Neurology Asia 2022; 27(2) : 511 – 513

A case of probable chronic inflammatory demyelinating polyradiculoneuropathy presenting as unilateral lumbosacral plexopathy

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Abstract

Focal chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) is rare but should be considered in the differential diagnosis of chronic progressive neuropathy affecting a single limb. We report here a 66-year-old male presented with progressive right lower limb weakness and sensory deficits in keeping with a lower right lumbosacral plexopathy. His electrophysiological studies, whilst the distal motor latency and conduction velocities were within normal limits, showed marked asymmetry of right tibial and peroneal nerves with minimum F wave latency significantly prolonged on the right (greater than 120% upper limit of normal and also compared to the opposite side). This raised possibility of proximal demyelination. MRI with contrast revealed a diffused thickening of the right lumbosacral plexus and proximal right sciatic nerve. A lumbosacral plexus biopsy would have been helpful but was not undertaken due to patient's preference. No alternative cause was detectable upon extensive investigation. Patient was initiated on intravenous immunoglobulin for probable focal CIPD and has remained stable over a short period of follow up. In conclusion, even though lower limb involvement has rarely been described in literature, focal CIDP should be considered as a differential diagnosis of patients with focal neuropathies including unilateral lumbosacral plexopathy.

Keywords: CIPD, demyelination, polyradiculoneuropathy, plexopathy, monomelic neuropathy

INTRODUCTION

There are uncommon variants of chronic inflammatory demyelinating (CIDP) including focal variants. Focal CIDP is rare however should be considered in the differential diagnosis of chronic progressive neuropathy affecting a single limb.[1-4] Diagnosis can be challenging due to clinical heterogeneity of CIDP. Electrophysiological study and nerve/plexus MRI are often helpful. The most commonly used diagnostic criteria is that of the European Federation of Neurological Societies/ Peripheral Nerve Society (EFNS/PNS) guideline.[5] Focal CIDP is a diagnosis of exclusion of other chronic demyelinating neuropathies including polyneuropathy, multifocal motor neuropathy and paraproteinemic demyelinating neuropathy. We present a case of probable CIDP presenting as unilateral lumbosacral plexopathy.

CASE REPORT

A 66-year-old healthy male presented with a 10-year history of progressive right lower limb weakness and sensory deficits. His right leg would give way occasionally with an exercise tolerance of 400 metres limited by fatigue. This was associated with radiculopathy and numbness extending from his right calf into the foot. On further history, he had given up work as a truck driver 20 years ago due to atraumatic lower back pain. Neurological examination of the right lower limb revealed normal tone and motor examination revealed MRC grade 5/5 hip flexion, 4/5 hip extension, 5/5 hip abduction, 5/5 hip adduction, 4+/5 knee flexion, 5/5 knee extension, 5/5 ankle dorsiflexion, 4/5 plantarflexion, 5/5 ankle inversion and 4/5 ankle eversion. Right knee and ankle reflexes were diminished with down-going plantar response. Sensation to light touch, pain and temperature were diminished below the right knee. Clinically, this was in keeping with a right lower lumbosacral plexopathy with predominantly sciatic nerve involvement.

Laboratory investigations encompassing inflammatory markers, autoimmune and vasculitis screen, myeloma and lymphoma workup, infective

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Date of Submission: 10 September 2021; Date of Acceptance: 21 October 2021

https://doi.org/10.54029/2022dkr

screen with HIV, hepatitis, syphilis and Lyme disease serology, and neuropathy screen such as antineuronal antibody, vitamin B6, B12, folate, thyroid function and diabetes screen were all unremarkable. Neurofascin and contactin 1 antibodies were however not evaluated. Cerebrospinal fluid (CSF) analysis was bland with leucocytes $2x10^9$/L, protein 0.42g/L (0.15-0.45), glucose 3.5mmol/L (2.8-4.2). CSF viral and mycobacterium were negative, and so were oligoclonal bands, flow cytometry and cytology. Contrast enhanced magnetic resonance imaging (MRI) revealed a diffused thickening of the right lumbosacral plexus and proximal right sciatic nerve (Figure 1).

Importantly, there was no enhancement or nodularity in the sciatic nerve. Neve conduction study was performed. Whilst the distal motor latency and conduction velocities were within normal limits, there was marked asymmetry of right tibial and peroneal nerves with minimum F wave latency significantly prolonged on the right (greater than 120% upper limit of normal compared to the opposite side) (Figure 2). This raised possibility of proximal demyelination. There was also markedly reduced right sural and superficial peroneal sensory nerve action potential indicating a postganglionic lesion, namely a lumbosacral plexopathy. Needle electromyography showed reduced recruitment and chronic denervation reinnervation changes in the right medial gastrocnemius. On the basis of electrophysiologic and supportive neuroimaging findings, with no alternative identifiable cause, the patient was initiated on a 4-weekly regimen of intravenous immunoglobulin (IVIG) for probable focal CIPD. He has remained stable over a period of 12 months.

