Multifocal Motor Neuropathy: A clinical case study

Madeline A Bone

Abstract

Multifocal motor neuropathy is a rare and chronic movement disorder associated with the progressive weakening of the patient’s limbs. This case study examines the clinical presentation, findings, treatment, and outcome of a female patient treated with intravenous immunoglobulin therapy. The case study illustrates the important role the neuroscience nurse plays in the care and treatment of a patient with a complex disorder.

Keywords: multifocal motor neuropathy, movement disorders, intravenous immunoglobulin.

Introduction:

This case study will examine a patient with a rare and disabling movement disorder. The primary objective of this essay seeks to critically analyse and discuss the neuroscience nursing care and interventions of multifocal motor neuropathy. Firstly, the patients’ clinical presentation, past medical history and the underlying pathophysiology of multifocal motor neuropathy will be comprehensively explored and the epidemiology, etiology and the patients’ risk factors will be identified. Secondly, relevant diagnostic investigations will be examined with the goal of identifying the interrelationship between the patient’s health background and risk factors. Lastly, the nursing management and treatment for the patient will be discussed with a holistic approach, taking into consideration the social, ethical and psychological effects of the disorder.

The key aspects that will be discussed in this paper will be addressed with the utmost importance placed on maintaining patient confidentiality.

Case Study:

A 65-year-old female, who will be referred to as Mrs Smith, was admitted to the ward with a history of multifocal motor neuropathy first diagnosed in 2010 after experiencing left hand wasting. A nerve conduction study this admission revealed motor block. She had a history of hypertension and a previous hip replacement, otherwise the patient was generally well. The patient’s current symptoms included limited use of both hands/fingers due to significant weakness, pain in hands, ulnar muscle wastage and bilateral foot drop. The patient had been receiving a maintenance dose 22.5g intravenous immunoglobulin (IV Ig) every 4 weeks with a loading dose of 120mg once a year. The current admission on the ward was for a five-day course of 120mg of IV Ig.

Multifocal motor neuropathy (MMN) is a slow, progressive motor disorder in which the demyelination of motor axons occurs (Nowacek & Teener, 2012). The likely etiology of MMN is an autoimmune attack on the motor nerves (Lawson & Arnold, 2014). MMN is an acquired disorder that is associated with elevated levels of antibodies to ganglioside GM1 (anti-GM1) and immunoglobulin M (IgM) antibodies (Léger, Guimarães-Costa & Iancu Ferruglia, 2015). Studies reveal the link between the presence of IgM anti-GM1 antibodies in approximately 50% of MMN patients (Vlam et al. 2015; Lawson & Arnold, 2014).

Questions or comments about this article should be directed to Madeline Bone
Email address: madelinebone96@gmail.com
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GM1 is found universally in the body, however is most abundant in peripheral motor nerves (Lawson & Arnold, 2014). GM1 is responsible for numerous functions including paranodal stabilisation and ion channel clustering (Lawson & Arnold, 2014). Disruptions to these functions results in conduction failure across the paranodal regions causing a cascading effect on the action potential propagation and leading to conduction failure in the peripheral nerves (Lawson & Arnold, 2014). The abundance of GM1 is found in the myelin of motor nerves compared to the sensory nerves, which may provide insight into why MMN does not affect sensory nerve function (Lawson & Arnold, 2014). The progressive nature of the disorder is due to autoantibodies binding to GM1 gangliosides, activating the classical complement system pathway and causing nerve damage by initiating the complement membrane attack complex within the peripheral nerves (Yuki, Watanabe, Nakajima & Spath, 2010).

