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Lauren R. Jackson

Otterbein University, lauren.jackson@otterbein.edu

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Progressive Multifocal Leukoencephalopathy in Multiple Sclerosis
Lauren Jackson BSN, RN
Otterbein University, Westerville, Ohio

Introduction

Progressive multifocal leukoencephalopathy (PML) is a rare and debilitating 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 immunosuppressive 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 deterioration of neurological function and is believed to be a result of self-antigen and an autoimmune response (Keating & McCormick, 2014). There are no cures for MS and the majority of cases are classified as relapsing-remitting (Makar & Dinak, 2013). The first-line medications used to treat MS show only a 10% reduction in relapses per year. Recently a new medication, Natalizumab has shown a 40% reduction in relapses 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 long-term users (more than 38 months) (Shebolt et al., 2011). There is no cure for PML and 30-50% 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 document patient diagnosis with MS, as MS is a fairly common neurological disorder with 1 in 200 Americans living with the disease. The risk for developing the disease (Multiple Sclerosis Society [MSS]), however, may be newly diagnosed or looking for new treatment options. Knowing the risk of PML, signs and symptoms and its effects can assist the APN and patient to make an informed decision about their treatment options.

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, fatigue, seizures, impaired memory and ataxia (Fitzgerald, Koralnik, & Wiendl, 2013).
• The disease continues to progress slowly over time with worsening symptoms, leading to death or neurological disabilities (NINDS, 2015). See Figure 1.

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 lesions that develop in the white matter of the brain can often be observed on MRI (see Figure 2). The signs and symptoms can correspond to the disease spread throughout the brain, and cerebrum spinal fluid can be analyzed for presence of the JC virus or antibodies (Berger et al., 2015).
• Patients with MS were more likely to develop immunosuppression therapy or another underlying immune disorder, develop PML at a rate of 13 to 1,000. The statistics are based on a time frame of 5 years (Fitzgerald, 2015).
• A development of PML plaques can lead to glial cell loss and demyelination leading to neurological signs and symptoms (White & Khalili, 2011).

Underlying Pathophysiology

• PML is thought to be caused by the JC virus. The JC virus was discovered after retinas were 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 & Khalili, 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 & Khalili, 2011). Despite the prevalence of JC antibodies, 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 & Khalili, 2011).
• The JC virus is transmitted during childbirth 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 the virus enters a latent stage 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 & Khalili, 2011). See Figure 2.
• The latent form of the JC virus can also be found in the bloodstream, particularly in B lymphocytes and can circulate in the brain (White & Khalili, 2011).
• Gliomas are released due to the autoimmune response in MS, causing a reactive cytokine cascade and destruction. A PML lesion on the brain can then develop due to the initial immunosuppression (White & Khalili, 2011).
• CD8 T cells play an important role in eliminating strains and controlling their spread within patients. Natalizumab suppresses T lymphocytes from crossing the blood-brain barrier, in an effort to control the demyelination in MS, which may prevent these cells from spreading the latent JC virus adding to the reactivation and development of PML (White & Khalili, 2011).
• The resulting development of PML plaques can lead to glial cell loss and demyelination leading to neurological signs and symptoms (White & Khalili, 2011).

Implications for Nursing Care

• The APN should be aware that any patient presenting with a history of MS and abrupt onset should be evaluated for PML of the brain. These patients may be newly diagnosed or looking for new medications for its treatment, causing the development of PML (Gorelik et al., 2011). PML affects glial cells and creates glial cell damage (National Institute of Neurological Disorders and Stroke [NINDS], 2015). Despite the prevalence of JC virus, there is still much that needs to be discovered to gain an understanding of the JC virus and PML (White & Khalili, 2011). Patients with MS are more likely to develop immunosuppression therapy or another underlying immune disorder, develop PML at a rate of 13 to 1,000. The statistics are based on a time frame of 5 years (Fitzgerald, 2015).
• Symptoms of MS relapse tend to develop quickly, over hours to days while PML symptoms occur more slowly (Fitzgerald, 2015).
• MS responds to steroid therapy or levels off, whereas PML continues to progress despite treatment (Fitzgerald, 2015). Patients with MS are stressed for a cure by a specialist, however APNs may encounter these patients in family practice or even in the acute care setting and need to be aware of PML and its effects. Symptom management and attempting to improve quality of life may be a vital part of the APN’s role, even dealing with a patient with PML.
• APNs in family practice may encounter newly diagnosed patients with MS and needs to be able to discuss the treatment options and rules/benefits to assist their client during that difficult time. Therefore, understanding of PML and its relation to MS is important.

Conclusion

The JC virus is a fairly common virus that many people are unaware of, but with the pathophysiology of the virus, this may affect its dormancy for lifelong. However, the virus can reactivate in certain immunosuppressive conditions and certain medications for its treatment, causing the development of PML (Gorelik et al., 2011). The APN should be aware that PML can develop in patients with MS and know what signs to look for and further testing is needed. If PML is suspected, fast action and referral is needed by the APN.

References


Figure 3 retrieved from: http://pmllconsortium.org/healthcare-professionals/diagnosis

Figure 3 retrieved from: http://pmllconsortium.org/healthcare-professionals/diagnosis

Figure 2 retrieved from: http://www.nature.com/ncr/journal/v47/529/tab/ncr2010.16#F1

Figure 1 retrieved from: http://teahub.io/articles/progressive_multifocal_leukoencephalopathy.php

Image 1 retrieved from: http://nethealthbook.com/articles/progressive_multifocal_leukoencephalopathy.php