MR IMAGING OF PERIPHERAL NERVE SHEATH TUMORS OF THE EXTREMITIES

Sirikan WONGSRISOONTORN MD[1], Pimjai SIRIWONGPAIRAT MD[1]
Suphaneewan JAOVISIDTH MD[1]

ABSTRACT

Two patients with peripheral nerve sheath tumors (PNST) of the extremities were presented, emphasizing on magnetic resonance (MR) imaging findings. In both cases, T2-weighted images showed inhomogeneously hypersignal intensity and contrast enhanced studies revealed intense enhancement predominantly at periphery. An oval shape or tapering of longitudinal end of the mass suggest its extension along longitudinally oriented structure such as neurovascular bundle.

Keywords : Nervous system; neoplasm Nerves; peripheral, MR

INTRODUCTION

PNST most frequently results from proliferation of neural supporting tissues and the two most common entities of PNST are neurofibroma and neurilemoma (schwannoma).[1] PNST can be found as a solitary lesion or as a part of clinical presentation of neurocutaneous syndrome e.g. neurofibromatosis.

Diagnosis of PNST while having many features suggestive of such syndrome is not difficult. But for a solitary PNST in the extremity, the diagnosis may be problematic. We reported 2 cases of PNST of the extremity emphasizing on their MR characteristics.

CASE REPORTS:

Case 1:

An eighty-seven-years-old female patient presented with a palpable, gradually enlarging mass in her right arm for 1 year with paresthesia along the course of ulnar nerve. Physical examination revealed a hard mass locating in posterior aspect of right arm. Plain film showed soft tissue mass corresponding to the palpable lesion without calcification or ossification. MR imaging revealed a 2.7x 2.7x 4.5 cm-size mass lesion, having well-defined margin with tapering of its inferior aspect, within the triceps muscle of the right arm (Fig. 1 a). On T1-weighted images, the mass showed slightly inhomogeneously isosignal with surrounding muscle (Fig. 1 b). On T2-weighted images, the mass exhibited inhomogeneously hypersignal; composing of peripheral hypersignal area with central mixed hyper- & hyposignal intensities (Fig. 1 c). After gadolinium administration; thick, irregular, intense enhancement of the mass was observed peripherally in fat suppressed T1-weighted images (Fig. 1 d). The ulnar nerve could not be well separated from the mass. No muscle atrophy, bone involvement, internal calcification, or signal void was detected. Surgery was

Department of Radiology, Ramathibodi Hospital, Faculty of Medicine, Mahidol University, Rama 6 Street, Bangkok 10400, Thailand.

performed and revealed a 3-cm mass with necrosis, with questionable its location within the ulnar nerve. Pathologic examination disclosed an oval-shape mass in the center of ulnar nerve trunk, with middle part of the nerve trunk blended into the deep part of the mass. Histologic diagnosis was schwannoma with focal hemorrhagic infarction.

Case 2:

A 42-year-old male patient presented with pain and ill-defined nodular mass in his right forearm for 8 months. He had no fever and there was no change in size of the mass as compared to the first detection. Ultrasonography disclosed a hypoechoic well-defined solid mass. MR imaging was performed later, and revealed a 2.0x 1.3x 1.2 cm-size well-defined oval-shape mass in right forearm, showing homogeneously slightly hypersignal intensity on T1-weighted images, inhomogeneously hypersignal intensity on T2-weighted images particularly at periphery (Fig 2), and inhomogeneously intense enhancement more at periphery after intravenous gadolinium administration. No associated muscle atrophy, bony involvement, internal calcification, cyst formation or signal void was detected. Excisional biopsy was performed and the section revealed a schwannoma.

al. MR imaging of extracranial nerve sheath tumors. J Comput Assist Tomogr 1992; 16: 448-453

