Multiple Neurilemomas in the Upper Extremity
A Series of Three Cases

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Abstract
Neurilemomas are the most frequently arising benign nerve tumors of the upper extremity and are also called Schwannomas. Generally, they present as solitary tumors, although multiple tumors are common. Regardless of number, they are usually found on the flexor surface of the forearm and hand, and multiple tumors are almost always located within a single major nerve, its branches, or both. We present three patients who had multiple neurilemomas; two patients had tumors within a single major nerve and its branches, and the third patient had an unusual occurrence of one tumor in the ulnar nerve and a second tumor in a branch of the median nerve.

Neurilemomas, also referred to as Schwannomas, are the most common neoplasms involving peripheral nerves. They comprise approximately 5% of all benign soft tissue tumors, and their incidence in the upper limb ranges from 3% to 19% of all neurilemomas. They generally involve nerves on the flexor surface of the forearm and are usually solitary although multiple tumors do occur. Neurilemomas are encapsulated nerve sheath tumors that arise from the Schwann cells of myelin sheaths. Nerve fascicles remain intact and are typically splayed over the surface of the tumor. Those fascicles can usually be preserved at surgery.

Case One
DW, a 41-year-old urologist, first examined more than 20 years ago, had a gradually enlarging mass on the volar aspect of his dominant right ring finger that had appeared 8 years earlier. He complained of numbness in the finger and also of pain in the area of the medial epicondyle of his elbow that began approximately one year earlier. His past medical history was significant in that he had prior excisions of two tumors in that same extremity. The first tumor was excised during childhood from the same right ring finger but at a more proximal site. The operation had been performed by his father, a general surgeon, who simply discarded the tissue and did not submit it for microscopic study. When the patient was 27, a second tumor was excised from the ulnar nerve of his right wrist and was reported to be a “neurilemoma.”

Clinical examination showed a nontender mass that measured 2.0 cm x 1.0 cm over the middle segment of the ring finger, directly over the ulnar digital nerve. The mass was distal to the operative site of the mass excised from that same finger by his father many years earlier. The second mass was nontender, but the patient reported numbness at the ulnar tip of the finger and two-point discrimination was slightly impaired (7 to 8 mm). There was a second mass that the patient was not aware of at the site of his elbow pain. This mass was at the cubital tunnel, directly over the ulnar nerve, where it passed between the two heads of the flexor carpi ulnaris muscle. The mass measured approximately 1.5 cm x 0.5 cm, and percussion over it produced distal paresthesias that radiated into the little finger.

At surgery, the tumor in the finger was within the ulnar digital nerve and the tumor at the elbow, within the ulnar nerve (Fig. 1). The nerve fascicles at both sites were splayed over the tumors and were preserved at surgery. Histological examination confirmed that both tumors were neurilemomas. Postoperatively, forearm pain and sensitivity over the ulnar nerve in that area disappeared. However, numbness in the...
ring finger persisted, and two-point discrimination increased to 11 to 12 mm.

The patient returned 9 years later, because he had noted two new masses, one on the volar-ulnar aspect of the ring finger distal to the site of the last operation and a second over the dorsum of the distal segment of the same finger, just proximal to the nail. At surgery, the mass on the volar side of the finger was within the ulnar digital nerve, distal to the distal interphalangeal flexion crease, and measured 0.3 cm x 0.3 cm. The dorsal mass did not appear to be associated with any nerve. Both tumors were excised, and the pathologist reported that both were neurilemomas. The patient remained symptom free for almost 10 years, except for slight numbness at the ulnar tip of the ring finger. He returned because of two additional masses (the seventh and eighth tumors in his right upper extremity), one on the ulnar side of his palm and the other in the little finger. At surgery, both tumors were intraneural, the palmar tumor within the common digital nerve to the ring and little fingers, and the little finger tumor within the ulnar digital nerve. Both were excised, and both were neurilemomas on microscopic examination. The patient has no disability; he continues his full time practice in urology and participates fully in leisure time activities.

Case Two

SF, a 31-year-old female secretary, presented with soft masses on the volar aspects of her dominant left hand and forearm. The palmar mass had been present for more than 13 years and had been diagnosed as a “ganglion” by her family physician. The forearm mass had appeared within the past 3 years and was associated with increasing numbness and paresthesias in the thumb and little fingers. That mass was located 3.0 cm proximal to the wrist flexion crease and measured 1.0 cm x 1.0 cm. It was nontender and there was no Tinel over it. However, percussion over the palmar mass that also measured 2.5 cm x 1.5 cm caused paresthesias that radiated into the middle finger.

At surgery, both masses were intraneural tumors, the palmar mass within the common digital nerve to the index and middle fingers, and the forearm mass within the median nerve (Fig. 2). Both tumors were excised and pathological study confirmed that they were neurilemomas. Postoperatively, there were no motor or sensory deficits.

