

# Median nerve schwannoma: A case and review of literature

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## ABSTRACT

We report a case of a median nerve schwannoma, a rare type of a benign tumor of Schwann cells that presents as a palpable and painful mass on the flexor aspect of the forearm. Schwannomas of the median nerve make up 0.1–0.3% of all hand tumors. Symptoms are caused by an entrapment syndrome resulting from the growing tumor. Pain is the most common complaint of schwannomas. Imaging studies include computed tomography and magnetic resonance imaging and ultrasound. It is difficult to differentiate schwannomas from neurofibromas solely on the basis of a radiological investigation. Tumors of the median nerve are diagnostically challenging and median nerve schwannomas are rare. Diagnostic pearls are described to facilitate a more accurate and timely diagnosis. These characteristics include mobility, Tinel's sign, S-100 histological staining, and Antoni patterns. With a correct diagnosis, the tumor can be extirpated with preservation of nerve function and a low risk of recurrence.

**Key words:** Median nerve, schwannoma, Tinel's sign

## Introduction

Schwannomas also known as neurilemmas are usually originated from Schwann cells located in the peripheral nerve sheaths. They are the most common tumors of the hand (0.8–2%).<sup>[1]</sup> Schwannomas also known as neurilemmas, are the most common benign tumors of the neural sheaths of the peripheral nerves.<sup>[2]</sup> Usually, the ulnar and peroneal nerve are involved while only 7% of schwannomas involve the median nerve.<sup>[3]</sup> Benign tumors involving peripheral nerves of the upper extremity are uncommon<sup>[4]</sup> and they cause difficulties in classification, clinical diagnosis, and treatment. In the upper limb, they may be mistaken for ganglia or carpal tunnel syndrome.<sup>[5]</sup> They usually grow slowly and appear as painless swellings for several years before diagnosed.<sup>[6]</sup> They usually present with solitary swelling along the course of the nerve (d), however multiple lesions may be present in cases of NF type 1, familial neurofibromatosis, and sporadic schwannomatosis.

Incidence is similar between both genders and the tumor is the most common in 3<sup>rd</sup> and 6<sup>th</sup> decades. Schwannomas are generally represented as an asymptomatic mass; however pain, numbness, and fatigue may take place with the increasing sizes of the tumor; however it is unusual for a schwannoma to exceed three centimeters in diameter (d) Electromyography, magnetic resonance imaging (MRI), and ultrasonography are helpful in the diagnosis. Surgical removal is usually curative.

## Case Report

A 45-year-old female presented with a painful swelling at the flexor aspect of forearm. In his history, she was aware of the swelling for 1 year, but the numbness was only present for the last 2 months. On clinical examination, there was a painful solid mass 3–4 cm in length, little sensitive to pressure, mobility restricted in both directions. Percussion over the mass produced a Tinel's like sensation along the median nerve. No history of significant trauma was recalled. The patient experienced mild numbness in the distribution of the median nerve in the palm, but no motor weakness or muscle atrophy was detected. No cutaneous pigmented lesions were found. The patient reported she first palpated a nodule 2 year ago that grew up very slowly to the current size, but becoming painful since last 2 months.

An ultrasound (US) examination revealed a 4 cm long, well-circumscribed mass in close proximity to the neurovascular bundle of the flexor aspect of the forearm. On MRI examination, lesion had a relatively homogenous low signal, slightly lower to the flexor tendon of forearm, on T1 images. On T1 fat-suppressed images, increased gadolinium enhancement

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10.4103/1793-5482.161178

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was noticed at the periphery of the mass with a nonenhancing low signal central area. No peritumoral edema was present and a line of fat surrounded the tumor. Based on the long history of the patient, and the clinical, US, and MRI features, the diagnosis of a nerve sheath tumor arising from the median nerve was supported and the patient was scheduled for excisional biopsy.

A longitudinal incision centered over the tumor bulk at the distal part of flexor aspect of forearm was performed [Figure 1]. The tumor was situated below the flexor tendon of forearm. The tumor had an eccentric position and was firmly attached to the median nerve. The decision to preserve the median nerve and perform a marginal tumor excision was undertaken. The epineurium was longitudinally incised and the soft tumor mass dissected from surrounding nerve fascicles [Figures 2 and 3]. The mass was encapsulated and removed totally [Figure 4]. Immediately postoperatively the patient not experienced any sensory loss and weakness of the muscles innervated by the median nerve.