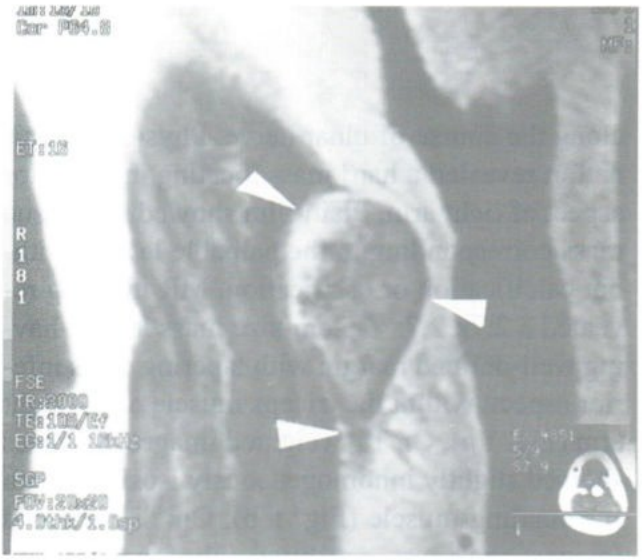

Figure 1 a-d. Case 1.

Fig. 1 A : T2-weighted sagittal MR image shows inhomogeneously hypersignal teardrop-shape mass (arrowheads). Inferior tapering of the mass indicates extension along longitudinally oriented structures.

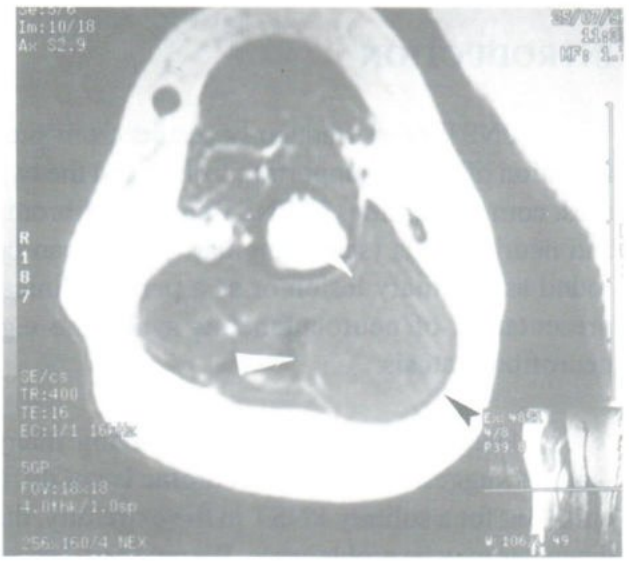

Fig. 1 B : T1-weighted axial MR image reveals the mass to be slightly inhomogeneously isosignal to surrounding muscles (arrowheads).

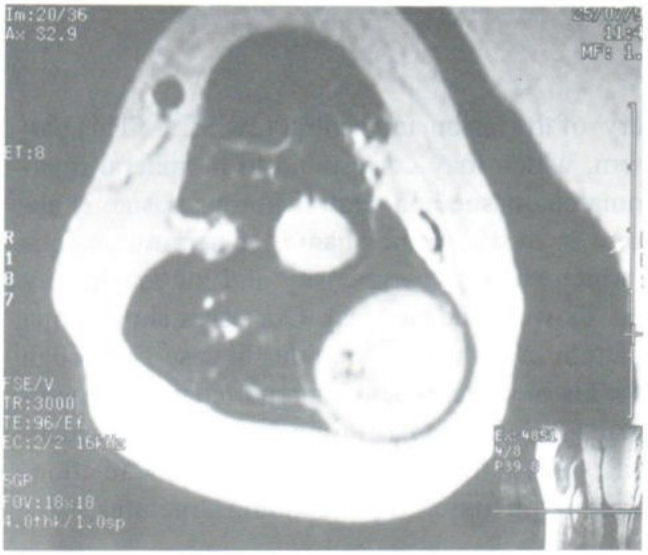

Fig. 1 C : T2-weighted axial MR image discloses peripheral hypersignal area whereas the central area shows mainly hyposignal intensity.

