Extensive multiple schwannomas of the median nerve: a rare case report

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ABSTRACT

Background: Schwannomas are benign peripheral nerve sheath tumors arising from Schwann cells and are uncommon in the upper limb. Median nerve schwannomas constitute approximately 0.1%-0.3% of all hand tumors, and extensive involvement of the median nerve by multiple schwannomas across multiple joints is exceptionally rare. Differentiating schwannoma from neurofibroma on imaging can be challenging.

Case Presentation: A patient presented with a painful palpable mass along the flexor aspect of the right forearm extending from the elbow to the distal interphalangeal joint. Clinical examination demonstrated a mobile mass with a positive Tinel's sign along the median nerve distribution. Magnetic resonance imaging revealed a longitudinally extensive lesion along the median nerve, but could not reliably differentiate a schwannoma from a neurofibroma. A core needle biopsy confirmed schwannoma with Antoni A and Antoni B areas and strong S-100 positivity. Complete surgical excision with preservation of the median nerve was performed, resulting in excellent functional recovery.

Conclusion: Extensive schwannomas involving the median nerve are rare and may present diagnostic challenges. Histopathological confirmation combined with meticulous nerve-preserving surgical excision allows accurate diagnosis and excellent functional outcomes.

Keywords: Median nerve, schwannoma, peripheral nerve sheath tumor, multiple schwannomas, surgical excision.

Type of Article: CASE REPORT **Specialty:** Oncology

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Key clinical message

A definitive diagnosis of schwannoma through preoperative core biopsy is essential for determining the appropriate surgical approach. Although complete tumor excision is the goal, careful nerve-preserving techniques are crucial to maintain neural integrity and function. The presence of extensive involvement of the median nerve should not preclude limb-salvage surgery aimed at preserving function.

Background

Schwannomas are benign peripheral nerve sheath tumors arising from Schwann cells and represent the most common benign tumors of peripheral nerves [1]. They are typically slow-growing, encapsulated lesions that arise eccentrically from the parent nerve and are usually solitary [2]. Although schwannomas may occur along any peripheral nerve, involvement of the median nerve is relatively uncommon, accounting for a small proportion of tumors affecting the hand and forearm.

Clinically, these tumors often present as slowly enlarging masses and may be associated with pain, paresthesia, or a positive Tinel's sign due to nerve compression. Radiological imaging, such as magnetic resonance imaging (MRI), helps delineate tumor extent and its relationship with surrounding structures; however, distinguishing schwannomas from other peripheral nerve sheath tumors, particularly neurofibromas, can be challenging [3].

We report a rare case of multiple schwannomas arising along the median nerve with extensive involvement from the elbow to the distal interphalangeal joint, successfully managed with nerve-preserving surgical excision.

Case Presentation

A 17-year-old girl presented with swelling in the flexor aspect of the right forearm for 2 years, associated with numbness and paresthesia for the last 2 months. There was no history of trauma or prior neurological disease. The patient had no personal or family history of neurofibromatosis (NF) or schwannomatosis.

Clinical examination

Two contiguous nodular fusiform swellings measuring approximately 14 × 4 cm were present along the flexor aspect of the forearm, extending from the antecubital fossa to the wrist and continuing to the palmar aspect of the ring finger. The masses were firm, mildly tender, and showed restricted mobility along the longitudinal axis. Percussion over the swelling elicited tingling in the median nerve distribution (positive Tinel's sign). Radial and ulnar arteries were palpable. No cutaneous stigmata of NF were observed.

