Case Report

Chronic inflammatory demyelinating polyneuropathy presenting with autonomic symptoms: a case report

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ABSTRACT

A 31 year old female presented with a one-year history of burning sensations, numbness and tingling feelings in her hands and feet. There was also excessive sweatiness of her hands and feet while experiencing these symptoms. Her symptoms were worse at night and during hot weather conditions and aggravated by contact with light clothing. She could not tolerate her shoes. She also complained of weakness in her distal lower limbs. Examination revealed prominent wasting of the thenar and hypothenar eminences, marked bilateral weakness of hand grip, palmar, and dorsal interossei. There was also the significant weakness of the dorsiflexor of the feet with allodynia. Vibration and joint position sense were impaired in the distal lower limbs. Ankle reflexes were diminished. Nerve conduction studies showed nerve conduction block in three nerves. She also had abnormalities from motor and sensory nerve conduction studies involving multiple nerves. She showed remarkable improvements in her symptoms after a course of pulsed oral dexamethasone therapy.

Keywords: Autonomic, Chronic, Demyelinating, Inflammatory, Polyneuropathy, Symptoms

INTRODUCTION

Chronic inflammatory demyelinating polyneuropathy is a chronic acquired immune mediated-mediated disorder of the peripheral nervous system. They are progressive, relapsing diseases that are potentially treatable. Globally, it is the commonest autoimmune demyelinating peripheral neuropathy with a prevalence of 1.2 to 7.7 per 100,000 and a slight male preponderance. The classic clinical presentation is that of a chronic symmetrical sensory and motor neuropathy with absent tendon reflexes. The autonomic nervous system is not affected in contrast to acute inflammatory demyelinating polyneuropathy (Guillain-Barre syndrome).

To the best of our knowledge, there has been only one previous report of this condition from Nigeria. We also highlight the importance of nerve conduction studies which are not readily available in resource-poor settings in making the diagnosis of this condition. The rapid resolution of her symptoms with pulsed oral dexamethasone as a treatment modality is also emphasized.

CASE REPORT

A 31-year-old female presented with a one-year history of burning sensations in her hands and feet. They were associated with intermittent episodes of numbness and tingling. There was also excessive sweatiness of her hands and feet while experiencing these symptoms. These symptoms were worse at night and during hot weather conditions. They were aggravated by contact with light clothing. She could not tolerate her shoes. She also
Complained of weakness in her distal lower limbs. No history of neck pain or low back pain. Her obstetric history was significant for three episodes of gestational diabetes mellitus. Blood sugar normalized in all instances after child delivery. Examination revealed prominent wasting of the thenar and hypothenar eminences, marked bilateral weakness of hand grip, palmar, and dorsal interossei. There was also the significant weakness of the dorsiflexor of the feet with allodynia. Vibration and joint position sense were impaired in the distal lower limbs. Ankle reflexes were diminished.

**Table 1: Motor nerve conduction study.**

<table>
<thead>
<tr>
<th>Nerve</th>
<th>Stimulation site</th>
<th>Amplitude (mv)</th>
<th>Latency (ms)</th>
<th>Conduction velocity (ms)</th>
<th>E-wave latency, (ms)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Median</td>
<td>Wrist</td>
<td>Right 7.84</td>
<td>Left 3.05</td>
<td>Right 3.05</td>
<td>Left 3.05</td>
</tr>
<tr>
<td></td>
<td>Elbow</td>
<td>Right 7.86</td>
<td>Left 7.75</td>
<td>Right 8.15</td>
<td>Left 7.75</td>
</tr>
<tr>
<td></td>
<td>Axilla</td>
<td>7.5</td>
<td>6.0</td>
<td>9.85</td>
<td>6.0</td>
</tr>
<tr>
<td>Ulnar</td>
<td>Wrist</td>
<td>16.96</td>
<td>9.39</td>
<td>2.22</td>
<td>2.1</td>
</tr>
<tr>
<td></td>
<td>Below elbow</td>
<td>4.16*</td>
<td>8.39</td>
<td>6.45</td>
<td>6.6</td>
</tr>
<tr>
<td></td>
<td>Above elbow</td>
<td>10.02</td>
<td>6.14</td>
<td>8.1</td>
<td>8.6</td>
</tr>
<tr>
<td>Peroneal</td>
<td>Ankle</td>
<td>7.85</td>
<td>8.61</td>
<td>3.6</td>
<td>3.3</td>
</tr>
<tr>
<td></td>
<td>Peroneal fossa</td>
<td>0.11*</td>
<td>-</td>
<td>12.55</td>
<td>-</td>
</tr>
<tr>
<td>Tibial</td>
<td>Ankle</td>
<td>9.07</td>
<td>11.66</td>
<td>6.2</td>
<td>6.2</td>
</tr>
<tr>
<td></td>
<td>Popliteal fossa</td>
<td>0.11</td>
<td>5.15</td>
<td>13.3</td>
<td>14.35</td>
</tr>
</tbody>
</table>

Key: *=deranged.