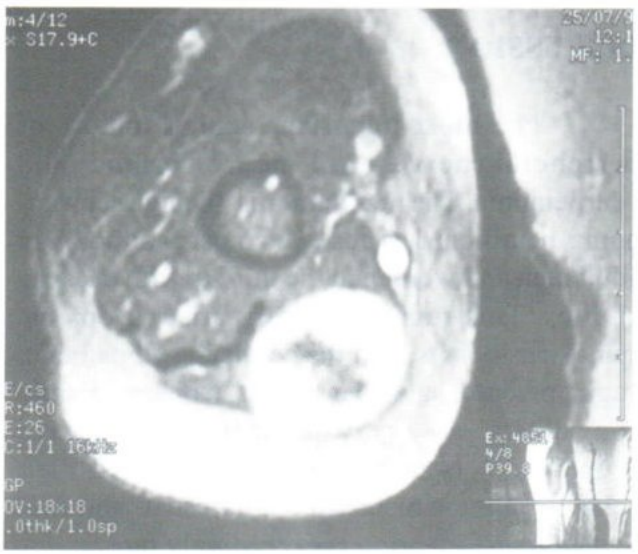

Fig. 1 D : Gadolinium enhanced fat-suppressed T1-weighted axial MR image exhibits thick, irregular peripheral enhancement.

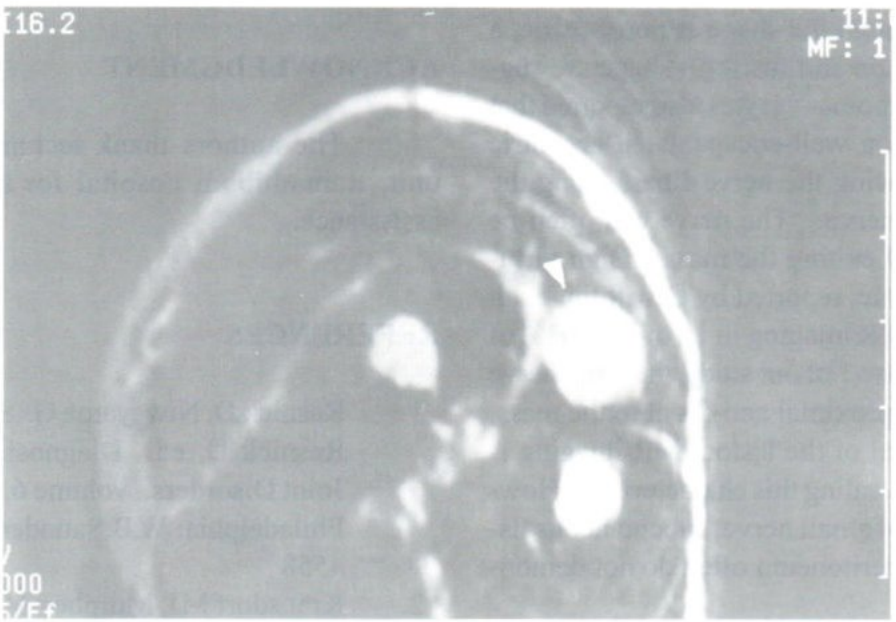

Figure 2 . Case 2.

Fig. 2 : T2-weighted axial MR image reveals the mass to have inhomogenously hypersignal on T2-weighted image predominantly at periphery (arrowhead).

DISCUSSION

The most important benign tumors of peripheral nerves are neurofibroma and neurilemoma. The neurilemoma (benign schwannoma, neurinoma) is observed most commonly in adult men and women in the third to fifth decades of life.[1] This tumor typically arises from the spinal nerve roots, and the cervical, sympathetic, vagus, peroneal, and ulnar nerves. It predominantly is a solitary, slow growing, and non-aggressive neoplasm.[1-3] Neurilemomas are benign in their behavior; tumor recurrence is unusual, and malignant transformation is exceedingly rare.[4]