Diagnostic assessment

Imaging

MRI demonstrated a well-defined, elongated, multilobulated lesion along the median nerve involving the

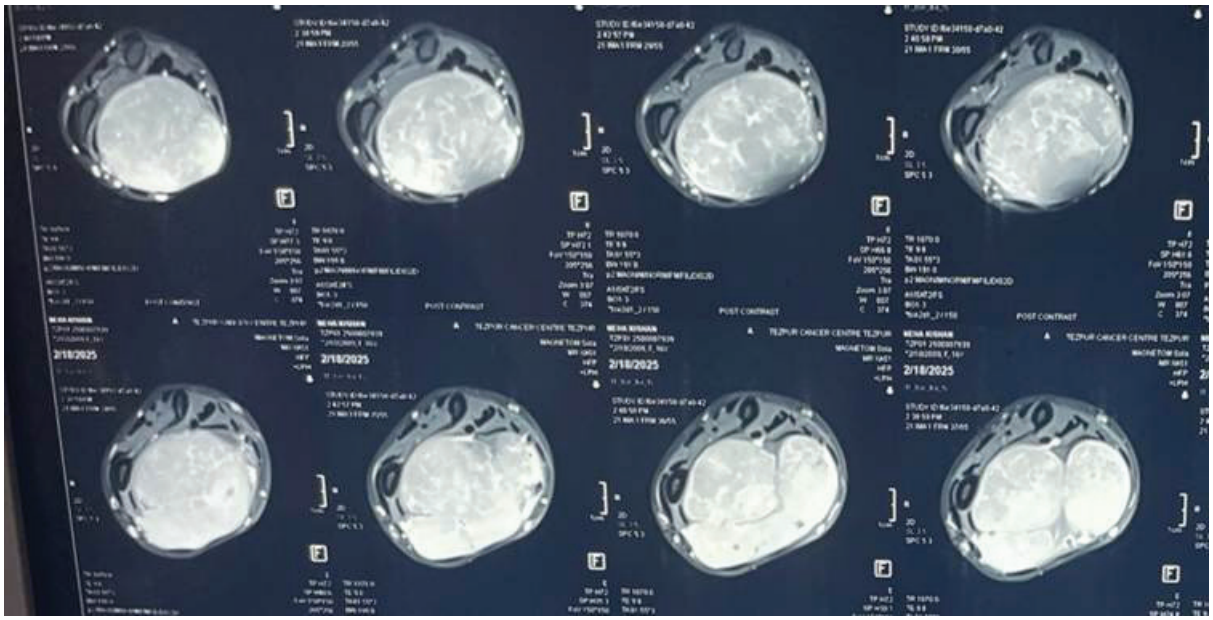

Figure 1. *MRI of the right forearm demonstrating a well-defined, elongated, multilobulated lesion along the course of the median nerve within the flexor compartment.*

