

Clinical Findings

Neural sheath tumors have characteristics that may suggest their true nature on physical examination. In particular, deep-seated lesions that have been present for a long time, or are located over the course of a nerve, or are laterally but not longitudinally mobile with respect to that nerve, are suggestive of a neural sheath tumor. One sign less frequently reported is pain or paresthesias in the distribution of the involved nerve upon percussion or other manipulation of the tumor. This sign was present in all of our neurofibroma patients who had a palpable mass and in all but one of those with schwannomas, and should indicate proximity of the lesion, whatever its nature, to nerve. Park, et al., described a functionally analogous sign with schwannoma of the vagus nerve wherein percussion of the mass would elicit cough.

Opinions differ with respect to the incidence of spontaneous pain syndromes in patients with peripheral nerve tumors. Neurofibromas more commonly present with pain, but experience is divided with regard to schwannomas, for which reported incidences of pain syndromes range from 0% to 100%. In the present series, the incidence of pain associated with neurofibromas was 47%; however, the incidence was 31% in schwannoma patients. An interesting observation was made by Heard, who noted that, among patients with radiating pain, the tumor was found within a “recognizable” nerve in 84% of cases. In those without radiating pain, only 29% of tumors were found within a nerve.

In the present series, prior biopsy or other attempt at removal was associated with a higher incidence of serious pain in patients who presented for secondary surgery. In addition, most of the patients who had new pain and paresthesias after tumor excision had undergone prior biopsy or attempts at removal.

Diagnostic Studies

Aspiration biopsy has been recommended as an initial testing procedure but has not gained widespread acceptance. Angiography has been generally used in cases of intracranial neural sheath tumors and has been advocated elsewhere, but we have not found it to be helpful in diagnosing the usual peripheral neural sheath tumor. Ultrasound has been advocated as a diagnostic procedure of choice, but no large series using this technique is available. Computerized tomography (CT) has been very effective for diagnosis of intrathoracic or intrapelvic tumors and is sensitive to the cystic change that frequently accompanies these tumors. It has also been praised as the diagnostic test of choice for tumors of the sacral nerve. Magnetic resonance (MR) imaging has similarly been helpful. It is capable of reliably imaging not only the tumor and its capsule (Fig. 3), but also the nerve from which the tumor arises (Fig. 4). Gadolinium enhancement has been of special value for intra-abdominal and/or pelvic tumors. Neither MR imaging nor CT of a single tumor, with or without enhancement, has been effective in differentiating a schwannoma from a solitary neurofibroma or one associated with von Recklinghausen’s disease, however. On the other hand, MR imaging does appear to be reliable in differentiating between tumors and vascular anomalies in patients with neurofibromatosis.

In addition, the MR image and/or CT scan can show the multifocal nature of a plexiform tumor or lesions located elsewhere that are suggestive of von Recklinghausen’s disease.

Plain radiographs may show enlargement of inter-
vertebral foramina or, less frequently, vertebral erosion such as might be seen with a large presacral neural sheath tumor. Occasionally, an intercostal or a plexus tumor of the lower trunk will also be apparent on a chest x-ray film. Three tumors in the present series were discovered in this manner. In summary, no imaging or radiographic test available at the present time differentiates neurofibroma from schwannoma nor identifies with certainty a malignant neural sheath tumor.

Electromyographic and even nerve conduction studies are usually normal with most neural sheath tumors. This certainly was the case in the present series of tumors with the exception of some primary nerve malignancies and tumors in which prior biopsy or removal had been attempted.

The Schwannoma vs. Neurofibroma Controversy

Benign nerve sheath tumors were referred to in the collective sense as "neurofibromas" from the time of their first description until the early 20th century. When experience with nerve tumors evolved from reports of single cases to series, it became the norm to distinguish between schwannomas (elsewhere referred to as "neurilemomas") and neurofibromas. Even von Recklinghausen's suggested separate entities.

Pathological studies of benign nerve sheath tumors of every type have sometimes shown a hazy distinction between schwannoma and neurofibroma, the most constant finding being a higher content of collagen or elastin in neurofibromas. Both tumor types have been implicated as being of Schwann cell origin and some of their histological similarities are apparent even on transmission electron microscopy. Special stains, immunochemical studies, and electron microscopic studies show some preferential uptake or changes in either schwannomas or neurofibromas. These special studies supplement light histological stains but are not, in our experience, diagnostic in themselves. For example, S-100 protein stains show uptake in the "Schwann-like" cells in both tumors, although such uptake may be absent in some of the spindle-shaped cells seen in neurofibromas. On the other hand, the neurofibroma is more likely to have cells resembling a perineurial cell or a perineurial fibroblast. Electron microscopic study may thus show differences in the numbers of pinocytotic vesicles and remnants of basement membrane as well as the amount of well-developed endoplasmic reticulum in the presumed cells of origin.

