Original Study

Benign Nerve Tumors of the Hand and the Forearm

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Abstract

We used a hand surgeon's 1978–1994 pathology reports to retrospectively review the incidence, preoperative and postoperative diagnoses, and presenting signs and symptoms of benign nerve tumors. Twenty-four (11.5%) of our series of 208 soft-tissue tumors of the hand and the forearm were benign nerve tumors. Nerve tumors were the third most common tumor after giant cell tumors of tendon sheath and inclusion cysts. Correct preoperative diagnosis was made in only 1 (4.2%) of the 24 cases. Schwannomas and neurofibromas were equally distributed (12 each), and 2 cases of neurofibromatosis (8.3%) were documented. Two (16.7%) of the 12 patients with schwannomas and 4 (33.3%) of the 12 patients with neurofibromas had neurologic symptoms. Six (85.7%) of the 7 digital tumors were dorsally located. In the literature, incidence of benign nerve tumors is much lower (ie, 1%-5%), and preoperative diagnosis consistently incorrect in our study. Incidence of neurologic symptoms (numbness, paresthesia) as presenting symptoms was higher in our study than previously documented. Although benign nerve tumors are most often located on the volar surface of the hand, 25% of the lesions we found were on the dorsal surface of the fingers.

The differential diagnosis for a soft-tissue tumor has 3 general categories: non-neoplastic lesions of soft tissue, benign tumors, and malignant tumors. Non-neoplastic lesions include foreign body granuloma, gout, pseudogout, epidermal inclusion cyst, mucoid cyst, and ganglia[1]; benign neoplasms include giant cell tumor of tendon sheath, fibroma of tendon sheath, lipoma, hemangioma, glomus tumor, schwannoma, and neurofibroma[1]; and malignant tumors include epithelial sarcoma, fibrosarcoma, malignant fibrous histiocytoma, primitive neuroectodermal tumors, malignant schwannoma, and cutaneous malignancies such as squamous cell carcinoma, basal cell carcinoma, and malignant melanoma.[1] As this differential diagnosis can be broad, and treatment approaches can vary according to diagnosis, it is helpful to have some knowledge of these entities with respect to clinical signs and symptoms and treatment.

In this article, we review the pathology and clinical findings of schwannomas and neurofibromas treated by a hand surgeon over a period of 16 years and retrospectively review all these cases. We sought to identify the clinical characteristics, locations, and preoperative diagnoses of nerve tumors of the hand and the forearm.

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Materials and Methods

We retrospectively reviewed all of a hand surgeon's pathology reports of soft-tissue tumors of the hand and the forearm at a single institution from 1978 to 1994. Twenty-four benign nerve cell tumors were then analyzed for preoperative diagnosis, presenting symptoms, age, sex, symptom duration, presence or absence of neurologic symptoms, and location.

One inclusion criterion for operation was a hand or forearm lesion of unknown etiology. Many of these cases were treated before use of magnetic resonance imaging (MRI) became commonplace in helping to diagnose and evaluate tumors, so in most cases the surgery was excisional biopsy. Another inclusion criterion was a symptomatic lesion of known etiology. Exclusion criteria were neurofibromatosis and ganglion cysts.

Surgical technique involved making a longitudinal incision, identifying the nerve proximally and distally to the lesion, and making the excision under magnification. Resection involved making it as complete as possible while maintaining as much of the normal nerve anatomy as possible. Nerve graft was not used. Fine-needle biopsy was not routinely performed before excision.

Results

Of the 208 soft-tissue tumors identified in the 16-year period, 24 (11.5%) were benign nerve tumors. Nerve tumors were the third most common tumor after giant cell tumors of tendon sheath and inclusion cysts. Preoperative