Case Three

GW, a 40-year-old male clerical worker, complained of ulnar-sided right wrist pain of approximately 1 year’s duration and occasional paresthesias in his ring and little fingers. The patient was also aware of a gradually enlarging mass on the volar surface of the mid-forearm.

On examination, the mid-forearm mass measured approximately 3.0 cm x 2.0 cm. It was nontender and there was no Tinel with percussion over it. On the volar-ulnar side of the proximal forearm, there was second much smaller mass over the course of the ulnar nerve at a site 5.0 cm distal to the medial epicondyle. The second mass was tender, but there was no Tinel with percussion over it. There were no sensory deficits in the hand, but there was marked weakness of the ulnar innervated intrinsic muscles. The median innervated intrinsic muscles and all extrinsic muscles were not affected. Radiographs of the forearm were negative, but magnetic resonance imaging (MRI) with contrast highlighted both masses. Electrodagnostic studies showed a focal neuropathy of the ulnar nerve at the site of the mass.

At surgery, the mid-forearm tumor was not within the median nerve, but was adjacent to it and attached by a thin nerve fiber (Fig. 3). It was encapsulated and easily excised. The more proximal tumor was within the ulnar nerve and its fascicles splayed over it. Those fascicles were preserved at excision of the tumor. Both tumors were neurilemomas. Postoperatively, there were no sensory deficits and the
patient recovered full power of the ulnar nerve innervated intrinsic muscles within 4 months.

Discussion

Neurilemomas are the most common neoplasms arising from neural tissue. They were first described by Verocay, in 1908, who termed them neurinomas. The name neurilemoma was later coined by Stoudt in 1935. Neurilemomas usually present as painless, round or oval masses on the flexor surfaces of upper extremities. They rarely measure more than 2.5 cm in diameter and are often misdiagnosed as cysts, ganglions, or lipomas. Neurilemomas are slightly more common in females and generally appear between the second to eighth decades. They are usually solitary tumors, although there are many reports in the literature of patients with multiple tumors that arise sequentially. In cases of multiple neurilemomas, their distribution is generally within one major nerve and its branches. This was the situation in our first two patients. The first patient has had a total of seven histologically confirmed neurilemomas that developed sequentially over a period of more than 15 years. An eighth tumor, the first he had when a child, was removed by his physician father, who did not have it pathologically examined. That tumor was probably a neurilemoma, since it involved the little finger of the same extremity as the other tumors. Rarely, multiple neurilemomas can arise in different major nerves, as in our third patient.

Neurilemomas arise eccentrically from nerves or are located centrally within nerves. Regardless of location, they are usually firm in consistency, with occasional focal areas of degeneration and cysts. Since they are encapsulated and arise from the cells of nerve sheaths and not from the nerve fibers themselves, they can usually be separated from the fibers when excised. This can be difficult for a large tumor within a digital nerve, since there are few fascicles at this location and the fascicles themselves may be severely attenuated. However, resection of a neurilemoma with the involved portion of the nerve is almost never required. Neurilemomas are composed of several types of cellular elements that characterize its histological appearance (Fig. 4). Those elements are Antoni A tissue, composed of closely packed spindle shaped cells arranged in a palisading net work; Antoni B tissue that is far less organized, consisting of myxoid tissue with cystic spaces and sparsely populated with cells; and Verocay bodies that are linear arrays of spindle cell nuclei. The three elements are not all always present in every tumor.

Neurilemomas must be differentiated from neurofibromas. Aside from café-au-lait discolorations that are common with neurofibromas, there are two important differences between the tumors. The first is that, unlike neurilemomas, neurofibromas are not encapsulated and the nerve’s fascicles are an integral part of the tumor. Excision is therefore impossible without damaging the fibers. It is for that reason that solitary neurofibromas are generally left undisturbed if nerve function has not been severely compromised and the patient is asymptomatic. The second important difference is that, unlike neurilemomas, neurofibromas have potential for malignant degeneration. This is more likely to occur when there are multiple tumors associated with Von Recklinghausen’s disease, where the incidence of malignancy is approximately 15%. Therefore, biopsy is recommended for a neurofibroma that enlarges or becomes painful, although malignant degeneration may not necessarily result in pain.
Since the treatment and prognosis are significantly different for neurilemomas and neurofibromas, careful inspection of these tumors at surgery is critically important. Proper treatment for a neurilemoma is excision and careful surgical technique; the aid of a microscope or loupe magnification will usually avoid any further damage to the nerve’s fascicles. Since neurilemomas are often multiple, generally arising as sequential tumors, patients should be alerted to this possibility, and periodic clinical examinations are advisable. The sequential development of additional tumors is usually within the same major nerve and its branches, but other tumors can also arise in different major nerves.

**Disclosure Statement**
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**References**