Predominant Upper Limb Chronic Demyelinating Polyneuropathy Associated with HBV Infection

Ioan-Cristian LUPESCU, Adriana Octaviana DULAMEA

Neurology Department, Fundeni Clinical Institute, Bucharest, Romania
“Carol Davila” University of Medicine and Pharmacy, Bucharest, Romania

ABSTRACT

Chronic inflammatory demyelinating polyneuropathy is an acquired, presumably immune-mediated peripheral neuropathy, characterized by symmetric sensory-motor involvement. Although most often idiopathic, it has been described in association with several disorders, sometimes improving under treatment. We present the case of a 57-year-old male who was admitted to hospital for paresthesias and muscle weakness affecting both upper limbs, initially only the hands, but with worsening and ascending progression during the last three years. The lower limbs were also involved but to a lesser extent. Electromyography indicated multifocal chronic demyelinating polyneuropathy with predominant upper limb involvement. Lumbar puncture showed a raised cerebrospinal fluid protein level. Laboratory samples revealed positive serology for HBV. Based on these, the diagnosis of chronic inflammatory demyelinating polyneuropathy with chronic hepatitis B was made. The patient received IVIG therapy and has since been coming periodically for IVIG sessions, with clinical and electromyographic improvement.

Keywords: chronic demyelinating polyneuropathy, hepatitis B.

INTRODUCTION

Chronic inflammatory demyelinating polyneuropathy (CIDP) is an acquired, presumably immune-mediated peripheral neuropathy that affects mainly the spinal roots, majorplexuses and proximal nerve trunks (1). It tends to have an insidious onset and a steadily progressive or relapsing course (2), and it is usually characterized by symmetric sensory-motor involvement (3), with muscle weakness affecting both proximal and distal limbs.

Related variants of this disorder have also been described such as multifocal acquired demyelinating sensory and motor neuropathy (MADSAM), multifocal motor neuropathy (MMN) and distal acquired demyelinating symmetric neuropathy (DADS) (3).

Although often idiopathic, CIDP is known to occur with several conditions (2, 4).
**CASE REPORT**

We present the case of a 57-year-old male who was admitted to hospital in 2015 for paresthesias and muscle weakness affecting both upper limbs. Symptoms began to develop three years before presentation at the level of both hands, with progressive worsening and proximal ascension.

The sensory-motor symptoms started affecting the lower limbs as well but to a lesser extent than the upper limbs.

Electromyography revealed multifocal chronic demyelinating polyneuropathy with predominant upper limb involvement.

Lumbar puncture was performed, showing slight increase of CSF protein content (0.4 g/L) with normal cellularity.

Blood samples were collected, including autoimmune, infectious and tumoral markers, vitamin B12 and Borrelia serology. Results showed elevated levels of cholesterol and triglycerides and, more importantly, positive markers for HBV. Apart from these, all other laboratory tests were within the normal range.

Abdominal ultrasound and chest x-ray were within normal range.

The patient received IVIG 75 g during five consecutive days and lipoic acid 600 mg q.d., under which there was a slight amelioration of symptoms. Also, he was sent to the Gastroenterology Department, where the diagnosis of chronic Hepatitis B was made and chronic treatment with Entecavir 0.5 mg q.d was initiated. In this setting, the diagnosis of **CIDP with chronic Hepatitis B** was established.

Since then, the patient has been coming periodically for IVIG sessions.

In 2016, he was admitted to hospital for worsening of symptoms, with motor deficit 4/5 MRC on proximal upper limbs, 0/5 MRC on distal upper limbs, 4/5 MRC on proximal lower limbs and 3/5 MRC on distal lower limbs.

A MRI of the brain and spinal cord was performed but it did not show any acute or demyelinating lesions.

We initiated corticotherapy with Methylprednisolone 16 mg q.d., under which symptoms improved, but because it interfered with HBV treatment efficacy and due to cortisone-induced osteoporosis, corticotherapy had to be interrupted after six months.

The patient has since improved both clinically and electromyographically, in 2017 showing motor deficit limited to upper limbs 4/5 MRC proximally and 2/5 MRC distally. Electromyography revealed only sensory polyneuropathy predominantly on upper limbs.

Hepatitis B viremia is still detectable and the patient is continuing antiviral treatment with Entecavir.

**DISCUSSION**

Clinical picture holds a special place in the assessment of patients with chronic polyneuropathy, because it can guide you and trick you at the same time.

Although rare, CIDP can present as a focal or multifocal neuropathy affecting one or both upper limbs (1, 5, 6). When occurring in the setting of chronic Hepatitis B or C, the main differential diagnosis is with vasculitis neuropathy, which classically manifests as mononeuritis multiplex, but can also take the form of a distal symmetrical sensory-motor neuropathy (1).
In this setting, electromyography becomes essential, as it distinguishes demyelination from the axonal damage seen with vasculitis.

Chronic inflammatory demyelinating polyneuropathy associated with other illnesses sometimes responds to treatment of the underlying disorder (2, 7). In our case, it is hard to tell whether the chronic treatment with Entecavir has influenced neurological symptoms on the long run. We do know that most of the improvements occurred after IVIG sessions and during corticotherapy.

Acknowledgments: The authors would like to thank dr. Draghici Mirela for performing the electromyograms, thus helping with diagnosis and follow-up.
Conflicts of interest: none declared.
Financial support: none declared.

References


5. Gorson KC, Ropper AH, Weinberg DH. Upper limb predominant, multifocal chronic inflammatory demyelinating polyneuropathy.

