



An atypical chronic inflammatory demyelinating polyradiculoneuropathy that radiologically mimicking neurofibromatosis: Case report

Cetin Kursad Akpınar^{a*}, Hakan Dogru^b, Kemal Balci^b, Murat Terzi^b

^aDepartment of Neurology, Vezirkopru State Hospital, Samsun, Turkey

^bDepartment of Neurology, Medical Faculty, Ondokuz Mayıs University, Samsun, Turkey

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ABSTRACT

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* Correspondence to:

Çetin Kürsâd Akpınar
Department of Neurology,
Vezirkopru State Hospital,
Samsun, Turkey
e-mail: dr_ckakpinar@hotmail.com

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Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) is an uncommon demyelinating disorder with a relapsing and remitting or continuously progressive course. Patients may have motor and sensory involvement, but generally motor involvement may be more prominent and more severe in lower extremities. CIDP is a treatable neuropathy that is challenging to diagnose and has a broad spectrum of presentations. When ranked by the descending frequency, postural tremor in the arms, peripheral nerve thickening, papilledema and facial or bulbar weakness, respiratory failure and autonomic dysfunction can be seen in patients with CIDP. Demyelinating neuropathy predominantly affects spinal roots, plexuses and proximal nerve trunks and thickened nerves can be palpable in about 10% of the patients. In patients with atypical presentation, the diagnosis of CIDP may be delayed and hypertrophic nerve roots have been reported in CIDP patients with delayed diagnosis. Magnetic Resonance Imaging (MRI) may be helpful in the diagnosis of CIDP by excluding the compressive or structural lesions that may lead to polyradiculopathy and MRI may also show hypertrophy of nerve roots and inflammatory processes in CIDP. We presented a patient with CIDP who had MRI findings of diffuse enlargement and mild enhancement of roots and extraforaminal segments of nerves in all segments.

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1. Introduction

Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) is an autoimmune disorder of the peripheral nervous system accompanied by symmetrical and progressive muscle weakness and/or sensory dysfunction in distal and proximal extremities (Rentzos et al., 2007). CIDP is characterized clinically by progressive or relapsing course (Pollard, 2002; Rentzos et al., 2007). The patients may have motor and sensory involvement, but generally motor involvement may be more prominent and more severe in lower extremities (Rotta et al., 2000). History of previous infection occurs in less than 10% of patients (Pollard, 2002). The reported prevalence of CIDP ranges from 1 to 2 per 100.000 population and is equal in both sexes (Rentzos et al., 2007). Demyelinating neuropathy predominantly affects spinal roots, plexuses and proximal nerve trunks and thickened nerves can be palpable in about 10% of the patients (Crino et al., 1993; Morgan et al., 1993; Matsuda et al., 1996;

Goldstein et al., 1996; Mizuno et al., 1998). When ranked by the descending frequency, postural tremor in the arms, peripheral nerve thickening, papilledema and facial or bulbar weakness, respiratory failure and autonomic dysfunction can be seen in patients with CIDP (Rotta et al., 2000; Pollard, 2002). Myelopathy caused by massive thickening of the peripheral nerves and symptomatic lumbar stenosis or vision loss caused by progressive pseudotumor cerebri are the other rare clinical features (Midroni and Dyck, 1996). Diagnostic criteria have been based on clinical features, electrodiagnostic findings, cerebrospinal fluid results, and nerve biopsy findings. In this paper, we present a case of CIDP with an atypical clinical history whose imaging findings are similar to neurofibromatosis.

2. Case

A 42-year-old male patient admitted with progressive paresthesia and weakness in upper and lower extremities.



Fig. 1. Midsagittal T2-weighted magnetic resonance imaging of cranium demonstrating periventricular hyperintense lesions

The patient had complaint of stumbling for the last five years and shuffling for one year. Medical history of the patient was unremarkable except for smoking and rare alcohol use. Neurological examinations of cranial nerves were normal except for fasciculations detected on his tongue. He had moderate quadriparesis (muscle strength in the upper and lower extremities were 4/5 and 5/5, respectively). Fasciculations were also observed in his right upper extremity. He had diffuse muscle atrophy with increased tone, absence vibratory sensation below the knee, reduced pinprick and proprioception distally in the lower extremities and brisk deep tendon reflexes. The patient had postural tremor in both hands. Due to progressive weakness that developed severe in the lower extremities, increased tonus and brisk deep tendon reflexes, initial diagnosis of the patient was multiple sclerosis.

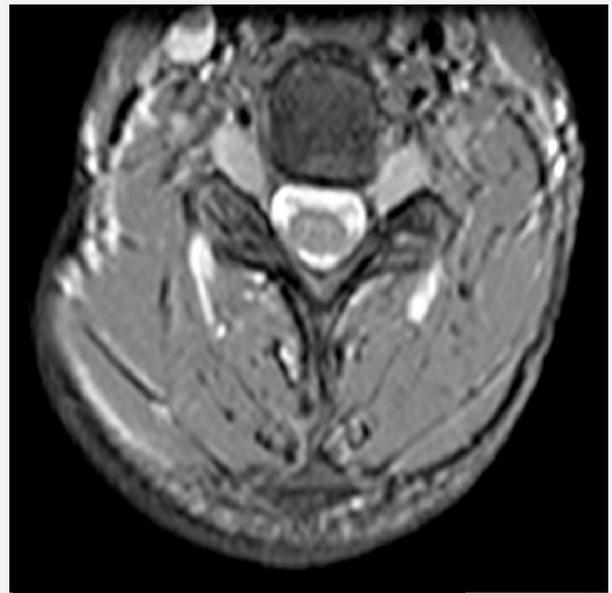


Fig. 2. Axial T2-weighted magnetic resonance image of the cervical and thoracic spine demonstrating significant nerve root enlargement bilaterally

In the Electroneuromyographic (EMG) evaluation of the patient conduction blocks showing demyelination were observed and symmetrical and severe sensorimotor polyneuropathy was detected. Cerebrospinal fluid (CSF) protein level was markedly elevated (643 mg/dL). No cell was found in CSF and IgG index was within normal limits. In laboratory investigations, no pathological findings except for low vitamin B12 level (171; 197-866) was detected. The investigations for brucella, syphilis, HIV and immunoelectrophoresis levels were within normal ranges. On T2-weighted brain Magnetic Resonance Imaging (MRI), a few corpus callosum located hyperintense lesions were observed. On cervical and thoracic T2 weighted MRI, hyperintense lesions, not causing expansion, were observed in C5, T1 and

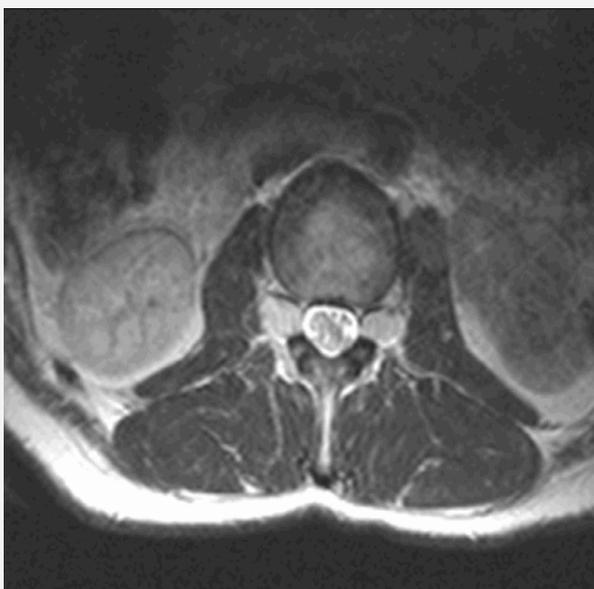


Fig. 3. Axial T2-weighted magnetic resonance image of the lumbar spine demonstrating significant nerve root enlargement bilaterally

