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Case Report

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Progressive multifocal leukoencephalopathy in HIV patient: A Rare Case Report.

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Abstract

Progressive multifocal leukoencephalopathy (PML) is a rare demyelinating disease caused by the JC virus, predominantly affecting immunocompromised individuals, including those with HIV/AIDS. This case report describes a 40-year-old HIV-positive female with a history of ART non-adherence, presenting with neurological deficits and diagnosed with PML. A 40-year-old Hindu female presented with multiple episodes of generalized tonic-clonic seizures over the past 10 days, blurring of vision, occasional diplopia in the right eye, fatigue, and right upper limb weakness with tingling sensation. She had been HIV-positive for 10 years and was on ART. On examination, she was conscious but confused. Neurological examination revealed decreased visual acuity in the right eye, mild confusion, attention and concentration difficulties, right upper extremity weakness (4/5 strength), positive Babinski sign on the right. MRI brain showed bilateral multifocal asymmetric white matter lesions. CD4 count was 80/mm³. She was treated with IV fosphenytoin, IV dexamethasone, and restarted on ART. Her condition improved, and she was discharged on oral levetiracetam with follow-up planned. This case underscores the importance of adherence to ART in preventing severe opportunistic infections like PML in HIV-positive patients.

Keywords:- Progressive multifocal leukoencephalopathy, Human immunodeficiency Virus, Antiretroviral therapy, Magnetic resonance imaging.

INTRODUCTION

Progressive multifocal leukoencephalopathy (PML) is a rare and often fatal demyelinating disease of the central nervous system caused by the reactivation of the John Cunningham (JC) virus in immunocompromised individuals.

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First identified in 1958, PML predominantly occurs in individuals with compromised immune systems. The predisposing conditions include haematological malignancies, organ transplants, or advanced Human Immunodeficiency Virus (HIV) infection. Among these, HIV-associated PML represents a significant challenge due to the high prevalence of immunosuppression in this population.[1]

The JC virus is a polyomavirus that remains latent in approximately 50-80% of the general population. In immunocompromised hosts (particularly those with depleted CD4+ T-cell counts below 200 cells/µL) the virus can reactivate. This reactivation may cause widespread demyelination in the central nervous system. This demyelination arises from the destruction of oligodendrocytes that results in progressive neurological deficits. [2] In the context of HIV, PML has become an important diagnostic consideration, especially as patients may present with neurological symptoms even before a formal diagnosis of AIDS. Clinical manifestations of PML are diverse and often include cognitive decline, motor weakness, visual disturbances, speech abnormalities and ataxia. The progression of symptoms is typically rapid, leading to severe disability or death in untreated cases.[3]

The diagnosis of PML depends upon combination of clinical, radiological, and laboratory findings. Magnetic Resonance Imaging (MRI) plays a central role, often revealing multifocal, confluent, hyperintense lesions in the white matter on T2-weighted sequences. These lesions lack significant mass effect or enhancement, distinguishing them from other central nervous system (CNS) pathologies. Confirmation of the diagnosis is typically achieved through polymerase chain reaction (PCR) detection of JC virus DNA in cerebrospinal fluid (CSF). PCR remains a test with high sensitivity and specificity for diagnosis of JC virus infection.[4]

Management of PML focuses on optimizing antiretroviral therapy (ART) to restore immune function with supportive care. Despite advances in ART, the prognosis of PML remains poor, with a high mortality rate and significant residual neurological impairment among survivors. Adjunctive treatments including anticonvulsants and corticosteroids may be employed based on individual clinical scenarios. Anticonvulsants, such as levetiracetam and fosphenytoin, are frequently used to manage seizures in these patients. The use of corticosteroids is more controversial and generally reserved for immune reconstitution inflammatory syndrome (IRIS) to mitigate inflammatory damage.[5]

Given the significant morbidity and mortality associated with PML, early recognition and intervention are important. This case report highlights a rare case of HIV-associated PML, emphasizing the clinical, radiological, and therapeutic challenges encountered in its management.

CASE REPORT

A 40-year-old Hindu female, diagnosed with HIV 10 years ago, presented with multiple generalized tonic-clonic seizures over the past 10 days. She reported blurring of vision and occasional diplopia accompanied by fatigue and weakness in the right upper limb with a tingling sensation. Her adherence to ART had been poor over the preceding months. Her spouse, also HIV-positive, had passed away a few years ago. There was no history of diabetes, hypertension, thyroid disease, or other chronic illnesses.

On examination, the patient was conscious but disoriented to time, place, and person. Her vital signs were stable, with a blood pressure of 122/80 mmHg, a pulse rate of 88/min, and a respiratory rate of 18/min. General examination revealed no pallor, icterus, cyanosis, clubbing, or lymphadenopathy. Neurological examination showed decreased visual acuity in the right eye, mild confusion, and difficulty in attention and concentration. Motor strength was 4/5 in the right upper limb and 5/5 in other extremities. Sensory examination revealed decreased pinprick and light touch sensation in the right upper limb, and dysmetria was noted on finger-to-nose testing. Gait was unsteady with a tendency to veer to the right.