Table. Clinical Features of Schwannomas and Neurofibromas

Patient	Age (y)	Sex	Location	Size (cm)	Symptom	Duration	Preoperative Diagnosis
Schwannomas							
1	51	F	Dorsal wrist	2.0	Pain	6 months	Lipoma
2	31	M	Volar wrist	1.5	None	Unknown	Giant cell tumor
3	43	M	Volar forearm	1.0	Pain	9 months	Lipoma
4	52	M	Volar forearm	1.5	Numbness	Unknown	Schwannoma vs neurofibroma
5	73	F	Volar hand	1.5	Pain	10 years	Ganglion cyst
6	79	F	Volar hand	2.0	Pain	5 years	Giant cell tumor vs neurofibroma
7	17	F	Dorsal finger	1.0	None	3 years	Hemangioma
8	22	F	R volar hand L dorsal finger	2.0	Enlarging mass	1 year	Lymphangioma
9	50	M	Dorsal finger	0.7	Pain	Unknown	Giant cell tumor
10	75	M	Volar forearm	2.0	Numbness	5 months	Unclear
11	61	M	Volar hand	1.0	Pain	4 months	Unclear
12	35	F	Volar forearm	1.5	Pain	2 years	Lipoma
Neurofibromas							
1	64	M	Dorsal finger	1.0	None	18 months	Giant cell tumor
2	41	M	Forearm	5.0	None	12 months	Lipoma
3	29	M	Dorsal finger	1.0	Pain	5 months	Ganglion vs giant cell tumor
4	42	F	Ulnar wrist	0.7	Numbness	Unknown	None
5	38	M	Web space	1.0	Pain	5 years	Ganglion vs giant cell tumor
6	61	F	Volar finger	2.5	Numbness	6 years	Ganglion
7	73	F	Volar hand	2.0	None	Unknown	Inclusion cyst
8	24	M	Volar wrist	3.5	Numbness	6 months	Ganglion vs schwannoma
9	44	M	Volar hand	2.5	Numbness	4.5 years	Inclusion cyst
10	53	F	Dorsal finger	1.0	Pain	Unknown	Unclear
11	60	F	Volar hand	1.5	None	Unknown	Unclear
12	6	F	Volar finger	1.0	Enlarging mass	Unknown	Ganglion vs hemangioma

diagnosis was consistently incorrect (Table): Correct preoperative diagnosis was made in only 1 (4.2%) of the 24 cases. Schwannomas and neurofibromas were equally distributed (12 each), and 2 cases of neurofibromatosis (8.3%) were documented. Two (16.7%) of the 12 patients with schwannomas and 4 (33.3%) of the 12 patients with neurofibromas had neurologic symptoms. Although most benign nerve tumors are located on the volar surface of the hand, 6 (85.7%) of the 7 digital tumors we found were dorsally located. Five (20.8%) of the 24 cases were lesions of the forearm, and 4 (33.3%) of the 12 schwannomas were of the forearm. All cases were followed up for a minimum of 1 year. There were no recurrences.

Discussion

True nerve tumors are of neuroectodermal origin. They are completely encapsulated by epineurium and thus have a true capsule. The 2 types of benign nerve tumors representing the vast majority of all benign nerve tumors are schwannoma and neurofibroma. Benign tumors of peripheral nerves have been documented to be relatively rare in the hand and forearm, representing less than 5% of tumors in this area.[2] Ganglion cysts are generally documented as the most common soft-tissue tumor in this region, followed by giant cell tumors of the tendon sheath.[1]

Often, the diagnosis is not made before surgery. In our series, patients were most commonly diagnosed with ganglion (5 cases), giant cell tumor (4), and lipoma (4). Although no tumor was malignant, malignancy remains a possibility. Given this possibility and the poor prognosis associated with poorly planned tumor surgery, care should be taken with such surgery, especially when malignancy is suspected. Hemostasis should be meticulous; the biopsy incision should be planned for possible later resection, including the original biopsy tract; and tumors should be handled as atraumatically as possible to limit possible tumor seeding.

Schwannoma

Schwannoma (ie, neurilemoma) is a benign encapsulated proliferation of Schwann cells involving peripheral nerves. After neurofibroma, it is the most common tumor[3] involving peripheral nerve. Schwannoma is usually a solitary tumor[4] along the course of a nerve, and it appears to have an ovoid or dumbbell shape when exposed surgically (Figure 1). It is extrinsic to the nerve proper, grows slowly, and is usually less than 3 cm in diameter. The lesion is well encapsulated, and the nerve fibers typically do not enter the tumor.

Histologically, schwannoma has the consistent appearance of a differentiated Schwann cell and has 2 components: highly ordered dense arrays of spindle cells (Antoni-A cells) and a hypocellular region of connective tissue with less organized spindle cells (Antoni-B cells). The Antoni-A cells have a characteristic palisading pattern formed by 2 rows of adjacent nuclei (“picket fence” appearance).