MMN is a chronic disorder that tends to mimic the more fatal form of movement disorders, motor neuron disease (Lawson & Arnold, 2014). A recent study revealed one third of patients are given an initial diagnosis of motor neuron disease (MND), with further testing eventually ruling MND out (Dimachkie, Barohn & Katz, 2013). MMN is considered a benign disease, however the functional impairment can have a serious effect on the patient’s quality of life (Cats et al. 2010). The prevalence of MMN is estimated to affect 1-2 in 100,000 individuals (Meuth & Kleinschnitz, 2010). A study has revealed the mean age for onset of symptoms is approximately 40 years old. (Meuth & Kleinschnitz, 2010). As in Mrs Smith’s case, the upper extremities are usually the first to be affected and most often the presenting symptom (Latov, 2014). The nerves that are most commonly affected are the ulnar, median and radial nerves, with patients presenting with difficulty extending the fingers and wrist and a reduced hand grip (Lawson & Arnold, 2014). This is in line with Mrs Smith’s initial symptoms. The patient currently has restricted movement in both hands as well as weakness in her arms and lower limbs. MMN causes muscle atrophy which is relatively mild in the early stages of the disease and worsens as the disease progresses (Berger, McCallus & Lin, 2013). The specific cause of this disorder is still not known. A known risk factor is gender, with men being twice as likely to develop the disorder than women (Sutedja, 2010). Mrs Smith does not have any past history or risk factors that would indicate that she would be at risk of developing the disorder.

To correctly diagnose Mrs Smith, she underwent nerve conduction studies. Nerve conduction studies are considered the gold standard, as the test can identify multifocal partial conduction blocks (Meuth & Kleinschnitz, 2010). Motor conduction blocks are defined as a decrease of action potentials recorded from a specific or group of muscles subsequent to proximal nerve stimulation as compared with distal nerve stimulation (Lawson & Arnold, 2014). MMN affects the multifocal partial conduction blocks on motor nerves, but not sensory nerves, which is why MMN affects the movement of the limbs but has no sensory impairment (Yuki, Watanabe, Nakajima & Spath, 2010). An international study has revealed 80% of MMN patients have conduction blocks detected in the ulnar nerves and 77% detected in median nerves (Cats et al. 2010). Mrs Smith underwent further diagnostic imaging including a computed tomography scan (CT) of the brain and comprehensive blood tests which showed no evidence of disease. However, the nerve conduction study revealed a motor block in the ulnar and median nerves which confirmed the diagnosis of MMN.

Mrs Smith had managed the disorder for nearly a decade with effective treatment options allowing her to maintain independence. The treatment Mrs Smith had been receiving for the past 6 years was intravenous immunoglobulin therapy (IVIg). MMN has no known cure, however therapies, such as immunoglobulin therapy, aim to reduce motor deficits, slow down ongoing axonal degeneration and promote remyelination (Léger, Guimarães-Costa & Iancu Ferfoglia, 2015). IVIg is a solution manufactured from human plasma protein (Australian Red Cross, 2018). It contains typical IgG antibodies with a broad spectrum of antibody activity and is used to treat patients requiring antibody replacement as well as autoimmune, hematological and neurological disorders (Australian Red Cross, 2018). The mechanism of action of IVIg is not fully understood, however it is thought the antibodies work against the mechanisms of the classical complement pathway in order to
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prevent the membrane attack (Stangel et al. 2016). IVIg neutralises pathogenic antibodies, inhibits antibody production by B cells and suppresses inflammatory mediators produced by T cells (Léger, Guimarães-Costa & Iancu Ferfoglia, 2015). Several studies prove that IVIg has a beneficial long-term effect on muscle strength, however it is not able to prevent a small decrease in muscle strength and increase of axon loss as the disorder progresses (Nobile-Orazio & Gallia, 2013; Léger et al. 2008). A retrospective study of 40 patients with MMN found that 68% of patients were dependent on long-term maintenance IVIg infusions to stabilise their motor condition (Léger, Guimarães-Costa & Iancu Ferfoglia, 2015). Most patients will become less responsive to IVIg therapy over time and will therefore require higher and more frequent doses to achieve the result (Lawson & Arnold, 2014). Similarly, another study revealed that patients who did not receive IVIg treatment suffered more severe weakness and had further progression of axon loss in comparison to those patients who received IVIg treatment (Jovanovich & Karam, 2015).