Investigations revealed a CD4 count of 80/mm³. Fundoscopy revealed primary optic atrophy. CSF analysis was normal and PCR for JC virus was not performed due to resource limitations.Routine laboratory parameters, including kidney and liver function tests, were within normal limits. MRI of the brain showed bilateral multifocal asymmetric white matter lesions, consistent with PML.

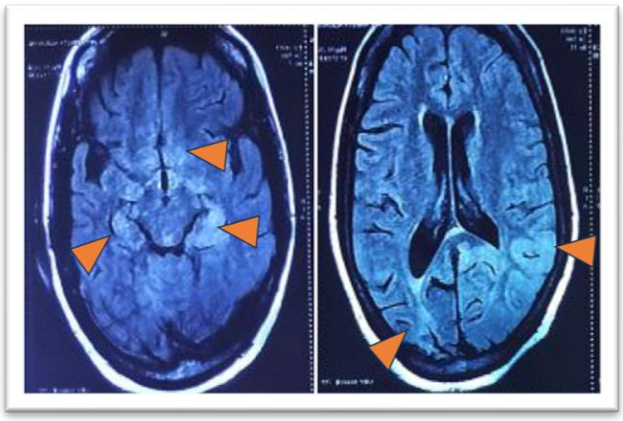

Figure 1:- Bilateral multifocal asymmetric white matter lesions on MR angiography consistent with Progressive multifocal leukoencephalopathy (PML).

The patient was managed with intravenous fosphenytoin for seizure control and intravenous dexamethasone for a week, alongside the reinitiation of ART with a tenofovir-lamivudine-dolutegravir (TLD) regimen. Gradual improvement in weakness and orientation was observed over seven days of hospitalization. She was discharged with levetiracetam 500 mg twice daily and advised regular follow-up.

DISCUSSION

This case underscores the diagnostic and therapeutic challenges of managing PML in HIV-infected individuals. The presentation of seizures, visual disturbances, and focal neurological deficits, combined with MRI findings, was consistent with advanced PML.[6] Early initiation of ART and supportive care facilitated clinical improvement, emphasizing the critical role of immune restoration in disease management.[7] Several similar cases have been reported in the literature.

Choudhary S et al reported a case study to describe the clinical presentation and diagnostic challenges of AIDS-related progressive multifocal leukoencephalopathy (PML), a rare neurological complication. The study detailed the case of a 40-year-old male presenting with left-sided hemiparesis and cranial nerve palsy. Diagnostic investigations, including MRI, revealed typical white matter lesions associated with PML, while cerebrospinal fluid analysis excluded other infections. The patient tested positive for HIV with a CD4 count of 120/µL, but polymerase chain reaction (PCR) testing for JC virus was unavailable. Based on clinical and radiological evidence, the diagnosis of AIDS-related possible PML was established. Despite initiating highly active antiretroviral therapy (HAART) and prophylaxis for opportunistic infections, the patient succumbed to the disease within one month.On the basis of these findings, the authors concluded that AIDS-related PML remains a rare but significant neurological condition in developing countries, emphasizing the need for improved diagnostic capabilities, such as JC virus PCR testing, and awareness of this condition among clinicians. They recommended early suspicion and prompt management in similar presentations.[8]

Shah R et al conducted a review study to analyze the imaging manifestations of progressive multifocal leukoencephalopathy (PML) in immunocompromised patients. The review focused on neuroimaging features, particularly those from magnetic resonance imaging (MRI), in populations including individuals with HIV/AIDS, organ transplant recipients, and patients on immunosuppressive therapies. Typical findings included multifocal, asymmetric white matter lesions without mass effect or significant enhancement, appearing hyperintense on T2-weighted and FLAIR sequences and hypointense on T1-weighted images. In HIV-positive patients, lesions were predominantly in the subcortical white matter, while non-HIV immunosuppressed patients showed greater cerebellar and brainstem involvement. Advanced imaging techniques such as diffusion-weighted imaging revealed variable diffusion patterns, and proton magnetic resonance spectroscopy showed decreased N-acetylaspartate with elevated choline levels. On the basis of these findings, the authors concluded that prompt recognition of these imaging patterns is essential for early diagnosis and management of PML and recommended heightened clinical vigilance for PML in at-risk patients presenting with new neurological symptoms.[9]

A high index of suspicion is crucial in patients with HIV presenting with neurological symptoms, given the broad differential diagnosis that includes opportunistic infections, primary CNS lymphomas, and demyelinating disorders. Early diagnosis through comprehensive clinical evaluation and appropriate imaging studies, such as MRI, is essential to identify the underlying pathology.[10]

Prompt management is vital to improve outcomes in these patients. In this case, the patient demonstrated significant clinical improvement

with the initiation of antiepileptic therapy and corticosteroids, alongside the resumption of ART. Furthermore, the case highlights the need for ongoing monitoring and follow-up to assess the patient's response to treatment, prevent recurrence and improve the overall quality of life. Regular follow-up ensures that any changes in the patient's neurological status can be promptly addressed, and adjustments to the therapeutic regimen can be made as necessary.

CONCLUSION

Managing HIV-associated PML requires a comprehensive approach involving early recognition, timely diagnosis, and prompt initiation of ART and supportive therapies. This case illustrates the critical importance of ART adherence and highlights the ongoing need for awareness and vigilance in diagnosing and managing this rare yet devastating condition.

Conflict Of Interest

None

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None

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