Multiple Schwannomas of the Median Nerve

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Abstract

Schwannomas originating from the Schwann cells of the nerve sheath, typically manifest as solitary, encapsulated outgrowths. These types of benign tumors are found in the head and neck region, the upper and lower extremities. Schwannomas usually present as solitary peripheral nerve tumors but they can also occur as multiple contiguous lesions in conditions like neurofibromatosis type 2-related schwannomatosis. However, there are very few reports in the literature about the involvement of a single nerve by multiple schwannomas without having any systemic pathology.

This case report discusses a rare instance of multiple schwannomas affecting the median nerve in a 30-year-old male. The patient presented with a 4-year history of a painful mass and intermittent paresthesia in his thumb, index, and middle fingers. Clinical examination revealed a lobulated mass along the median nerve, confirmed by magnetic resonance imaging (MRI), which showed multiple multilobulated lesions encasing the nerve. Microsurgical excision was performed, with histopathological analysis confirming the diagnosis.

Postoperatively, the patient experienced complete symptom resolution, and no recurrence was noted at the 18-month follow-up.

This case highlights the diagnostic and therapeutic challenges of managing multiple schwannomas in the absence of systemic pathology. It underscores the importance of considering such occurrences in patients with peripheral nerve tumors and demonstrates the value of MRI for preoperative assessment and planning.

Keywords

- schwannoma
- median nerve
- peripheral nerve tumor

Introduction

Schwannomas or neurilemomas, originating from the Schwann cells of the nerve sheath, typically manifest as solitary, encapsulated outgrowths.[1] These types of benign tumors are found in the head and neck region, the upper and lower extremities.[1,2] Although benign peripheral nerve tumors of the upper and lower extremities are extremely uncommon,[3] schwannomas constitute one of the major types.[2,4,5] In the extremities, the ulnar and peroneal nerves are more commonly affected than the median nerve. The incidence reported for solitary median nerve schwannoma is around 5% whereas for multiple lesions, it is still not reported in the literature.[5–8]

Schwannomas usually present as solitary peripheral nerve tumors but they can also occur as multiple contiguous lesions in conditions like neurofibromatosis type 2 (NF2)-related schwannomatosis. However, there are very few reports in the literature about the involvement of a single nerve by multiple schwannomas without having any systemic pathology.[9]

In this article, we report a case of median nerve schwannomatosis, describing the clinical presentation, imaging,

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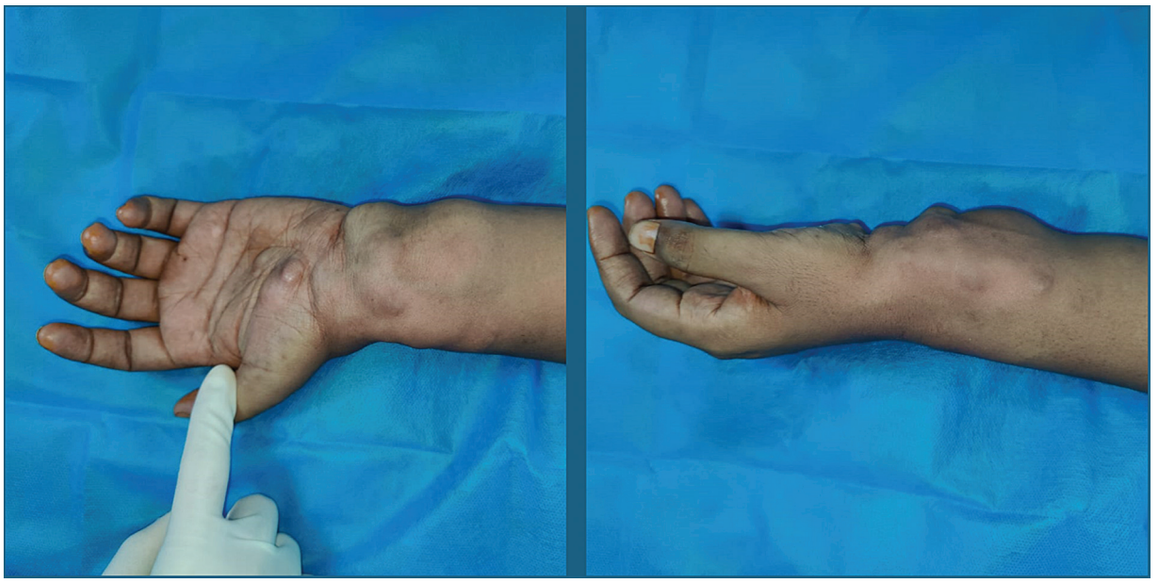

