7-2016

Progressive Multifocal Leukoencephalopathy in Multiple Sclerosis

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Recommended Citation
Jackson, Lauren R., "Progressive Multifocal Leukoencephalopathy in Multiple Sclerosis" (2016). Master of Science in Nursing (MSN) Student Scholarship. 135.
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Introduction

Progressive multifocal leukoencephalopathy (PML) is a rare and deforming disease caused by the JC virus and results in demyelination of oligodendrocytes and glial cells, leading to plaque development on the brain and neurological problems (Gorelik et al., 2011). PML develops in individuals who have an immune disorder and who have been treated with immunomodulatory medications. Such individuals include those affected with Multiple Sclerosis (MS) (Gorelik et al., 2011). MS is a disease that involves inflammation of the central nervous system and causes a progressive degeneration of neurologic function and is believed to be a result of self-antigen and an autoimmune response (Keating & McCormack, 2014). APNs with no care for MS and the majority of cases are classified as relapse-remitting (Mayer & Dunsack, 2013).

The first-line medications used to treat MS show only a 30% reduction in relapses per year. Recently a new medication, Natalizumab, has shown a 68% reduction in relapse rates per year (Baldwin & Hogg, 2013). While Natalizumab shows promise in the treatment of MS, the drug has shown to increase the risk of development of PML, especially in young people under 36 months of age (Sobocinski et al., 2011). There is no cure for PML and 30-40% of patients who develop the disease die within the first few months of diagnosis. Those that survive, are typically left with severe neurological damage (National Institute of Neurological Disorders and Stroke [NINDS], 2015).

As an Advanced Nurse Practitioner (APN), the author will encounter patients diagnosed with MS, who are at a high risk for developing the disease (National Multiple Sclerosis Society [NMSS]). Therefore, an understanding of PML and its relation to its use in patients with MS is needed (Fitzgerald, 2015). By understanding PML, the APN should be aware that PML can develop in patients with MS and needs to be able to discuss the treatment options and rules/benefits to assist their client during this difficult time. Therefore, an understanding of PML and its relation to MS is important.

Signs and Symptoms

Signs and symptoms of PML can vary greatly depending on the area of the brain that is affected (Berger et al., 2015). Patients may initially present with distinct behavioral or cognitive changes that are reported by family members (Fitzgerald, 2015). Other neurological symptoms that can occur include hemiparesis, lethargy, aphasia, anxiety, seizures, impaired memory and ataxia (Fitzgerald, Keating, & McCormack, 2014). The disease continues to progress slowly over time with worsening symptoms, leading to death or neurological deficits (NINDS, 2015). See Figure 1.

Underlying Pathophysiology

PML is thought to be caused by the JC virus. The JC virus is discovered after tetanus was observed on the bodies of oligodendrocytes. Although much has been learned about the virus, there is still a lot that needs to be discovered to gain a better understanding of the JC virus and PML (White & Khalil, 2011). The JC virus is thought to affect 66-92% of the general population, as evidence by JC antibody present in populations studied, and is acquired in childhood (White & Khalil, 2011). Despite the prevalence of JC antibody, the actual occurrence of PML is low leading to the conclusion that the virus is re-activated, after a period of latency, in immunosuppressed individuals (White & Khalil, 2011). The JC virus is transmitted during childhood and is thought to have initial portal of entry into the tonsils and GI tract. The body’s immune system fights the virus, defeats the virus, and then the virus enters a latent state within the tonsils, bone and/or kidneys. The majority of people infected with the latent JC virus will never know the virus is there, unless they develop an autoimmune disorder or take immunosuppressive medications (White & Khalil, 2011). See Figure 2.

The latent form of the JC virus can also be found in the bloodstream, primarily as lymphocytes and can circulate in the brain (White & Khalil, 2011). Glycoproteins are released due to the autoimmune response in MS, causing a reaction that had prior transmission and replication. A PML lesion on the brain can then develop due to the initial immunosuppression (White & Khalil, 2011).

CD4 T cells play an important role in eliminating strains and controlling those that remain (Bossolasco et al., 2014). Natalizumab suppresses T lymphocytes from crossing the blood-brain barrier, as an effort to control the demyelination in MS, which may prevent these cells from supporting the latent JC virus adding to the reactivation and development of PML (White & Khalil, 2011). The resulting development of PML plaques can lead to glial cell loss and demyelination leading to neurological signs and symptoms (White & Khalil, 2011).

Significance of Pathophysiology

The pathophysiology is significant in that many patients diagnosed with MS and attempting or undergoing immunosuppression therapy may also have the antibodies for the JC virus which increases their risk of reactivating the infection, resulting in PML. Therefore, screening and close observation is warranted in the patient with MS. The pathophysiology of the disease assists in diagnosing. The lessons that develop in the white matter of the brain can be observed on MRI (see Figure 3). The signs and symptoms can occur gradually as the disease spreads throughout the brain, and careful spinal fluid can be analyzed for presence of the JC virus or antibodies (Berger et al., 2015).


Conclusion

The JC virus is a fairly common virus that many people are unaware of (National Multiple Sclerosis Society). However, those with MS are at a higher risk for developing the disease (National Multiple Sclerosis Society). Therefore, an understanding of PML and its relation to its use in patients with MS is needed (Fitzgerald, 2015). By understanding PML, the APN should be aware that PML can develop in patients with MS and knows what signs to look for and what further testing is needed. If PML is suspected, fast action and referral is needed by the APN.