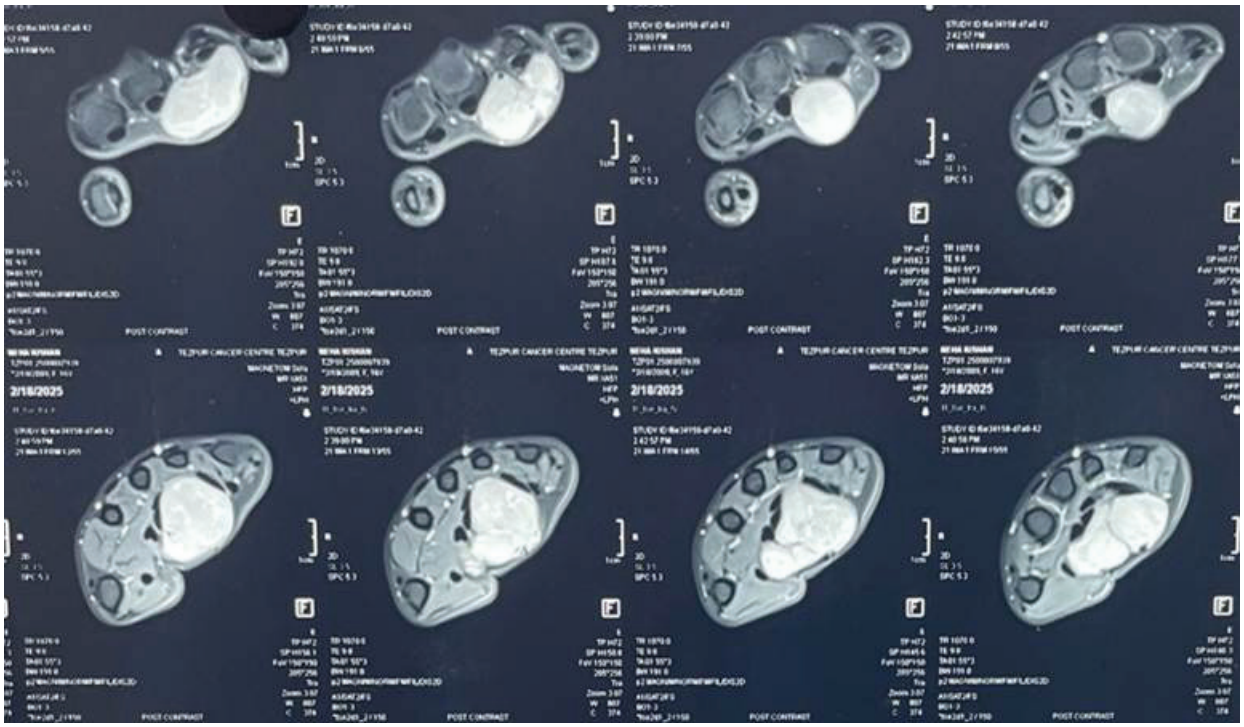

Figure 2. *MRI of the wrist and hand showing distal extension of the lesion along the median nerve into the palmar aspect of the hand.*

flexor compartment of the forearm with extension to the wrist and palm. The lesion appeared hypointense on T1-weighted images and hyperintense on T2-weighted images with areas of cystic degeneration (Figures 1 and 2).

Histopathological examination

Core needle biopsy revealed spindle cells arranged in palisading patterns. Immunohistochemistry (**IHC**) demonstrated strong **S-100 positivity** with a Ki-67 index <5%, confirming schwannoma.

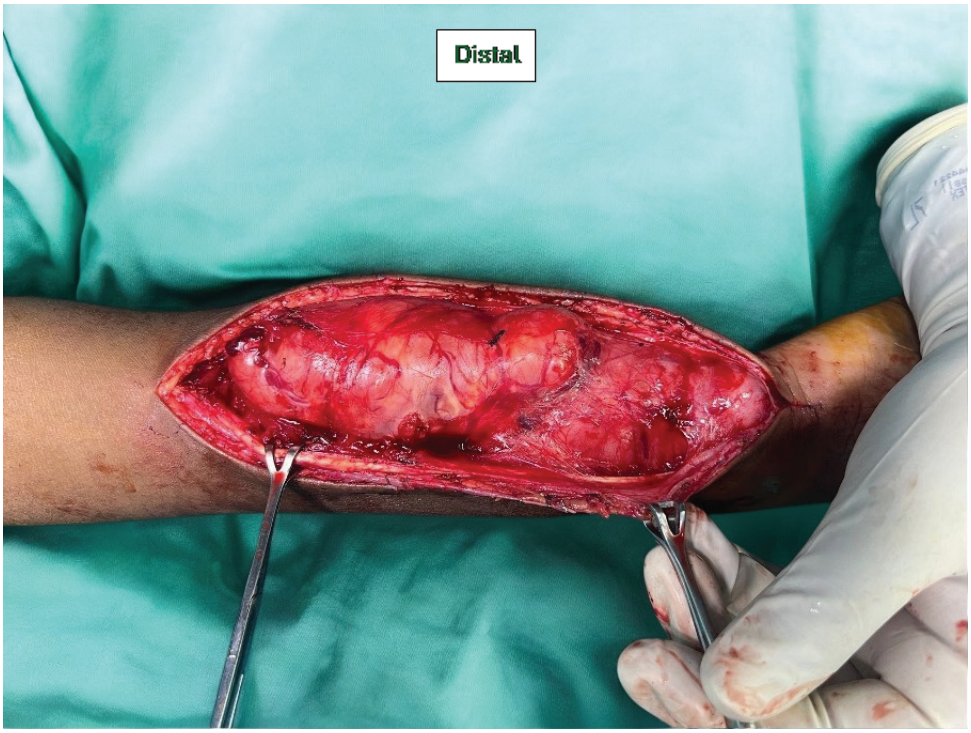

Figure 3. *Intraoperative photograph demonstrating exposure of the tumor beneath the flexor tendons within the flexor compartment of the forearm.*