Fig. 1 Clinical preoperative images showing a grossly enlarged, lobulated, firm mass noted on the volar aspect of the distal forearm and proximal hand.

histological findings, surgical outcomes, and functional results. We also discuss the diagnostic and treatment challenges associated with these tumors and the rationale behind the approach.

Case Report

A 30-year-old, right-handed male, advocate by profession, presented with a history of a painful mass in the volar aspect of his left forearm and proximal hand for over 4 years. He presented with complaints of intermittent pain upon touch along with a tingling sensation in the thumb, index, and middle fingers. No history of trauma was reported by the patient.

On examination, a grossly enlarged, lobulated, firm mass measuring 20×7 cm was noted on the volar aspect of the distal forearm, extending into the proximal hand (►**Fig. 1**). The mass has firm consistency with restricted mobility and tenderness on palpation. A positive Tinel sign was observed with paresthesia present along the distribution of median nerve. There were no cutaneous pigmented lesions found anywhere on the body. No family history of NF was reported by the patient.

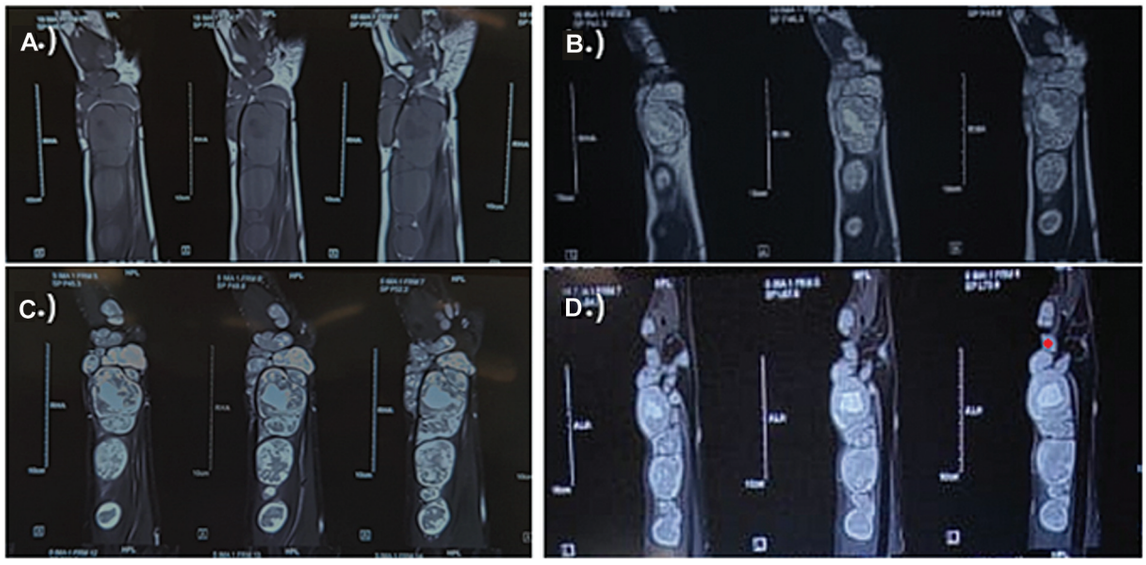

Fig. 2 Magnetic resonance imaging (MRI) of left upper limb demonstrating hypointensity on T1-weighted images in coronal (**A**) and sagittal plane (**B**) and heterogeneous hyperintensity on T2-weighted images in coronal (**C**) and sagittal plane (**D**).