The histopathology result demonstrated an ovoid tumor measuring 3.5 cm × 4 cm. The center of the mass was solid and firm. Histologically, the characteristic features include the presence of alternating Antoni A and Antoni B areas. Antoni A area is composed of spindle-shaped Schwann cells arranged in interlacing fascicles. There may be nuclear palisading. In between two compact rows of well aligned nuclei, the

cell processes form eosinophilic Verocay bodies [Figure 5]. Mitotic figures may be present. Antoni B area consists of a loose meshwork of gelatinous and microcystic tissue. Large, irregularly spaced, thick-walled blood vessels are noted in Antoni B area. These may contain thrombus material in the lumina. Immunohistochemistry shows S-100 stain was positive. The final pathology report was consistent with schwannoma. At 6 months follow-up the wound is well healed with no pain upon palpation. The patient returned to full labor work as a farmer.

## Discussion

Schwannomas are rare tumors.<sup>[7]</sup> They are usually solitary and benign lesions; however, they can be multiple suggesting an underlying tumor predisposition syndrome<sup>[8]</sup> and may be associated with neurofibromatosis type 1 and schwannomatosis.<sup>[9]</sup> Diagnosis of a schwannoma in the preoperative period is challenging because of the slow growth and paucity of symptoms. Diagnostic accuracy is crucial to maintaining the integrity of the nerve involved and to properly plan the appropriate surgical intervention. Schwannomas can be asymptomatic or can produce pain, a positive Tinel's sign or a Tinel's like sensation and sensory alterations.<sup>[3,10]</sup> The tumors are transversely mobile but immobile longitudinally, likely due to their nested intraneural location. Schwannomas share many features with other soft tissue tumors and are frequently misdiagnosed due to similarities. Differential diagnosis should

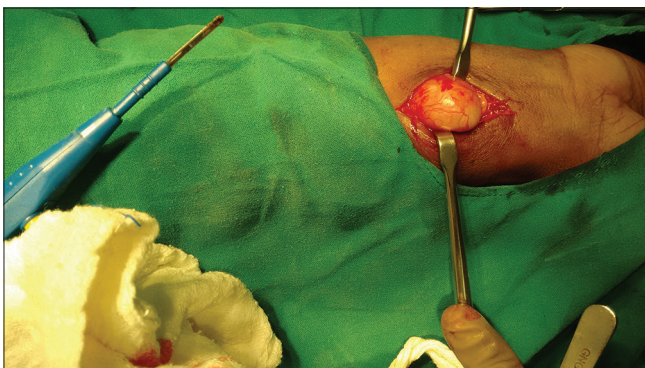


Figure 1: Schwannoma over flexor aspect of forearm

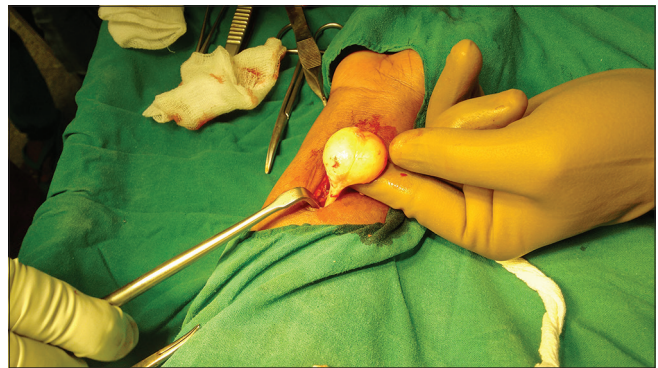


Figure 2: Schwannoma intact with median nerve

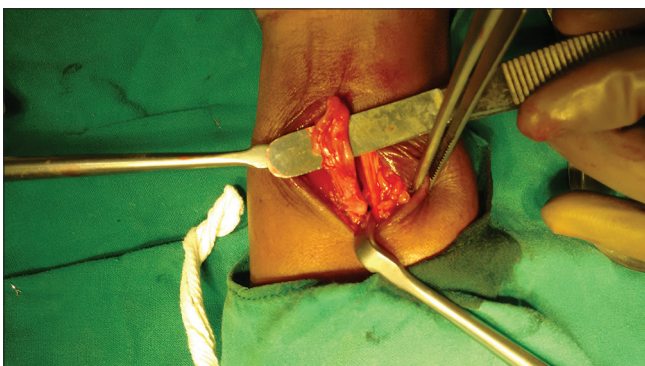
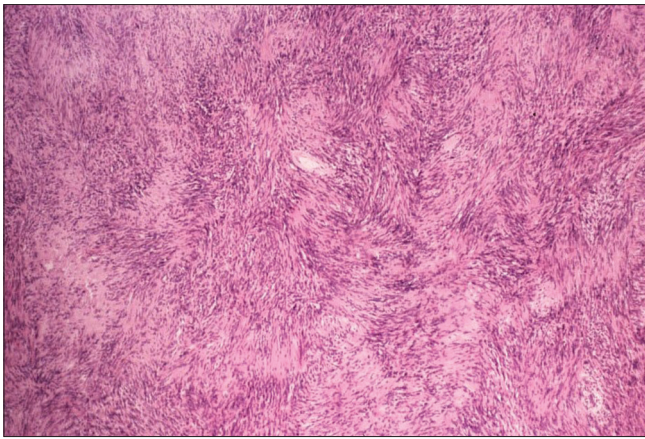