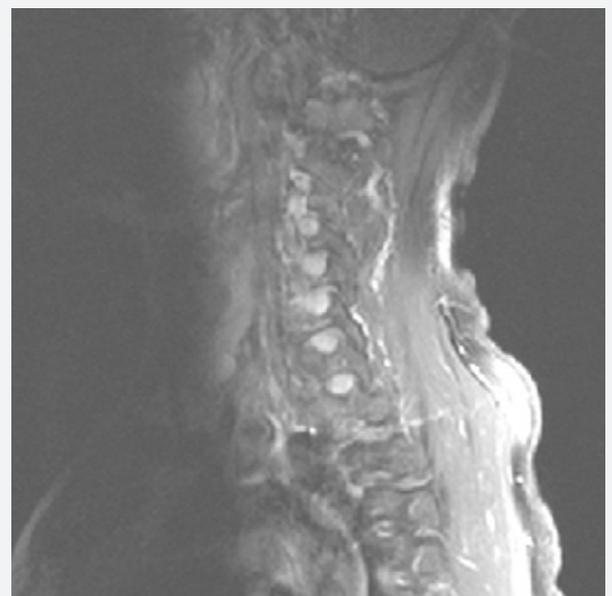


Fig. 4. Midsagittal T2-weighted magnetic resonance imaging of the cervical spine demonstrating nerve root hypertrophy in multiple cervical levels

T3 segments. MRI examination of the spine revealed diffuse thickening of foraminal and extraforaminal segments of all the spinal nerves bilaterally in cervicothoracic and lumbosacral regions (Fig. 1, 2, 3, 4). There was no nodularity on thickened nerves and the radiological interpretation of these lesions were considered to be consistent with neurofibromatosis. The patient was diagnosed CIDP and received intravenous immunoglobulin treatment with a dose of 0.4 g/kg/day for five days. After being discharged from the hospital, the patient received steroid treatment with a dose of 1 mg/kg and a significant improvement in the clinical findings of the patient was observed at the end of the third month of the treatment.

3. Discussion

The clinical presentation and course may vary in CIDP and unlike Guillain-Barre syndrome, muscle weakness progress slowly with or without remissions over months (Pollard, 2002; Rentzos et al., 2007). CIDP is characterized by segmental demyelination and remyelination of peripheral nerves and nerve roots and is known to cause hypertrophic neuritis (Duggins et al., 1999). Central invasion of the hypertrophic nerve roots may also cause spinal cord compression (Schady et al., 1996).

In such patients, the diagnosis of CIDP may be delayed due to atypical clinical features of upper motor neuron lesions (such as brisk deep tendon reflexes, spasticity). In our patient, the initial diagnosis was multiple sclerosis because of progressive weakness that developed severe in

the lower extremities, increased tonus and brisk deep tendon reflexes. MRI may be helpful in the diagnosis of CIDP by excluding the compressive or structural lesions that may lead to polyradiculopathy and MRI may also show hypertrophy of nerve roots and inflammatory processes in CIDP (Crino et al., 1993; Morgan et al., 1993; Goldstein et al., 1996; Midroni and Dyck, 1996; Mizuno et al., 1998). Duggins et al (1999) demonstrated that all patients with spinal root and plexus hypertrophy had a relapsing–remitting course and a significantly longer duration of disease than those without hypertrophy.

Our patient also had at least five years duration of complaints and on MRI of the cervicothoracic and lumbosacral spine, the nerve roots were prominently enlarged leading the radiologist to suspect neurofibromatosis. Mizuno et al. (1998) also showed nerve damage and subsequent hypertrophic nerve development due to demyelination between relapses in patients not receiving treatment.

In cases with CIDP, though not always, abnormal contrasting and hypertrophy are observed in cauda equina and lumbar nerve roots. In the literature, a few CIDP cases with well-circumscribed and contrasted lesions displaying expansion in cervicothoracic and lumbosacral foramina have been reported. In the differential diagnosis of nerve root hypertrophy, CIDP should be considered in the differential diagnosis beside neurofibromatosis, Guillain-Barre syndrome, neoplastic lesions, lymphoma, meningeal carcinomatosis, sarcoidosis, and CMV polyradiculopathy.

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