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Surgical outcome of isolated benign peripheral nerve sheath tumors without neurofibromatosis

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Abstract

Background Benign peripheral nerve sheath tumors (BPNSTs) include schwannomas and neurofibromas. About 10% of soft tissue sarcomas are malignant peripheral nerve sheath tumors (MPNSTs), which are invasive and aggressive tumors. These can happen occasionally or after radiation exposure. Up to 90% of schwannomas are made up of differentiated neoplastic Schwann cells. Malignant transformation of schwannomas is rare.

Methods We collected the medical records of all patients (including their family histories), performed comprehensive physical and neurological assessments, and checked for the presence of a Tinel-like sign, as well as screening for neurofibromatosis (NF) signs. Magnetic resonance imaging (MRI), nerve conduction studies, and ultrasound were done for all cases.

Results We have operated on 21 patients with age range 29–52 years. The mean age was 39.4 years. 14 of these patients were females and 7 were males. The presenting symptoms were just swelling at nerve site in 14 patients, spontaneous pain at the nerve sites in 7 patients, and sensory deficit at the nerve distribution sites in 9 patients. The other 12 patients were sensory intact and only 7 patients had motor deficit. Postoperatively all patients had improved motor and sensory deficit and none of intact patients were worsened. Preoperatively we did MRI to show important nearby vascular structure anatomical abnormalities and we ordered nerve studies to all patients that showed abnormalities, which was only in 9 patients. Gross total resection was done in 18 patients and other 3 cases had partial resection to avoid sensory and motor deficits. The pathological analysis revealed 11 schwannomas and 10 neurofibromas. With 1 year follow up there was no recurrence in any patients.

Conclusion Benign peripheral nerve sheath tumours are safely resected without increased sensory and motor deficits after surgery and with improve clinical outcome with no recurrence on follow up.

Keywords Benign, Excision, Neurofibroma, Neurofibromatosis, Schwannoma

Introduction

Some soft tissue tumors known as peripheral nerve sheath tumors (PNSTs) originate from components of the peripheral nervous system, such as Schwann cells, and perineural cells [1]. Benign peripheral nerve sheath tumors (BPNSTs) include schwannomas and neurofibromas. About 10% of soft tissue sarcomas are malignant peripheral nerve sheath tumors (MPNSTs), which are invasive and aggressive tumors. These can happen

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occasionally or after radiation exposure [2, 3]. Up to 90% of schwannomas are made up of differentiated neoplastic Schwann cells. They impact individuals of all ages and are solitary and intermittent. These are slow-growing, encapsulated tumors that may exhibit cystic degeneration or calcifications [1, 4]. Malignant transformation of schwannomas is rare. Asymptomatic lesion of the skin or subcutaneous tissue of the head and neck, or in the flexor surfaces of the limbs is a common presentation of schwannomas [1]. Soft, skin-colored papules or tiny subcutaneous nodules are the outward manifestations of neurofibromas. Most typically, the tumor involves the epidermis; medium-sized nerves are affected less frequently. Cutaneous neurofibroma rarely causes pain and usually asymptomatic. About 8–13% of patients with neurofibromatosis type 1 (NF1) often develop MPNSTs, and they are typically linked to NF1, an autosomal dominant disorder [1, 4, 5]. Surgical resection remains the treatment of choice of BPNSTs because they may cause neural compromise, rapid growth, and pressure they apply to nearby structures. In asymptomatic cases, observation is accepted because the benign nature of the tumor [6]. However, to diagnose a symptomatic PNSTs we depend on clinical examination, ultrasound, and MRI scan. It is crucial to understand the anatomical relationship between healthy nerve fascicles and BPNST in order to remove the tumor completely, to reduce symptoms, and to maintain nerve function [7, 8]. Schwannomas grow inside the nerve, causing thickening of the nerve. As the tumor is encapsulated the nerve bundles do not penetrate it, but intertwist it, which allows enucleation of the tumor [9]. There is no studies that showed the surgical outcomes after excision of isolated BPNSTs without neurofibromatosis, especially in long follow up period. In our study we assessed the outcome including neurological deficits, and other complications of isolated benign peripheral nerve sheath tumors without neurofibromatosis, and the surgical outcome after excision.