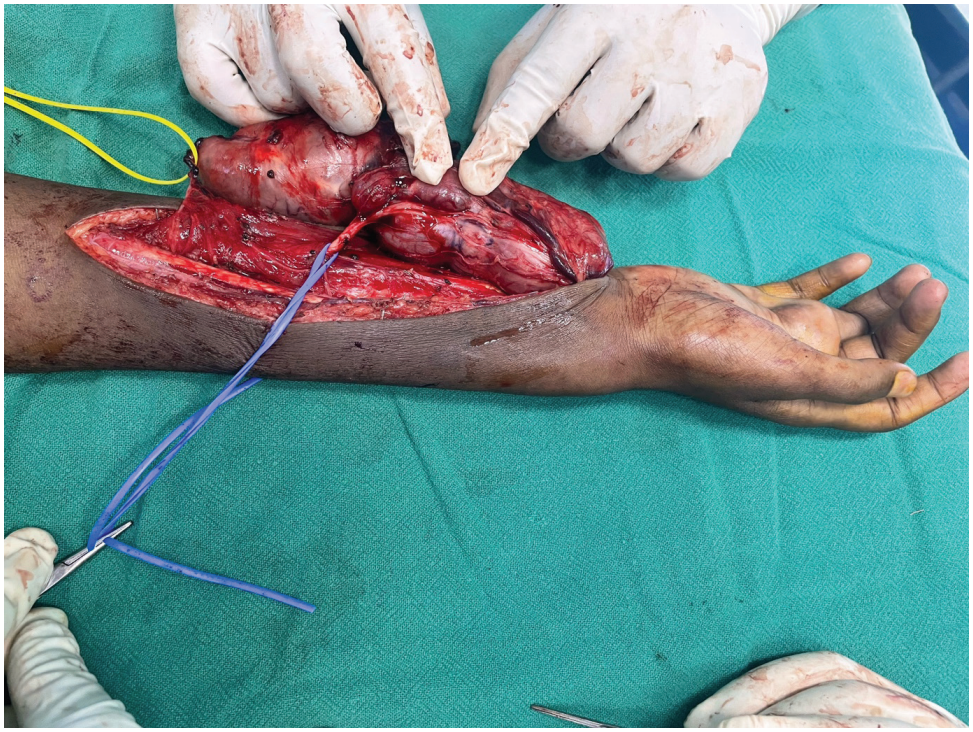

Figure 4. *Intraoperative view showing the tumor arising eccentrically from the median nerve with displacement of surrounding nerve fascicles.*

Treatment and intervention

Surgical procedure

The patient underwent complete surgical excision of the tumor with preservation of the median nerve. The operation involved careful dissection under 2.5× magnification using a surgical loupe. A longitudinal incision had been centered over the tumor bulk, extending from the elbow to the wrist. The incision was deepened to the deep fascia. A plane was created between the flexor carpi radialis and the palmaris longus muscles (Figure 3). The median nerve was identified below the radial attachment of the flexor