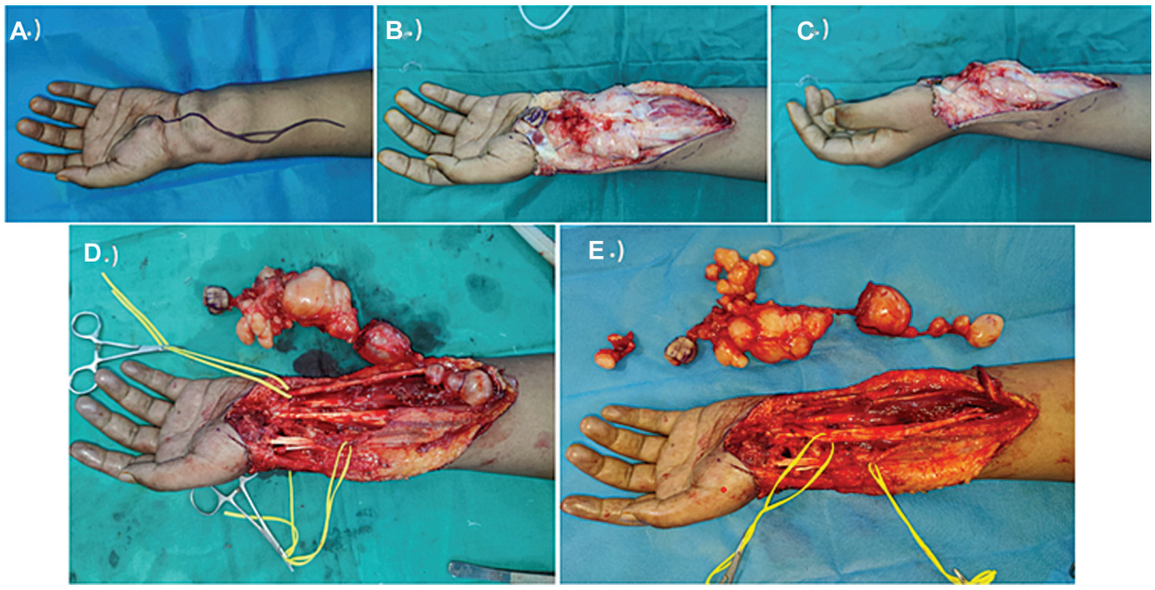

Fig. 3 Intraoperative images showing (**A**) curvilinear incision. (**B**, **C**) Multilobulated tumor arising from the median nerve sheath. (**D**) Careful dissection and separation of the tumor from the median nerve. (**E**) Showing completely excised tumor specimen from the median nerve.

Contrast-enhanced magnetic resonance imaging (MRI) was done to characterize the lesion, which showed multiple, diffusely enhancing, multilobulated, interconnected round-to-oval masses involving the mid to distal forearm and wrist in the subcutaneous and muscular plane. Some lesions were insinuated into the flexor compartment of the carpal tunnel. The lesions were heterogeneously hyperintense on the T2-weighted sequence and isointense on the T1-weighted sequence with diffuse postcontrast enhancement. Combined, the lesions occupied approximately 18 to 19 cm in length, with the largest measuring 3.9 × 3.8 cm (►**Fig. 2**). The lesions were abutting and compressing the median nerve in the mid and distal forearm, with focal encasement of the median nerve in the mid-forearm.

Based on clinical and radiological findings, the patient was planned for a microsurgical exploration. A curvilinear incision was made extending from the palmer aspect of the proximal hand to the mid-lateral forearm (►**Fig. 3**). The median nerve and the associated tumor were identified using a nerve stimulator and were isolated. Under high resolution, longitudinal incision was made on the epineurium and dissection of the soft tumor mass from the neighboring nerve fascicles was done carefully. The tumor was removed, and the specimen was sent for histopathological examination.

