CASE REPORT

A Case Report on Isolated Ancient Ulnar Nerve Schwannomas with a Daughter Lesion

Prerna Anup Patwa[1], Rohan Kumar Singh[1], Gaurav V. Mishra[1], Suresh V. Phatak[1], Rajasbala Pradeep Dhande[1]*

[1]Department of Radio-diagnosis, Jawaharlal Nehru Medical College, Sawangi, Meghe, Wardha-442001 (Maharashtra) India

Abstract:

Peripheral Nerve Sheath Tumours (PNSTs) are the primary neurogenic tumours of the nerve sheath and they are divided into benign and malignant. Benign PNSTs are schwannomas and neurofibromas. Schwannoma is often seen associated with Neurofibromatosis type II and has imaging features overlapping neurofibroma. In this article we discuss the various imaging features like target sign, fascicular sign, split fat sign and characteristics useful for diagnosis of the lesion.

Keywords: Peripheral Nerve Sheath Tumour, Schwannoma, Neurofibroma, Ulnar Nerve

Introduction:

Peripheral Nerve Sheath Tumours (PNSTs) are neurogenic tumours arising from the nerve sheath outside the central nervous system. They are classified broadly into two types - benign and malignant. The benign type largely includes schwannoma and neurofibroma, apart from those some rare lesions in this category includes perineurioma, a lesion arising from the perineural cells and hybrid type of nerve sheath tumour where the tumour contains morphological characteristics of any of the two of the above mentioned lesions [1].

Schwannomas, also known as neurilemmoma is one of the two benign peripheral nerve sheath tumours which needs to be distinguished from its other common counterpart the neurofibroma. These lesions comprise 5% of all the benign peripheral nerve sheath tumours and are seen equally in males and females especially of 2nd to 4th decade [2]. Here, in this article we will be discussing the imaging characteristics of the peripheral nerve sheath tumours of the extremity and the characteristics that help in differentiating schwannomas from neurofibromas.

Case Report:

A 16 year old female patient came to the Acharya Vinoba Bhave Hospital, Wardha to evaluate a small soft tissue swelling along the lateral aspect of the elbow. The swelling was of a size of a pea initially and gradually increased to the current size which was 2-folds larger. There was no history of trauma or restriction of movement. There is minimal tenderness over the swelling and there were no signs of inflammation. An initial assessment was done with a radiograph which revealed a doubtful soft tissue lesion on the lateral aspect of the elbow. MRI was performed, which revealed a well-defined fusiform lesion of altered signal intensity was noted at the postero-medial aspect of proximal end of the elbow along the ulnar nerve. The lesion was eccentric and showed intense peripheral enhancement with central hypointense area of

necrosis on post-contrast scans. The lesion appeared hyperintense to the adjacent muscles on T1-weighted, T2-weighted images and Short Tau Inversion Recovery (STIR) images. Target sign, T2 hyperintense rim sign and split fat sign were seen on T2-weighted images. Another similar intensity lesion was noted posterior to the medial epicondyle. With the following imaging characteristics a provisional diagnosis of schwannoma was made as the lesion. The patient was operated on and the lesion was resected completely. On histopathological examination the diagnosis was confirmed as schwannoma. The typical Verocay bodies showing the hypercellular Antoni A areas and the hypocellular areas as Antoni B cells.

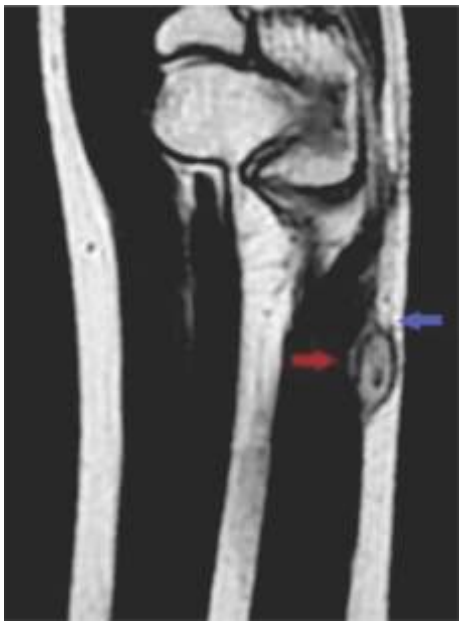

Fig. 1: Coronal T2WI Image showing a Fusiform Lesion along the Ulnar Nerve with T2 Hyperintense Rim Sign (Maroon Arrow) and Split Sign (Blue Arrow)