DISCUSSION

Although lower limb involvement has rarely been described in literature, we suspect our patient has likely focal CIDP manifesting as unilateral lumbosacral plexopathy. The patient reported slowly progressive symptoms with motor and sensory involvement, limited to a single limb. Despite a decade-long history of these symptoms, the electrophysiological study demonstrated the fibular and tibial compound muscle action potential amplitude were only mildly reduced than what would be expected in a primary axonal process. While the distal motor latency and conduction velocity in the right fibular and tibial nerves were within normal limits, there was a significant F wave prolongation compared to the opposite site compared to previous study done 10 years ago. This suggests a more proximal

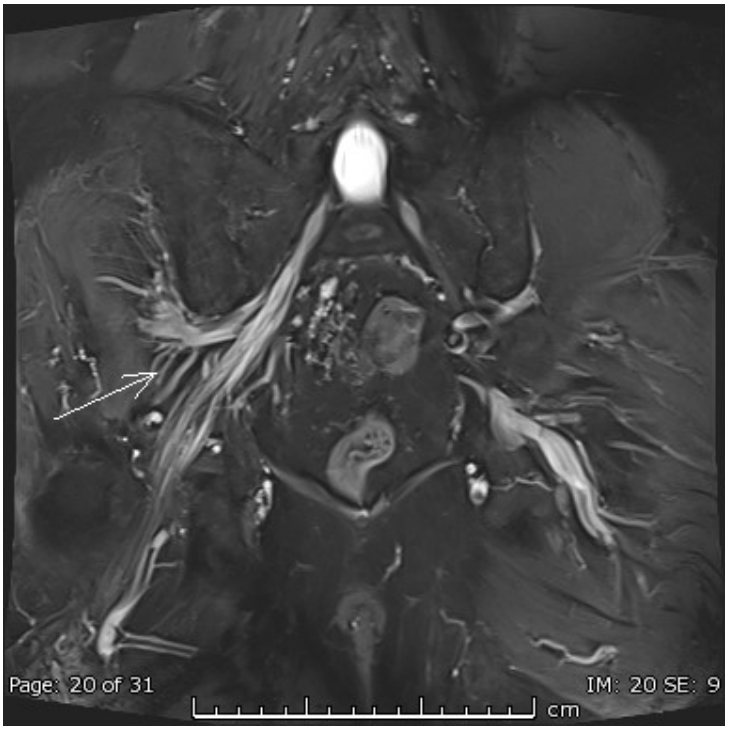

Figure 1. Contrast MRI showing diffused thickening of plexus (arrow).

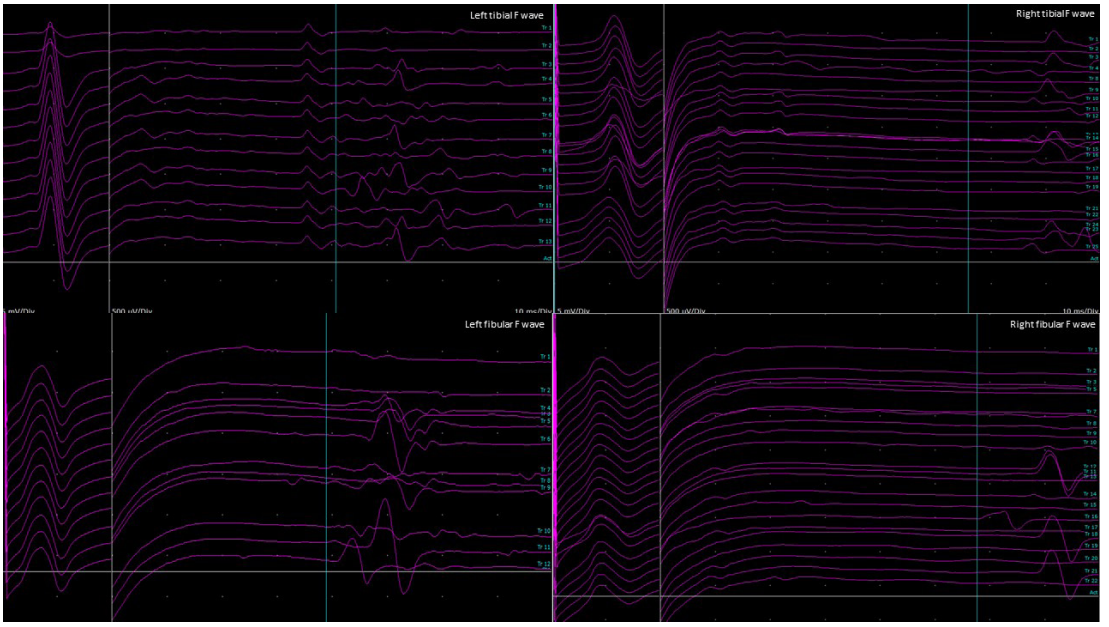

Figure 2. F wave latencies of left vs right tibial and fibular nerves on NCS. F wave latencies were significantly prolonged exceeding 120% in the right tibial and fibular nerves indicating a proximal demyelination.

demyelinating process, i.e. at lumbosacral plexus. Alternative etiologies were excluded through extensive investigations. A lumbosacral plexus biopsy was not performed due to patient's preference.

A recent case series of focal CIDP by Benoit *et al*. included 18 cases with demyelinating plexus neuropathy of which majority had brachial plexopathy (67%), with lumbosacral plexus involvement being less common.[6] These tended to have a benign course and when treated, most responded to IVIG (75%). MRI played an important role in establishing diagnosis as changes were noted in all patients with plexus involvement (nerve root thickening/ increased STIR signal in the involved plexus) as was the case in our patient. Somatosensory evoked potential was also helpful but only 39% with focal demyelinating plexus neuropathy fulfilled the EFNS/PNS criteria for definite CIDP. This is not surprising as EFNS/PNS criteria for CIDP relies on demyelination in the distal segment and can be negative when limited to sensory nerves. The value of plexus biopsy is not always justifiable given its invasive procedural risk.

This case adds to the limited literature regarding focal CIDP particularly with lower limb involvement and highlights its diagnostic challenges. Focal CIDP should be considered as a differential diagnosis of patients with focal neuropathies including unilateral lumbosacral plexopathy given it is treatable.

DISCUSSION

Financial support: None

Conflicts of interest: None

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