CASE REPORTS

PLEXIFORM NEUROFIBROMATOSIS AND THE "TARGET SIGN" ON MRI

PLEKSİFORM NÖROFİBROMATOZİS VE MRI' DA "HEDEF PATERNİ"

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SUMMARY: *T2-weighted MR imaging (MRI) of soft tissue tumors of neural origin may show round lesions with a central hypointensity and a hyperintense rim resembling a target. This "target sign" on T2-weighted MR imaging is helpful in differentiating neurofibromas from malignant peripheral nerve sheath tumors. Here, we report a five-year-old boy with lower limb plexiform neurofibromatosis and discuss the "target sign" on MRI.*

Key Words: *Plexiform Neurofibromatosis, MR, Target Sign.*

ÖZET*: Nöral kaynaklı yumuşak doku tümörleri, T2-ağırlıklı MR görüntülerinde santrali hipointens çevresi hiperintens hedefe benzer yuvarlak lezyonlar oluşturabilir. T2-ağırlıklı MR görüntülerinde izlenen "hedef paterni", nörofibromaların malign periferal sinir kılıfı tümörlerinden ayrılmasında yardımcıdır. Bu çalışmada alt ekstremite yerleşimli pleksiform nörofibromatozisli beş yaşındaki bir çocuk sunulmuş ve MR incelemesindeki "hedef paterni" tartışılmıştır*

Anahtar Kelimeler: *Pleksiform Nörofibromatozis, MR, Hedef Paterni.*

INTRODUCTION

Malignant degeneration of peripheral neurofibromas is the leading cause of cancer death in patients with type one neurofibromatosis (NF1). Therefore, it is important to distinguish benign from malignant nerve sheath tumors. MR imaging is the modality of choice in diagnosis and for defining the extent of soft tissue tumors (1). MRI has been widely used to evaluate nerve sheath tumors (2). A "target sign" on T2-weighted images is one of the MR findings that support the benignity of peripheral nerve sheath tumors. The "target sign" is defined as increased signal intensity in the peripheral zone and decreased signal intensity in the central part of nerve sheath tumors (3).

CASE REPORT

A five-year-old boy with NF-1 was first admitted to hospital because of a one-week history of right knee pain. On physical examination, there were seven cafe-au-lait macules in the abdominal region and on the patient's back, the largest of which was 0.5 mm in diameter. His right lower limb was larger than his left.

On X-ray roentgenograms, there was pelvic asymmetry. The femur, tibia and fibula were larger on the right side with associated soft tissue density.

A diagnosis of NF-1 is established when two or more of the following findings are present: six or more 5 mm or larger cafe-au-lait spots, one plexiform neurofibroma or two or more neurofibromas of any type, axillary or inguinal region freckling, optic nerve glioma, two or more hamartomas of the iris, and one or more diagnostic osseous lesions. Osseous lesions are

found in 30-80% of patients and usually affect the anterolateral part of the tibia and fibula. Focal gigantism, and solitary or multiple cyctic lesions may be included. Periosteal dysplasia, subendotelial sclerosis and medullary signal changes are other manifestations of NF-1 (4).

An MR examination was performed on a 1 Tesla MR unit (Signa, HiSpeed, GE Medical Systems, Milwaukee, WI, USA) and included the following sequences: pre- and post-gadolinium T1-weighted axial and coronal images and T2-weighted axial and sagittal images. T1-weighted coronal images revealed a huge mass, consisting of multiple tubular structures, extending from the inguinal region to the ankle, on the right side. On T1-weighted axial and coronal images, the central portion of the mass was isointense with muscle, while the peripheral zone was hypointense. Following an intravenous gadolinium injection, the central portion of the lesion showed marked contrast enhancement (Fig. 1, 2). On T2-weighted axial and sagittal images, the central zone of the mass was isointense with muscle, and the peripheral zone was hyperintense when compared with the fat (Fig. 3).