The biphasic or alternating pattern of Antoni-A and Antoni-B cells is characteristic of schwannoma.[5] This alternating pattern and encapsulation can differentiate schwannoma from neurofibroma. Clinically, they most commonly occur equally in males and females and between the ages of 20 and 50. According to Strickland and Steichen,[2]

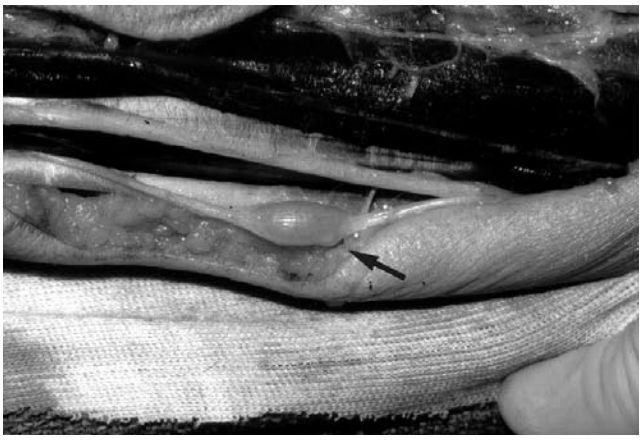

Figure 1. Schwannoma of the dorsal sensory branch of the ulnar nerve (arrow). Note ovoid and well-encapsulated appearance. Neurofibromas are more intimately associated with the nerve and are not easily separated from it. This clinical appearance helps in differentiating schwannomas from neurofibromas.

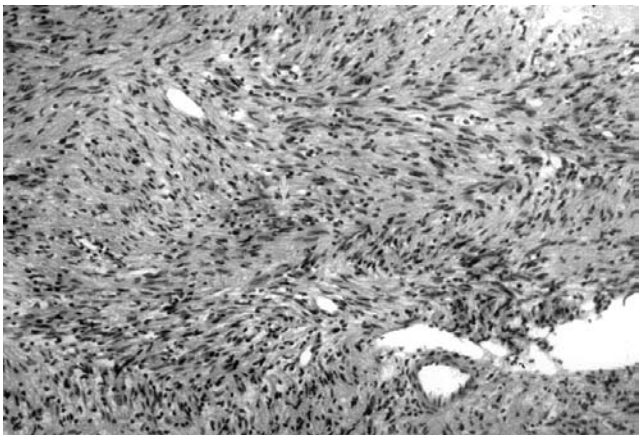

Figure 2. Microscopic section of schwannoma shows acellular areas between rows of opposing nuclei—characteristic of Verocay bodies.

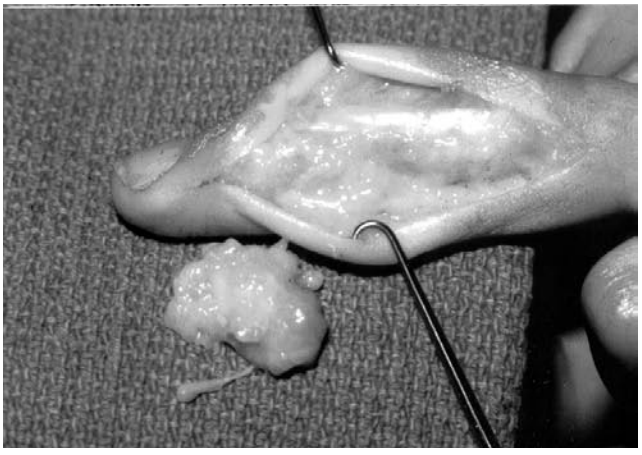

Figure 3. Plexiform growth pattern typical of neurofibromatosis.

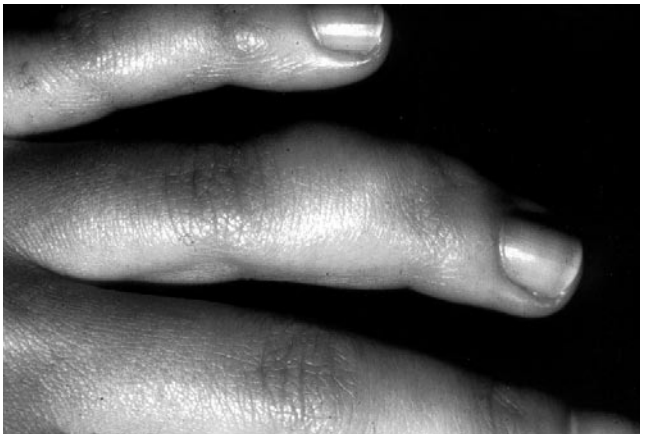

Figure 4. Irregular fusiform swelling in plexiform neurofibroma.

presentation is usually as a painless mass, though pain and paresthesia can occur.

