

Ulnar nerve Schwannoma: Case Reports and Litterature Review

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Abstract

Introduction

Schwannomas are benign tumors that most commonly arise from peripheral nerve sheath and consist of 5–8% of soft tissue neoplasm.^{1,2,3} it is a non–invasive tumor covered by epineurium.² Schwannomas are benign tumors of the nerve sheath common in adults. Schwannomas could be found in any age groups and tends to be developed at in periode of 30– 60 years. It very slowly grows, and neither influenced by gender or race.^{1,2} Mostly these tumors are solitary, found in the upper extremities rather than lower ones with a ratio of 2:1, and normally found in the volar surface.² Signs and symptoms are not obvious, leading to frequent misdiagnosis. Based on literature only a quarter of all diagnostic tests provide accuracy. The choice of treatment is tumor resection.

Case Report

A 49–years old male, Malay, complain a lump in the medial aspect of the right arm since 4 years. There is no history of fever, weight loss, or previous trauma. He feels pain in his right upper arm that extends up to his finger. On physical examination, it found a mass on the medial aspect of the forearm measuring 5 x 3 cm, firm and solid boundaries. There is no muscle weakness. MRI show soft tissue lesions arises among the muscles of the flexor digitorum superficial and palmaris longus running out through the course of the right ulnar nerve (Fig 1).

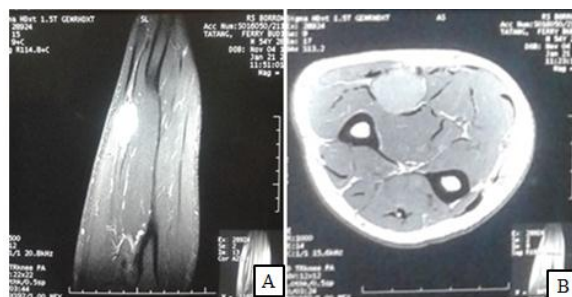


Fig 1. A) Sagittal view MRI contrast shows a solid oval mass enhances homogeneously running out the course of right ulnar nerve. B) Coronal view MRI show soft tissue lesions arises among the muscles of the flexor digitorum superficial and palmaris longus

During resection found tumors are derived from ulnar nerve, encapsulated and well defined. Ulnar nerve clearly identified. Resection was done through intracapsular approach to avoid iatrogenic ulnar damage.(Fig 2) No neurological deficit post-operatively. Histopathology result revealed mass composed of hyper- and hypocellular areas, spindle-shaped, hyperplastic, palisade structures with monotonous oval cell nucleus. Stroma filled with hyperplastic fibrocollagen tissue, this is the features of schwannoma.(Fig.2F)

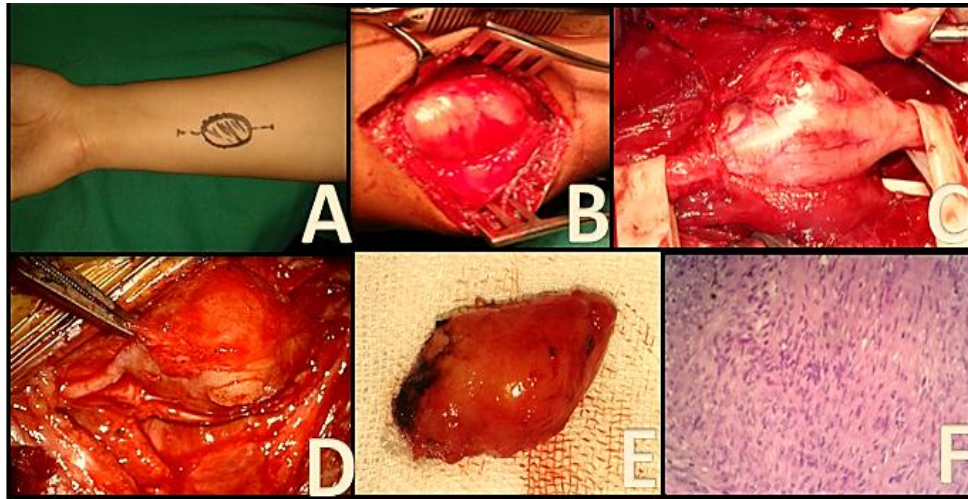


Fig 2. A) Linear incision is designed to access the lump; B) Mass beneath the surface of pronator teres muscle; C) Dissection and identification of proximal and distal of Schwannoma; D) Intracapsular resection of Schwannoma; E) Tumor mass macroscopy F) Histopathology: mix typed Schwannoma Antoni A and B.

Discussion

Schwannoma is a very rare peripheral tumor found in clinical practice, this tumor remains the most common tumor of the nerve sheath, particularly found in wrists as well as in hand. This kind of tumor found in 5–8% of all benign soft tissue neoplasm.^{3,5} Schwannoma is a well encapsulated benign tumor that grows slowly and often asymptomatic.⁶ It also has a lot in common with other soft tissue tumors, thus often misdiagnosed. Accurate diagnostic is very important to maintain the integrity of the nerves involved and plan an appropriate surgical intervention. This tumor is often found as a mobile mass without muscle weakness. Positive Tinel's sign is found in majority of cases. The differential diagnosis includes neurofibromas, ganglion cysts, malignant tumors, lipomas, and xanthomas.^{2,7}

Schwannomas is difficult to be distinguished with neurofibroma clinically.⁸ It can be distinguished with fine-needle aspiration biopsy.⁹ Malignant mass shows distinct signs from a benign Schwannoma. It often misdiagnosed in the early stages. Malignant tumor is fixed, firm, manifesting pain at rest and motor weakness.¹⁰ Motor weakness might be found depends on the tumor site, and even though a benign tumor exceed of 2.5 cm. Kehoe and colleagues (1995) have analyzed 88 tumors of peripheral nerves, found that only one was accurately diagnosed as Schwannoma prior to surgery.^{3,11}

Hence, MRI is essential in instituting a diagnosis of nerve sheath tumor, the use of Gadolinium contrast T1 and T2 on MRI provide a very useful technique in diagnosing of Schwannomas. Imaging featuring a round or oval tumor mass, eccentric to the nerve, capsule, isolated, and non-invasive, Koga and colleagues (2007) found the presence of the target sign provides specificity of 100% and sensitivity of 59%. Target sign referred to the intensity of contrast on the central of tumors. Histologically schwannoma show Antoni A pattern shows a low cell concentration whereas Antoni B area shows high cell, which is the hallmark of Schwannomas. Tumor cells histology staining using of protein S-100 is considered very immuno-positive, specific to Schwannoma, excluding neurofibroma.^{1,8,13}

As the comparison, neurofibroma is encapsulated, surrounding involved nerve and cannot be removed surgically and left the nerve undamaged, thus, nerve grafting to restore its function is often required.^{3,7,8} In Schwannoma, nerve fascicles can be separated surgically to avoid post operative neurological deficits.⁷ This emphasizes the importance of accurate preoperative diagnosis. Despite the structural differences in soft tissue tumors, it remains difficult to distinguish through imaging techniques. The most commonly affected peripheral nerve is the median nerve.¹⁴ Schwannomas usually recurred at the same location. Surgical resection is recommended due the degree of severity as well as its recurrency rate is quite low. Neurological deficit may develop following tumor resection because of dissection and retraction of involved nerve. However, its normal function usually return in a few months.²

Conclusion

Schwannoma is a very rare peripheral extremity tumor and often diagnosed following histopathology exam. Tumor resection with no neurologic deficit is the treatment of choice with low recurrence rate.

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