BULLETIN OF
THE NEW YORK ACADEMY
OF MEDICINE

VOL. 61, NO. 7 SEPTEMBER 1985

BENIGN NERVE TUMORS OF THE UPPER EXTREMITY

DEAN S. LOUIS, M.D., AND FRED M. HANKIN, M.D.
Department of Surgery
University of Michigan Hospitals
Ann Arbor, Michigan

SOFT tissue tumors of the hand and upper extremity, except for ganglions, are infrequent in most surgical practices.[10,13,15] Benign tumors involving peripheral nerves of the upper extremity are therefore uncommon, and often present as diagnostic and treatment challenges. Recognition of the characteristic behavior of these tumors is an important step toward their successful management.

Neurofibromas and neurilemmomas are the most frequently encountered of these tumors. The exact incidence of these tumors, however, is unclear. Money[11] in 1950 surveyed 360,672 hospital admisssions over a 30-year period and reported three neurilemmomas involving the upper extremities, and only nine neurofibromas involving the limbs (these were not further classified into upper and lower extremity). Butler,[3] in a 1960 personal series of 437 hand tumors, found four neurofibromas and three neurilemmomas. Stack[15] in 1960 reported three neurofibromas and three neurilemmomas in a series of 300 hand tumors. Boyes[2] in 1970 reported a series of 394 hand tumors, in which he found 13 neurofibromas. Strickland and Steichen[18] in

1977 reported six tumors of nerve origin in a series of 689 hand and arm tumors. The incidence in their series was slightly greater than 1%. The exact diagnosis in all their cases was not revealed. Thus, not only are tumors of neural origin extremely rare but they represent less than 5% of all tumors involving the hand.[3,10,11]

We shall review benign nerve tumors involving the upper extremity and give examples of their clinical presentation from our 15 years of clinical experience to illustrate major points.

NEURILEMMOMA

Neurilemmomas are one of the two commonest tumors of nerve origin involving the upper extremity. Thought to arise from Schwann cells of the peripheral nerve,[16,17] they usually present as a solitary swelling along the course of the nerve. These lesions may be painful if bumped, but usually present as an asymptomatic enlargement of the functionally intact nerve. A neurilemmoma can usually be moved from side to side, or at 90 degrees to the axis or path of the peripheral nerve involved.[4,12] With careful surgical technique and magnification, a neurilemmoma can be removed from the nerve sheath proper without creating any neurological deficit. When a patient with a neurilemmoma presents with a neurological deficit, it is due to extrinsic compression of the tumor mass upon the peripheral nerve. It is unusual for neurilemmomas to exceed three centimeters in diameter (Figure 1). These tumors are most often solid and may vary in color, although they usually are yellowish-gray. Histologically, these tumors have a biphasic pattern with tightly packed spindle cells described as Antoni-A tissue, and with looser, less cellular patterns described as Antoni-B type cells (Figure 2). In some areas of these tumors, spindly cells may be arranged in palisades like a picket fence. These Verocay bodies are quite characteristic of the tumor.[5,16] Malignant degeneration has not been described in this tumor, and recurrence following appropriate extirpation is rare.

NEUROFIBROMA

Neurofibromas, like neurilemmomas, are thought to arise at least in part from Schwann cells, which have the capacity to act as collagen-producing facultative fibroblasts.[5] Neurofibromas, unlike neurilemmomas, are not always solitary, but may occur as multiple lesions and in association with von Recklinghausen's disease (Figure 3).[17] Neurofibromas, in contrast to neu-

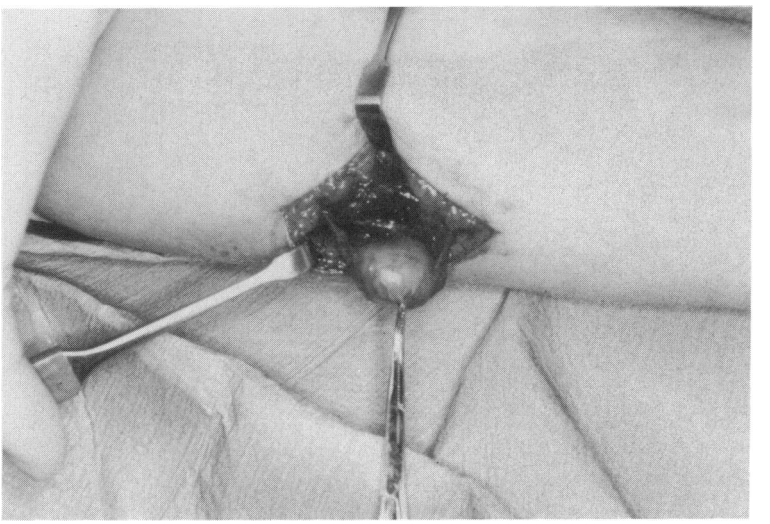

Fig. 1. This is the typical encapsulated appearance of a neurilemmoma with extensions of tissue at either end where it arises from the ulnar nerve. It was removed without creating any neurological deficit.