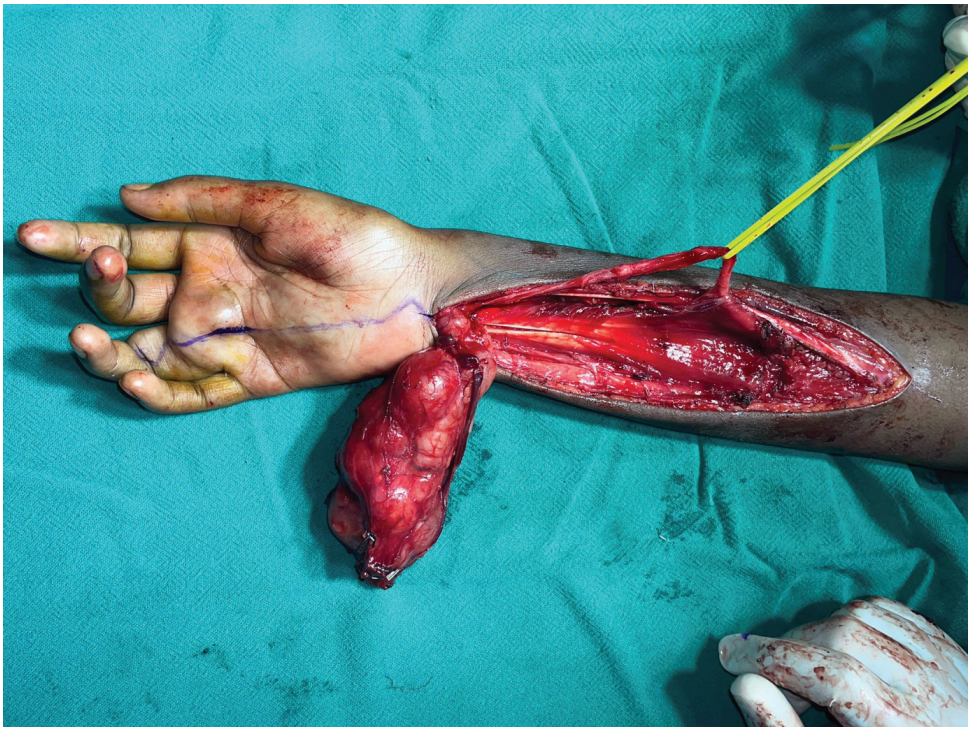

Figure 5. *Intraoperative dissection of the wrist and palm demonstrating extension of the tumor along the median nerve with preservation of adjacent neurovascular structures.*

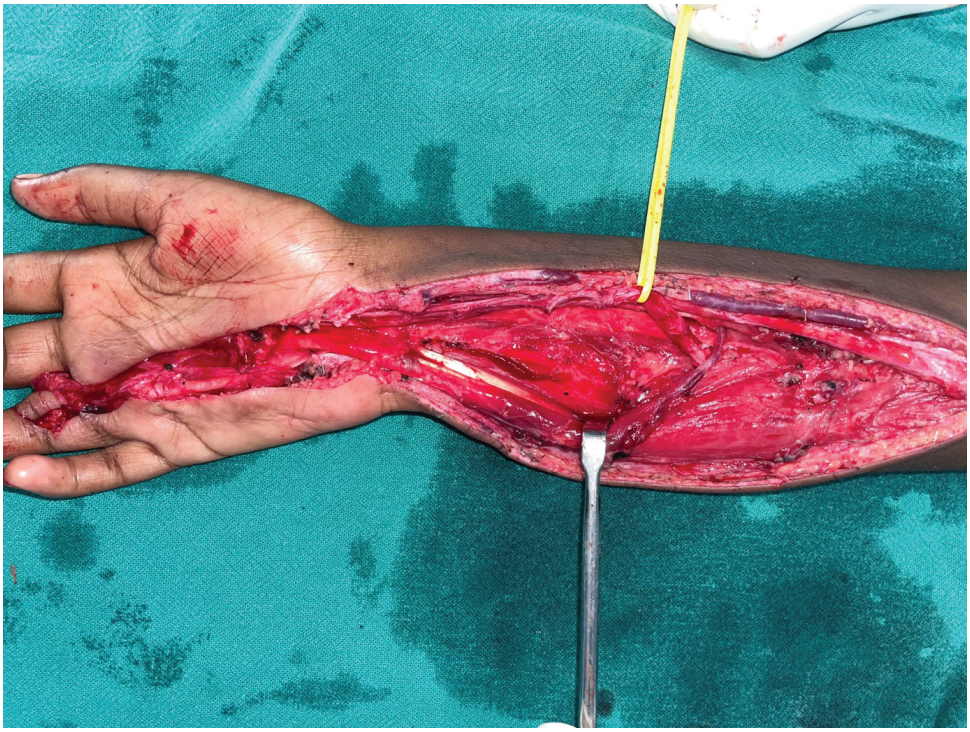

Figure 6. *Final intraoperative view after tumor excision showing intact median nerve branches with preservation of neural continuity.*

digitorum superficialis. The tumor was firmly adherent to the median nerve and had an eccentric location (Figure 4). Using fine scissors, the soft tumor mass was separated from the surrounding nerve fascicles as well as median nerve branches (anterior interosseous nerve as well as muscle branches) after the epineurium was cut longitudinally (Figure 5). The incision was later extended over the wrist. Meticulous dissection was performed without the use of a tourniquet. We believe that identifying minute vessels is better done this way, which is important for

