

An Unusual Presentation of Giant Schwannoma of Hand: A Case Report

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ABSTRACT

Peripheral nerve sheath tumors are uncommon. Peripheral nerve tumors (PNTs) constitute less than 5% of all hand tumors. Schwannomas, also known as neurolemmas, are benign tumors of the peripheral nerve sheath arising from Schwann cells. The median nerve schwannomas account for 0.1-0.3% of all cases. Here is a case of a seventeen-year-old male who presented with a painless lump on the thenar aspect of the left hand simulating lipoma clinically. The patient underwent various imaging modalities including Magnetic Resonance Imaging (MRI) which suggested a peripheral nerve sheath tumour most likely Schwannoma. The lesion was excised under the regional block and was confirmed as Schwannoma histologically.

Keywords: *Lipoma; Magnetic Resonance Imaging; Neurilemmoma; Nerve Sheath Neoplasms*

INTRODUCTION

Schwannomas or neurolemmas are encapsulated benign tumors of the peripheral nerve sheath arising from the Schwann cells. Their presence in the upper limbs is reported in only 0.8-2 % of the cases. All major nerves of the upper limb can be affected with tumors of the ulnar, radial, and median nerves being most commonly encountered. Diagnosis, classification, and treatment of the nerve sheath tumor of the extremity can be challenging due to their rare prevalence as well as the slow growth of lesions without symptoms.

CASE REPORT

A 17-year-old male, presented with progressively increasing painless fluctuant swelling on his left

hand (Figure 1a). Examination revealed a well-defined mobile lump of size 8.0 x 6.0 cm in the thenar aspect. Tinel's sign was negative. Ultrasonography showed well-defined homogeneously hypoechoic lesions with minimal internal vascularity. The median nerve was identified at the level of the carpal tunnel but couldn't be identified separately at the site of the lesion suggesting proximity of lesion with the nerve. The Plain X-ray of the hand was unremarkable.

Contrast-enhanced Computed Tomography (CT) (Figure 1b) showed a heterogeneously enhancing iso-dense lesion (similar attenuation of the muscle) on the palmar aspect superficial to the tendon of flexor digitorum superficialis (FDS) and

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flexor digitorum profundus (FDP). No extension into the joint was noted. Carpal bones appeared normal with no evidence of erosion and sclerosis.

Further in MRI (Figure 1c, d), the lesion appeared as a well-circumscribed ovoid structure displaying homogenous and isointense signal (relative to muscle) on the T1 sequence and mildly heterogeneous with a signal intensity greater than fat in the T2 and STIR sequence. FDS and FDP muscle tendon was seen separate from the lesion. The lesion was noted adjacent to the course of

the median nerve without evidence of invasion/enlargement of the nerve.

The patient underwent total excision of the lesion that showed grey white glistening capsule on the outer surface (Figure 2a) The lesion was located adjacent to the median nerve without evidence of enlargement/ invasion of the nerve. The postoperative period was uneventful without neurological deficit and the patient was subsequently discharged.

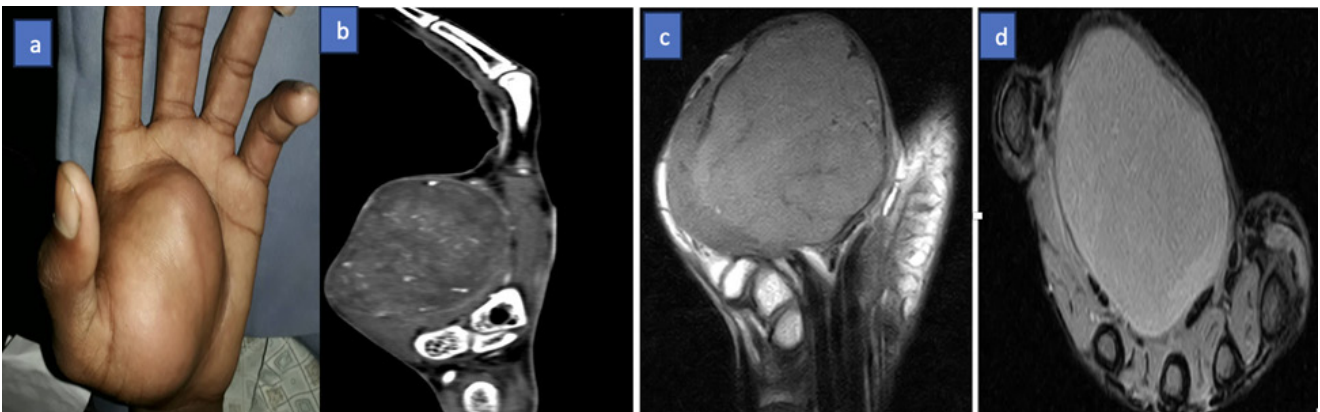


Figure 1: a) swelling in the left hand which was non-tender, and soft in consistency. b) Contrast-enhanced CT scan sagittal section showed encapsulated lesion with heterogeneous enhancement without invasion into the adjacent bone and tendon. c, d) T2 weighted coronal and axial image showing well-defined T2 iso-hyperintense lesion with T2 hypointense capsule.

Histopathology: Histopathological examination confirmed the tumor to be schwannoma. Microscopic examination (Figure 2b,c) revealed predominantly cellular Antoni A areas as well as hypocellular Antoni B areas. Hypocellular

areas showed hyalinization, edema, sclerosis, and focal myxoid change. However, the Verocay bodies, mitosis, and necrosis were not observed. Unfortunately, S100 protein staining was not performed in our case.

