Magnetic Resonance Tractography in Malignant Peripheral Nerve Sheath Tumor

Ravindra Bhimrao Kamble, Sanjeev Jayateerth Mudakavi, Raghvendra Seetaram, Diwakar Nagraj
Department of Radiology and Neurology, Vikram Hospital, Bengaluru, Karnataka, India

Correspondence: Dr. Ravindra B Kamble, Vikram Hospital, #71/1, Millers Road, Bengaluru - 560 052, Karnataka, India.
E-mail: drravindra31@rediffmail.com

ABSTRACT

Malignant peripheral nerve sheath tumors (MPNSTs) are rare soft tissue neoplasms, which occur in isolation or due to the malignant transformation of benign neurofibroma. Conventional features of MPNST are very well described. We intend to present diffusion tractography findings in a case of MPNST.

Key words: Magnetic resonance imaging; peripheral nerve; tractography; tumor

Introduction

Peripheral nerve sheath tumors are either benign or malignant. Initially for malignant peripheral nerve sheath tumor (MPNST), various terminologies such as malignant schwannoma, malignant neurilemmoma, neurofibrosarcoma, and neurogenic sarcoma were used. To avoid confusion, World Health Organization coined the term MPNST.[1] These tumors may arise *de novo* from peripheral nerves or may arise from preexisting neurofibroma.[2] There are many publications describing the imaging features of benign and malignant peripheral nerve tumors, but magnetic resonance imaging (MRI) with tractography features of MPNST is described in very few cases.[3-5]

We report a case of MPNST in a young 25-year-old patient with conventional MRI features along with diffusion tractography findings.

Case Report

A 25-year-old male patient presented to our hospital with numbness and paresthesia of the left foot which was episodic initially and gradually increased in frequency for a period of 6 months, with the progressive involvement of thigh and leg. Lately, he had also noticed thinning of the left calf muscles. Initial MRI investigations of the lumbar spine and pelvis done elsewhere were normal.

He then presented in our hospital for persisting numbness. On examination, he had a mild swelling in the posterior aspect of the left thigh, which was tender and Tinel's sign was elicited clinically. Patient also had multiple swellings over the body since childhood. Nerve conduction and electromyographic studies suggested sciatic nerve lesion in the left thigh. Screening ultrasound scan showed a large fusiform, hypoechoic mass in the intermuscular plane in the posterior mid-thigh region on the left side (not shown). Conventional MRI sequences showed well-defined lobulated mass involving sciatic nerve with tapering ends. The lesion was hypointense on T2-weighted and isointense to muscle on T1-weighted with a split fat sign suggesting intermuscular origin [Figure 1]. Nerve fascicles were swollen and were well appreciated (fascicular sign). Perilesional edema was seen in the inferior aspect of the lesion with hyperintense signals in hamstring muscles [Figure 2]. Few uninvolved sciatic nerve fibers were seen splayed by the mass lesion [Figure 3].

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Diffusion tractography (TR 2900, TE 94, 30 different directions, Bandwidth 1302) was done and after postprocessing on Neuro 3D (three-dimensional) software, fractional anisotropy (FA), and apparent diffusion coefficient (ADC) values were calculated in the proximal nerve and in the tumor. Tractography images showed splayed sciatic nerve fibers at the periphery of the lesion as shown in Figure 4. FA values in the sciatic nerve above the lesion were 288 ± 85.5 and ADC values were 1662 ± 139.5. FA values in the tumor were significantly reduced (164 ± 59) with no change in ADC values as compared to the proximal nerve (1670 ± 108). Patient underwent biopsy with debulking of the lesion. Histology showed it to be high-grade neoplasm with nests and sheets of malignant round cells which are suggestive of a high-grade MPNST with primitive neuroectodermal tumor differentiation.