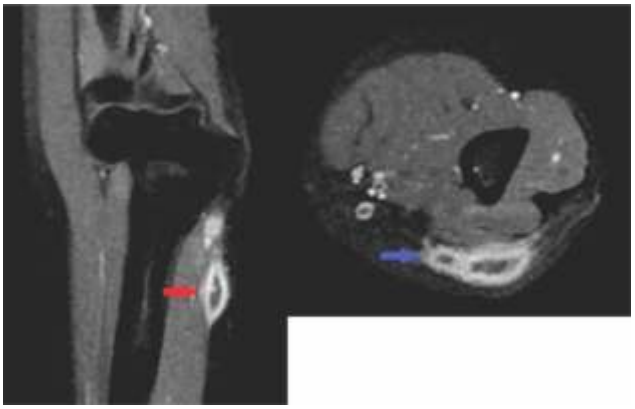

Fig. 2: T1WI Coronal (a) and Axial (b) Image showing Isointense Lesion with Slightly Hypointense Centre (Red Arrow) and a Daughter Lesion (Blue Arrow) Noted the Ulnar Nerve

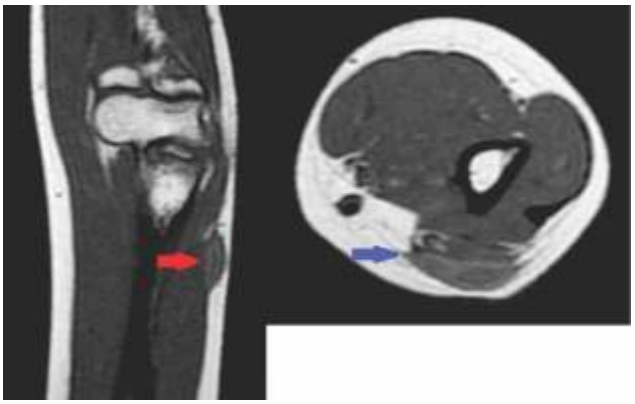

Fig. 3: T1WI Contrast Coronal (a) and Axial (b) Image showing Intense Peripherally Enhancing Lesion with Central Hypointensity Suggesting Necrosis (Red Arrow) and Daughter Lesion (Blue Arrow) Noted the Ulnar Nerve

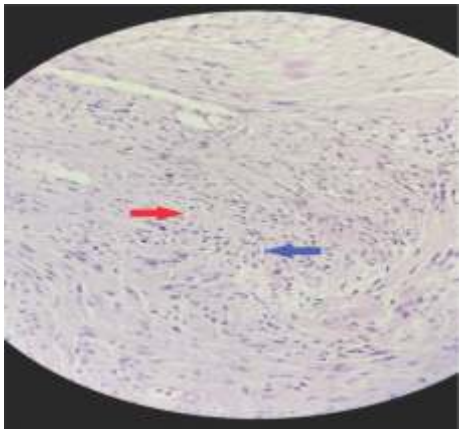

Fig. 4: Histopathological Image of the Resected Tumour showing Verocay bodies (Red Arrow) and Palisading (Blue Arrow)

Discussion:

PNST are divided into two types: schwannoma and neurofibroma. A peripheral nerve is composed of Schwann cells made of myelin and carry neuronal signals from the nodes of Ranvier down to the muscles. At cellular level both these tumours contain cells which are related to the normal Schwann cells, while at pathologic review these lesions are composed of epineurium and hence contain true capsules. Schwannomas are found to be eccentric to the affected nerve which is often displaced and appears as a peripheral mass. If present, this feature is especially helpful in distinguishing neurofibroma from schwannoma on the radiological images. As schwannomas grow inside the epineurium, it forms a capsule and hence can be resected en bloc. While neurofibromas are tumours arising from the fascicles of a nerve and do not create a capsule, so cannot be separated from the nerve. Both tumours are closely associated with nerves and as the tumours grow in size there is mass effect to the nerve fascicles causing pain and numbness [3-4]. While schwannomas are benign peripheral nerve sheath tumours that can be found anywhere in the body, head and neck are the most common location of these tumours, followed by the flexor compartment of the extremities. Although they are found sporadically and are usually solitary with no associated with Neurofibroma Type 1 (NF-1); multiple schwannomas are often found to be associated with NF-1. In head and neck, these lesions can occur in any of the peripheral, cranial and autonomic nerves; vestibulocochlear nerve being the commonest and accounting for the majority of cerebellopontine angle tumours. It is also associated with NF-2, especially when present bilaterally. Spinal schwannomas are seen as dumbbell shaped tumours arising from and causing widening of the neural canal [5-6].

When dealing with Schwannomas of the extremities, they are the most common solitary peripheral nerve sheath tumours. These tumours peculiarly are round to oval, slow-growing painless masses, they are firm in consistency and present with paraesthesia of the associated nerve. Flexor surface is most commonly affected in the upper limb as this compartment consists of the larger nerves of the lot [5].

Schwannomas are known to be affecting an age group of 20 to 40 years with no gender predilection [1]. These lesions are benign with complete recovery on surgical resection. Schwannomas do not recur after treatment and rarely undergo malignant transformation [6]. Schwannomas which are present since a long time owing to its characteristics of painless mass and due to its slow growing nature undergo calcifications, hyalinization and cystic degeneration and have distinguishing features on imaging are called ancient schwannomas [1].

Table 1: Features to Differentiate Peripheral Nerve Sheath Tumors (PNST)

Signs/Features	Schwannoma	Neurofibroma	Malignant PNST
Location of Tumor wrt nerve	Eccentric	Central	Central
Target sign	Present	Present (common)	Absent
Fascicular sign	Present	Present	Absent/Occasional
Split fat sign	Present	Present	Absent
T2 hyperintense rim	Present	Rare	Absent
Intratumoral cyst	Common	Rare	Occasional

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MR imaging is a modality of choice for diagnosis of these lesions. The two PNSTs can be very well differentiated on MRI by certain imaging features/signs, although a final diagnosis is often made by histopathological examination. On imaging, these lesions are classically eccentric to the nerve, fusiform shaped with tapering end. These lesions show low to intermediate signals on T1-weighted images, high signals on T2-weighted images and are easier to detect when the lesion arises from the anatomic location of the nerves of origin. On T2-weighted images these lesions can be either homogeneously hyperintense or can show "target sign" with central hypointensity due to fibrous stroma and peripheral hyperintensity due to myxomatous component. Target sign, although more common in neurofibroma, is also seen in schwannoma. And our case demonstrated the sign well. If the lesion appears heterogeneous, it suggests degeneration of the lesion which is most often cystic and is seen most commonly in schwannoma than neurofibroma [3].

"Fascicular sign" is a peculiar sign seen in neurogenic tumours and normal nerves and represents benign nature of the lesion, although occasionally these are focally present in the malignant variety of the same. Multiple ring-like hypointense foci within a hyperintense lesion on T2-weighted are nothing but the fascicular bundles."T2 hyperintense rim" is a thin rim of T2 hyperintensity seen most commonly in schwannomas. "Split fat sign" is the presence of fat at the upper and lower poles of the mass seen on T1-weighted images and suggests that the lesion is in the intramuscular planes. It is a characteristic of benign lesions as malignant lesions will obliterate this sign due to its infiltrative nature.

