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A case of chronic inflammatory demyelinating polyneuropathy presented with unilateral ptosis

Sadegh Izadi[1], Sina Karamimagham[2], Maryam Poursadeghfard[3]

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Abstract

Chronic Inflammatory Demyelinating Polyneuropathy is an autoimmune disease with progressive and relapsing courses. The main clinical presentations are diffuse deep tendon hyporeflexia or areflexia and symmetric proximal-distal muscles weakness. Myasthenia gravis is also an immune mediated disease with fluctuating ocular and bulbar symptoms and sometimes weakness. Although both myasthenia gravis and chronic inflammatory demyelinating polyneuropathy are immune mediated disorders, clinical presentations are obviously different in the two diseases.

Herein, we will report a case of chronic inflammatory demyelinating polyneuropathy who presented with isolated unilateral ptosis. Initially, the patient was managed as ocular type of myasthenia gravis, but after progression to general limb weakness and areflexia, the diagnosis of chronic inflammatory demyelinating polyneuropathy was made. Although unilateral ptosis is a typical feature of myasthenia gravis, it may be seen as the first presentation of chronic inflammatory demyelinating polyneuropathy as well which mimics myasthenia gravis disease.

Keywords: Blepharoptosis, Chronic Inflammatory Demyelinating Polyradiculoneuropathy, Myasthenia gravis.

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Introduction

Chronic Inflammatory Demyelinating Polyneuropathy (CIDP) is an acquired and autoimmune disease which involves both sensory and motor peripheral nerves (1). It is a lifelong disease with progressive and relapsing courses. The main clinical presentations are diffuse hyporeflexia or areflexia and symmetric proximal-distal muscles weakness. It occurs mainly between 40 to 60 years of age with mild preference for males. It rarely presents in the atypical form as unilateral or multifocal or distal involvement (2). There are frequent reports about spinal nerve roots hypertrophy in CIDP resulting in spinal canal st enosis (3). Facial nerve palsy and ophtalmoplegia were also reported in CIDP (4,5). Here we will report a rare presentation of undiagnosed CIDP (isolated and unilateral ptosis) which mimicked Myasthenia Gravis (MG) at first from Shiraz, south of Iran.

Case presentation

A 55 year old lady was referred to Motahhari clinic, an outpatient center affiliated to Shiraz University of Medical Science, with a history of isolated left eye ptosis without significant fluctuation during the day for 3 months. She had no history of diplopia or weakness. At that time general and neurological examinations were normal. With clinical impression of Myasthenia Gravis complementary evaluations were started. Magnetic Resonance Imaging (MRI) of the brain, Thyroid Function Tests

1. MD, Assistant Professor of Neurology, Department of Neurology, Medical School, Shiraz University of Medical Sciences, Shiraz, Iran. Izadisad@sums.ac.ir
2. MD, Fellowship of Neonatology, Department of Pediatric, Medical School, Shiraz University of Medical Sciences, Shiraz, Iran. karamimagham@yahoo.com
3. (**Corresponding author**) MD, Assistant Professor of Neurology, Department of Neurology, Medical School, Shiraz University of Medical Sciences, Shiraz, Iran. poursadegh@sums.ac.ir

and edrophonium chloride (tensilon) test were normal. Nerve Conduction Velocity (NCV) and Electromyography (EMG) did not show any significance and Repeated Stimulation Test (RST) of the Orbicularis Oculi muscle showed about 10% decremented pattern, so with possibility of myasthenia gravis oral form of pyridostigmine bromide (mestinon) was prescribed.

She showed no response to medication and gradually progressed to both lower extremities weakness (muscle power 4-/5) mainly in the proximal muscles in a symmetric pattern and general hyporeflexia (deep tendon reflex 1/2) was also appeared about two months later. Repeated NCV study and EMG showed significant increased distal latency and decreased conduction velocity in demyelinating range for the left posterior tibial and left ulnar nerves with conduction block in the right posterior tibial nerve. All F waves were prolonged. These data met criteria for CIDP. To confirm these findings, Lumbar Puncture (LP) was performed. Cerebro-Spinal fluid (CSF) analysis revealed a protein level of 150 mg/dL without any abnormal White Blood Cell (WBC). Serum protein and immunoglobulin electrophoresis were within normal value. Repeated RST did not reveal any significant decremented pattern. Therefore, she was diagnosed as CIDP and managed with Intravenous Immunoglobulin (IVIG). Relative improvement was accessed and she was discharged with oral prednisolone in a good condition.

Discussion

CIDP is a lifelong and treatable autoimmune polyneuropathy (6). It usually presents with limbs weakness and infrequently cranial nerves are affected as well, but isolated unilateral ptosis especially as a first complaint is not a usual symptom in CIDP. There are a few reports in the literature to describe cranial neuropathy as CIDP relapse. Husain and his colleague introduced a diabetic patient with CIDP who had presented with progressive arms and legs weakness superimposed by lower motor neuron type facial palsy 3 months later (4). In 2009 and 2010, two patients were introduced with CIDP and ophthalmoplegia. The first patient was a 40 year old woman diagnosed with CIDP at 25 years of age; she later experienced relapse with ptosis and third nerve palsy (7). The second one had controlled CIDP for 2 years and developed to isolated oculomotor nerve palsy as a relapse of her disease (8). In another report in 2002, a patient firstly presented with unilateral ocular ptosis and isolated adduction deficit and 2 years later was diagnosed with CIDP "(9). In all the above patients, in addition to limbs weakness, ocular manifestation was a prominent feature of CIDP.

What is noticeable in our patient is the initial presentation with isolated unilateral ptosis misleading to a wrong MG diagnosis. MG is an autoimmune disease with mainly ocular manifestation. The unique feature of MG is fluctuation and diurnal variation within 24 hours. MG sometimes overlaps with other autoimmune diseases such as diabetes mellitus, rheumatoid arthritis, hyperthyroidism and Sjögren (10,11). In 1999, a nineteen year old boy was reported who simultaneously had MG and CIDP, two diseases with antibody mediated ethiology (12). In our patient, all investigations for MG including edrophonium chloride test, RST and empirical therapy with pyridostigmine bromide were negative. So diagnosis of concomitant MG is very unlikely.

CIDP responds well to IVIG and prednisolone. Plasma exchange is also another choice to control symptoms (13). This patient improved after starting Intravenous Immunoglobulin and oral prednisolone.

Conclusion

CIDP is an acquired multifocal and segmental demyelination in the peripheral nerves that sometimes affects cranial nerves in isolation as a relapse or even as a first manifestation. But isolated unilateral ptosis at the beginning of disease is a very rare manifestation that could mislead physicians to an incorrect diagnosis.

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