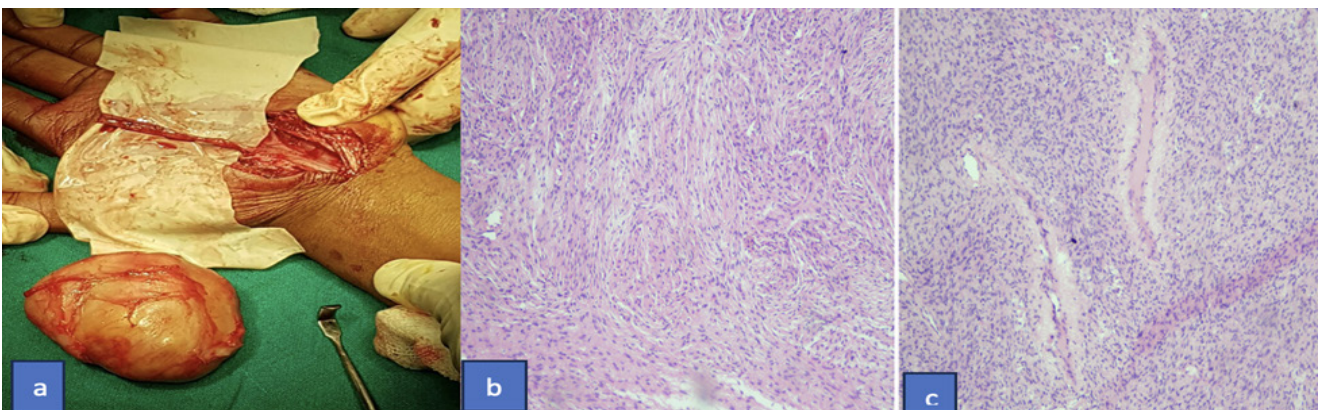


Figure 2: a) Intraoperative image showing the well-defined lesion with intact median nerve after the excision of the lesion. b and c) Palisading cells in Antoni A zone and loosely meshed cells in Antoni B zone

DISCUSSION

Schwannoma was first described by Verocay in 1908, followed by Stout in 1935 who suggested the term neurilemmoma as the tumor arises from the nerve sheath and Schwann cells. Schwannomas can originate from a peripheral, cranial, or autonomic nerve in any part of the body. They are less common in the upper and lower extremities (0.8-2%) and are most frequently observed between the ages of 30-60 years. 90% of all cases of Schwannomas are solitary and sporadic; however, 10% of Schwannomas occur at multiple sites.^{1,2}

Ganglion cysts, hemangiomas, lipomas, giant cell tumors of the tendon sheath, glomus tumors, and nerve sheath tumors are the most common hand lesions. Clinical diagnosis of Schwannoma is rarely straightforward due to a wide differential diagnosis; the closest differential being neurofibroma. In contrast to neurofibromas, which are fusiform masses within the nerve that are closely linked to the nerve fascicles, Schwannomas are encapsulated, eccentrically located tumors that displace the nerve fascicles and tend to occur more often in mixed nerves rather than in pure motor or sensory nerves.^{3,4}

Schwannomas clinically presents as a lump without any symptoms or as discomfort and paresthesia. Pain is usually caused by compression of the neighboring nerve, which can lead to Wallerian degeneration. Schwannomas are mobile only in the longitudinal plane along the course of the affected nerve. If a sensory or mixed nerve is affected, Tinel's sign-shooting paresthesia in the affected nerve's distribution occurs upon percussion of the tumor.⁵

As ultrasonography offers dynamic information about the lesion and its relationship to the adjacent structure, it is the initial imaging modality of choice. From a surgical perspective, it is crucial to distinguish between Schwannoma and neurofibroma. This can be done by using contrast-enhanced Magnetic Resonance Imaging (MRI) to determine the tumor's origin and location in relation to surrounding structures. These lesions show homogenous iso-hypointense

on T1-weighted images, while on T2-weighted images, they show high signal intensity. It is the fat surrounding the lesion that causes the "split fat sign" on T1 weighted images along the long axis of the affected limb. "Target signs" which consist of a central low-intensity signal (fibrous) and a peripheral high-intensity area (myxoid), are seen in 50% of cases. Target signs are typically observed in cases of Neurofibroma, while Schwannoma frequently exhibits fascicular signs. Additionally, calcification, necrosis, and cystic cavitation are more frequent in Schwannoma than in Neurofibroma.^{6,7}

The definitive diagnosis of Schwannoma depends on the characteristic histopathologic features and specific immunohistochemical markers. S-100 protein is specific for schwannomas and helps to rule out neurofibromas. The histological features known as Antoni A and Antoni B are unique to schwannoma.⁸

The preferred treatment in schwannoma is total surgical excision of the mass without damaging the paternal nerve. Even though schwannoma can be diagnosed based on certain radiological features, MRI cannot determine whether the mass can be entirely enucleated during surgery.

Recurrence of Schwannoma is rare. Malignant transformation of median nerve schwannoma is uncommon; there are currently no reports of it in the literature. In the largest published series, no malignant transformation was reported at mid-term follow-up of benign solitary Schwannomas.^{10,11}

CONCLUSION

Because of the low prevalence of the lesion, diagnosis of the lesion can be challenging. So, any swelling along the distribution of the nerve, Schwannoma should be kept in differential diagnosis and should be properly evaluated with radiological findings before excision of the swelling.

CONFLICT OF INTEREST

None

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