

AN INTERESTING CASE OF PERIPHERAL NERVE TUMOUR - ULNAR NERVE SCHWANNOMA

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ABSTRACT

Schwannomas are benign tumours of schwann cell origin, and can involve cranial nerve, spinal nerve roots, intercostal nerve, posterior mediastinum, retroperitoneum. Solitary peripheral nerve tumours are uncommon. They can involve a peripheral nerve partially or completely. They usually present as an innocuous solitary mass in the extremities in 3rd to 5th decade of life, with or without associated neurological symptoms. Schwannomas are the commonest benign tumours detected in the peripheral nerves representing approximately 8% of all soft tissue neoplasms. They are non-invasive, arise from the peripheral nerve sheath, encapsulated by epineurium. In this article, we present a 30 year old female diagnosed as peripheral ulnar nerve tumour. Early diagnosis and surgical treatment helped us in preventing permanent nerve damage.

KEYWORDS : Benign Schwannoma, Peripheral Nerve Tumour, ulnar Nerve Schwannoma, Ancient Schwannoma, Antoni A And Antoni B Areas.

INTRODUCTION:

Schwannomas are the commonest benign tumours detected in the peripheral nerves representing approximately 8% of all soft tissue neoplasms. There are non-invasive, arise from the peripheral nerve sheath, encapsulated by epineurium. There is no sex or racial difference but there exist a 2:1 occurrence in favour of upper limbs comparing to lower while mostly develop during the 3d and 6th decade of life. They are slow growing, usually misdiagnosed as lipomas until they become painful. Early diagnosis is crucial before these tumours become oversized and can create surgical problems. The symptoms appear to be nonspecific which helps patients delay and sometimes doctor's delay. These benign tumors are easily mistaken for other entities such as lipomas, neurofibromas, hemangiomas, and synovial cysts. Pathologically Schwannomas are benign encapsulated neoplasms of schwann cell. Microscopically they are composed of two cell patterns Antoni type A and Antoni type B. Schwannomas usually arise from a single fascicle and grow circumferentially displacing the intact nerve fascicles and are located eccentrically on the nerve root.

CASE REPORT:

A 30 year old female presented to us with a gradually increasing swelling over the medial and inner aspect of her right arm over the biceps muscle, for the past one year. She noticed numbness over her fourth and fifth fingers. The patient experienced mild numbness at the distribution of the ulnar nerve at the palm but no motor weakness or muscle atrophy were detected. patient noticed Significant enlargement and concomitant paraesthesias during the last one month. No history of significant trauma was recalled. On examination we found a single soft swelling measuring about 5X4 cm over the medial inner aspect of right arm. The transillumination test was negative, and the swelling was freely mobile along the axis perpendicular to the right upper limb, and it was immobile along the longitudinal axis of the limb. Tinnel's sign was positive over the swelling. Xray of the right arm revealed a vague shadow. Ultrasound was done which revealed an elongated soft tissue lesion of size 4.8*2.1*2.3cm with tapered ends noted in medial aspect of right mid arm in intamuscular plane suggestive of ulnar nerve sheath tumour. MRI of right shoulder with upper limb done which revealed a well defined fusiform heterogeneous T2 Hyperintense lesion along neurovascular bundle in medial aspect of mid third and traversing along ulnar nerve -suggestive of peripheral nerve sheath tumour. Blood

investigations were within normal limits. Nerve conduction study was done which revealed right ulnar neuropathy-sensory, motor, axonal predominantly. The nature of the disease explained to patient and we proceeded with surgical resection of the tumour. Intraoperative findings were tumour encapsulated over the ulnar nerve. The tumour was excised in toto without damaging the nerve bundle and sent for HPE. HPE revealed the tumor consisted of varying hypertrophic peripheral nerve fascicles, showing a plexiform pattern, and mainly composed of Antoni A areas that consisted of dense fascicular and interlacing proliferation. post operative period was uneventful.



Fig 1 Clinical Picture Showing Swelling And Ulnar Nerve Damage To The Fingers

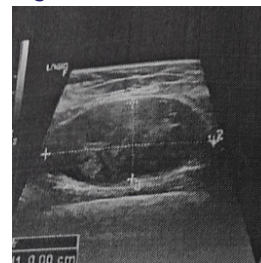


Fig 2 Ultrasound Was Done Which Revealed An Elongated Soft Tissue Lesion Of Size 4.8*2.1*2.3 Cm With Tapered Ends Noted In Medial Aspect Of Right Mid Arm In Intamuscular Plane Suggestive Of Ulnar Nerve Sheath Tumor