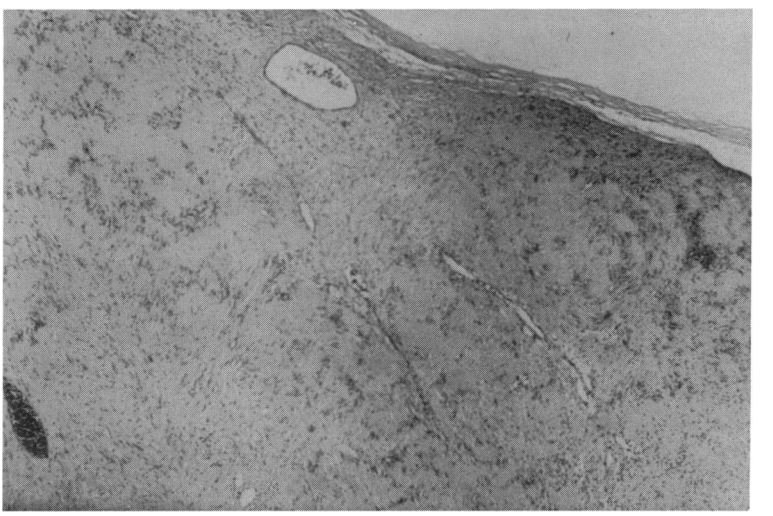

Fig. 2. The encapsulating sheath of this neurilemmoma can be seen in the upper right hand corner of this photomicrograph. Alternating, loosely arranged Antoni-B cells can be seen in the bottom right portion of this picture, whereas the densely compacted palisading spindle cells of the Antoni-A type can be seen just beneath the encapsulating sheath.

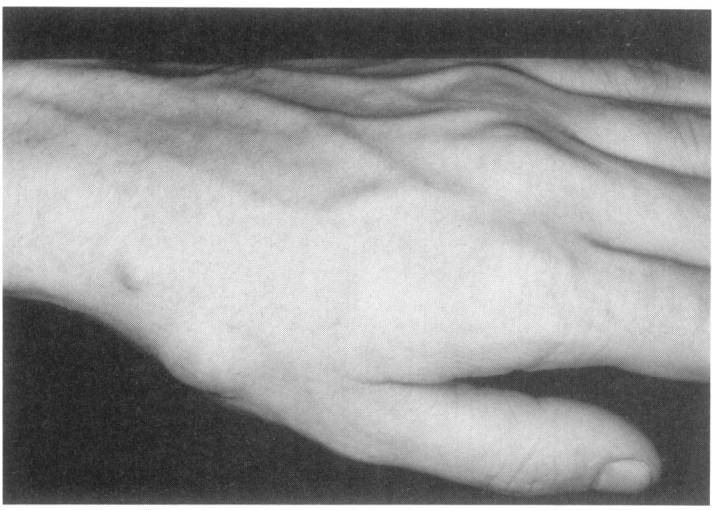

Fig. 3*a*. This is the rather characteristic appearance of neurofibromas occurring in a patient with von Recklinghausen's disease. Lesions involve tissue just beneath the skin as well as the skin itself.

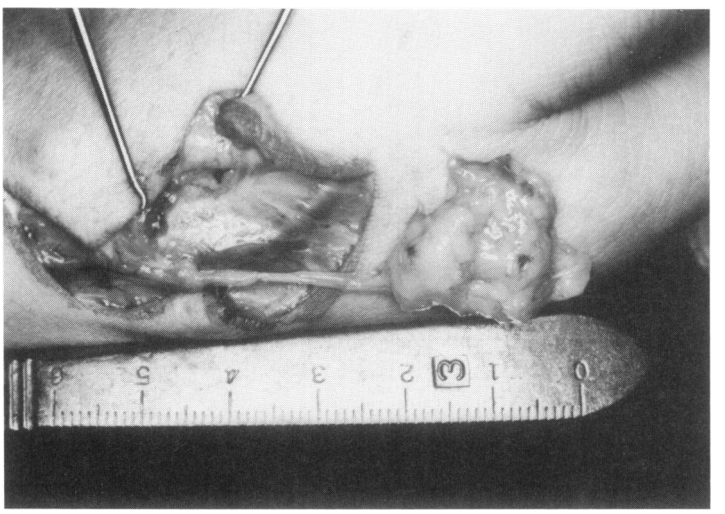

Fig. 3*b*. The dense involvement of this branch of the radial nerve is apparent. Excision of the tumor may create significant problems for the patient if a major branch of a peripheral nerve is involved (see text).

rilemmomas, are found within the substance of the nerve, and thus it is not possible to enucleate them from the fascicles. Therefore, if such a tumor is symptomatic, it may be necessary to excise it segmentally and to perform a nerve graft (Figures 4*a* and 4*b*). Such an operative decision may be of great clinical significance to the patient if a major peripheral nerve is involved. Microscopically, the histological pattern varies considerably, but the most common form is of spindly shaped Schwann cells intermixed with a collagenous matrix. There are, however, varieties that resemble the Antoni-A tissue, as seen in the neurilemmoma. Multiple variations are possible, depending somewhat on the site and the age of the lesion.[5] When a neurofibroma is associated with von Recklinghausen's disease, it may be necessary to excise the lesion if it becomes painful or rapidly increases in size. Enneking has stated that the incidence of malignant degeneration is 10% in patients with neurofibromatosis, but less than 1% in individuals with solitary neurofibromas.[4]

Benign Intraneural Tumors of Non-Neural Origin

Fibrofatty infiltrations—hamartoma of the median nerve. Since 1964 scattered reports have described this entity.[9,14,19,20] The median nerve at the wrist (Figure 5*a*) and in the palm is most frequently involved and becomes infiltrated by fibrous and fatty tissue. Histologically, there is thickening of the connective tissue supporting structures and infiltration of the nerve by fat (Figure 5B), which is not a normal constituent of neural tissue. The presentation is usually in childhood or young adult life, and symptoms are highly variable. The mass may be asymptomatic or motor and sensory abnormalities may be present. The currently recommended management for this rare tumor is decompression of the carpal tunnel and adjacent involved sites. The long-term follow-up of these patients is not clearly established at this point. They do not all do well. In one small series of patients, progressive deterioration in neurological function occured with the passage of time.[9] The use of microsurgical techniques has allowed the removal of some of this tissue as the fascicles have been separated from it. However, this is a tedious and potentially harmful procedure. There have been no reports of malignant degeneration in this tumor.

Intraneural hemangiomas. Intraneural hemangiomas have been reported at a variety of sites.[1,6,7,8] Figure 6 depicts a hemangioma being removed from within the substance of the ulnar nerve. These lesions are most baffling, in that they frequently do not present as discrete masses, but rather

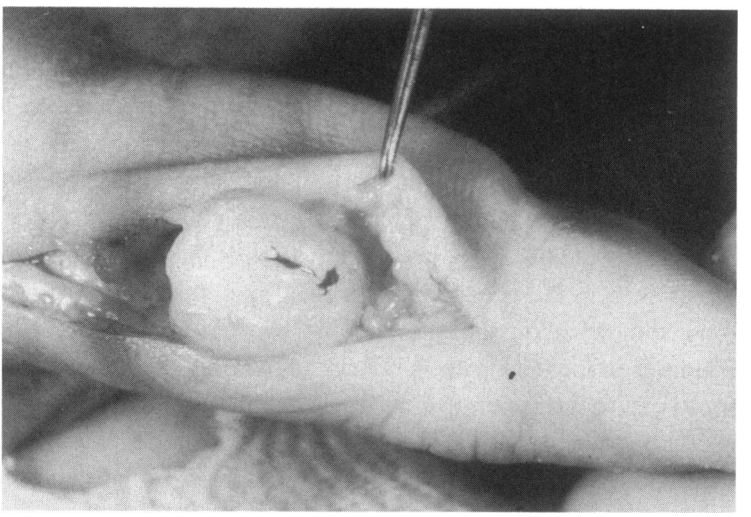

Fig. 4*a*. This neurofibroma involved the radial digital nerve on the index finger of this patient. The individual fascicles of the nerve could not be separated from tumor tissue and therefore the mass was removed and a nerve graft used to bridge the defect.

