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A Rare Presentation Of HIV As Progressive Multifocal Leukoencephalopathy

Dr. Arun Kumar. S[1], Dr.Sakthivel.V [2], Dr.Deepak.E.R[3]

[1]Associate Professor, Department of General Medicine, Vinayaka Mission's Medical college &Hospital, Karaikal, India; ORCID Id: https://orcid.org/0000-0002-0326-9473
[2]Head of the department& Professor, Department of General Medicine, , Vinayaka Mission's Medical college & Hospital, Karaikal, India;
[3] Postgraduate, Department of General Medicine, Vinayaka Mission's Medical college & Hospital,Karaikal, India;

Email Id: [1]avsarun.kkl@gmail.com, [2]drshruthisakthi@rediffmail.com, [3]deepak.ross@yahoo.com

ABSTRACT:
Progressive multifocal leukoencephalopathy (PML) is a rare and severedemyelinating diseasewhich occurs in immunocompromised patients. A 51-year-old male presented to us with a difficulty in walking and loss of sensation on left side, he was provisionally diagnosed to be a case of cerebrovascular accident. His serology tests came as positive for HIV. Further work up with imaging as well as laboratory testing was done and he was proved to be a case of progressive multifocal leukoencephalopathy which is rarely reported in Indian literature. Our patient unfortunately passed away after 3 days. This case is a reminder that one should keep an open mind for multiple as well as rare possibilities while dealing with a patient of neurological deficit.

Keywords: JC virus; demyelinating diseases; progressive multifocal encephalopathy

1. INTRODUCTION

Progressive multifocal leukoencephalopathy is a relentless disease which causes demyelination of the central nervous system. It is caused by a polyoma JC virus and occurs typically in immunocompromised patients.[1] Here we report a 51-year-old male who presented with difficulty in walking and loss of sensation on left side, he was provisionally diagnosed to be a case of cerebrovascular accident. Laboratory results showed the patient to be HIV positive. Further work up was done and we arrived at a diagnosis of HIV related PML. We are reporting this case as AIDS presenting as PML has been very sparingly been reported in Indian literature.

Case report:

A 51-year-old male, a driver by profession with history of chronic alcohol consumption presented with an acute onset of difficulty in walking along with reduced sensation over the left side of his body for the past 2 weeks. The patient gave a history of generalized itching, loss of appetite and weight along with occasional bouts of fever. The patient was poorly built, had oral candidiasis and multiple hypopigmented plaques over his limbs. His vitals at time of admission were normal. A detailed examination of the central nervous system showed intact higher functions; however, he had reduced power of 4/5 in left upper limb, 2/5

in left lower limb which was hypertonic with exaggerated deep tendon reflexes. Sensory modalities of touch and pain was reduced in left side, whereas vibration and temperature were lost. Plantar showed extensor response on both sides. Examination of the cardiovascular and respiratory system were normal. An ophthalmology opinion suggested the patient to have gaze palsy. A specialist dermatologist opinion confirmed that the patient had multiple plaques over both his limbs along with positive auspitz sign suggesting the patient to have psoriasis. An initial diagnosis of a cerebrovascular accident was contemplated.

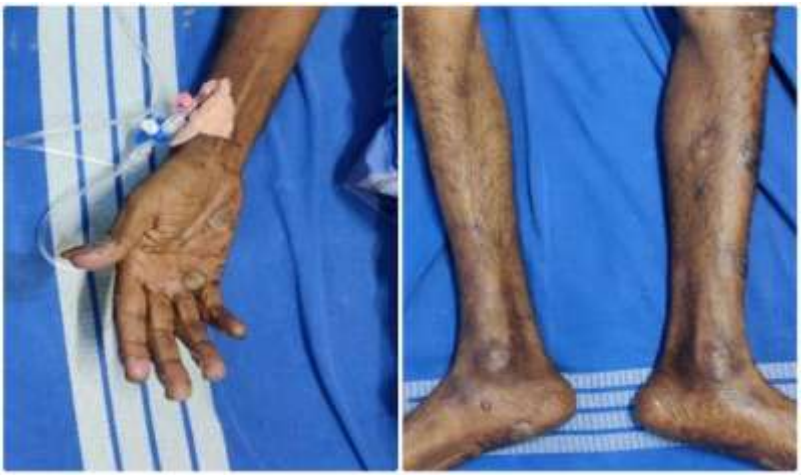

Figure 1 : The patient was in an ill nourished state with a history of significant weight loss

Figure 2 : Multiple plaques were present over the patients upper and lower limbs. Auspitz sign was positive.

