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Case Report

Role of MRI in the Diagnosis of CIDP Patients with Inconclusive Core Criteria

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Abstract

Summary

Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) is a clinical entity caused by inflammatory process mediated by immune reaction that involves nerve roots, plexuses and peripheral nerve trunks. In its typical form, CIDP produces symmetrical proximal and distal muscle weakness, sensory involvement and reduced or absent deep tendon reflexes. CIDP is usually progressive at least for 8 weeks, although it can occur in a relapsing–remitting pattern.

We report a case of forty year old diabetic patient presenting with 7-8 months history of weakness of all four limbs (both proximal and distal) with sensory involvement in the form of numbness (glove and stocking pattern) and tremor in bilateral hands. On evaluation, he was diagnosed as a case of CIDP with supporting radio-images with inconclusive electro diagnostic and cerebrospinal fluid (CSF) findings and responded well to the management.

Keywords: Chronic inflammatory demyelinating polyradiculoneuropathy, diabetes mellitus, lower motor neuron paralysis, European Federation of Neurological Societies, Peripheral Nerve Society.

Background

CIDP is a clinically heterogeneous entity occurring as a relapsing or progressive disorder. [1] The European Federation of Neurological societies/Peripheral Nerve society (EFNS/PNS) criteria assist in the diagnosis of CIDP by increasing the accuracy, which include Magnetic Resonance Imaging MRI) findings such as hypertrophy of the nerve roots or plexuses [2] however only a few studies have reported such MRI abnormalities in patients with possible CIDP. [3]

Here we report a case of CIDP not fulfilling core criteria where MRI findings helped in the diagnosis and treatment.

Case Presentation

A diabetic patient "in his 40s" presented to Neurology Outpatient Department with 7-8 months history of weakness of bilateral lower limb (both proximal and distal) with sensory involvement in the form of numbness distally in upper and lower limbs. He had no history suggestive of any cranial nerve or bowel bladder involvement. He was on oral anti diabetic drug with poor control. He had no significant history of peripheral neuropathy in the family.

On examination he had power 4/5- in proximal upper limb with normal distal upper limb power while lower limb proximal power was 3/5 with distal dorsi flexion having MRC grade of 2/5 while rest of the power in lower limb was 5/5.His deep tendon reflexes were absent with affected primary sensory modalities such as light touch, pain, temperature, joint position and vibration sensation while his plantars were bilateral flexor. Remaining neurological examination was unremarkable. On the basis of progressive weakness of more than 8 weeks with sensory involvement and areflexia of all four limbs he was diagnosed clinically as a case of possible CIDP. On routine tests his HbA1C was 12.5

percent, serum vitamin B12 was 162.5 pmol/l. Other investigations including HIV, HbsAg, anti HCV, serum Veneral disease and research laboratory (VDRL) came negative. [Table 1].

Table 1: showing various laboratory parameters.

Investigations	Findings	Range
Hemoglobin	10.3 gm/dl	13-17
Total leukocyte count	4020 cells/mm3	4000-10000
TSH	2.14 mIU/ml	74.-100
Blood sugar fasting	97 mg/dl	70-140
Blood sugar post prandial	130 mg/dl	
HbA1c	12.5 %	
Serum vitamin B12	162.5pmol/l	5.1-165
HIV 1 and 2	Non-reactive	
HbsAg	Negative	
Anti HCV	Non-reactive	
Serum VDRL	Negative	
Urine culture and sensitivity	Sterile	
CSF	Proteins- 67.5 mg/dl Sugar- 70 m/dl Cell count- 06 cells (86% polymorhs, 14% mononuclear), few red blood cells	

His nerve conduction study (NCS) revealed Motor nerve conduction showing reduced amplitude, increased latency and decreased conduction velocity of bilateral median and ulnar nerve with bilaterally non recordable peroneal and tibial nerve. Median, ulnar and sural sensory study was non recordable. His cerebrospinal fluid analysis revealed mild protein elevation (67.5 mg/dl) with normal sugar and cell count. After his NCS and CSF finding the diagnosis was inconclusive. Due to ongoing progression of the symptoms it was planned to rule out CIDP due to the fact that there is high prevalence of this reversible entity in diabetics. So we conducted MRI Dorsal spine with contrast which showed hyper intensity and enhancement of spinal lumbar nerve root that supported the diagnosis of CIDP. [Figure 1].

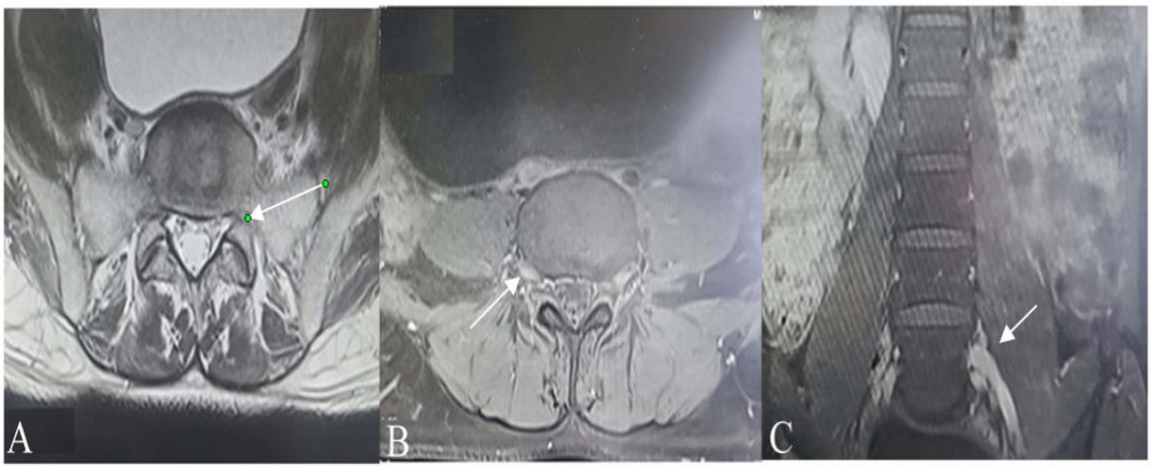

Figure 1:
(A) axial T2 weighted image with (arrow) showing mild to moderate thickened nerve root.
(B) axial T1 weighted image with contrast (arrow) showing hyperintense and thickened spinal nerve root.
(C) coronal T1 weighted image (arrow) with contrast showing enhancement of spinal lumbar nerve root.

Differential diagnosis

Metabolic Neuropathy like diabetes mellitus and vitamin B12 deficiency, Infective Neuropathy like chronic active hepatitis and HIV, Hereditary Neuropathy.

Treatment

Our patient was treated with four days of intravenous methyl prednisolone without any significant improvement in his Medical Research Council (MRC) power grade of 3/5 in proximal lower limb and he was remain non-ambulatory. He

was started on intravenous immunoglobulin (2gm/kg body weight) over 5 days. Our plan is to repeat ivig every 3 weeks with maintenance dose of 1 gm/kg body weight and reassessing response after 6 weeks.

Outcome and follow up

His power improved to MRC grade 4/5 and the patient is doing all the routine activities unaided however he still has not returned to his work.

Discussion

CIDP comprises a group of immune-mediated neuropathies with varied clinical presentations and electro-diagnostic features. Early recognition of these treatable disorders is important as delay in diagnosis result in significant disability and morbidity. There are various publications that have drawn attention to the high rate of CIDP misdiagnosis. [4] These diagnostic errors result in use of inappropriate treatments for longer duration with risk of secondary axonal changes. Radio imaging like MRI studies, although performed in many studies with definite CIDP fulfilling EFNS/PNS criteria; it may be helpful in clinically possible CIDP with inconclusive electrophysiological and CSF findings. Multiple studies were carried out to evaluate MRI in CIDP however most studies evaluated patients having definite CIDP. [Table 2].

Table 2: Comparison of the current study with other published studies

Author	Year	MRI Assessment
Adachi et al [5]	2011	Brachial and lumbar plexus: 66.7 percent patients have high signal intensity on STIR with contrast enhancement in 31.6 percent
Goedee et al [6]	2017	Enlargement of brachial plexus in 37 percent with T2 hyperintensity in 57 percent patients.
Shibuya et al [7]	2015	Nerve enlargement seen in 88 percent of the patients on patients on Magnetic resonance Neurography
Jongbloed et al[8]	2017	Brachial plexus abnormalities detected in 45 percent of patients.

Jeanne Thirounin et al [9] successfully treated 30 out of 75 patients (40 percent) of the possible CIDP patients with supportive features like axonal loss, nerve biopsy, plexus MRI in the absence of EFNS/PNS criteria. In our study the clinical feature was consistent for CIDP in the absence of EFNS/PNS criteria with insignificantly raised CSF protein level. Our patient was treated with 500 milligram pulse dose of intravenous methyl prednisolone (MPS) for 4 days without any improvement in sensory, motor and/or Rankin scale and hence switched to IVIG treatment. Our treatment response to intravenous methyl prednisolone is corroborating with retrospective multicentre study conducted by Van Liverloo et al that showed 43% patients did not respond to MPS. [10]. Our patient showed significant response to IVIG which is in concordance with a large Italian retrospective study that showed 78% response rate to IVIG. [11].This clinical scenario has not been reported in literature to the best of our knowledge. Hence, it is important to properly diagnose CIDP patients not meeting EFNS/PNS criteria by using supportive criteria like MRI of the plexuses.
Nowadays, MRI is usually not performed that plays a crucial role in the diagnosis of CIDP where electro diagnostic and CSF findings are inconclusive.

Take home messages

- Assessment of nerve root enhancement by MRI is useful in the diagnosis of clinically probable CIDP in the absence of core diagnostic criteria.

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