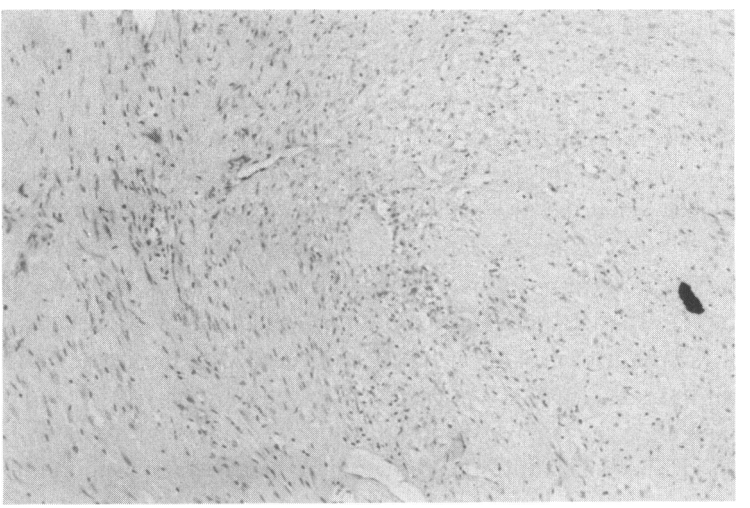

Fig. 4*b*. The dense collagenous matrix of a typical neurofibroma from the specimen removed in Figure 4*a* is seen quite well on the left hand side of this photomicrograph. The loose and myxomatous arrangement on the right hand side of this slide does, however, resemble the Antoni-B type cells seen in neurilemmomas.

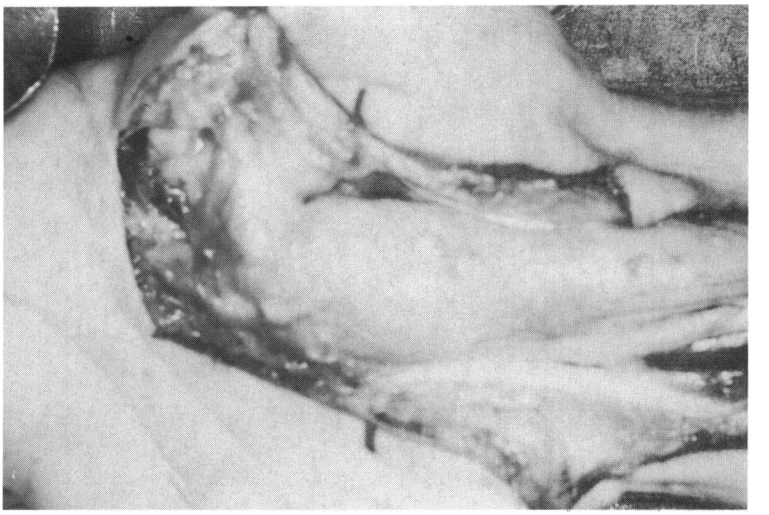

Fig. 5*a*. This clinical photograph demonstrates a lipofibroma of the median nerve. The thumb is at the top of the picture. There is massive enlargement of the nerve just at the base of the thenar muscles.

with obscure symptoms, such as pain interpreted as arising deep within the limb. Occasionally a localized swelling may be seen. Removal of the aberrant vascular tissue usually relieves the symptoms completely.

Lipomas. Figure 7 depicts an intraneural lipoma which presented as a mass in the wrist of a 35-year-old woman. This lesion was tender to palpation, and on percussion pain radiated along the median nerve distribution. Intraneural lipomas and lipofibromas are discrete, noninfiltrating and easily removed.[15]

Summary

Benign tumors involving peripheral nerves of the upper extremity are rare. They may arise from neural tissue, i.e., neurilemmomas and neurofibromas, or may consist of abnormalities of adjacent tissues, such as blood vessels, i.e., intraneural hemangiomas. Lipofibromas and lipomas are composed of tissues that are not constituents of normal nerves. Recognition and appreciation of these benign tumors is important in that radical treatment is not necessary, and in most cases function may be preserved or restored by appropriate surgery.