Immediately in the postoperative period, marginal skin discoloration was noted, which improved after a few dressings (►**Fig. 4**). There was minor sensory loss in the median nerve distributed area of the palm, with no muscle weakness, which improved after 2 weeks. After 3 months postsurgery, the wound had healed well with no pain upon palpation. At one and a half years, the patient had a complete resolution of symptoms with no signs of recurrence.

Gross pathology examination revealed multiple solid, lobulated, grayish-white encapsulated tumors with the largest one measuring 7.0 × 6.0 cm. Histologically, the tumor exhibited a classic benign nerve sheath tumor pattern, with hypercellular areas of palisading nuclei bundles (Antoni

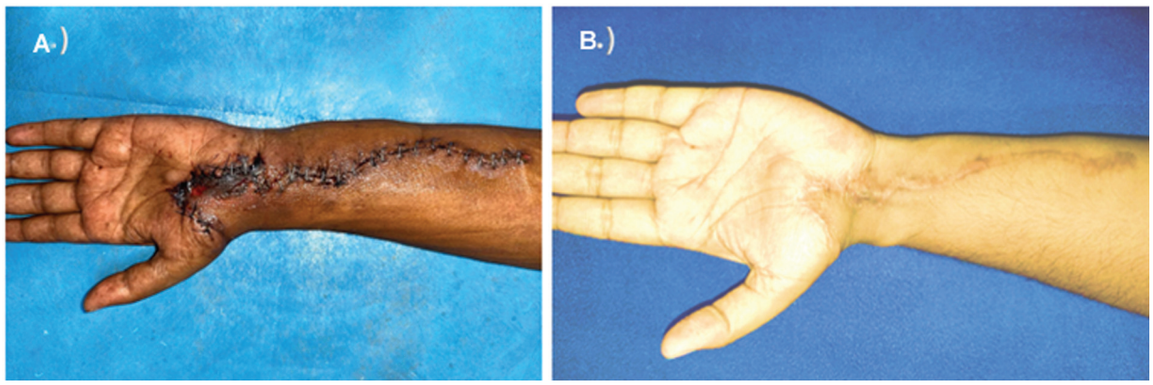

Fig. 4 Postoperative images after 10 days (**A**) and 2 months (**B**) of operation.

A tissue) and areas with myxoid changes, loose stroma, and fewer cells (Antoni B tissue). Immunohistochemistry examination showed positivity for S-100, consistent with schwannoma.

Discussion

Benign tumors arising from the peripheral nerves are relatively rare in the upper extremities, with schwannomas being the most prevalent one.[2,4,5] Typically, they manifest as solitary nerve tumors, although they can manifest as a group of continuous lesions in systemic conditions such as NF2 and/or schwannomatosis.[1,2] However, in literature, there are isolated instances reported describing multiple schwannomas involving a single peripheral nerve without any systemic pathology.[9]

Schwannomas (neurilemomas) primarily affect individuals in their third and sixth decades of life, without a significant gender predilection. While they often present as asymptomatic masses, symptoms such as pain, numbness, and fatigue may emerge as the tumor enlarges. However, schwannomas usually do not exceed 3 cm in diameter.[6,10] Consequently, diagnosing these tumors in the upper extremity poses a challenge due to their slow growth, often resembling focal peripheral compression neuropathies like carpal tunnel syndrome and cubital tunnel syndrome.[1,5,6,10] Other common differential diagnoses that may complicate the clinical picture include neurofibroma, ganglion cyst, lipoma, and fibroma.[11,12] Therefore, one must contemplate the possibility of median nerve tumor while encountering such cases. If there is presence of any motor deficit, malignancy should also be ruled out before arriving at a diagnosis.[11]