The diagnosis of plexiform neurofibromatosis was made based on the clinical and MRI findings. The histopathologic examination of the biopsy material obtained from

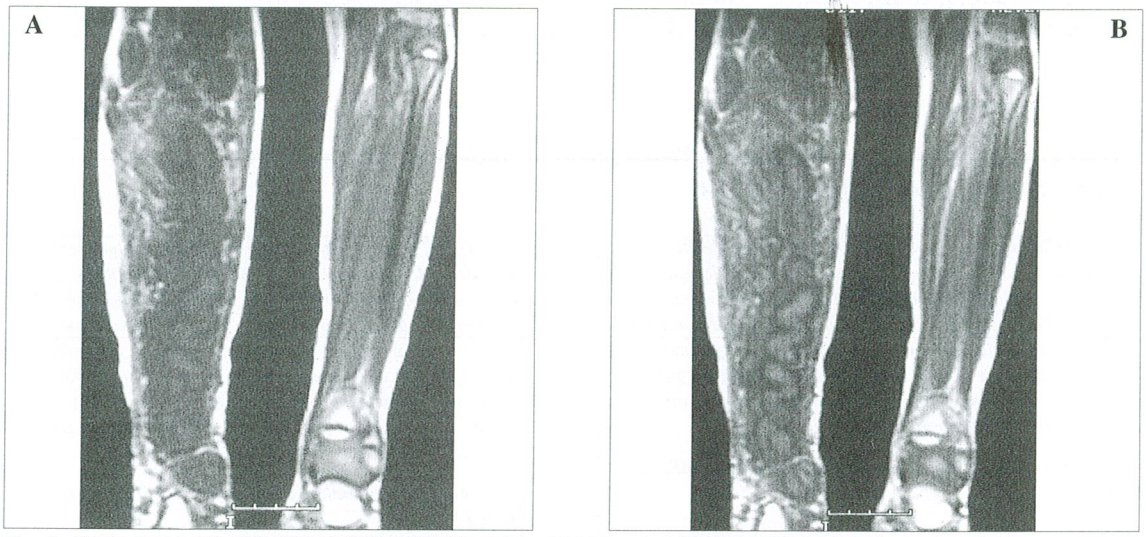


Fig. 1: Coronal pre- (A) and postgadolinium (B) T1-weighted MR images. The central zone of the lesion shows more contrast enhancement than the peripheral regions (B).

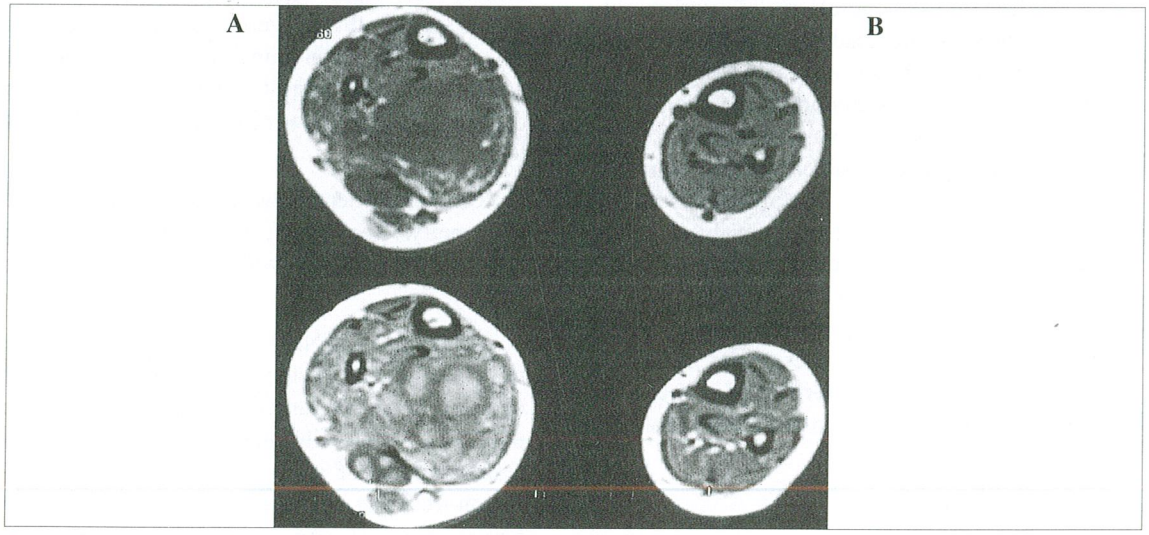
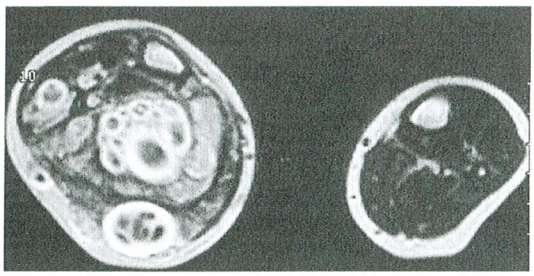
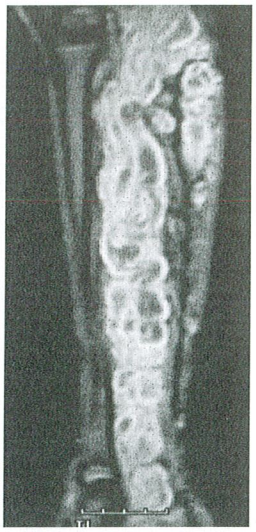