Patients and methods

The aim of our study was to assess outcome including neurological deficits, and other complications of isolated peripheral nerve sheath tumors without neurofibromatosis and assess the surgical outcomes after excision. Our study is a retrospective case study which was conducted at Neurosurgery department of menoufia university hospitals and neurosurgery private clinics. Sample size was dependent on all patients with peripheral nerve sheath tumors who were treated surgically in 2019–2022 in neurosurgery department of menoufia university hospitals and were followed up for 6 months for recurrence rate. They will be 21 solitary peripheral nerve sheath patients. The data collection was obtained through a

self-designed questionnaire approved by specialists in the Neurosurgery Department of Menoufia University. We collected data from participants, who fulfill our eligibility criteria, and, in the setting, we chose, just after admission of the patient to Neurosurgery Department of Menoufia University Hospitals or Neurosurgery private clinics. Depending on the participant consent and after 4–6 weeks for incision inspection then at 3 and 6 months from the surgery. Preoperatively we obtained the medical histories of all patients (including their relatives), conducted thorough physical and neurological examinations, and checked for a Tinel-like sign and NF signs [10] before surgery. MRI, (Fig. 1) nerve conduction studies, and ultrasound were performed for all cases. We usually recommended excisional biopsy to remove lesions that appeared benign on imaging if they were symptomatic or grew over time. We used nerve conduction to locate the nerve fascicles displaced by the tumor and to confirm the continuity of the original nerve fascicle. Statistical analysis was performed with SPSS version 11.0 statistic software package.

Surgical technique

The surgical technique for removing schwannomas and neurofibromas, the most common benign PNSTs, depends on how they relate to the nerve fascicles (Fig. 2). Schwannomas originate from a single fascicle and displace the others around the capsule (Fig. 3). Neurofibromas involve multiple fascicles that intermingle with the tumor. Schwannoma, the tumor is exposed by a long incision along the nerve and the displaced fascicles are separated from the capsule with blunt instruments. The tumor is then isolated from its parent fascicle at both ends and excised (Fig. 4). Alternatively, for large tumors, the capsule can be opened, and the tumor contents can be removed first, followed by the capsule itself. In neurofibroma, the incision and exposure are similar to schwannomas, but the dissection of the tumor from the fascicles is more meticulous and careful. The patients are evaluated after 4–6 weeks for wound healing and after 6 months for tumor recurrence. Donner et al. [11] published an article in 1994, where they first reported that minimal deficits could result from excising most neurofibromas, if the surgery is done along the fascicles and the nerve anatomy is identified at both ends of the tumor. Unlike the schwannoma, the neurofibroma affects some of the nerve's fascicular structure. The entry and exit sites of the tumor usually have more than one fascicle, and these are typically larger than those in schwannomas. The tumor is removed by separating, moving, and splitting the tumor capsule from the adjacent and sometimes attached nerves, while preserving the tumor capsule (Figs. 5, 6). The fascicular pattern at both ends of



Fig. 1 MRI showing benign peripheral nerve sheath tumour of radial nerve



Fig. 2 Swelling appearing on median nerve site

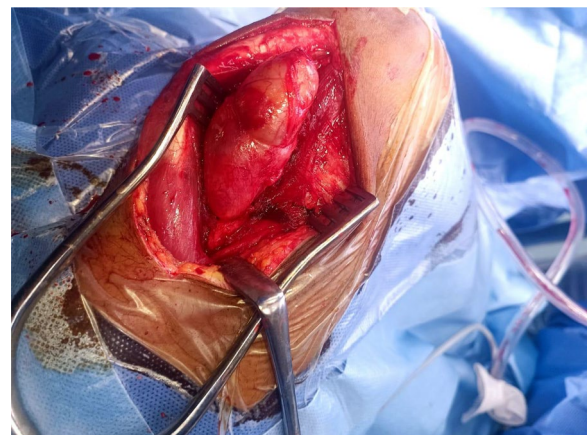


Fig. 3 Schwannomas originate from radial nerve a single fascicle and displace the others around the capsule

the tumor is then determined, and the mass is gradually peeled off from the central part by working from one end to the other alternately.