Management usually consists of excisional biopsy. Because the nerve fibers do not enter the tumor, complete excision is usually possible. When the lesion is not encapsulated, or when the nerve fibers enter the tumor, the possibility of malignancy or another diagnosis should be considered.[3] Diagnosis can be confirmed by the presence of alternating Antoni-A and Antoni-B cells as well as Verocay bodies, which are acellular areas between rows of opposing nuclei (Figure 2). Recurrences after excision are unusual. Stout[4] reported 50 cases with no recurrence after simple excision. Removal is typically possible without damage to the surrounding nerve. Malignant degeneration of these tumors is rare.[2]

Neurofibroma

Neurofibromas are benign nonencapsulated tumors growing from Schwann cells. However, unlike schwannomas, they have a wider, more disordered spectrum of cellular pathology involving portions of the peripheral nerve fiber. Neurofibromas tend to involve the central portion of the nerve and may be difficult to dissect free without damaging nerve fibers. Because neurofibromas occur within the nerve and are not extrinsic to it, as schwannomas are, they cannot be enucleated from the nerve fibers (unlike schwannomas).[6]

Neurofibromas occur in 2 basic disease patterns: sporadic and in association with neurofibromatosis. Localized neurofibromas are thought to be solitary lesions not associated with systemic disease. Approximately 90% of neurofibromas are localized lesions[4]; the rest are associated with neurofibromatosis. This distinction becomes important because of the higher likelihood of malignancy of neurofibromas in neurofibromatosis.

Neurofibromatosis (ie, von Recklinghausen disease) is a neurocutaneous syndrome that affects the skin, the nervous system, the musculoskeletal system, and the eyes in various manifestations. Neurofibromatosis 1 involves a genetic defect of chromosome 17, encoding for a tumor suppressor gene, NF1, which is transmitted in an autosomal-dominant pattern. Clinical signs include café au lait spots, axillary/inguinal freckling, optic glioma, Lisch nodules (iris hamartomas), osseous lesions, and neurofibromas.

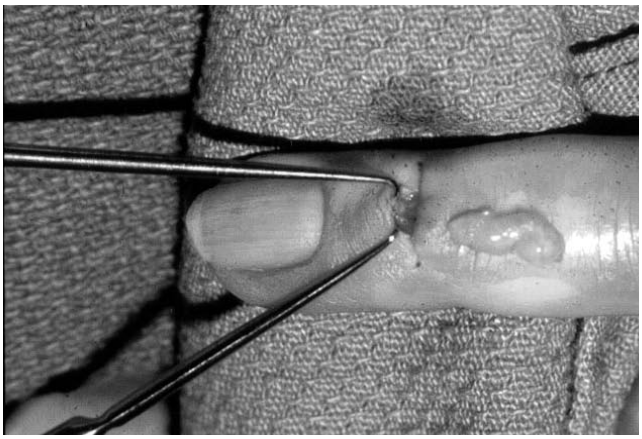

Figure 5. Excised plexiform neurofibroma with wormlike appearance. An incorrect preoperative diagnosis of mucous cyst was made, and the proper surgical principles of oncology were not followed. A longitudinal incision should have been considered.

Neurofibromas often occur in a plexiform growth pattern, which is pathognomonic of neurofibromatosis (Figure 3). Plexiform growth is multicentric and involves the same nerve over an extended area, resulting in irregular thickening of the nerve (Figure 4). Neurofibromas can involve an entire extremity, and they have a classic "bag of worms" appearance (Figure 5). Skeletal abnormalities occur in up to 40% of patients[5] with the disease and can manifest as long-bone bowing, scoliosis, and nonossifying fibromas.