Schwannomas tend to have both Antoni A and B tissue. The Antoni A portion of the tumor is relatively cellular with spindle- or eel-shaped cells, some of which may form palisades and become Verocay bodies. Antoni B tissue is a less compact or loose arrangement of spindle cells in a clear mucinous matrix. The spindle-shaped cells of a neurofibroma are separated by a more myxomatous stroma than is usually seen in a schwannoma. Distorted Schwann cell-axon complexes with both myelinated and unmyelinated axis cylinders are also evident. Since the neurofibroma has a more myxocollagenous matrix than a schwannoma, a stain for mucopolysaccharide such as alcian blue will be more intense in the neurofibroma. Such a stain is not, however, easy to standardize even in a given laboratory. On the other hand, stains for reticulum tend to be more positive in neurofibromas than schwannomas, and a Bodian stain is more likely to show axons.

It was believed by early authors that neurofibromas were "unencapsulated" lesions. This characterization was based upon histological discovery of nerve fiber elements within the tumor and not upon gross appearance or resectability. Our surgical experience indicates that most neurofibromas have a capsule and that fascicles are often peripherally enclosed in its layers. Thus, most neurofibromas should not be designated as unencapsulated tumors.

We have often encountered disagreement between frozen- and permanent-section histological diagnosis. The pathologists at our institution frequently deal with neural sheath tumors, and it is believed that basing a
Neural sheath tumors of major nerves

Surgical Management of Schwannomas and Neurofibromas

The argument for resection of schwannomas is a clear one (Fig. 5). These shiny, yellowish-white tumors are encapsulated, "extrafascicular" lesions in every sense. 

Despite this characterization, one can almost always find a small fascicle entering and exiting the proximal and distal poles of the tumor. This fascicle does not transmit NAP's, and can be sectioned so that the tumor can be totally removed. There is usually no difficulty encountered in their dissection unless the tumor is massive or prior surgery has been performed. 

Our operative approach involves extracapsular excision, but some authors have performed intracapsular enucleation with or without subsequent removal of the capsule with excellent results. 

Outcome is uniformly good for those patients who have complete excision of the tumor at first operation. However, large pelvic schwannomas are exceptions. We have encountered six of these lesions ranging from 10 to 33 cm in largest dimension. The difficulty presented by these tumors rests in the fact that they grow anteriorly, obscuring the location of intact pelvic plexus elements that lie behind them. Our usual method of dealing with this problem is by intracapsular removal of the tumor with the Cavtron ultrasonic surgical aspirator followed by dissection of the capsule away from the parent nerve once the course of the nerve is defined. We performed subtotal tumor excision in two elderly patients with very large tumors.

Decision-making regarding the management of schwannomas is difficult because the natural history of untreated schwannomas is largely unknown; however, the risk of malignant degeneration appears to be remote. Since most of these patients present without motor deficits, the decision to operate is based largely on expected improvement of pain, the presence of space-occupying symptoms, or rarely, cosmesis. Results after excision have usually been favorable with each of these symptom complexes.

In contrast to schwannomas, the proper treatment of neurofibromas has been controversial for some time. As early as 1930, surgeons commented that some neurofibromas within large nerve trunks were resectable without damaging the trunk itself; yet within the same theses, they described these lesions as unencapsulated. This concept has enmeshed itself into surgical theory; even as recently as 1991, authors have warned against attempted resection or have advocated en bloc resection of neurofibroma with involved nerve and end-to-end or graft repair based on frozen-section histology. Still others have advocated biopsy and observation for signs of increasing neurological deficit before resection is attempted.

A potential form of treatment for neurofibromas is intracapsular enucleation. This may be a necessary initial step in excision of very large tumors, but in smaller tumors the treatment is likely to leave fragments behind. In NF or von Recklinghausen's disease patients, there is a 15% risk of malignant degeneration of these tumors, so this approach is not without theoretical risk. By comparison, the incidence of malignant degeneration of solitary neurofibromas independent of von Recklinghausen’s disease is unknown but is considered to be quite low. Some of the largest series to date concerning management of benign nerve sheath tumors have advocated as the initial treatment radical excision of the tumor from its parent nerve. These series, however, have concentrated on experience with NF patients with peripharyngeal, nasal, submucosal, or subcutaneous tumors that did not involve large nerve trunks. 

Treatment of these tumors by wide excision is probably acceptable, but deeper tumors that involve major neural structures require a less radical approach if function is to be maintained. Patients with NF-1 have a characteristic gene defect on chromosome 17 and are...
especially prone to multiple growths. Treatment of each of these by wide resection and grafting would be crippling.