Results

We have operated on 21 patients with the age range 29–52 years. The mean age was 39.4 years. 14 (67%) of these patients were females and 7(33%) were males. Nerve distribution sites are shown in Table 1. The presenting symptoms were just swelling at the nerve sites in 14 (67%) patients, spontaneous pain at the nerve sites in 7 (33%) patients, and sensory deficit at the nerve distribution sites in 9 (43%) patients. The other 12 (57%) patients were sensory intact. Only 7 (33%) patients had motor

deficit. Postoperatively all patients had improved motor and sensory deficit and none of the intact patients were worsened. Preoperatively we ordered an MRI scan to show anatomical abnormalities of important nearby vascular structure. We ordered nerve studies to all patients that showed abnormalities and they were only 9 (43%) patients. Unfortunately, we have no nerve stimulator in our facility, therefore we did not use it. Gross total resection was done in 18 (86%) patients and the other 3 (14%) cases had partial resection to avoid sensory and motor deficits. The pathological analysis revealed 11 (52%) schwannomas and 10 (48%) neurofibromas. With 1 year follow up there was no recurrence in any patients.

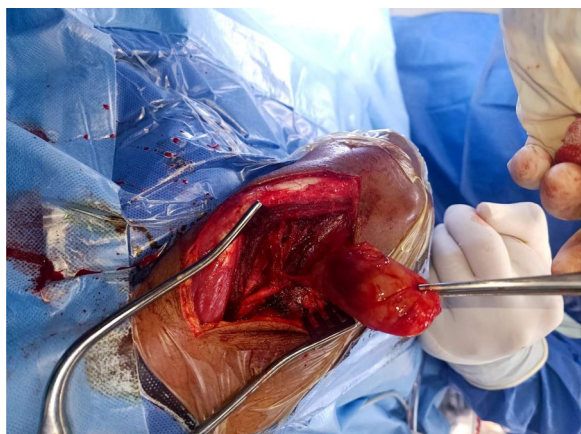


Fig. 4 The tumor is then isolated from its parent fascicle at both ends and excised

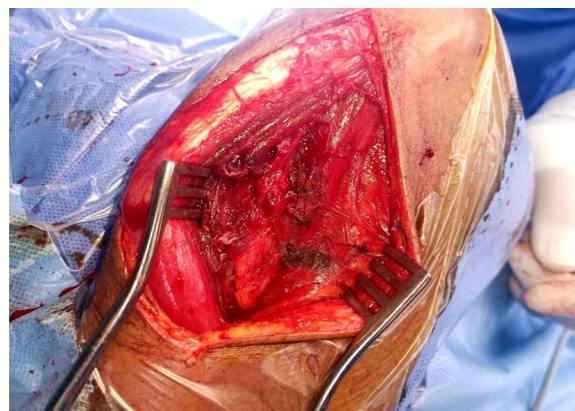


Fig. 6 Preserving nerve fibres after dissection of the tumour from radial nerve



Fig. 5 Preserving nerve fibres after dissection of the tumour from median nerve

Discussion

In this study, the number of female patients was twice the number of male patients. Although in literature there was no sexual predominance [12].

Schwannomas are benign tumors that arise from the nerve sheath and typically grow slowly without causing pain. However, if they involve large nerves, they may cause discomfort or pain. On gross examination, schwannomas are small (usually < 5 cm), well-defined, and encapsulated by the epineurium. The shape of the

Table 1 Distribution of the tumours nerve sites

Nerve site	Count of nerve site
Tibial	4
Radial	4
Median	3
Ulnar	3
Sciatic	3
Femoral	2
Common peroneal	2
(Blank)	
Grand Total	21

tumor depends on the size of the nerve it originates from. If the nerve is small, the tumor is spindle-shaped and may resemble a neurofibroma; if the nerve is large, the tumor is more spherical [13].

In similar study, patients who had previously underwent attempted resections and preoperative biopsy had a significantly increased risk (41%) for developing postoperative neurologic deficits when compared with patients who presented with de novo tumors (15%) [14]. In our study all patients with deficits had improved outcomes after surgery.

On previous study, gross total resection (GTR) was achieved in 85% of patients. [15]. This was the same result in our study. We avoided GTR in some cases to prevent sensory and motor deficits.

