7-2018

Chronic Inflammatory Demyelinating Polyneuropathy

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Introduction

- The topic was selected to educate health care professionals on the complex pathophysiological process of chronic inflammatory demyelinating polyneuropathy (CIDP).
- CIDP is an autoimmune disease of the peripheral nervous system in which the myelin sheath surrounding nerves is attacked, leading to improper conduction of nerve impulses to peripheral nerves (Greer & Wolf, 2015).
- There are three courses CIDP can take; progressive, relapsing-remitting, or monophasic (Schaik et al., 2016, p.2).
- Regardless of clinical course, initial presentation is nonspecific and gradual (Alabdali et al., 2017, p.223).
- Progressive course: unrelenting progression of symptoms with limited to no improvement or stabilization.
- Relapsing-remitting course: periods of exacerbations with increased symptoms accompanied by periods of remission.
- Monophasic: one occurrence of symptoms that is treated and resolves (van Shaik et al., 2016, p. 3).
- Multiple sources debate whether CIDP is over or under-diagnosed, but prevalence is currently estimated at 1 to 2 per 100,000 adults (Reynolds, Sachs & Stavros, 2016, p.32).
- CIDP is more common in males than females, and is most commonly diagnosed between ages 40-60 (Reynolds, Sachs & Stavros, 2016, p.32-33).
- CIDP has treatments available, but not all courses of disease may be corrected with available treatments, and progressive accumulation of disability may occur (van Shaik et al., 2016, p. 3).
- The majority of patients with CIDP require long-term management and treatment (van Shaik et al., 2016, p. 3).

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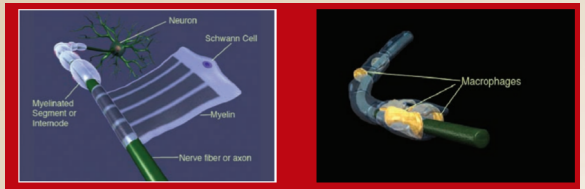
Presentation of Case

- A male between 40 and 60 years of age noticed gradual onset of bilateral weakness of his lower extremities, then numbness, and lastly difficulty walking over the course of several months.
- He presented to his physician, who performed a thorough neurological assessment and nerve conduction studies (NCS). His physician also completed an MRI and spinal tap to rule out other potential diagnoses. After obtaining test results, his physician confirmed his diagnosis of CIDP. (Kang et al., 2013, p.1-2)
- He was initially treated with steroids, but did not show improvement, so he was started on a maintenance therapy of IVIG every three weeks.



Figures 2 & 3: Myelinated Nerve Fiber & Nerve Fiber Demyelination

Retrived from: GBS/CIDP Foundation International. (2018). CIDP chronic inflammatory demyelinating polyneuropathy. *GBS/CIDP Foundation International.* Retrieved from https://www.gbs-cidp.org/support/foundation-publications/

Signs & Symptoms

- CIDP creates neurological disability which sometimes leads to decreased functional status and decreased quality of life (Alabdali et al., 2017, p.223).
- Diagnosis of CIDP is based on thorough history and neurological assessment, as well as the presence of demyelination on nerve conduction studies (Alabdali et al., 2017, p.224).
- Cerebral spinal fluid will show elevated protein levels, and MRI imaging will show enhanced nerve roots (Alabdali et al., 2017, p.224).
- Symptoms present in three categories, sensory, motor, and autonomic dysfunction (Alabdali et al., 2017, p.223).
- Motor symptoms typically present first and include weakness, especially in lower limbs, loss of deep tendon reflexes, unsteady gait, and fatigue.
- Sensory symptoms include numbness, paresthesia in a "stocking-glove" pattern, diminished sensation, and burning sensations (Reynolds, Sachs & Stavros, 2016, p.32-34).
- Autonomic dysfunction related to CIDP was previously thought to be rare, but one study showed up to 86% of patients with CIDP experience autonomic dysfunction (Pasangulapati, 2017, p. 276).
- Autonomic symptoms include orthostatic hypotension, exercise intolerance, palpitations, dizziness, weakness, and vision disturbances (Adamec et al., 2016, p.1163).
- Characterized by a slow progression of symptoms that reaches its peak greater than 2 months after symptom onset (Ripellino, Fleetwood, Cantello, & Cristoforo, 2014).