Preoperative Diagnosis. The preoperative diagnosis was consistently incorrect; only 1 (4.2%) of the 24 cases received the correct preoperative diagnosis. Many of these cases were treated before MRI was routinely used as a diagnostic aid, and MRI was not performed before surgical excision in any of our cases. Although the preoperative diagnosis was consistently incorrect, nerve tumor was considered a possibility in the differential diagnosis in most cases. Kang and colleagues[10] reported 6 of 9 cases correctly identified with preoperative MRI and 0 of 14 cases correctly diagnosed without preoperative MRI. Holdsworth,[11] reporting a similar difficulty, obtained correct preoperative diagnoses in only one third of cases. In addition, White[12] reported 5 (11.1%) of 45 cases receiving correct preoperative diagnoses. These results underscore the importance of maintaining a high index of suspicion for nerve tumors. Nerve tumors appear bright on T_2-weighted images and moderately bright on proton-density-weighted images.[10] In addition, a capsule may be evident with schwannomas. We recommend using MRI to evaluate these lesions when the diagnosis is unclear.

Role of Needle Biopsy. Fine-needle biopsy may also be used to evaluate forearm lesions. Fine-needle aspiration was not routinely used in our series. We aspirate larger hand and forearm tumors to differentiate a solid tumor from a ganglion. Only recently has fine-needle aspiration become the standard at our institution. A needle biopsy is performed on most extremity tumors, excluding those of the hand, before a decision is made regarding definitive treatment.

"[When surgery is warranted] One must weigh the consequences of malignant degeneration of a neurofibroma against the potential for nerve damage during excision."

Whereas malignant transformation is rare in isolated neurofibromas, it has been reported to occur in 2% to 13% cases of von Recklinghausen disease.[7] When symptoms warrant, treatment for isolated neurofibromas is excision. However, when the diagnosis is clear and excision of nerve fibers is necessary, a small amount of tumor may be left behind, given the low potential for malignant transformation and recurrence, as suggested by Healey and McCormack.[7] One must weigh the consequences of malignant degeneration of a neurofibroma against the potential for nerve damage during excision.

Findings in This Series and in the Literature

In our series, nerve tumors were the third most common diagnosis of soft-tissue tumors in the hand and the forearm. Incidence as reported in the literature has ranged from 1.8%[8] to 4.9%[9] of all tumors of the hand. In a review of 689 tumors of the hand and the forearm, Strickland and Steichen[2] found 6 nerve tumors (0.87%). Thus, incidence is higher in our series than in previous reports. Schwannomas and neurofibromas were equally distributed.

Clinical Symptoms. Several authors have described nerve tumors as painless and not tender. Stoudt[13] felt that the most common presentation was incidental, with clinical symptoms absent. In our series, incidence of symptomatic tumors was higher: 75.0% (9/12 schwannomas) and 58.3% (7/12 neurofibromas). The most common presentation symptoms were pain (58%) and neurologic dysfunction (16%) for schwannomas and paresthesia (33%) and pain (25%) for neurofibromas.

Neurologic Symptoms. Incidence of neurologic symptoms on presentation has been lower in previous series. Holdsworth[11] reported 2 (11.1%) of 18 nerve tumors presenting with paresthesia; we reported a slightly higher incidence of neurologic symptoms, 2 cases (16.7%) in 12 patients with schwannomas and 4 cases (33.3%) in 12 patients with neurofibroma. No patient in our series presented with objective motor weakness. White,[12] however, reported paresthesia and hypesthesia in 24 (53.3%) of 45 patients with schwannoma and in 1 (22.2%) of 45 patients

with motor weakness, though that review was not confined to the upper extremity. In our series, the higher incidence of neurologic symptoms with neurofibromas most likely represents the more intimate association between neurofibromas and the nerve, as neurofibromas tend to involve the central portion of the nerve and are not extrinsic to the nerve.

Ganglion Cysts. A weakness of this study was the low incidence of ganglion cysts in our series. Based on a literature review, Bush[14] reported an incidence of ganglia in tumors of the hand and wrist between 33% and 56%. In this series of 208 soft tissue tumors of the hand and forearm, the incidence of ganglia was 15%. This low incidence reflects our reluctance to operate on ganglia, given their benign nature and likelihood of recurrence. We tend to see more problems caused by ganglion excision than by the ganglions themselves. Our basic approach to hand and wrist ganglions is to "aspirate, splint, procrastinate, and then reaspirate." Zubowicz and Ishii[15] found a cure rate of 85% in multiple aspirations. This use of aspiration as treatment rather than surgery explains the relatively higher incidence of nerve tumors in our series. Nerve tumors were third in incidence of tumors of the hand and the forearm, behind inclusion cysts and giant cell tumors, and nerve tumor incidence was similar to but slightly higher than lipoma incidence.