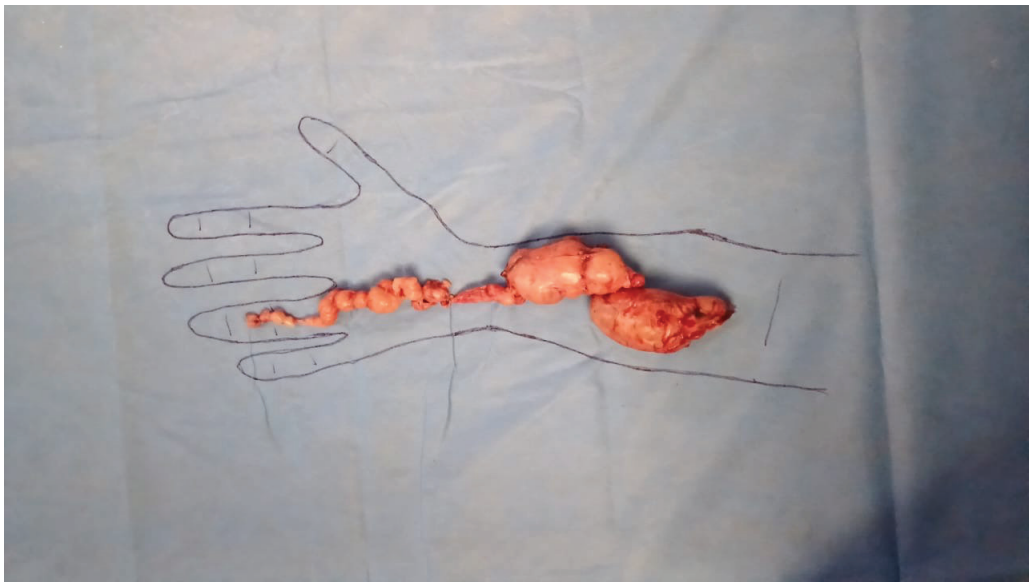

Figure 7. *Gross specimen of the excised tumor demonstrating encapsulated nodular schwannomas removed from the median nerve.*