Our surgical approach to neurofibromas has been to consider them "intrafascicular tumors" and, by definition, tumors that require division of one or more parent fascicles for complete removal (Fig. 6). At operation these tumors do, however, exhibit a capsule. Between the layers of this capsule, it is not unusual to find intact nerve fascicles that may be separated from the tumor proper with microsurgical technique. The easiest method of identifying these fascicles is careful dissection at the proximal pole of the tumor. The fascicle(s) that gives rise to the tumor itself and are embedded within its substance are also found at this end. In comparison to the schwannoma, the tissue entering and exiting a neurofibroma is either a solitary but larger fascicle than is seen with a schwannoma or, in some cases, is actually two or three small, separate fascicles. Similar fascicle(s) are identified at the distal pole exiting the tumor, and division of both the proximal and distal fascicle(s) is necessary for a gross total removal of the tumor. Our experience with this potentially hazardous step has been good. These fascicles usually did not transmit an NAP, and their division has not been implicated in a demonstrable loss of function postoperatively (Fig. 7). Histological study of the entering fascicle has usually shown a disordered array of small to moderate-sized axons with poor myelination. Study of the exiting fascicle(s) has shown a degenerative picture with, on occasion, only fine and poorly myelinated axons (Fig. 8).

**Plexiform Neurofibromas**

Plexiform neurofibromas involve entire nerves or great lengths of them growing both intra- and extrafascicularly. They do not exhibit a capsule; rather, they appear to grow along tissue planes, similar to supportive-cell tumors like gliomas of the central nervous system. These features of a plexiform lesion make total tumor removal incompatible with maintenance of function. Our results with operative removal of these tumors are not good. Subtotal removal of some lesions has appeared to provide some amelioration of pain, but the tumor left behind has usually continued to grow. None of our patients has died as a result of plexiform neurofibroma, but the natural course with or without surgery has tended toward a complete palsy of the involved nerve. Our experience with radical removal and replacement of the resected segment with grafts has also been poor. This approach has been reported elsewhere with similar results. By the time a plexiform tumor has grown sufficiently to produce symptoms, it involves a segment of nerve that is many inches long. Resection leaves a gap that is associated with poor return of function after graft repair.
Incisional Biopsy of Neural Sheath Tumors

Many of the neural sheath tumors in this series were treated initially by incisional biopsy with or without debulking of the tumor. Results from secondary operation on these tumors are worse than those of procedures on unoperated tumors. Prior operation for biopsy or partial removal leads to obscuration of tissue planes and thus a difficult dissection. The incidence of decreased motor function with resection of a previously nonoperated schwannoma was 4% while that in the reoperated group was 19%. For neurofibromas, the incidence was 7% and 23%, respectively, for the two groups. A similar experience in smaller series has been cited elsewhere.15,30

One argument favoring biopsy of neural sheath lesions concerns its use in ruling out malignancy.18 However, malignant changes are not always spread throughout the tumor, so biopsy results at one or even multiple sites could be misleading. Furthermore, the gross appearance of malignant and benign tumors is fairly constant; rarely does one mimic the other.22,32,39,45 The malignant neural sheath tumor is usually quite firm, very adherent to adjacent structures, and usually destroys or badly distorts the internal architecture of the nerve of origin. Most of the malignancies that we have encountered were suspected preoperatively because of rapid increase in tumor size, progressive neurological loss, or, occasionally, imaging studies that suggested local invasion. Under these circumstances, timely referral to an experienced surgeon for definitive intervention makes the necessity for biopsy less urgent. In the event of suspected malignancy based on observed local invasiveness, however, incisional biopsy is indicated prior to instituting definitive therapy.

Conclusions

With the exception of the rare plexiform tumors, benign nerve sheath tumors should be considered removable; however, neurofibromas have been believed by many to be unresectable due to a misleading histological appearance that suggests invasiveness. Our results indicate that neurofibromas as well as schwannomas can be removed, even from large nerve trunks, with an acceptable risk of injury to the nerve. Excision of solitary neurofibromas is possible, especially if an intrafascicular approach at both poles of the tumor and repeated NAP recording are used. Fascicles entering and leaving the tumor substance are usually nonfunctional, and their division usually does not lead to increased neurological deficit. The functional risk of resection of neurofibromas is greater in patients with von Recklinghausen's disease. A similar procedure is warranted nonetheless because of the usual severity of the presenting symptoms and the risk of malignant degeneration in symptomatic tumors in these patients.

If the tumor is removed at the first surgical exposure,
excision is accomplished more easily and the long-term functional outcome is greatly improved. For these reasons, we advocate that surgical biopsy of tumors suspected to be of benign neural sheath origin be deferred. Total excision undertaken without prior biopsy or subtotal removal, if done by an experienced surgeon, appears to provide the best outcome overall.

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