Case Report

Acute Onset of Chronic Inflammatory Demyelinating Polyneuropathy: A Case Report

Jie Wu, Chunling Song, Yanqiu Han, Shanji Nan

Abstract

Acute onset chronic inflammatory demyelinating polyneuropathy (A-CIDP) with facial weakness and mild respiratory muscle weakness in the three episodes is rarely reported. We describe a Chinese man with A-CIDP who experienced weakness in both upper limbs as well as facial muscles, and his condition deteriorated in three different consultations over the span of 14 weeks. Patients with A-CIDP were more likely to have prominent sensory signs rather than motor signs and were less likely to have autonomic dysfunction, facial weakness, or need for mechanical ventilation. Our patient had facial weakness in the first and second episodes, and mild respiratory muscle weakness in the first and third episodes; hence, our patient was most likely an A-CIDP case different from those cases in the literature. Early recognition of A-CIDP in patients with apparent GBS is important so that proper diagnostic and therapeutic management can be initiated promptly.

Keywords: Chronic inflammatory demyelinating polyneuropathy; Guillain-Barre syndrome

Introduction

Chronic inflammatory demyelinating polyneuropathy (CIDP) is an acquired immune-mediated inflammatory disorder of the peripheral nervous system which has clinical similarities to acute inflammatory demyelinating disease of the peripheral nerves called Guillain-Barre syndrome (GBS) [1, 2]. Sixteen percent of patients with acute CIDP (A-CIDP) experience rapid deterioration within 8 weeks followed by a chronic course, while 6-16% of treated patients with GBS have one or more deteriorations after initial improvement that is typically described as treatment-related fluctuations (TRFs) [3, 4]. Here we report a case of a 49-year-old male patient with A-CIDP presenting with apparent GBS.

Case Report

A 49-year-old male was admitted to the hospital on November 1, 2012 for syndrome of weakness in the facial muscles and progressive loss of sensation that began in his upper limbs and progressed distally. One day prior to admission, the patient suffered weakness in his legs, and walked slowly with an unsteady gait. He also presented with slurred speech and dysphagia. According to the patient, he had intermittent diarrhea for a month before the onset of the syndrome.

Physical examination findings and electromyogram (EMG) characteristics are summarized in Table 1. The patient developed mild dyspnea on his second day of admission. On day 3, a lumbar puncture was performed and the results showed increased protein content (Table 1).

The patient was initially diagnosed with GBS, stage II hypertension (moderate to severe), and viral hepatitis. Owing to financial constraints, the patient could not afford gamma globulin replacement therapy or plasma exchange. He opted for steroid treatment instead. He took methylprednisolone (60 mg orally once daily) for 5 days followed by tapering of the dose by 5 mg every 2 days. The patient was given an intramuscular injection of vitamin B1, B12 and mouse nerve growth factor. His symptoms improved progressively, and the lower limb muscle strength recovered to the point where he could carry heavy loads with a normal gait. The patient’s speech returned to normal, and his dysphagia and dyspnea subsided. The sensation in all his limbs was normal. He had intermittent sensation loss on his back. He was discharged after 8 days, while still taking a daily dose of 50 mg methylprednisolone, 100 mg vitamin B1, and 500 μg methylcobalamin. The dose of methylprednisolone was gradually reduced until cessation by 5 mg every 2 days.

Fourteen weeks after the first episode, the patient drank 10 mL of alcohol. He experienced slurred speech, weakness in his lower limbs, and inability to walk. He was readmitted to the hospital on February 1, 2013.

