

[CASE REPORT]

Magnetic Resonance Neurography in a Patient with Distal Neuralgic Amyotrophy

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Abstract:

The pathophysiology of neuralgic amyotrophy (NA) remains to be elucidated. However, high-resolution magnetic resonance imaging and ultrasound sonography have provided new insights into the mechanism underlying the development of NA and its diagnosis. We report a case of idiopathic distal NA with hyperintensity and thickening in the inferior trunk extending to the posterior and medial fasciculus of the left brachial plexus, which was detected by magnetic resonance neurography (MRN) with diffusion-weighted whole-body imaging with background body signal suppression (DWIBS). The abnormal signal intensity diminished after the improvement of symptoms following corticosteroid treatment. MRN with DWI can help diagnose distal NA and evaluate the post-therapeutic response.

Key words: neuralgic amyotrophy, magnetic resonance imaging, magnetic resonance neurography, diffusion-weighted whole-body imaging with background signal suppression, distal

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Introduction

Neuralgic amyotrophy (NA) or Parsonage-Turner syndrome is characterized by acute severe pain followed by patchy paresis or atrophy in the upper extremities (1, 2). Immunotherapy, such as corticosteroids in the acute phase of NA, can shorten the duration of pain and functional recovery (3, 4). Thus, an early diagnosis is essential.

Magnetic resonance neurography (MRN) with diffusion-weighted whole-body imaging with background body signal suppression (DWIBS) can visualize the peripheral nerves of the whole body. We previously reported enlarged nerve diameters and diffusion limitations in chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) patients with peripheral nerve impairment (5).

In the present study, we report a case of idiopathic distal NA (dNA), in which high-signal thickening of the brachial plexus - which resolved with treatment - was observed on MRN with DWIBS in the acute phase of the disease.

Case Report

A 21-year-old healthy right-handed man was admitted to our hospital due to acute severe and intolerable pain in the left arm, hand, and fingers (fourth and fifth) followed by progressive weakening of his finger muscles over a 2-week period. The pain occurred at rest and was alleviated by keeping the extremity at elbow flexion and shoulder adduction. Moreover, the pain was exaggerated at night, causing sleep disturbance, and did not respond to the usual analgesic treatment. He reported no history of vaccination, smoking, trauma, or infection. His medical and family histories were unremarkable. A neurological examination showed mild weakness of the left wrist flexors, the extensor pollicis brevis (EPB), the second and third flexor digitorum profundus (FDP), and the abductor pollicis brevis (APB), and moderate to severe weakness of the left first dorsal interossei (FDI), the abductor digiti minimi (ADM), the fourth and fifth FDP, and the extensor digitorum (ED). He did not show any muscle weakness in the left triceps brachii, brachioradialis, or

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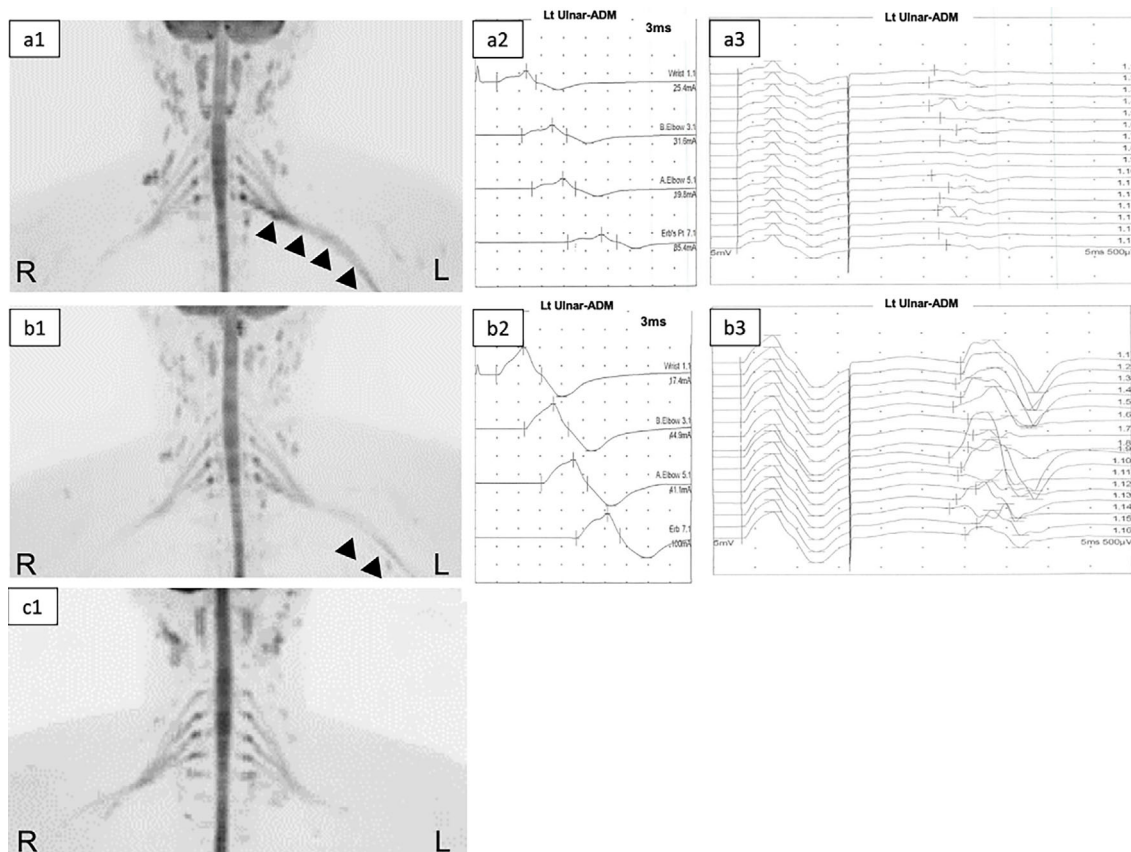