Pathophysiology of CIDP

Underlying Pathophysiology

- CIDP symptoms are a result of immune-mediated destruction of peripheral nerves.
- Acts on both the cellular and the humoral pathways of the immune system.
- "Activated T lymphocytes invade the peripheral nervous system through derangement of the blood brain barrier. Once within the peripheral nervous system these activated T cells generate pro-inflammatory cytokines and produce cytotoxic activity against myelin" (Reynolds, Sachs & Stavros, 2016, p.32).
- T cells are the main antagonists of myelin sheath with "T cells, especially T helper 1 (Th1) and T helper 17 (Th17) on one side and T regulatory (T reg) on the other" (Ripellino et al., 2014).
- While pathogenesis is not fully understood, macrophages, cytokines, and complement are also involved in degradation of myelin (Ripellino et al., 2014).
- Immune response potentially targets antigens on peripheral nerves, such as myelin protein zero, myelin basic protein, connexin 32, and gangliosides (Reynolds, Sachs & Stavros, 2016, p.32-33)
- Activated T cells survive apoptosis as a result of an altered Fas pathway function (Ripellino et al., 2014).
- "The myelin sheath is composed of various proteins, such as myelin protein zero, which account for more than 50 percent of the total membrane protein in human peripheral nervous system myelin" (Köller et al., 2003).

Significance of Pathophysiology

- It is likely that many subtypes of CIDP exist (Reynolds, Sachs & Stavros, 2016, p.32).
- More research is needed to pinpoint the exact antigens targeted in CIDP, specifically for each subtype of CIDP (Reynolds, Sachs & Stavros, 2016, p.32).
- Approximately 70% of patients respond to immunomodulation therapy, but as patients have different subtypes, some patients are difficult to treat and research pinpointing specific proteins linked to CIDP is necessary (Ripellino et al., 2014).
- In the future, treatments may be specifically designed for patients on a case by case basis after identification of the pretense of specific proteins such as CNTN1 and NF155 (Reynolds, Sachs & Stavros, 2016, p.34).

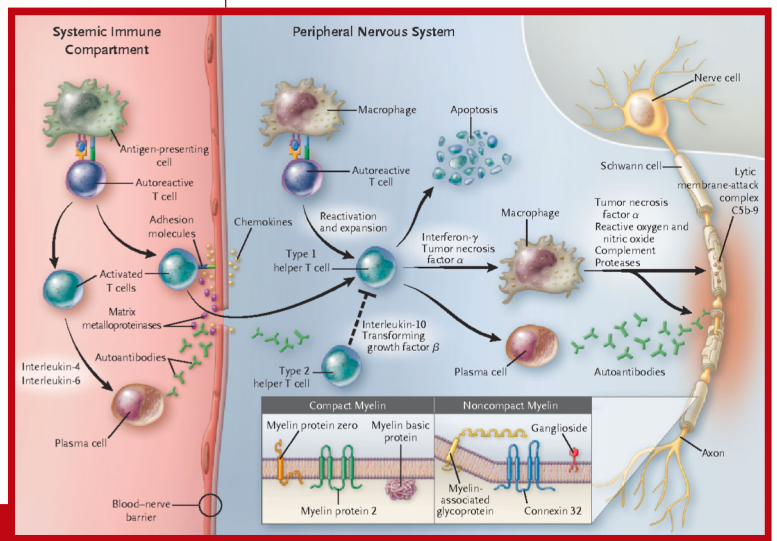


Figure 3: Immunopathogenesis of CIDP

Retrived from: Köller, H., Kieseier, B.C., Jander, S., & Hartung, H.C. (2003). Chronic inflammatory demyelinating polyneuropathy. *The New England journal of medicine*, 352 13, 1343-56.

Treatment

- Pharmacological immunosuppression with medications such as azathioprine, methotrexate, and cyclophosphamide (Ripellino et al., 2014).
- Intravenous or subcutaneous immunoglobulin administered approximately every three weeks (van Shaik et al., 2016, p. 3).
- Intravenous or oral steroids (van Shaik et al., 2016, p. 3).
- 70% of patients respond to immunotherapy (Ripellino, Fleetwood, Cantello, & Cristoforo, 2014).

Implications for Nursing Care

- Complete thorough neurological assessments.
- Involve physical and occupational therapy in management of weakness and sensation abnormalities and the impact of symptoms on activities of daily living.
- Educate patient and family members on CIDP.
- Encourage a healthy and well-balanced diet
- Monitor progression of disease process (GBS/CIDP Foundation International, 2018).

Conclusion

- CIDP is a complex autoimmune demyelinating disorder that commonly impacts patient's quality of life and activities of daily living (Alabdali et al., 2017, p.224).
- More research is needed to improve diagnosis and treatments for specific subtypes of CIDP, which will improve patient outcomes (Ripellino et al., 2014).

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