Physical examination findings and EMG characteristics are summarized in Table 1. On day 5, the patient underwent lumbar puncture. The results showed increased protein content (Table 1). Examination for anti-ganglioside antibody spectrum 2 (GM1, GM2, GM3, GD1a, GD1b, GT1b, and GQ1b) was negative (Euroimmun). Based on his clinical features and disease progression, the patient was differentially diagnosed...
Table 1. Findings for Physical Examination and Cerebrospinal Fluid

<table>
<thead>
<tr>
<th>Episode</th>
<th>Speech</th>
<th>Face</th>
<th>Upper limbs</th>
<th>Lower limbs</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Dysphasia</td>
<td>Bilateral facial palsy, incomplete eyelid closure with gap of 3 mm</td>
<td>Muscle strength* graded 4; absence of reflexes in bilateral biceps and triceps tendons</td>
<td>Absence of reflexes in bilateral biceps and triceps tendons</td>
</tr>
<tr>
<td>2</td>
<td>Hoarse voice</td>
<td>Facial palsy</td>
<td>Muscle strength* graded 4 with weaker grip strength in left hand, absence of reflexes in bilateral biceps and triceps tendons</td>
<td>Absence of all tendon reflexes, hypalgesia (10 cm distal to the knees)</td>
</tr>
<tr>
<td>3</td>
<td>Dysphasia</td>
<td>Absence of reflexes in bilateral superficial peroneal and Achilles tendon, reduced reflex in the left patellar tendon</td>
<td>Muscle strength* graded 4; absence of all tendon reflexes, hypalgesia (10 cm distal to the knees)</td>
<td>Absence of reflexes in bilateral tibial nerve H-reflex.</td>
</tr>
</tbody>
</table>

Muscle strength* graded according to the manual muscle test (MMT) grading system. 

- Pandy test: +
- Pandy test: ++
- Pandy test: +++
- Positive
- (-) Absence of all tendon reflexes; hypalgesia (10 cm distal to the knees)
- (-) Absence of all reflexes in peripheral sensory and motor nerve endings; an absence of the bilateral tibial nerve H-reflex.

Discussion

Ruts et al [5] suggested A-CIDP should be suspected when a patient with GBS deteriorates after 9 weeks from the onset or when deterioration occurs three or more times. In this case, the patient’s condition deteriorated over 14 weeks after the first onset and the deterioration occurred three times; hence, he was diagnosed with A-CIDP according to the key diagnostic features described by Ruts et al [5]. In addition, Dionne et al performed a retrospective review on 30 patients with acute inflammatory demyelinating polyneuropathy (AIDP) and 15 patients with A-CIDP and revealed that infectious prodrome was rarely correlated with A-CIDP; patients with A-CIDP were more likely to have prominent sensory signs rather than motor signs. In their studies, 53.3% A-CIDP patients presented with signs of sensory ataxia, impaired vibration perception, and stocking-glove hypalgesia, which was consistent with the symptoms experienced by our patient. Nevertheless, Dionne et al also reported that patients with A-CIDP were less likely to have autonomic dysfunction, facial weakness, or need for mechanical ventilation [6]. Ruts et al performed a prospective longitudinal study on patients with GBS-TRF and with A-CIDP in order to distinguish these two diseases in the early phase of disease. Their results revealed, at all time points, A-CIDP group was less severely affected than GBS-TRF group in level of weakness and severity. A-CIDP patients did not
need artificial ventilation, had significantly less cranial nerve dysfunction, and tended to have more CIDP-like electrophysiologic abnormalities. In Rut’s study, only one out of eight A-CIDP patients appeared cranial nerve dysfunction (VII). However, in this case, our patient had facial weakness in the first and second episodes, and mild respiratory muscle weakness in the first and third episodes; hence, our patient was an A-CIDP case different from those cases reported in the previous literatures.

Early recognition of A-CIDP in patients with apparent GBS is clinically difficult, but important because it can lead clinicians to initiate steroid therapy. While A-CIDP requires maintenance therapy after the initial response to steroid treatment, adjuvant immunosuppressive therapy is commonly used to lower the steroid dose, thereby reducing side effects [4].

Acknowledgments

The authors would like to thank Medjaden Bioscience Limited for providing assistance in the preparation of this manuscript.

Conflicts of Interest

No potential conflicts of interest relevant to this article were reported by all authors of the study.

References