Routine investigations revealed an elevated ESR, mildly raised liver enzymes, normal electrolytes, normal renal function test, normal ECG and chest X-ray. His HIV card test came to be positive which was confirmed by ELISA and western blot. The absolute CD4 count was 140/µL. His MRI of brain showed multiple ill-defined T2/Flair hyperintense lesions in bilateral frontal lobes, bilateral parietal lobes, left temporal lobes, left occipital lobe, bilateral basal ganglia and right mid brain.

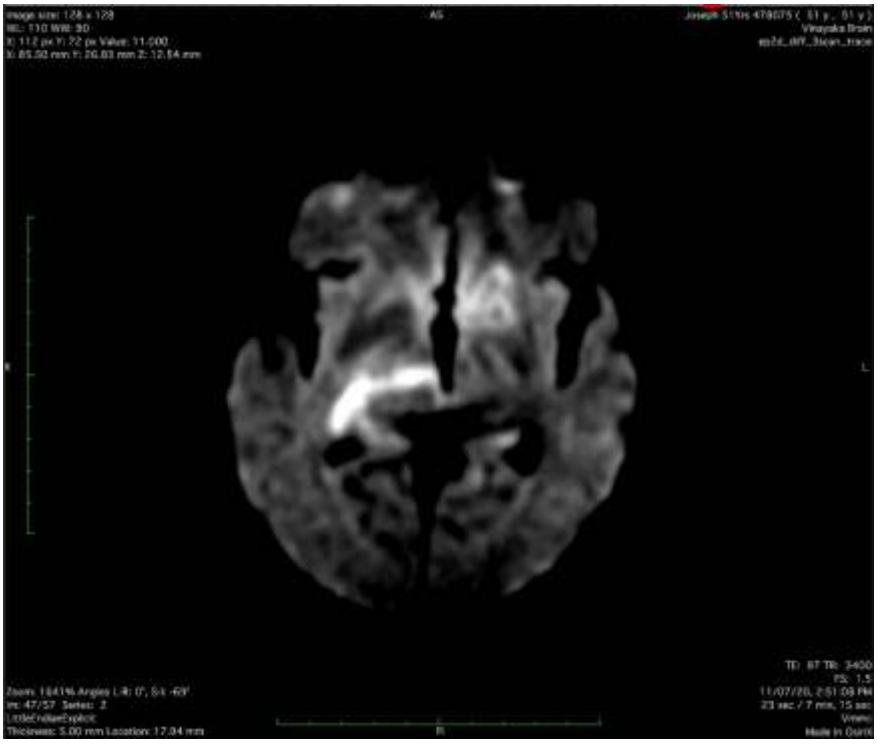

Figure 3 : Axial T2/FLAIR images shows assymetrical patchy areas of high signal intensity involving the right temporal region with multiple punctate high T2 signal lesions around the periventricular and subcortical white matter

