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HIV Myelopathy with Pml: A Rare Presentation

MEDICAL SCIENCE

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ABSTRACT

Introduction: Spinal cord myelopathy is often present in patients with AIDS as a part of neurocognitive disorder. Vacuolar myelopathy is characterized by subacute onset and presents with gait disturbances, ataxia, spasticity and bladder-bowel dysfunction. . Progressive Multifocal leucoencephalopathy(PML) is characterized by altered mental status, motor deficits, limb & gait ataxia, and visual symptoms such as hemianopia and diplopia.

Case History: We describe a case of 20 years old male, a known case of HIV, presented with acute onset right upper limb and left lower limb weakness which gradually progressed to quadriparesis and bladder-bowel involvement, altered sensorium and loss of consciousness.

Discussion & Conclusion: All investigations were done to rule out all possible clinical conditions. Patient was diagnosed as having HIV myelopathy with PML: a rare presentation. Both of these conditions are progressive and fatal & regular treatment with ART can prevent them.

INTRODUCTION

Spinal cord disease or myelopathy is present in 20% with AIDS, often as a part of HIV Associated Neurocognitive Disorders. 90% with HIV myelopathy have some evidence of dementia. There are three main types. The first one is vacuolar myelopathy, a condition pathologically similar to SACD of cord, as with pernicious anemia, characterized by subacute onset and often presents with gait disturbances, predominantly ataxia and spasticity may progress to include bladder and bowel dysfunction. Physical findings include exaggerated deep tendon reflexes and extensor plantars. Upper-extremity function is usually normal. MRI of the spine is usually normal. CSF examination may show nonspecific abnormalities, such as protein elevation. Abnormal sensory evoked potentials may precede clinical symptoms of myelopathy. Aggressive antiretroviral therapy can lead to improvement of symptoms

The pathogenesis of vacuolar myelopathy is unknown, but may be related to abnormal transmethylation mechanisms induced by the virus and/or cytokines.

Second form involves dorsal columns presenting as pure sensory ataxia. Third form is also sensory presenting with paraesthesias and dysesthesias of lower extremities. AIDS myelopathy most often occurs in late stages of AIDS; most patients die within six months of developing symptoms of myelopathy.

Progressive multifocal leukoencephalopathy (PML) is a severe demyelinating disease of the central nervous system that is caused by reactivation of the polyomavirus JC (JC virus). Prior to the widespread use of highly active antiretroviral therapy (HAART), PML was recognized as a major opportunistic infection associated with AIDS in adults, with a prevalence of 1 to 5 percent. PML usually manifests with subacute neurologic deficits including altered mental status, motor deficits (hemiparesis or monoparesis), limb ataxia, gait ataxia, and visual symptoms such as hemianopia and diplopia. Most patients with HIV infection and PML are profoundly immunosuppressed with CD4-positive T-cell counts <200 per mm^3. Initial symptoms may vary greatly from patient to patient, depending on the location of their lesions in the central nervous system white matter. PML typically spares the optic nerves and the spinal cord. The disease course of PML is usually progressive and fatal. The median survival of patients without HIV infection is only a few months. Survival in the era of highly active antiretroviral therapy is longer, but survivors are often left with severe neurologic sequelae.

CASE HISTORY

A 20 year old male, a known case of HIV infection detected two years back and started on ART (TLE) since then, presented with complaints of acute onset right upper limb and left lower limb weakness.

There was no history of fever, headache, nausea, vomiting, altered sensorium, convulsion, cough with expectoration, breathlessness, bowel bladder incontinence, difficulty in speech or vision, giddiness, involuntary movements, tingling, numbness or pain in limbs. There was no history of trauma, head injury, major surgery and any addiction such as alcohol. He was on ART since 2014 (ZLN), but he was noncompliant for ART (defaulter).

On clinical examination, patient showed increased tone in affected limbs, exaggerated deep tendon reflexes, bilateral extensor plantars, reduced power more in right sided upper limb and left lower limb, decreased fine touch sensation over right arm, bilateral sustained ankle clonus, Romberg's positive and no nystagmus with normal pupil and light reflex.

Hematological investigations were all normal including complete blood counts, liver function tests, renal function tests, ESR, serum vitamin B12 happened to be adequate.

CSF analysis and MRI brain with contrast were done. CSF analysis came out to be inconclusive with mild leucocytosis (lymphocytosis) (5 cells/microlitre), mildly raised proteins and normal glucose and ADA levels. MRI brain was normal. Other investigations like HCV, HBsAg, Toxoplasma IgM status, CMV, HSV, RPR, VDRL tests were all negative. Serum homocysteine and CPK total were normal. CD4 count was 154.

Meanwhile patient showed gradual deterioration as far as neurological status was concerned, showing involvement of both upper and lower limbs now, with increased tone and lead pipe rigidity, bowel bladder incontinence developed too. EMG-NCV and MRI cervical spine with contrast were done which happened to be a normal.

Patient also showed occasional slow responses as delayed speech and motor response, inappropriate smiling, difficulty in sustained comprehension, gradually leading to speech loss, spastic quadriparesis, altered sensorium followed by loss of consciousness.

He was initially managed with higher antibiotics ART was restarted. Treatment thereafter remained mainly supportive with antibiotics, supportive drugs and physiotherapy.

A repeat MRI brain was done after 10 days which showed changes of PML in bilateral Centrum Semiovale. Patient gradually deteriorated to vegetative state requiring mechanical ventilation and died of acute cardiorespiratory arrest.

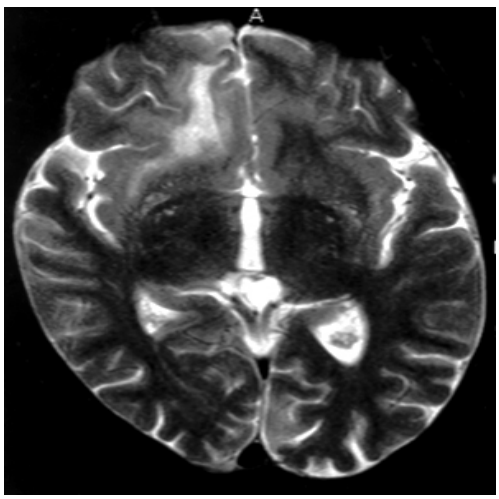

Figure-1: A sharply demarcated hyperintense lesion without mass effect is seen in this T2-weighted image in the subcortical right frontal white matter suggestive of PML

DISCUSSION & CONCLUSION

Patient presented with primarily neurological symptoms with no history of fever or convulsions or any other features of meningitis negating any possibility of infective etiology supported by normal blood counts, renal and liver function tests, ESR, negative viral markers (CMV, HCV, HBsAg, HSV), negative IgM toxoplasma, RPR, VDRL. Also CSF analysis and MRI brain were normal. Normal serum vitamin B12 levels ruled out possibility of SACD. Normal CPK total levels ruled out HIV associated myopathy. Normal homocysteine level ruled out possibility of CV stroke.

Normal EMG-NCV ruled out HIV associated peripheral neuropathy. Cervical Myelopathy was ruled out by normal MRI cervical spine. The patient's clinical profile and the result of various sequential investigations led to the diagnosis of HIV myelopathy, a part of HIV associated neurocognitive disorder (HAND) and PML.

HIV myelopathy is a diagnosis of exclusion in patients with HIV. Regular treatment with ART can prevent PML & HIV Myelopathy. So, awareness regarding ART is necessary among HIV patients. Patients, who have been started on ART, should remain compliant with their treatment.

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