Strengths of This Study. Several of the previous investigators reviewed nerve tumors only of the hand. Our study is unique in that it includes tumors of both the hand and the forearm. Twenty percent of the tumors (and 33% of the schwannomas) were in the forearm. Another strength of this study is its large number of cases, which represent all the pathology reports for hand and forearm tumors at one institution.

Symptom Duration; Mass, Size, and Location. Symptom duration ranged from 6 months to 10 years (means, 30.6 months for schwannomas, 32.4 months for neurofibromas). Several patients had symptoms for several years. Seddon[16] reported that schwannoma size usually does not exceed 2.5 cm, though larger tumors have been reported. Seddon's finding is consistent with ours (largest schwannoma, 2.0 cm). Although benign nerve tumors can occur anywhere, in our series the volar surface of the hand was the most common location, followed by the volar forearm (consistent with previous findings[11,12,17]). Of note, however, 6 (85.7%) of the 7 digital tumors were dorsally located.

Conclusions

Benign nerve tumors, such as schwannomas and neurofibromas, are relatively uncommon entities, representing approximately 11.5% of benign soft-tissue tumors in one surgeon's 16-year experience. Schwannomas can be distinguished from neurofibromas by the former's lack of potential for malignant degeneration and lack of plexiform growth or multiplicity of growth sites (present in neurofibromas and characteristic of neurofibromatosis). Benign nerve tumors most commonly presented with pain or paresthesia. Although the most common location of benign nerve tumors is the volar surface of the hand, 25% of the lesions we found were on the dorsal surface of the fingers. Preoperative diagnosis was consistently incorrect. MRI or fine-needle aspiration should be used in evaluation. In the majority of cases, once a diagnosis is clear, the entity can be treated with simple excision.

Authors' Disclosure Statement

No benefit in any form has been received or will be received from a commercial party related directly or indirectly to the subject of this article. No funds were received in support of this study. The authors report no actual or potential conflicts of interest in relation to this article.

References

1. Plate AM, Lee SJ, Steiner G, Posner MA. Tumorlike lesions and benign tumors of the hand and wrist. *J Am Acad Orthop Surg.* 2003;11:129-141.
2. Strickland JW, Steichen JB. Nerve tumors of the hand and forearm. *J Hand Surg Am*. 1977;2:285-291.
3. Idler RS. Benign and malignant nerve tumors. *Hand Clin.* 1995;11:203-209.
4. Stout AP. Tumors of peripheral nerves. *J Bone Joint Surg Am.* 1958;40:959.
5. Weiss SW, Goldblum JR, eds. Benign tumors of peripheral nerves. In: *Enzinger and Weiss's Soft Tissue Tumors.* 4th ed. St. Louis, Mo: Mosby; 2001:1111-1207.
6. Louis DS, Hankin FM. Benign tumors of the upper extremity. *Bull N Y Acad Med.* 1985;61:611.
7. Healey JH, McCormack RR. Nerve tumors. In: Bogumill GP, ed. *Tumors of the Hand and Upper Limb.* New York, NY: Churchill Livingstone; 1993:205-222.
8. Butler ED, Hamill JP, Seipel RS, deLorimier AA. Tumors of the hand. *Am J Surg*. 1960;100:293-303.
9. Stack HG. Tumours of the hand. BMJ. 1960;1:919-212.
10. Kang HJ, Shin SJ, Kang ES. Schwannomas of the upper extremity *J Hand Surg Br.* 2000;25:604-607.
11. Holdsworth BJ. Nerve tumours in the upper limb. A clinical review. *J Hand Surg Br. 1985;1(2):236-238.*
13. Stoudt AP. The peripheral manifestations of the specific nerve sheath tumors. *Am J Cancer.* 1935;24:751-796.
14. Bush D. Soft tissue tumours of the forearm and hand, In: Mackin EJ, ed. *Rehabilitation of the Hand and Upper Extremity.* 5th ed. St. Louis, Mo: Mosby; 2002:955-970.
15. Zubowicz VN, Ishii CH. Management of ganglion cysts by simple aspiration. *J Hand Surg Am.* 1987;12:618.
16. Seddon HJ. *Surgical Disorders of Peripheral Nerves.* Baltimore, Md: Williams & Wilkins; 1972.
17. Phalen GS. Neurilemomas of the forearm and hand. *Clin Orthop.* 1976;114:219.

This paper will be judged for the Resident Writer's Award.