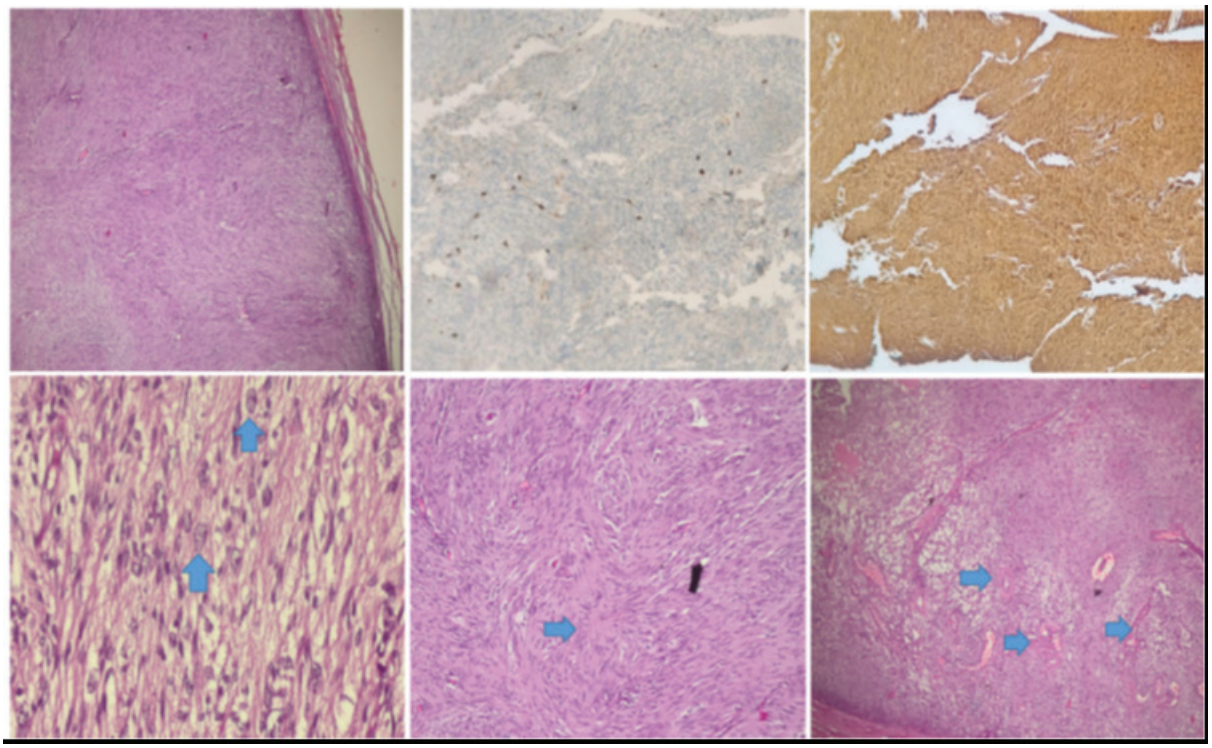

Figure 8. *Pre- and post-op histopathological images. (A). Low-power scanner view (×4) demonstrating an encapsulated, well-circumscribed tumor composed of spindle cells arranged in intersecting fascicles (upper left). (B). Immunohistochemical staining showing a low Ki-67 proliferation index (~5%) despite the presence of occasional bizarre nuclei (upper middle). (C). IHC demonstrating diffuse S-100 positivity in the tumor cells, supporting the diagnosis of schwannoma (upper right). (D). High-power view (×40) demonstrating areas of ancient change characterized by scattered atypical and bizarre nuclei (blue arrows), representing degenerative changes (lower left). (E). Low-power view (×10) showing intersecting fascicles of spindle cells with formation of a Verocay body (blue arrow), characterized by nuclear palisading and an intervening anuclear fibrillary zone (lower middle). (F). Low-power view (×10) showing numerous hyalinized vessels within the tumor, a characteristic feature frequently observed in schwannomas (lower right).*

preserving vasa nervosa. Staged incisions were made on the palmar aspect of the hand, extending to the ring finger in a zig-zag pattern. The flexor retinaculum was cut with a blade, and the median nerve was identified as it travelled posteriorly toward the middle finger as well as the lateral aspect of the ring finger. Posteriorly, below the flexor retinaculum's attachment on the wrist's medial aspect, the ulnar nerve, as well as the ulnar artery, were identified in Guyon's canal and preserved. The tumor did not arise from the ulnar nerve fibers, although its extension was toward the medial aspect of the ring finger. The tumor was completely removed, preserving all nerve fibers to the ring finger (Figures 6 and 7). The wound was closed in layers, and a 14 Fr suction drain was placed, which was removed on postoperative day 1.

Outcome

In the immediate postoperative period, on-table assessment following reversal of neuromuscular blockade with vecuronium demonstrated an optimal functional outcome, with improvement in the preoperative deficits likely caused by nerve compression (Supplementary 1 video).
Video link: https://youtube.com/shorts/fM07lHBpFpI

Final histopathology report

Grossly, there was an encapsulated white-grey soft tissue mass with yellow areas. Microscopically, the section shows an encapsulated tumor composed of compact hypercellular areas of spindle cell fascicles with ill-defined cytoplasm. Occasional areas of ancient change with atypical cells and blood vessels with thickened hyalinized walls were noted. This suggests a cellular schwannoma with focal ancient change (Figure 8).

Postoperative care

The patient was discharged on postoperative day 1. She received physical rehabilitation on an outpatient basis in our dedicated oncorehab unit.

