

# Effect of Intravenous Immunoglobulin on Bilateral Sixth Nerve Palsy in Chronic Inflammatory Demyelinating Polyneuropathy: A Case Study

Behnam Safarpour Lima<sup>1</sup>, Sepideh Paybast<sup>2</sup>, Mahtab Ramezani<sup>3</sup> and Mohammad Sistanizad<sup>4\*</sup>

<sup>1</sup>Department of Neurology, Imam Hossein Medical and Educational Center, Shahid Beheshti University of Medical Sciences, Tehran, Iran

<sup>2</sup>Neurology Resident, Department of Neurology, Imam-Hossein Hospital, Shahid Beheshti University of Medical Science, Tehran, Iran

<sup>3</sup>Department of Neurology, Loghman Hakim Hospital, Shaheed Beheshti University of Medical Science, Tehran, Iran

<sup>4</sup>Department of Clinical Pharmacy, Shahid Beheshti University of Medical Sciences, Tehran, Iran

\*Corresponding author: Mohammad Sistanizad, Department of Clinical Pharmacy, Shahid Beheshti University of Medical Sciences, Tehran, Iran, Tel: 00989122784895; E-mail: sistanizadm@sbm.ac.ir

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## Abstract

Chronic Inflammatory Demyelinating Polyneuropathy (CIDP) is an immune mediated demyelinating disorder, which involves both sensory and motor peripheral nerves. Cranial neuropathy is a rare manifestation in CIDP patients.

In this study, we describe a case of relapsing CIDP presented with acute onset bilateral sixth nerve palsy after being in remission for eight years.

A 23-year-old man, with symmetric paresis of lower limbs that gradually evolved to upper limbs over one year since he was 15, was referred to our neurology clinic. Electro-diagnostic study revealed sensory and motor demyelinating polyneuropathy. He received intravenous immunoglobulin (IVIG) followed by corticosteroids as maintenance therapy and had an excellent response to therapy. At his recent presentation (2016), he revealed sudden onset horizontal diplopia and bilateral abduction deficit, which was consistent with bilateral sixth nerve palsy. Brain magnetic resonance imaging (MRI) was normal and lumbar puncture showed elevated protein with normal cell count.

The presentation of CIDP can be highly diverse and heterogeneous. This report presents a rare manifestation of CIDP in a patient who had a dramatic response to IVIG.

**Keywords:** Chronic inflammatory demyelinating polyneuropathy; Sixth nerve palsy; Ophthalmoplegia

## Introduction

Chronic Inflammatory Demyelinating Polyneuropathy (CIDP) is an autoimmune disorder which mainly involves the myelin sheath of peripheral nerves (sensory and motor) [1-3]. Damage to the myelin sheath in CIDP leads to heterogeneous clinical pictures, usually involving chronic progressive weakness in more than one limb. CIDP is more common in men and young adults [3]. The prevalence of CIDP ranges from 1.9 to 8.9 persons per 100,000 and its incidence is 1.6 persons per 100,000 per year [4].

It is a lifelong disease with progressive and relapsing courses [1]. The main clinical presentations are diffuse hyporeflexia or areflexia and symmetric proximal-distal muscle weakness, tingling or numbness and fatigue [1-3]. Atypical presentations of CIDP are unilateral weakness, distal or multifocal limb involvement. Cranial neuropathy in CIDP is uncommon: based on some case studies, the prevalence ranges from 3 to 15 percent and the facial nerve is the most affected nerve [4,7]. Although there are several cases of CIDP with ophthalmoplegia in the literature [4], to the best of our knowledge simultaneous bilateral sixth nerve palsy has not been reported.

Herein, we describe a case of CIDP presented with a new attack of bilateral sixth nerve palsy due to the relapse of CIDP after being in remission for 8 years. Fortunately, he showed a remarkable response to IVIG therapy.

## Case Presentation

### Patient's demographics, history and physical exam

The patient was a 23-year-old man with a history of gradual developing symmetric paresis of his lower limbs, which progressed to upper limbs. Over one year (in 2008), he had become completely disabled and bedridden. After being evaluated in our clinic, on the basis of electro-diagnostic study and elevated cerebrospinal fluid (CSF) protein with normal cell count, he was diagnosed with CIDP. Eventually, he was treated with IVIG followed by corticosteroids as maintenance. He had an excellent response to therapy and was able to walk again.

At his recent presentation in January 2016, he revealed sudden onset horizontal constant diplopia. Other complaints, such as ptosis, periorbital pain, visual loss, color-vision impairment, vertigo and headache and bulbar symptoms were not noted. His neurological examination revealed a partial left sixth nerve involvement and a complete right sixth nerve involvement and areflexia in lower limbs, not associated with motor or sensory symptoms and signs.

### Patient's laboratory and diagnostic data

A lumbar puncture (LP) was performed and showed opening CSF pressure of 10 cmH<sub>2</sub>O, normal white blood cell count and glucose, and elevated protein. Routine blood biochemistry tests were normal. His brain MRI, with and without gadolinium, and brain magnetic resonance venography (MRV) were unremarkable. His chest and abdominopelvic CT scan was normal. EMG\_NCS findings were suggestive of CIDP.

Again, IVIG (25 grams daily for five days) was instituted. On the third day of therapy, remarkable improvement was observed in the paresis of the left lateral rectus. On the fifth day, right lateral rectus

paresis showed modest improvement. The patient was a candidate to receive monthly IVIG. Currently, he is asymptomatic with monthly IVIG therapy.

## Discussion

CIDP is mainly a peripheral nerve autoimmune disorder. Although central nervous system demyelinating lesions in conjunction with peripheral involvement have been reported, cranial neuropathies, particularly with oculomotor involvement, are not a common manifestation of CIDP [4]. However, in 2015 a case of recurrent isolated unilateral sixth nerve palsy in relapsing remitting CIDP was reported, while our patient presented with simultaneous bilateral sixth nerve paresis, which occurred eight years after the onset of motor and sensory involvements.

The NCS findings in this patient demonstrate that there can be discordance between the clinical and electro-diagnostic findings in patients with chronic inflammatory demyelinating polyneuropathy [4].

## Conclusion

Clinical presentation in CIDP can be diverse and heterogeneous. This report expands the clinical spectrum of CIDP presentations, along with EMG\_NCS findings, CSF protein increases and a dramatic response to IVIG.

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