Figure. a1: Magnetic resonance neurography (MRN) with diffusion-weighted whole-body imaging with background body signal suppression (DWIBS) showed abnormal thickening and hyperintensity in the lower trunk of the left brachial plexus with distal extension of the signal intensities. a2, a3: A nerve conduction study and F-wave study of the left ulnar nerve prior to treatment showed a decreased amplitude and F-wave frequency. b1: After immunotherapy, MRN with DWIBS showed decreasing enlargement in the left inferior trunk, but the signal intensity in the posterior fasciculus of the brachial plexus remained mildly increased. b2, b3: The decreased amplitude and F-wave frequency showed an improvement following immunotherapy. c1: A DWIBS image of a 36-year-old healthy man shows a normal high signal intensity in the spinal cord and brachial plexus nerves.

wrist extensors. Slight atrophy of the ADM and interossei muscles was observed. Dysesthesia and hypesthesia were noted on the left C7-Th1 dermatomes, most prominently in the C8 distribution. Morley's test, Allen's test, and Wright's test did not show any abnormalities.

The results of blood tests and a cerebrospinal fluid analysis were within the normal limits. A nerve conduction study showed mild decreased compound muscle action potential (CMAP) without the decrement of conduction velocity, sensory nerve action potential, or F-wave frequency in the left ulnar nerve (Figure a2, a3). The patient did not give his consent for needle electromyography due to severe pain. Cervical magnetic resonance imaging (MRI) showed no noticeable abnormalities. MRN with DWIBS revealed significant enlargement and an increased signal intensity of the inferior trunk extending to the posterior and medial fasciculus of the left brachial plexus (Figure a1) in comparison to a healthy control (Figure c1). Thus, dNA was diagnosed on the basis of the clinical course and the neurological, neurophysiological, and MRN findings.

Intravenous methylprednisolone (1 g/d, 3 days) was administered, switched to oral prednisolone (30 mg/d), and then gradually tapered with the dosage reduced by 5 mg every 2 weeks until it reached 20 mg and 2 mg every 2 weeks until it reached 10 mg. At present, we are slowly tapering off prednisolone with the dosage reduced by 1 mg every 2 weeks. The severe pain disappeared, and the muscle strength improved during the treatment, with the exception of slight weakness of the FDI and ADM. Slight hypesthesia in the ulnar nerve distribution persisted. At approximately 3 months after treatment, MRN showed decreasing enlargement in the left inferior trunk (Figure b1), while the signal intensities of the posterior fasciculus of the brachial plexus remained mildly increased. The nerve conduction study showed an improvement in the CMAP and in the F-wave frequency (Figure b2, b3).

Discussion

Our case showed weakness and mild atrophic change with

minor sensory disturbance restricted to the forearm, hands, and fingers. This was preceded by acute severe pain that became exacerbated at night and at rest. Although cervical radiculopathy is the most critical differential diagnosis for dNA, C8 (FDI, ADM, FDP, EPB, and ED), C7 (wrist flexor), and Th1 (APB) myotomes were involved. A sensory disturbance was observed in the C7-Th1 regions during the early course of the illness. Cervical MRI and X-ray did not show any abnormalities. The nerve conduction study showed mild left ulnar nerve involvement. The findings of MRN with DWIBS supported the abnormalities of the inferior trunk and the posterior and medial fasciculus in this patient. These clinical, electrophysiological, and radiological findings supported the diagnosis of dNA (1-3).

Typical patients with NA show paresis and atrophy, predominantly in the shoulder muscles, corresponding to the involvement of the upper and middle plexus. A dNA where weakness is limited to the forearm, hand, and finger muscles is rare (3, 6), although the hand and finger muscles were involved in 26.3-36.3% of all patients (3). MRN with DWIBS enabled the visualization of the entire brachial plexus and provided evidence of the involvement of the inferior trunk and posterior and medial fasciculus in our patient with dNA. This was similar to the schematic representation in a seminal review (2). Whether the abnormal signal in this case is specific for NA is controversial; however, a typical clinical course, as shown in this case, is considered to be suggestive of NA. Although some reports showed nerve thickening of the brachial plexus and peripheral nerves in CIDP and Charcot-Marie-Tooth disease by MRN (7, 8); however, it is possible to differentiate based on the clinical symptoms, the disease course, and the electrophysiological findings.

The early administration of steroids in the treatment of NA is expected to ameliorate the pain and recover muscle strength (1, 2, 4). The early evaluation and management of the condition is crucial for avoiding subsequent complications.

The pathophysiology of NA remains poorly understood. Previous imaging studies demonstrated that inflammation rarely occurred in the brachial plexus (9); however, no reports have described the findings of MRN with DWIBS. The abnormal MRN with DWIBS findings in this study can be associated with inflammation. However, the pattern of weakness and sensory disturbance was well explained by mononeuritis multiplex rather than brachial plexitis. Muscle

weakness in NA shows a patchy distribution and all of the muscles innervated by the same nerve are not necessarily involved equally. Our patient had muscle weakness in the ECD and EPB, which are innervated by the radial nerve, but no weakness in brachioradialis, triceps brachii, or wrist extensors. Further studies, including high-resolution MRI and an ultrasound investigation (10), are necessary to elucidate the gap between the brachial plexus findings and the clinical presentation.

Conclusion

MRN with DWI is helpful for the diagnosis of dNA and the evaluation of the post-therapeutic response.

The authors state that they have no Conflict of Interest (COI).

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