There are only a few reports concerning MR imaging findings of PNST, especially of the extremities.[5,6] The size of PNST are variable and there is no clue of diagnosis. Data from 2 series,[7,8] comprising 22 benign and 8 malignant peripheral nerve sheath tumors, also showed great variability in size. Although the shape is nonspecific, a well-defined margin and fusiform shape are suggestive of neurilemoma.[6,9] It was also reported that neurilemoma has a well-encapsulated eccentric nature and displacing the nerve fibers along the long axis of the nerve.[9] The nerve can often be seen entering and exiting the mass, a sign virtually pathognomonic, reported by Cerofolini et al. to be present on MR imaging in 16 of 17 (94%) of their cases.[10] In case 1 of our study, the ulnar nerve can be identified proximal and distal to the mass, but not at the level of the lesion. MR imaging is of great help in revealing this characteristic. However, the lesions of small nerve, subcutaneous tissue and the retroperitoneum often do not demonstrate this finding.[3]

Regarding the internal signal intensity (SI), most of PNST reveal relatively low SI on T1-weighted MR images and high SI on T2-weighted images.[11] On T2-weighted images and on T1-weighted images following intravenous administration of gadolinium, a target pattern may be identified,[1,6] consisting of increased SI at the periphery of the lesion and central low SI.[1,12] This pattern, which may correspond to peripheral myxomatous tissue and central fibrous tissue, is absent in cystic, hemorrhagic, or necrotic lesions (which show hyperintensity and inhomogeneity on T2-weighted images). Our cases show target pattern in both, more obvious in case 1 although the histologic examination reported internal hemorrhagic infarction. MR imaging also allows identification of the nerve trunk of origin and relationship of the tumor to surrounding structures.[1]

In conclusion, MR characteristics of peripherally increased SI on T2-weighted images and on T1-weighted images after contrast injection, an oval shape or tapering longitudinal end of the lesion, and demonstration of nerve entering and exiting the mass should suggest the diagnosis of PNST.

ACKNOWLEDGMENT

The authors thank technicians of MRI unit, Ramathibodi hospital for their technical assisstance.

REFERENCES

1. Resnick D, Niwayama G. Soft tissues. In: Resnick D, ed. Diagnosis of Bone and Joint Disorders. Volume 6. Third edition. Philadelphia: W.B.Saunders, 1995: 4549-4558
2. Kransdorf MJ, Murphey MD. Neurogenic tumors. In: Kransdorf MJ, Murphey MD, eds. Imaging of Soft Tissue Tumors. Philadelphia: W.B.Saunders, 1997: 235-254
3. Sundaram M, Sharafuddin MJA. MR imaging of benign soft tissue masses. Magn Reson Imaging Clin N Am 1995; 3: 620-621

4. Carstens H, Schrodt G. Malignant transformation of a benign encapsulated neurilemoma. Am J Clin Pathol 1969;51:144-146
5. Murphey MD, Smith WS, Temple HT. Imaging of malignant peripheral nerve sheath tumors. Radiology 1996;201 (P): 359-360
6. Smith WS, Murphey MD, Temple HT. Imaging of benign peripheral nerve sheath tumors with pathologic correlation. Radiology 1996;201 (P): 360
7. Levine E, Huntrakoon M, Wetzel LH. Malignant nerve-sheath neoplasms in neurofibromas: Distinction from benign tumors by using imaging techniques. AJR Am J Roentgenol 1987;149:1059-1063
8. Stull MA, Moser RP Jr, Kransdorf MJ, et al: Magnetic resonance appearance of peripheral nerve sheath tumors. Skeletal Radiol 1991;20:9-13
9. Suh J-S, Abenoza P, Galloway HR, Everson LI, Griffiths HJ. Peripheral (extracranial) nerve tumors: Correlation of MR imaging and histologic findings. Radiology 1992;183:341-346
10. Cerofolini E, Landi A, DeSantis G, et al: MR of benign peripheral nerve sheath tumors. J Comput Assist Tomogr 1991;15: 593-597
11. Burk DL, Brunberg JA, Kanal E, et al. Spinal and paraspinal neurofibromatosis: Surface coil MR imaging at 1.5 T. Radiology 1987;162:797-801
12. Varma DGK, Moulopoulos A, Sara AS, et al. MR imaging of extracranial nerve sheath tumors. J Comput Assist Tomogr 1992;16:448-453