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A CASE REPORT OF PROGRESSIVE MULTIFOCAL LEUKOENCEPHALOPATHY IN A PATIENT OF HIV

General Medicine

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ABSTRACT

Progressive Multifocal Leukoencephalopathy (pml) Is A Rare And Fatal Viral Disease Occurring Commonly In Immunocompromised Patients. Pml Is Characterized By Multifocal Areas Of Demyelination Of Varying Size Distributed Throughout The Brain But Sparing The Spinal Cord And Optic Nerves.patients Often Present With Visual Deficits, Dementia, Confusion,altered Mental Status,personality Changes, Weakness Including Hemi- Or Monoparesis, Ataxia And Seizures.diagnosis Of Pml Is Frequently Based On Mri Findings .the Most Commonly Associated Conditions With Pml Are Aids,hematologic Malignancies, Transplant Recipients And Chronic Inflammatory Disease.we Here Present You A Case Of Pml In A 63 Year Old Hiv Positive Male Patient ,who Was Not Compliant To Haart.

KEYWORDS

LEUKOENCEPHALOPATHY, JC VIRUS, DEMYELINATING DISORDER, HIV,

INTRODUCTION

Case Report

We present a case,of a 63 years old male brought to our tertiary care hospital by his wife and elder son for complaints of confused mental state for 1 month,intermittent fever for 15 days and inability to walk and stand for 7 days.

Patient was a retired government school teacher by occupation. Patient consumed a mixed diet and was a chronic alcoholic for the past 20 years.The patient was diagnosed with retroviral disease (HIV) 4 years prior but was not compliant to HAART medications.

On clinical examination the patient was confused, afebrile, averagely built. Vitally patient had a pulse of 94/min regularly, with all peripheral pulses palpable. Patient had a blood pressure of 110/80 mm HG and SPo2 of 98% on room air and respiratory rate of 16/min. Patient had no pallor cyanosis, icterus, clubbing, lymphadenopathy or edema .

Systemic examination:

Cardiovascular examination- Both heart sounds were normal with no audible murmur.

Respiratory system examination-Breaths sounds were normally and equally heard over both lung fields, with no adventitious sounds.

Neurological examination –Patient was confused, not oriented to time place and person and did not obey oral commands.He had bowel and bladder incontinence and had gradually become aphasic over a duration of last 20 days.He accepted oral food and fluids only when fed by another person.There was no visible muscle wasting.He had spasticity in all four limbs.Plantar reflex in both lower limbs was extensor. The deep reflexes namely biceps, triceps, supinator, knee jerk, ankle jerk had +3 grade response.Patient had no signs of meningismus and involuntary movements.

Per abdomen examination: .No tenderness/guarding/ rigidity were present on palpation.Bowel sounds were normal.

MRI brain of the patient showed multiple variable sized subcortical and deep white matter lesions in bilateral fronto parietal lobes. No mass effect was noted. Patient was diagnosed with PML radiologically.

DISCUSSION:

Progressive Multifocal Leukoencephalopathy (PML) is a rare and fatal viral disease that is characterized by progressive inflammation of the white matter of the brain at multiple locations . An estimated 2-7% of people with HIV disease will develop PML. The cause of PML is a type of polyomavirus called the JC virus (John cunnigham virus) that resides in latent form in 70 – 90% of the adult population worldwide. The virus can cause persistent asymptomatic infection in approximately one third of the adult population and only causes disease when the immune system has been severely weakened. It occurs almost exclusively in people with severe immune deficiency, such as transplant patients on immunosuppressive medications, patients receiving certain kinds of chemotherapy or AIDS. The virus usually remains latent, typically only causing PML in immunocompromised patients. PML is a demyelinating disease, in which the myelin sheath covering the axons of nerve cells is gradually destroyed, impairing the transmission of nerve impulses. .PML usually affects the white matter of the brain (cortex).PML may also involve the basal ganglia and deep white nuclei as there are white matter fibers in these regions also.PML has more rapid progression compared to other demyelinating diseases. The median survival of patients with PML is 6 months and in 9% of patients, survival exceeds 1 year. MRI offers superior sensitivity in the detection and characterization of the lesions though the diagnosis may be suspected by CT scan. The pattern may be unilateral, but more often bilateral and asymmetrical. The CSF biochemistry and cytology is normal with mild elevation in protein and/or igG may be found. In CSF, detection of JC virus by PCR is diagnostic.

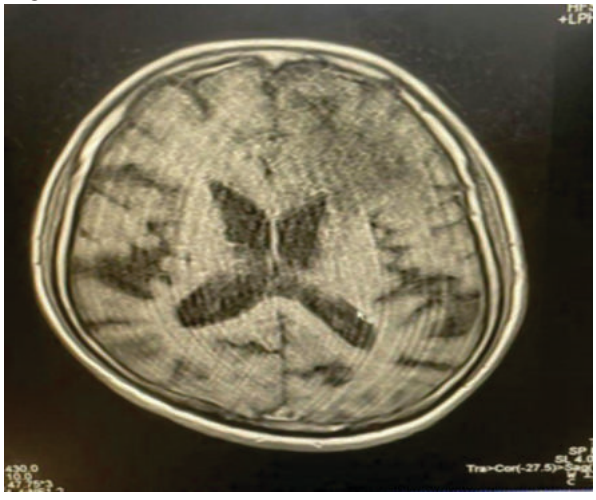

Figure 1- T1 Wt image shows low signal intensity areas in bilateral fronto parietal lobe.Figure 2- T2 Wt image shows high signal intensity areas in bilateral fronto parietal lobe.

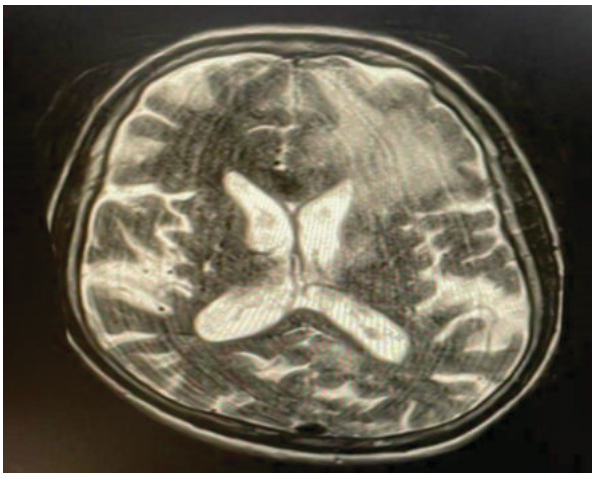

Figure 2- T2 Wt image shows high signal intensity areas in bilateral fronto parietal lobe.

CONCLUSIONS

Progressive multifocal leukoencephalopathy is a rare but fatal condition which most commonly occurs in immunocompromised patients. There is no specific antiviral agent available for JC virus infection and the existing treatment goal is to restore the host immune system.In HIV positive patients treated with HAART, 1 year survival is approximately 50 % although 80% of survivors have significant neurologic sequelae.Although institution of HAART enhances survival in HIV positive PML patients,the associated immune reconstitution in patients with an underlying opportunistic infection such as PML may also result in a severe CNS inflammatory syndrome(IRIS) associated with clinical worsening.A number of drug agents such as Cidofovir, Cytarabine, Mefloquine,Pembrolizumab etc are still under trials.Physiotherapy and nursing care are important besides drug therapy**.**

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