Figure 3: Median nerve after excision of schwannoma



Figure 4: Encapsulated schwannoma specimen





**Figure 5:** Histology of schwannoma with its characteristics pattern

include neurofibroma, ganglion cysts, malignant tumors, lipomas, and xanthomas.<sup>[6,11]</sup> Neurofibromas, in particular, cannot be distinguished from schwannomas on physical examination. The symptoms appear to be nonspecific, which adds to difficulty in diagnosis. The slow growth pattern of benign nerve tumors, allows for adaptation of the nerve function to the pressure effects.<sup>[12]</sup> Thus, although schwannomas can rarely induce impaired motor function, neural tumors producing motor deficits should always raise a high suspicion of malignancy.

On MRI, gadolinium enhanced T1-weighted and T2-weighted MRIs are particularly useful in diagnosing schwannomas. Koga *et al.* (2007) found the presence of the target sign to be 100% specific and 59% sensitive for the tumors. The target sign is the contrasting central and peripheral intensities demonstrated on the images. On pathologic analysis, a schwannoma has a true capsule composed of epineurium. The hallmark of schwannomas is the alternating pattern of Antoni A and B areas. Antoni A is cellular areas with spindle cells and nuclear palisading forming Verocay bodies, whereas Antoni B is hypocellular areas in a myxoid background. Histological staining reveals a strongly positive S-100 protein that is specific for schwannomas and helps to rule out neurofibromas. Imaging shows the tumors as round or oval, eccentrically located in relation to the nerve, encapsulated, isolated, and noninvasive.<sup>[13]</sup> In comparison, neurofibromas are nonencapsulated and intimately surround the nerve. They cannot be surgically removed without damaging the connected nerve, often necessitating nerve grafting to repair functioning. Schwannomas, on the other hand, can be separated surgically from the nerve fascicles avoiding neurologic deficits.<sup>[14]</sup> This emphasizes the importance of a correct preoperative diagnosis. Despite the structural differences of soft tissue tumors, they are difficult to distinguish with imaging. Domanski *et al.* (2006) aspirated 116 different schwannomas, and results were not sufficient for diagnosis for about 44% of the cases.<sup>[15]</sup> Extirpation of the intraneural schwannoma can be challenging. Sterile tourniquet dissection is recommended and assists in

visualization. Identification of the nerve proximal and distal to the tumor is the first important step to reducing injury and traction neuropraxia. Identification of the individual splayed nerve fascicles as they spread over of the tumor is critical in determining the entry through epineurium. A longitudinal incision is created between the splayed fascicles down to the tumor sheath. Once the outer layer of the tumor is identified, a plane can be developed between the more superficial fascicles and the tumor wall. Slow, deliberate, circumferential dissection with a “peanut” and Littler scissors facilitates delivery of the tumor. Once the tumor is removed, the nerve is inspected for injury, the tourniquet is released, and precise hemostasis is achieved. Repair of the epineurium is not required and the longitudinally split muscle is repaired loosely over the nerve. Immediate postoperative hand therapy is instituted.

It is uncommon for schwannomas to recur in identical locations. Das *et al.* (2007) found that surgical removal of schwannomas was successful in alleviating preoperative symptoms, while maintaining nerve functioning in 89% of their cases.<sup>[13]</sup>

## Conclusion

Here we present a case of schwannoma of the median nerve. Schwannomas are rare peripheral nerve tumors that have important diagnostic and radiographic features. MRI typically reveals the target sign of biphasic contrast of peripheral and central regions and distinct encapsulation displacing the intimately associated nerve fascicles. These tumors are transversely mobile and longitudinally immobile, have a positive Tinel’s sign, and exertional dysaesthesia or pain. Surgical resection must be approached with caution to protect nerve function and continuity. Surgical resection is associated with good outcomes. The recurrence rate is low.

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**How to cite this article:** Padasali PS, Shankaregowda VS, Kshirsagar SD. Median nerve schwannoma: A case and review of literature. *Asian J Neurosurg* 2015;10:212-5.

**Source of Support:** Nil, **Conflict of Interest:** None declared.