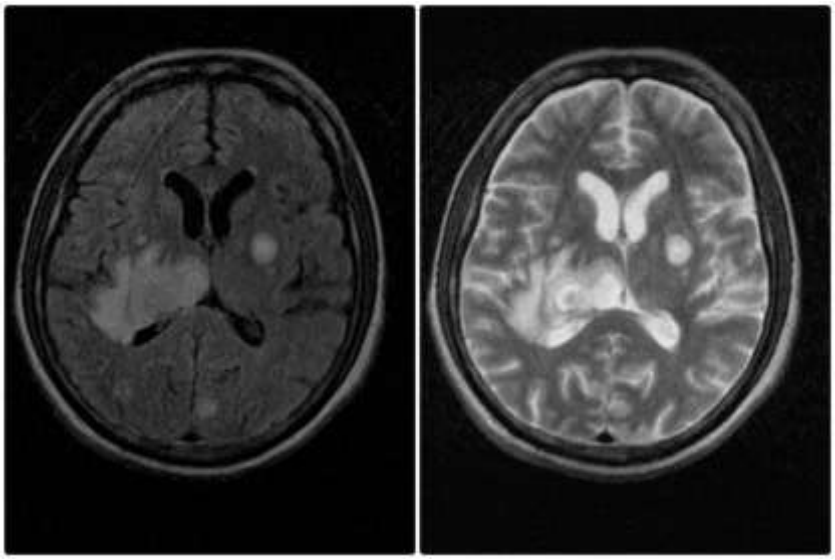

Figure 4 : Peripheral patchy diffusion restriction is seen in DWI images

Cerebrospinal fluid analysis was mostly unremarkable, apart from a mildly elevated protein. Cultures were negative. PCR of CSF was done for the John Cunningham (JC) virus and it was positive. Patient was treated conservatively with empirical antibiotics, antihistamines, IV fluids and physiotherapy. During his stay in the hospital his sensorium deteriorated. After diagnosis of PML he was referred to the nearest antiretroviral center where HAART was initiated along with other supportive medication and care, but unfortunately, he passed away after 3 days.

2. DISCUSSION

Progressive multifocal leukoencephalopathy (PML) is a demyelinating disease which is rare and usually fatal. The causative agent was identified as the human polyomavirus (JCV) in 1967. It was named as JC virus in the year 1971 from the initials of the first case (John Cunningham) from whom the virus was first isolated.[2,3]

Since the AIDS epidemic, the incidence of opportunistic infections has increased notoriously. In case with cellular immunodeficiencies, reactivation of the latent JC virus can occur ultimately resulting in PML. PML is usually seen in HIV patients and they account for up to 85 percent of all cases. Thus far, the established clinical manifestation of the JC virus is PML. In developed countries the disease has been recognized in around 2 to 4 percent of HIV patients in the pre-HAART period.[4–6]

The JC virus causes destruction of the myelin producing oligodendrocytes in the central nervous system which produces the neurologic signs and symptoms of the disease. The important pathologic features are the intranuclear inclusion in oligodendrocytes (which are particles of the JC virus) and large multilobulated nuclei. The clinical presentation of PML consists of progressive focal neurological dysfunction manifesting commonly as monoparesis, hemiparesis, aphasia, dysarthria, ataxia or visual field defects. The patient may have an altered mental status with confusion, dementia or even coma. Seizures have been reported but the incidence is less (<10%). There are however, no features suggestive of systemic infection or raised intracranial pressure.[7]

The reactivation of JCV does not only cause PML but has also been known to cause JC virus associated granule cell neuronopathy [8], JC virus encephalopathy [9] or JC virus associated meningitis.[10]

The stages to arrive at a diagnosis of PML consist of Clinical suspicion, Radiological identification and Etiological confirmation (by CSF/tissue analysis).[11] The first step depends on the progression of focal neurological symptoms in the setting of an immunocompromised state. The next step involves neuroimaging (MRI is preferred) which show white matter lesions in areas corresponding to the deficit. Due to demyelination, it appears hyperintense on T2 sequences and hypointense on T1 sequences (which indicate the destruction of the white matter). The later aids in distinguishing PML from other pathologies such as HIV-1 encephalopathy which has more diffuse central white matter changes on the T1 sequences. In PML there is usually no or minimal contrast-enhancement and there is no mass effect which differentiates it from cerebral toxoplasmosis and primary CNS lymphoma.[12] The etiological diagnosis is made by the detection of JCV DNA in the cerebrospinal fluid (by PCR). The sensitivity of this method is around 72-92% while the specificity is around 92-100%.[13]

Numerous strategies have been tried for the treatment of the JC virus, but most of them were unsuccessful. The only therapy which has been proved effective is HAART which has increased the survival rate. The reported 1-year survival with HAART is around 39-56%.[14,15]

3. CONCLUSION

JC virus-related PML should always be suspected in a HIV patient who presents with sudden progressive neurologic manifestations and lesions in white matter on MRI brain scan. Awareness of this entity and early diagnosis are essential considering the extremely poor prognosis.

4. DECLARATION OF PATIENT CONSENT

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Conflict Of Interest: The authors declare that there is no conflict of interests regarding the publication of this manuscript

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