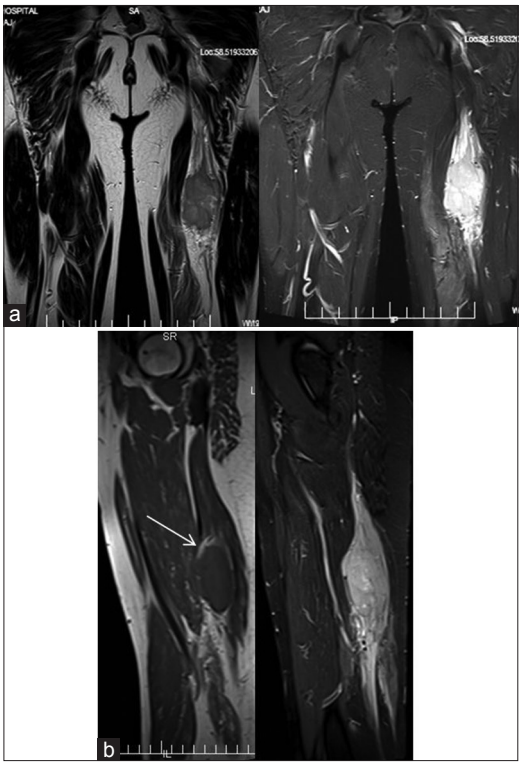

Figure 1: Coronal image in (a) (T2-weighted left and proton density (PD) fat sat right) and sagittal in (b) (T1-weighted [T1W] left and PD fat sat right) showing fusiform mass lesion with lobulated contours and split fat sign seen in T1W sagittal image (white arrow)

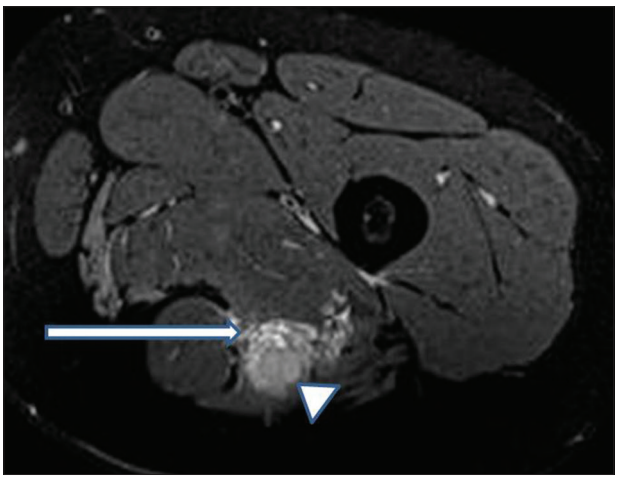

Figure 3: Proton density fat sat axial image showing splayed fibers (long arrow) and swollen fibers due to tumor (arrowhead)

Discussion

The incidence of MPNST is 0.001% in the general population.[1] These tumors most commonly affect individuals in 20–50 years of age[2] although cases have also been reported in the elderly. These are rare tumors and form about 5–10% of all soft tissue sarcomas and tend to occur more commonly in neurofibromatosis.[6]

Magnetic resonance imaging is a best imaging modality for these neuronal soft tissue tumors. However, after the introduction

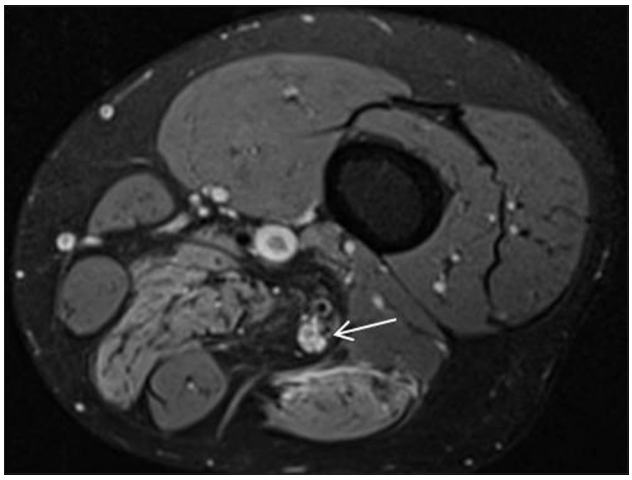

Figure 2: Axial proton density fat sat image showing selective hamstring muscle hyperintense signal changes with hyperintense sciatic nerve (white arrow)