Follow-up and outcomes

Short-term outcomes

A short-term follow-up was conducted 2 weeks after surgery, during which staplers and sutures were removed. Sensory neural functions were not impaired.

Long-term follow-up is planned with periodic clinical examination to monitor for recurrence and assess neurological function. Although recurrence after complete excision of a schwannoma is uncommon, continued surveillance is recommended.

Discussion

Schwannomas are benign peripheral nerve sheath tumors that typically occur as solitary lesions. Extensive involvement of the median nerve by multiple schwannomas spanning several anatomical compartments is exceptionally rare and poses unique diagnostic and surgical challenges [4,5].

The present case demonstrates an unusually long-segment involvement of the median nerve extending from the elbow to the distal interphalangeal joint. Most reported cases describe isolated schwannomas located at the wrist or forearm, whereas extensive lesions involving multiple compartments of the upper limb are rarely described [6]. Although the presence of multiple schwannomas has historically been referred to as schwannomatosis, current classification systems define schwannomatosis as a distinct entity within the NF spectrum requiring specific clinical or genetic criteria. In our patient, the lesions were confined to the median nerve without evidence of systemic disease or genetic confirmation; she does not fulfil the updated 2022 criteria for NF-2-related schwannomatosis as described by Plotkin et al. [7] Therefore, this presentation is more appropriately interpreted as multiple schwannomas arising along the median nerve rather than true schwannomatosis.

Preoperative diagnosis of peripheral nerve sheath tumors can be challenging. Clinically, these lesions may present as slowly enlarging masses associated with pain, paresthesia, or a positive Tinel's sign. Imaging modalities such as MRI are useful for demonstrating tumor extent and its relationship to adjacent structures, although distinguishing schwannomas from neurofibromas solely on imaging may be difficult [8]. Histologically, schwannomas are characterized by a well-defined capsule derived from epineurium and demonstrate alternating Antoni A and Antoni B patterns, with strong S-100 immunoreactivity [9].

From a surgical perspective, schwannomas can usually be excised while preserving the parent nerve because they arise eccentrically from the nerve sheath. Careful identification of proximal and distal nerve segments, magnified dissection, and meticulous separation of the tumor capsule from the splayed nerve fascicles are essential steps to avoid neurological injury. Recurrence following complete excision is uncommon, and most patients experience significant symptomatic improvement with preservation of nerve function. Das et al. [10] reported improvement of preoperative symptoms with maintained nerve function in approximately 89% of patients undergoing surgical excision of schwannomas.

Conclusion

Complete surgical excision with preservation of the parent nerve remains the treatment of choice for median nerve schwannomas. Careful microsurgical dissection allows tumor removal while maintaining neural integrity and functional outcome. Even in cases with extensive involvement of the median nerve, nerve-preserving surgery can achieve excellent symptomatic and functional recovery.

Supplementary Material

Video available here:
https://youtube.com/shorts/xWtiu_vde9w

What's new?

The authors describe an unusual case of extensive multiple schwannomas involving the median nerve, extending from the elbow to the distal interphalangeal joint across several anatomical compartments. The case highlights the diagnostic challenges, the role of preoperative tissue diagnosis, and the feasibility of meticulous nerve-preserving surgical excision resulting in excellent functional recovery.

List of Abbreviations

IHC	Immunohistochemistry
MRI	Magnetic resonance imaging
NF	Neurofibromatosis

Conflict of interest

The authors declare that they have no conflicts of interest.

Funding

This research did not receive any specific grants from funding agencies in the public, commercial, or not-for-profit sectors.

Consent for publication

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Ethical approval

Patient perspective

The patient reported significant relief of symptoms following surgery and was satisfied with the cosmetic and functional outcomes.

Data availability

Data will be available on request due to privacy/ethical restrictions. The data that support the findings of this study are available on request from the corresponding author. The data are not publicly available due to privacy or ethical restrictions.

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Summary of case

1	Patient (gender, age)	17-year-old female
2	Final diagnosis	Schwannoma
3	Symptoms	Numbness and paraesthesia
4	Medications	None (managed surgically)
5	Clinical procedure	Surgical excision of the mass
6	Specialty	Oncology