The effectiveness of IVIg treatment is dependent on how soon after diagnosis treatment is commenced. A recent international study found that 94% of the 88 MMN patients examined responded positively to IVIg treatments (Jovanovich & Karam, 2015). The 6% of MMN patients who did not respond to the treatment had been diagnosed with the disorder later and their symptoms had become advanced (Jovanovich & Karam, 2015). Furthermore, the dosage strength and frequency of infusions plays a significant role in the effectiveness of treatment (Léger et al. 2008).

Mrs Smith was admitted to the ward for a dose of 120g IVIg over five days. The Clinical use of Intravenous Immunoglobulin in Australia Guidelines recommend IVIg be given initially at a 2g/kg dosage over two to five days (Australia Red Cross, 2018). This ratio accurately correlates with Mrs Smith’s weight and the prescribed dose of 120g of IVIg. A loading dose is common practice among practitioners internationally and is then followed by maintenance doses of 0.4-1g/kg every two to four weeks (Australia Red Cross, 2018; Schaika et al. 2006). Mrs Smith had been receiving maintenance doses of 22.5g IVIg every four weeks, however in the last few months she had reported her symptoms becoming more severe. In response, her IVIg dose was increased to 24.5g.

As a registered nurse, working within the neuroscience department, progressive motor disorders are not uncommon and the registered nurse needs to be able to provide the highest quality of care for these patients. It is important the neuroscience nurse understands the pathophysiology behind the disorder, as well as the correct administration of IVIg. IVIg must be given under strict protocols, ensuring the six patient rights are completed with two nurses and cross checks of the product, dose and rate. Reactions to IVIg are most likely to occur within the first hour of the infusion, therefore the patient must be closely monitored and the patient’s vital signs checked regularly (Australian Red Cross, 2018). Common adverse reactions the neuroscience nurse must be aware of include headaches, chills or fever, nausea and vomiting. Mild allergic reactions may occur such as skin rash and mild changes to heart rate or blood pressure. The neuroscience nurse must be aware of severe reactions that may occur during transfusion such as; anaphylaxis, haemolytic, thromboembolic events or aseptic meningitis (Hahn et al. 2013). IVIg can have a negative effect of the patient’s renal function, therefore Mrs Smith’s renal function should have been checked prior to commencing the infusion and continued to be monitored over the course of five days (Lawson & Arnold, 2014).

Along with closely monitoring the patient’s vital signs during and after the IVIg infusion; the role of the neuroscience nurse is to accurately monitor upper and lower limb strength, movement and sensation. The neuroscience nurse must also help with mobility and meal assistance. It is the role of the neuroscience nurse caring for a patient with MMN to advocate for that patient and provide a multidisciplinary approach to care, involving the physiotherapist and a social worker or discharge planner if the patient is struggling to manage with their disability. Mrs Smith lived alone and was mobilising with a four-wheel walker. An occupational therapist offered to assess her living conditions to implement further aids within the house, however she declined the services. Patients with MMN are faced with
not only the physical challenges that are attributed to the disorder, but the psychological and social effects can negatively affect the patient's quality of life. In 2016, a large international study revealed 75% of MMN patient's felt exhausted and left with no energy to complete day time tasks, as well as 59% stating they were embarrassed by their limitations (Katz, Lewis & Spatafora, 2017).

Reflection:
Caring for patients such as Mrs Smith allows for improved clinical skills and an in depth knowledge of IVlg infusions, which will benefit future patients and provide fellow colleagues with a resource of information regarding the expected benefits, risks and nursing interventions that are associated with IVlg infusions.

Conclusion:
MMN is a rare disorder that purely effects motor function. However, due to the disabling and incurable nature of the condition, patients may struggle with emotional and psychological issues. The correlation between MMN risk factors and Mrs Smith's medical history was weak, with the patient's age being the only known risk factor for her developing the disorder. There is strong evidence to suggest that early treatment of MMN is the most important factor in long term functional outcome. Fortunately, Mrs Smith was treated early and has been responding well to IVlg treatment.

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Conflict of interest
The author declares no conflicts of interest.

References:


