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A CASE STUDY ON CHRONIC INFLAMMATORY DEMYELINATING POLYNEUROPATHY

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ABSTRACT
Investigations ruled out myelopathy and neuropathy after patient initial presentation of being unable to move both lower limbs left more than right for the previous six months. With no reports of fever, convulsions, or back pain, both the left and right lower limbs' mobility steadily decreased. Given the clinical picture, polyneuropathy was probably the cause. After several negative testing, it was determined that it was Chronic Inflammatory Demyelinating Polyneuropathy. The start of definitive therapy led to a successful recovery.

Keywords: *Myelopathy, Neuropathy, Seizures, Polyneuropathy*

INTRODUCTION

Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP), a disorder affecting the peripheral nerves and nerve roots, causes sensory deficits and limb weakness (Vallat *et al.*, 2010). CIDP is regarded as an immune-mediated illness, although its pathophysiology and aetiology are still unknown (Nobile-Orazio, 2014, Koller *et al.*, 2005).

Since there are many different clinical manifestations of CIDP, at least 15 sets of diagnostic criteria have been created to encompass the complete range of CIDP and its variant forms (Hughes R *et al.*, 2001, Bromberg, 2011). The criteria from the European Federation of Neurological Societies and Peripheral Nerve Society (EFNS/PNS) from 2010 are now the most frequently accepted criteria to confirm the diagnosis of CIDP. They are based on a mix of clinical and electrodiagnostic characteristics. Immunoglobulins, corticosteroids, and plasmapheresis are proven cures for CIDP (Oaklander, 2017). Typically, these medicines only have temporary, limited clinical effects. The majority of CIDP patients need on going care for years or even decades (Oaklander, 2017). Typically, these medicines only have temporary, limited clinical effects. The majority of CIDP patients need on going care for years or even decades. Therefore, CIDP is a debilitating condition that has a significant impact on patients and patient-related health care expenses (Mahdi-Rogers and Hughes, 2014, Mygland and Monstad, 2001, Chio *et al.*, 207, Lunn *et al.*, 1999, McLeod *et al.*, 1999. Guptill *et al.*, 2014). The population-based burden and associated health expenses, however, are not understood. We must evaluate the incidence and prevalence of CIDP in order to make this determination.

CASE

A 70-year-old man was hospitalised after complaining that for the previous six months, he had been unable to move both lower limbs more to the left than to the right. Both the left and right lower limbs gradually lost their ability to move.

There were no additional symptoms from the patient, such as fever, convulsions, or back pain. There were no other pertinent past medical histories or family histories found.

He suffered a leg injury at work when a large object fell on his left leg. He was able to walk normally after his procedure. After a few days, he lost the ability to lift his left leg, which ultimately spread to his right leg as well.

In conclusion, the patient's condition changed in the two weeks after admission from subjective lower limb weakness and back discomfort to proximal and symmetrical objective weakness. He did not initially complain of upper limb weakness, but proximal and symmetrical arm weakness later appeared. No sensory abnormality was discovered, and all of the reflexes were found to be symmetrically decreased.

The subsequent weeks saw a steady decline in our patient's condition, which peaked around day 45 after admission. The patient exhibited acute bilateral upper limb paralysis and no motion in his lower limbs at this time. There was no sensory abnormality found, although reflexes were proportionately weaker.

Investigations

The initial blood tests demonstrated a leucocytosis of 18.2×10^9/L and a neutrophilia of 13.4×10^9/L. There was a hyponatraemia of 120mmol/L, and C-reactive protein was 48. No ECG or chest radiography was performed.

There were a lot of normal or negative investigations. Since the haematinics and blood glucose levels were normal, diabetes mellitus- or B12/folate deficiency-related neuropathy was ruled out. Multiple measurements of creatine kinase levels revealed that it peaked at 155 units/L. Myeloma screening, cerebrospinal fluid cytology, Ca19-9, carcinoembryonic antigen (CEA), alpha-fetoprotein (AFP), and antineuronal antibodies (anti-Yo, anti-Hu, and anti-Ri) were all within normal ranges. A CT thorax–abdomen–pelvis detected no malignancy. Other normal or negative tests included: antineutrophil cytoplasmic antibody (ANCA), antinuclear antibody (ANA), urinary porphobilinogen levels, lead levels, urinary organic acids, acetylcholine receptor antibodies (myasthenia gravis), ganglioside Q1b antibodies (Miller-Fisher syndrome), voltage-gated potassium channel antibodies (encephalitis, etc), antibodies against neurofascin-155, neurofascin-186/140, contact in associated protein 1 (CASPR)/contactin-1 (some subgroups of CIDP).

The only positive biochemical investigation was the cerebrospinal fluid protein of 1.36 g/L (0.15–0.45 g/L).

Finally, nerve conduction studies (NCS) confirmed a mixed motor and sensory demyelinating and axonal peripheral neuropathy which, along with the above test results, led to the diagnosis of CIDP.

Treatment

Two cycles of intravenous immunoglobulin (IVIG) therapy were given to our patient. He regained some strength, although it was hard to tell whether it was real or just a coincidence. He did, however, respond more clearly to intravenous glucocorticoid therapy (methylprednisolone), and he was ultimately transferred to a hospital for neurorehabilitation with lowering oral steroid doses.

Outcome and Follow-Up

Following diagnosis, the patient steadily acquired strength over the course of three months, first moving themselves in wheelchairs and later completing unaided transfers. He was able to navigate short distances with crutches three months after being released, but his release from the rehabilitation facility was repeatedly postponed because of recurring UTIs and hyperglycaemia brought on by the use of steroids.

DISCUSSION

Chronic Inflammatory Demyelinating Polyneuropathy (CIDP) is a rare disorder of the peripheral nerves characterized by progressively worsening sensory loss and weakness accompanied by loss of reflexes. It is one of several diseases and syndromes marked by persistent chronic immune-mediated inflammatory response. These include demyelinating syndromes with paraproteins, POEMS (polyneuropathy, organomegaly, endocrinopathy, monoclonal protein, skin changes) syndrome, multifocal motor neuropathy, MADSAM neuropathy, and demyelinating neuropathy secondary to systemic disorders such as infection or diabetes mellitus. About 1-2 new cases of CIDP are reported annually for every 100,000 people. Although the underlying pathology of CIDP is poorly understood, it is known to involve both cellular and humoral autoimmunity. The myelin that protects the nerves is damaged, which results in CIDP. CIDP is neither self-limiting nor spontaneous. 30% of CIDP patients will eventually become wheelchair dependent if left untreated. A large proportion of disability can be avoided with early detection and appropriate care. According to current thinking, the immune system

of the body, this ordinarily protects itself, views myelin as foreign and attacks it. Myelin is a crucial part of the peripheral nervous system. It wraps around the nerve axon similar to how insulation encircles an electrical wire. The spinal cord's nerves branch out to the rest of the body, stimulating muscle contraction and transmitting sensory information back to the nervous system from receptors in the skin and joints. Myelin allows electrical impulses to efficiently travel along the nerve axon. These electrical impulses are slowed or lost when myelin is injured or destroyed, and messages transmitted from the brain are disrupted and may never reach their intended destination. It is yet unclear what triggers this process. Tests such as nerve conduction and EMG (usually showing a demyelinating neuropathy), spinal fluid analysis (usually showing elevated protein with normal cell count), blood and urine tests (to rule out other disorders that may cause neuropathy and to look for unusual proteins). Clinically, CIDP cases can be categorised as "typical" or "atypical"; typical CIDP is a symmetrical polyneuropathy that affects both the proximal and distal muscles equally, whereas atypical CIDP includes "distal acquired demyelinating symmetric" (DADS) and multifocal acquired demyelinating sensory and motor neuropathy. According to medical opinion, it is crucial to take neurological factors into account if a patient presents with a fall or leg weakness. The progression of symptoms over two to 2–4 weeks suggests GBS, whereas CIDP will have a slower natural history (more than 8 weeks from onset to nadir). Usually, prominent sensory signs such as ataxia and impaired sensation favour CIDP. Proximal limb weakness favours CIDP. It can be challenging to accurately diagnose CIDP in practise. Initial CIDP presentations can vary. Some uncommon cases only exhibit sensory neurology, while other cases exhibit severe nerve root enlargement that results in compressive pathology.

CONCLUSION

There is still a potpourri of unanswered concerns despite the fact that clinical research in CIDP has addressed crucial issues in the management of patients with CIDP. These primarily focus on diagnosis and treatment, but they also cover related aspects like epidemiology and economic burden of the disease. Due to its heterogeneous presentation (both clinical and electrophysiological) and the shortcomings of clinical, serologic, and electrophysiologic diagnostic criteria, CIDP is extremely uncommon but under-recognised and under-treated. If diagnosed early, it is advised to begin early treatment to avoid loss of nerve axons. However, many individuals continue to have symptoms including residual numbness, weakness, tremors, fatigue and other symptoms which can lead to morbidity and diminished quality of life over the long run.

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