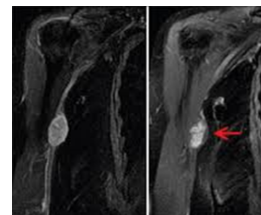


Fig 3 Mri Image Showing Hyperintense Peripheral Nerve Sheath Tumor

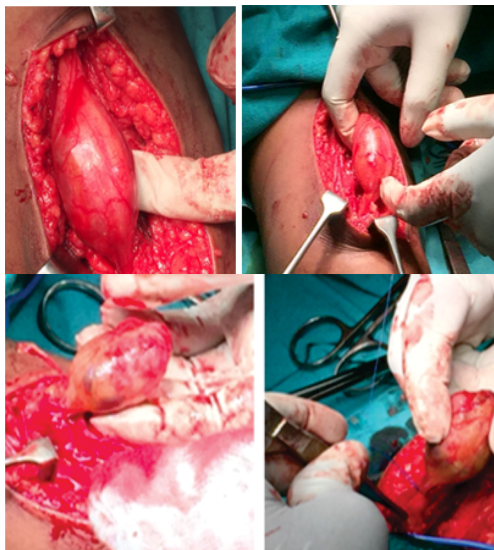


Fig 4 & 5 Intraoperative Pictures Showing Tumor

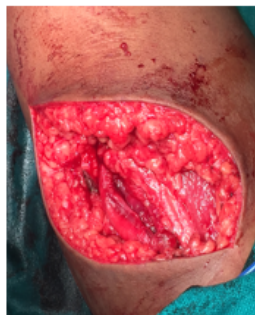


Fig 6 Ulnar Nerve Bundle Intact After Removal Of Tumour

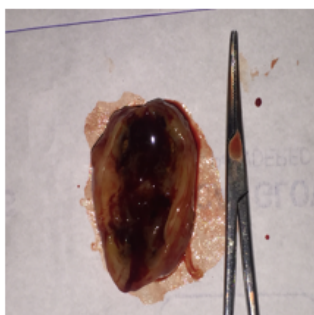


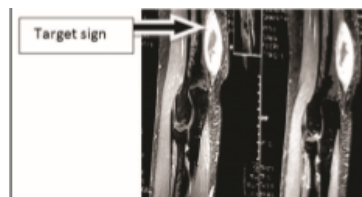
Fig 7 Specimen Showing Schwannoma

DISCUSSION:

Schwannomas are benign tumours of schwann cell origin, and can involve cranial nerve, spinal nerve roots, intercostal nerve, posterior mediastinum, retroperitoneum. Solitary peripheral nerve tumours are uncommon, 1-3 and an accurate preoperative clinical diagnosis may not be possible in a fair proportion of patients.⁴ The Schwannomas can involve a peripheral nerve partially or completely. They usually present as an innocuous solitary mass in the extremities in 3rd to 5th decade of life, with or without associated neurological symptoms. There is no sex predilection. Pathologically Schwannomas are benign encapsulated neoplasms of schwann cell. Microscopically they are composed of two cell patterns Antoni type A and Antoni type B. Schwannomas usually arise from a single fascicle and grow circumferentially displacing the intact nerve fascicles and, are located eccentrically on the nerve root. Multiple Schwannomas are rare and have usually been reported in association with neurofibromatosis 2 (NF2), or schwannomatosis. Diagnosis at times may be challenging and a careful clinical assessment and even technical investigations like Magnetic resonance imaging, CT and angiography may fail to indicate the correct preoperative diagnosis.^{5,6} The important clinical differentials that are often confused with a benign solitary schwannoma of an extremity include traumatic neuroma, neurofibroma, lipoma, cold abscess muscle hernia, haemangioma, and synovial cyst etc.⁶ Clinically it is extremely difficult to differentiate schwannoma from a neurofibroma, however intraoperative appearance of the tumour mass, its gross morphological characteristics and the subsequent histopathological and immuno-histochemical examination can settle the diagnosis in almost all the cases. Few authors have reported worsening of the nerve function following surgical

enucleation of these tumours⁴ but, fortunately majority of the benign peripheral nerve schwannomas can be safely and easily separated from the peripheral nerve trunk without damaging the peripheral nerve trunk as in our case. Schwannomas represent 5% to 8% of all soft tissue neoplasms, and because of this rarity they usually pose a challenge to have correct preoperative diagnosis. Commonly schwannomas present as sporadic solitary peripheral nerve mass and, when multiple are usually seen in association with NF2 or schwannomatosis. They present as isolated mass in the head neck region or over the volar and extensor aspects of extremities. Failure to timely detect these tumours may lead to loss of significant neurological functions of the patient. The upper and lower extremity ratio of schwannoma is 2:1 as seen in major series. In case of doubt on histology, Immunohistochemical examination using S100 is very helpful to differentiate schwannomas that stain positive for S100, from rest of the conditions. An appropriate clinical history and examination followed by investigations like Ultrasonography, MRI and electromyographic studies may sometimes help in a correct preoperative diagnosis, Majority of solitary benign peripheral nerve schwannoma can be enucleated safely without any structural damage to the nerve trunk.

Benign solitary schwannomas have excellent prognosis and, unless incompletely excised there is no risk of recurrence. Magnetic resonance imaging is the imaging modality of choice that can identify the nerve of origin, encapsulation, position of the nerve trunk in the tumour mass, tumour dimensions, and perilesional edema, and the characteristic target sign.



A correct surgical plane is critical to prevent any undesirable damage to the intact nerve fascicles and prevent postoperative neurological deficit. In our patient we did complete enucleation of the right ulnar nerve schwannoma, which was turned out to be benign cellular schwannoma on histopathological examination. Six weeks after surgery patient had fully regained normal sensations in her right inner one and half fingers, and she was highly satisfied with her surgical outcomes. At her last follow-up 2 months after surgery, she is doing well all her routine activities.

CONCLUSION:

Ulnar nerve schwannoma cases without permanent neuropraxia is less in literature. Careful physical examination and imaging features raise the diagnostic suspicion of schwannomas, and histopathologic features are the mainstay for the definitive diagnosis of these tumors. This case is being presented to highlight that when a soft tissue swelling with features of numbness, pain a differential diagnosis of peripheral nerve sheath tumour should strike in the back of the mind of a surgeon.

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