Benign histology characterizes schwannomas and neurofibromas, but they can recur locally. The literature reported recurrence rates for BPNSTs between 1.3 and 35.9% [16–20]. A precise diagnosis is crucial for the treatment of any tumor. Sondack and Chang [21] emphasize that a well-executed biopsy is an essential

first step in a multimodality approach to treatment, as poorly done biopsy can worsen the patient's condition.

There was no recurrence in our study after 1 year follow up. The recurrence rate is very low in other similar studies [22–24].

Geisinger [25] has explained the limitations of fine-needle aspiration biopsy of soft-tissue tumors in a clear way. The sample may not reflect the tumor and be inadequate to make a diagnosis.

A schwannoma is a type of benign tumor that grows from the nerve sheath. When it occurs in the arms or legs, it usually has some typical features. The tumor forms a lump that can be moved sideways, but not up and down along the limb. It is tender to touch and causes tingling sensations in the nerve territory when tapped, similar to Tinel's sign. This sign is very helpful for identifying a schwannoma. The nerve function is usually preserved unless the tumor is in a tight space. A painful nerve lump with worsening nerve damage is more likely to be a cancer than a schwannoma. [26].

MRI proved to be very useful for us. [27] The schwannoma appears as a capsule-like structure that is off-center from the nerve axis and pushes the fascicles to one side. We believe that the heterogenous appearance reflects the mixed tissue composition of the schwannoma and the presence of cysts. However, Moser and Parrish [28] warn that radiological features are not reliable indicators of benignity or malignancy of soft-tissue tumors in most cases. This crucial limitation should be strongly emphasized. They further state that plain radiography should be correlated with cross-sectional imaging before making a final interpretation of CT or MR findings. Filler et al. [29] described magnetic resonance neurography as "tissue selected imaging aimed at identifying and evaluating characteristics of nerve morphology, internal fascicular pattern, longitudinal variations in signal intensity and caliber, and connections and relations to other nerves or plexuses".

Ultrasonography became more refined and can help with better diagnosis. Martinoli et al. [30] showed a difference between lesions from outside or inside the nerve, and between tumors and neuromas. Chan [31] used ultrasound to show the internal structure of the nerve in a remarkable way.

Schwannomas grow outside the nerve bundles from which they originate, but neurofibromas are mixed with many nerve bundles and are harder to remove completely, regardless of how easy they are to reach. This could be a reason for this result [26].

Nerve trunks in the limbs can be clinically diagnosed easily and the optimal treatment is to excise the tumor while maintaining nerve function. Although, biopsy is necessary in cases where the diagnosis is uncertain.

Conclusion

Benign peripheral nerve sheath tumors are safely resected without increased sensory and motor deficits after surgery and with improved clinical outcome with no recurrence on follow up.

Abbreviations

NF	Neurofibromatosis
NFI	Neurofibromatosis Type I
PNSTs	Peripheral nerve sheath tumors
BPNSTs	Benign peripheral nerve sheath tumors
MPNSTs	Malignant peripheral nerve sheath tumors
MRI	Magnetic resonance imaging
GTR	Gross total resection

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Not applicable.

IRB approval

We take an approval from the IRB Committee of Faculty of Medicine of Menoufia University. Approved Number: 1/2024 NEUS 9-2.

Author contributions

MN: writing, reviewing, and editing, supervision, project administration. MD: validation, writing, original draft preparation. AS: conceptualization, methodology, software, visualization. HA: resources, investigation. All authors read and approved the final manuscript.

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Availability of data and materials

The data that support the findings of the study are available from Menoufia University Hospitals.

Declarations

Ethics approval and consent to participate

Approval was taken from the Institutional Review Boards (IRB) of the Menoufia faculty of medicine. IRB approval number and date is 1/2024 NEUS 9/2. Add to this consent was taken from neurosurgery Department of Menoufia University and an informed Consent was taken in which each participant has been informed of all aspects of the study and have the right to give up as they wanted. All data are confidential and were used only for the purpose of the study. The principal investigator and the co-investigators will be committed to the ethical principles that have their origin in the "Declaration of Helsinki".

Consent for publication

Informed consent was taken from all participants who agreed to participate and publish.

Competing interests

The authors declare that they have no competing interests.

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