**Table 2: Sensory nerve conduction study.**

<table>
<thead>
<tr>
<th>Nerve</th>
<th>Stimulation site</th>
<th>Amplitude (ms)</th>
<th>Latency (ms)</th>
<th>Conduction velocity (ms)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Median</td>
<td>Wrist</td>
<td>12.56</td>
<td>2.6</td>
<td>38.81</td>
</tr>
<tr>
<td></td>
<td>Finger II</td>
<td>21.89*</td>
<td>2.45</td>
<td>41.94</td>
</tr>
<tr>
<td>Ulnar</td>
<td>Wrist</td>
<td>1.38</td>
<td>20.05</td>
<td>1.22</td>
</tr>
<tr>
<td>Radial</td>
<td>Wrist</td>
<td>22.85</td>
<td>17.03</td>
<td>1.8</td>
</tr>
<tr>
<td>Sural</td>
<td>Calf</td>
<td>2.44</td>
<td>8.39*</td>
<td>9.55</td>
</tr>
</tbody>
</table>

Key: *=deranged.

Magnetic resonance imaging of both the cervical and lumbosacral spines were normal. Antinuclear antigen and anti-double stranded DNA were negative. Renal indices were normal. HIV I and II serology were negative. Fasting blood sugar was within normal limits. Nerve conduction study results are shown in Table 1 and 2. She had abnormalities with sensory nerve action potentials and compound motor action potentials involving multiple nerves. There was also multiple nerve conduction block on stimulation of three nerves (both median, right peroneal and right tibial) as evidenced by a change in the configuration of the proximal compound motor action potential when compared to the distal. These are highly suggestive of an acquired sensorimotor neuropathy of the demyelinating type, with chronic inflammatory demyelinating polyneuropathy as the most plausible diagnosis. Needle electromyogram revealed normal findings in all muscles tested. She was commenced on a six-month course of pulsed dexamethasone therapy. It was administered as a single 40 mg dose taken daily for five days the month. This was repeated till she completed six cycles of therapy.

**DISCUSSION**

Our initial diagnostic consideration in this lady was a degenerative spine disease to exclude a form of diabetes-associated peripheral neuropathy. However, her cervical and lumbosacral neuroimages were both normal. Though she had a normal fasting blood sugar, her previous obstetric history led to the consideration of an impaired glucose tolerance (IGT) neuropathy. IGT neuropathy is a predominantly distal symmetrical sensory polyneuropathy occurring in persons with impaired glucose tolerance. She was initially commenced on a five-day course of intravenous methylprednisolone (400 mg). She also received pregabalin for the distressing neuropathic pain.
However, her symptoms did not improve after a 1-month follow-up. The presence of excessive sweating at the hands and feet depicts autonomic nerve damage. Autonomic nerve damage is due to small fiber nerve involvement. The features of classical CIDP are due to large fiber nerve damage. Nerve conduction performed at this time revealed classic features of chronic inflammatory demyelinating polyneuropathy. The neuropathological criteria for diagnosis of CIDP was proposed by Nicolas et al. The nerve conduction studies must show one of the following abnormalities in at least three different nerves: conduction block or temporal dispersion in at least 3 different nerves with demyelination evidence in at least one nerve or conduction block or temporal dispersion in at least two different nerves and abnormal conduction values in at least one nerves or conduction block or temporal dispersion in one nerve and abnormal nerve conduction values in at least two other nerves or abnormal conduction values in three different nerves. Our patient had nerve conduction abnormalities in three nerves with conduction block three nerves.

Nerve conduction studies are the most important investigation for this condition as it can distinguish between an axonal and demyelinating neuropathy. Nerve biopsy is not very useful for making a diagnosis of this disorder. The sural nerve which is commonly biopsied is a distal sensory nerve which is not usually affected in the pathologic process. The brunt of nerve dysfunction falls on the motor fibres, nerve roots and proximal trunk. The extent of autonomic nervous involvement could not be ascertained as we lack diagnostic facilities to do so. In one previous report, a rectal biopsy showed axonal degeneration and depletion of postganglionic noradrenergic fibers in the rectal mucosa.

She reported dramatic and progressive improvements in burning sensations, tingling and pricking following pulsed oral dexamethasone therapy. This improvement was noticed after the second treatment cycle. She was unable to do a follow-up nerve conduction study due to financial constraints. A follow-up nerve conduction study would have showed improvements in conduction blocks, sensory and compound motor action conduction velocities. The cardinal objectives of treatment are to reduce symptoms and improve functional status. Therapies that have been shown to be of proven benefits from large randomized controlled trials include pulsed oral dexamethasone, intravenous immunoglobulin, plasma exchange, and prednisolone. Treatment with steroids is limited by adverse side effects (hypertension, diabetes, cataract, osteoporosis). However, dexamethasone was well tolerated in this patient. The use of intravenous immunoglobulins though effective is limited by its high cost. Plasma exchange provides rapid improvement in symptoms but deterioration may occur soon after wards.

CONCLUSION

Chronic inflammatory demyelinating polyneuropathy is not a commonly diagnosed disorder in our practice. Autonomic variants are even rarer. A high index of suspicion is important in making the diagnosis. This would have been impossible without the performance of a nerve conduction study. The utility of nerve conduction studies for the diagnosis of peripheral neuropathies cannot be overemphasized.

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Ethical approval: Not required

REFERENCES