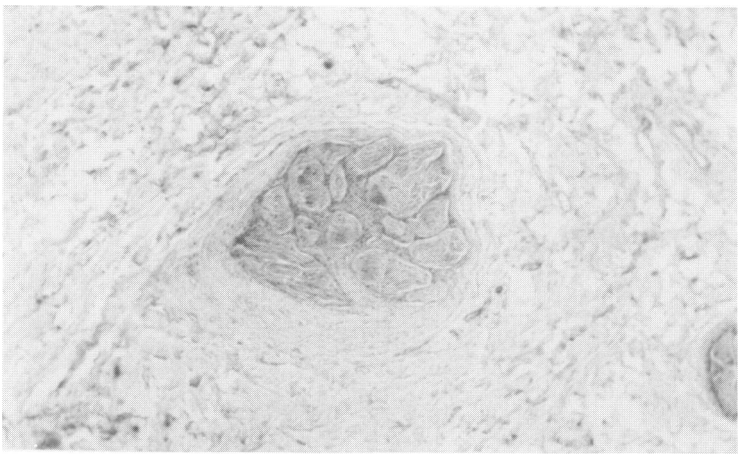

Fig. 5*b*. This photomicrograph is from the biopsy of the lesion seen in 5*a*. Enlargement of the supporting layers of the endoneurium and the perineurium are apparent in the central portion of this picture. The surrounding loose areolar tissue, composed of fibrous tissue and fat, is characteristic of this tumor.

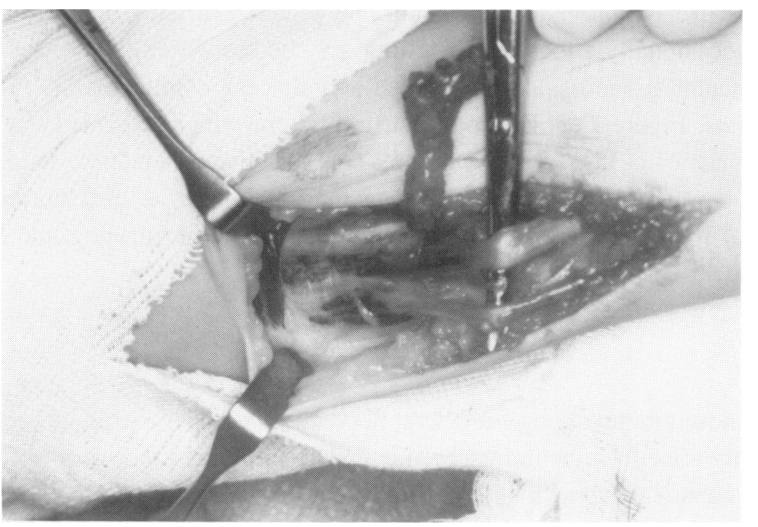

Fig. 6. An intraneural hemangioma involving the ulnar nerve at the wrist is seen in this figure. The ulnar nerve is lying over the hemostat in the right portion of the picture. Next to the hemostat, lying on the skin at the top of the picture, is the angiomatous tissue which has been delicately dissected from within the substance of the nerve. Additional hemangiomatous tissue may be seen between the two retractors on the left hand portion of the photograph.

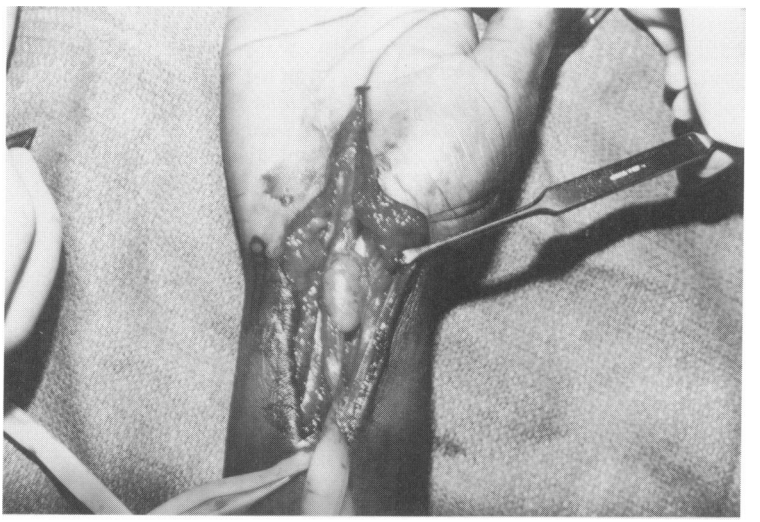

Fig. 7. A well encapsulated lipoma of the median nerve at the wrist is seen in this photograph. It was easily removed from within the substance of the nerve without creating any neurological deficit. Unlike the lipofibroma of the median nerve which is intertwined within the substance of the nerve and extremely difficult to remove, the lipoma can be easily shelled out.

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