Fig. 2: Axial pre- (A) and postgadolinium (B) T1-weighted MR images. The central portion of the lesion shows marked contrast enhancement (B).

A





B

Fig. 3: Axial (A) and sagittal (B) T2-weighted MR imaging shows the target sign . The central part of the lesion is hypointense (isointense to muscle) and the peripheral zone of the lesion is hyperintense.

the popliteal fossa also supported the diagnosis of plexiform neurofibromatosis. The patient was treated with conservative therapy.

DISCUSSION

It is clinically difficult to distinguish benign from malignant nerve sheath tumors. The optimal treatment of malignant peripheral nerve sheath tumors is surgical excision. However, excision of benign neurofibromas, which contain intermingled neural elements and Schwann cells, usually compromises the affected nerve. Furthermore, blind biopsy of large complex tumors may be subject to sampling errors. Thus, a test that could differentiate benign from malignant lesions would prevent unnecessary morbidity (5). MRI is the modality of choice in diagnosis and for differentiating benign and malignant peripheral nerve sheath tumors.

On T2-weighted MR images, the central zone of plexiform neurofibromas is hypointense (isointense to muscle), and the periphery has equal or higher signal intensities when compared with the fat. This depends on the higher collagen content of the central zone and the higher proteoglican content, which is rich in water, of the peripheral regions. Suh et al. defined this pattern as the target sign (3).

Lin et al. have described the sonographic equivalent of the target sign on MRI (6). In their report, numerous lesions demonstrating a hyperechoic central region and a hypoechoic periphery in a patient with plexiform neurofibromatosis were defined. The hypoechoic outer zone, which is correlated with fluid content, corresponded to the high signal peripheral zone on T2-weighted MRI examination. The hyperechoic central zone, histologically correlated with a dense fibrocollagenous center, corresponded to the low signal central zone on T2-weighted MRI examination. (6). The CT equivalent of the target sign has also been mentioned in numerous reports (7).

Suh et al. found that seven of 10 neurofibromas demonstrated the target sign , not seen in their series of neurofibrosarcomas (3). Varma et al. reported the target sign in 12 of 23 neurofibromas and schwannomas and in zero of nine malignant nerve sheath tumors (8). Bhargava et al. reported the target sign in all 12 neurofibromas but in only one of the neurofibrosarcomas (5). Hence, although not conclusive, the target sign may be a supporting finding in differentiating benign and malignant nerve sheath tumors.

In conclusion, the target sign on MRI may be a basis for differentiation of benign and malignant nerve sheath tumors.

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