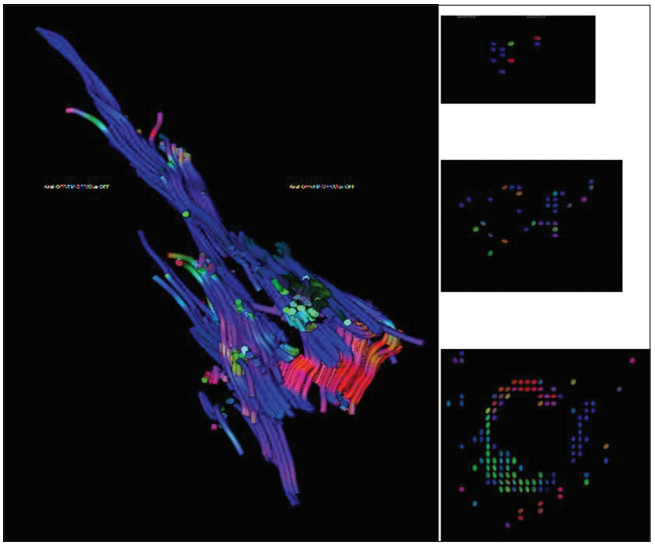

Figure 4: Tractography image showing splayed nerve fibers around the tumor

of high-resolution MR neurography technique in 1980, the imaging of nerve fibers and its lesions has been significantly revolutionized. This technique includes conventional anatomic two-dimensional and 3D images as well as functional MRI using diffusion based sequences (diffusion-weighted imaging and diffusion tensor imaging [DTI]).[5,7,8]

There are various features described on conventional imaging to differentiate between benign and MPNSTs. Fusiform shape with tapering ends with hyperintense signals on T2 and hypo to isointense on T1 suggests benignity.[3] In addition, benign tumors have well-defined margins with split fat and target signs.

Malignant tumors, however, are larger in size with irregular margins and loss of nerve continuity.[4] Intratumoral cysts/hemorrhages and adjacent soft tissue changes in the form of perilesional edema-like zone on T2 are also suggestive of malignant masses.[9]

On gadolinium-enhanced images, the presence of heterogeneous nodular peripheral enhancement is seen in malignant PNSTs whereas, focal central enhancement supports benignity.[10] Sometimes, heterogeneity and calcifications can be seen in benign tumors mimicking malignancy.[11]

Thus, conventional imaging can show both benign and malignant patterns in same lesion adding to the diagnostic dilemma. In our case, we had split fat sign and uninvolved fibers suggesting benignity as well as malignant features like large size with perilesional edema- like zone.

To overcome these limitations of conventional imaging, DTI (tractography) can be an important tool to facilitate the diagnosis. Tractography demonstrates normal or partial discontinuity of the nerve tracts in the benign tumors and partial to near-total discontinuity in malignant tumors.[5]

Other parameters such as ADC and FA are useful for tumor characterization on DTI. Low ADC and FA are indicative of malignant pathology.[5,12] Low FA and ADC are due to widening of the space between the axon and its covering due to the axonal blockade, vascular congestion, and distal Wallerian degeneration.[12]

In the present case, diffusion tractography showed peripheral displacement of few nerve fibers with few others showing partial discontinuity. ADC values in the lesion were similar to the nerve proximal to the lesion. However, FA values were significantly lower suggesting malignancy. Though, splaying of nerve fibers on tractography is generally indicative benign lesion, it can also be seen in malignant tumors as observed in our case.

Conclusion

Conventional imaging alone may be inadequate for characterizing nerve tumors, addition of DTI facilitates diagnosis, ADC values may be normal in malignant nerve sheath tumors, splaying of nerve fibers may not always represent benign tumors and low FA values should be considered as prime indicator of malignancy.

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Conflicts of interest
There are no conflicts of interest.

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