These signs are the tell-tale signs of neurogenic tumours and highly suggest their presence with proper characterisation of the said lesions providing guidance for a final diagnosis [2, 3, 7] (Table 1). In case of malignant PNSTs, the lesions are more infiltrative, greater (> 5cm in size) and are heterogeneous in signal intensity due to necrosis. Tell-tale signs of the PNSTs are often absent in the malignant form and they show perilesional oedema, bony destruction and often have solid enhancement pattern, peripheral enhancement when areas of central necrosis is present [6, 8, 9]. These lesions require a more aggressive treatment adjuvant to resection and timely follow up to look for recurrence although rare.

Other modalities of imaging of these masses are ultrasonography and computed tomography. On ultrasonography, these lesions appear as a well-defined hypoechoic echotexture lesion which is seen in continuity with the nerve and is eccentric in location to the nerve. On computed tomography, a homogenous mass lesion is noted along the anatomic location of the nerve. Variations are seen in case of ancient schwannomas which are associated with long standing nature and hence degenerations. There can be dystrophic calcifications, cystic changes and necrotic areas within the lesion [1].

Conclusion:

As discussed above, both the PNSTs show target sign, fascicular sign and split fat sign. Schwannomas are eccentric, whereas neurofibromas are more central in location in the nerve; they show homogeneous enhancement, schwannomas show a peripheral enhancement and another key differentiating sign is the T2 hyperintense rim which is seen in schwannoma and is absent in neurofibroma. Tracing the anatomical location thoroughly and recognising the characteristics of the lesions is important to distinguish the two as it guides the physician to aid the appropriate management.

References

1. Lin J, Martel W. Cross-sectional imaging of peripheral nerve sheath tumors: characteristic signs on CT, MR imaging, and sonography. *Am J Roentgenol* 2001; 176(1):75-82.
2. Kakkar C, Shetty CM, Koteshwara P, Bajpai S. Telltale signs of peripheral neurogenic tumors on magnetic resonance imaging. *Indian J Radiol Imaging* 2015; 25(4):453.
3. Beaman FD, Kransdorf MJ, Menke DM. Schwannoma: radiologic-pathologic correlation. *Radiographics* 2004; 24(5):1477-1481.
4. Albert P, Patel J, Badawy K, Weissinger W, Brenner M, Bourhill I, *et al.* Peripheral nerve schwannoma: a review of varying clinical presentations and imaging findings. *J Foot Ankle Surg* 2017; 56(3):632-637.
5. Crist J, Hodge JR, Frick M, Leung FP, Hsu E, Gi MT, *et al*. Magnetic resonance imaging appearance of schwannomas from head to toe: a pictorial review. *J Clin Imaging Sci* 2017; 7:38.
6. Chee DWY, Peh WCG, Shek TWH. Pictorial essay: imaging of peripheral nerve sheath tumours. *Can Assoc Radiol J* 2011; 62(3):176-182.
7. Yun JS, Lee MH, Lee SM, Lee JS, Kim HJ, Lee SJ, *et al*. Peripheral nerve sheath tumor: differentiation of malignant from benign tumors with conventional and diffusion-weighted MRI. *Eur Radiol* 2021;31(3): 1548-1557.
8. Kragha KO. Malignant peripheral nerve sheath tumor: MRI and CT findings. *Case Rep Radiol* 2015; 2015:241259.
9. Yu Y, Wu J, Ye J, Chen M. Radiological findings of malignant peripheral nerve sheath tumor: reports of six cases and review of literature. *World J Surg Onc* 2016; 14:142.

Author for Correspondence:

Dr. Prerna Anup Patwa, Department of Radiodiagnosis, Jawaharlal Nehru Medical College, Sawangi, Meghe, Wardha-442001, Maharashtra
Email: prernapatwa17@gmail.com Cell: 9869786498

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