MRI is frequently used to characterize these lesions and to find out any minute lesions that may have been missed on clinical examination. MRI typically depicts schwannomas as oval masses eccentrically located on the nerve.[5] Notably, schwannomas are encapsulated, whereas neurofibromas are not. The schwannomas exhibit hypointensity on T1-weighted images and heterogeneous hyperintensity on T2-weighted images.[13] These signal characteristics often correlate with histological features.[14] The heterogenicity appears due to the presence of cystic degeneration and hemorrhage inside the tumor. Antoni A areas, which are cellular and contain collagen, appear hypointense on both T1 and T2 images, while Antoni B areas, which contain a water-rich myxoid matrix, appear hypointense on T1 and hyperintense on T2 images. In T2 images, the target sign can be seen due to the central location of Antoni A areas (hypointense) surrounded by Antoni B areas (hyperintense).[5,14] Koga et al[15] reported that the target sign is 100% specific and 59% sensitive. Ultrasonography provides dynamic images, offering insights into the relationship between the nerve and musculotendinous structures.[12]

The slow growth of the tumor allows the nerve to gradually adapt to the changing pressure effects. As a result, the interval between the onset of symptoms and the surgical intervention can vary significantly, ranging from a few months to several years.[12] Surgical excision is recommended for all symptomatic tumors and tumors with a rapid increase in size utilizing microsurgical approach (►**Fig. 3**). The objective is to create a surgical window in the epineurium by circumferential dissection separating the tumor from the healthy nerve tissue, thus avoiding disruption of the nerve fascicles. Since schwannomas are well encapsulated and eccentrically placed, therefore a complete excision can be achieved without causing any significant nerve dysfunction unlike neurofibromas and primary malignant nerve sheath tumors.[14] If there is any minor nerve dysfunction following schwannoma excision, there could be three possible reasons. First, neuropraxia due to nerve handling during excision, which typically recovers in few weeks. Second, loss of small nerve fibers traversing the capsule during the opening of the capsule. Third, since schwannomas originate from neural sheaths, excision of the fascicles surrounding the sheath can result in minor deficits. In some cases, schwannomas may demonstrate an infiltrative pattern, necessitating fascicle resection during tumor excision. In a case series by Akambi Sanoussi and Dubert,[16] inseparable fascicles required resection in 8 out of the 14 cases they enrolled. Mizushima[17] reported that around 50% of cases develop neurological deficits postoperatively[18] and this risk appears to be even higher in plexiform subtypes. Recurrence following total excision is rare.[11,14,19]

Histological examination of schwannoma exhibits a well-circumscribed lesion with collagenous capsule. The tumor typically displays a mixture of compact Antoni A and less cellular Antoni B areas.[10] Antoni A areas consist of interlacing fascicles of spindle-shaped Schwann cells, often forming nuclear palisading known as Verocay bodies,[5] while Antoni B areas contain haphazardly distributed cells in a loose myxoid matrix. Immunohistochemistry reveals expression of S-100 protein and pericellular collagen type IV aiding in the differentiation of schwannoma.[19] A high K_i-67 index ($\geq$ 20%) is suggestive of malignant transformation.[20]

To the best of our knowledge, this represents the first documented case report of multiple interconnected schwannomas involving the median nerve of such significant size without having any systemic pathology.

Limitation

Due to the patient's unwillingness, genetic testing for conditions like NF2 or schwannomatosis was not conducted. Genetic analysis could help determine whether the condition is isolated or part of a systemic pathology.

Conclusion

Although schwannomas typically present as solitary tumors, there are rare instances where multiple schwannomas can occur along the same peripheral nerve. Identifying all such lesions preoperatively by doing careful clinical examination is crucial. Preoperative MRI scans play a significant role in this process by potentially preventing the need for additional surgeries due to undetected masses that could cause persistent symptoms. Treatment often involves excision using a

microsurgical approach, with a very low likelihood of recurrence.

Authors' Contributions

S.D.: Formal analysis and writing - original draft. A.A.: Conceptualization, data curation, formal analysis, and writing - reviewing and editing. R.T.: Data curation and formal analysis.

Ethical Approval

The authors declare that the procedures were followed according to the regulations established by the Clinical Research and Ethics Committee and to the Helsinki Declaration of the World Medical Association.

Conflict of Interest

None declared.

Acknowledgment

None.

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