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Case Report
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An Interesting Case of Infratentorial PML Mimicking Lateral Medullary Infarct

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Abstract

Introduction: Progressive Multifocal Leukoencephalopathy (PML), caused by John Cunningham (JC) virus, affects up to 2% to 7% of HIV positive patients. It is postulated to be a reactivation of JC virus in a severely immunocompromised state, to manifest as a multifocal inflammatory white matter disease of the brain, commonest in supratentorial region. We present a case of subacute ataxia and bulbar weakness in an undiagnosed HIV patient, mimicking a lateral medullary infarct.

Case Details: A 57 year old patient, not previously undiagnosed to have HIV infection, presented with slurring of speech and imbalance, 3 weeks after COVID-19 pneumonitis. MRI brain showed diffusion restriction with corresponding FLAIR hyperintensities in right lateral medulla. However, patient had progressive worsening of dysarthria and sensorium, warranting us to dig deeper and unmask HIV disease and a low CD4 cell count, thus helping us clinching the diagnosis of infratentorial presentation of PML.

Conclusion: Isolated medullary involvement in PML is an even rarer presentation of this rare disease seen in immunocompromised individuals.

Introduction

PML is a lytic infection of oligodendrocytes and astrocytes seen in an immunocompromised host, caused by reactivation of JC virus [1]. The neurological deficits correspond to areas of demyelination in the brain. The most frequently affected regions are the cerebral hemispheres at the periventricular and/or subcortical white matter [2]. Infratentorial presentation is rare and when occurs, is seen typically in the middle cerebellar peduncles and pons. Isolated medullary involvement is even rarer (5% to 7%) [2]. We present a rare case of infratentorial PML mimicking a lateral medullary syndrome in an undiagnosed HIV patient.

Case Presentation

57 year old male, no known comorbidities, presented with dysarthria, dysphagia and imbalance for 5 days. Patient had a past history of COVID pneumonia 3 weeks ago and was treated with oxygen with mask and remdesivir.

Patient had scanning speech and dysphagia, more to liquids. Examination revealed right gaze evoked nystagmus with right limb ataxia, with impaired tandem gait and swaying towards right side. MRI brain revealed multifocal areas of diffusion restriction and FLAIR hyperintensity involving the right half of the lateral medulla, left frontal and parietal lobes and right parietal lobe.MRI angiogram was normal (Figure 1).

2D echo did not reveal any abnormality, and patient was discharged on dual anti-platelets and statins with a Ryle's tube.

After 3 weeks, patient presented with worsening of dysphagia and imbalance with visible tachypnea, pooling of oropharyngeal secretions and hypoxia. Patient needed elective intubation for airway protection. Repeat MRI showed expansion of right medullary lesion with FLAIR hyperintensity with peripheral diffusion restriction and a new frontal hyperintensity, involving subcortical U fibers with a sharply demarcated peripheral border and an ill-defined inner border, without contrast enhancement (Figure 2).

HIV by ELISA was detected to be positive with CD4 count 155 cells/ml. Serum toxoplasma IgM was negative. CSF showed protein 94 mg/dl, sugar 53 mg/dl with parallel blood sugar 98 mg/dl with 2 cells, no atypical cells were noted.

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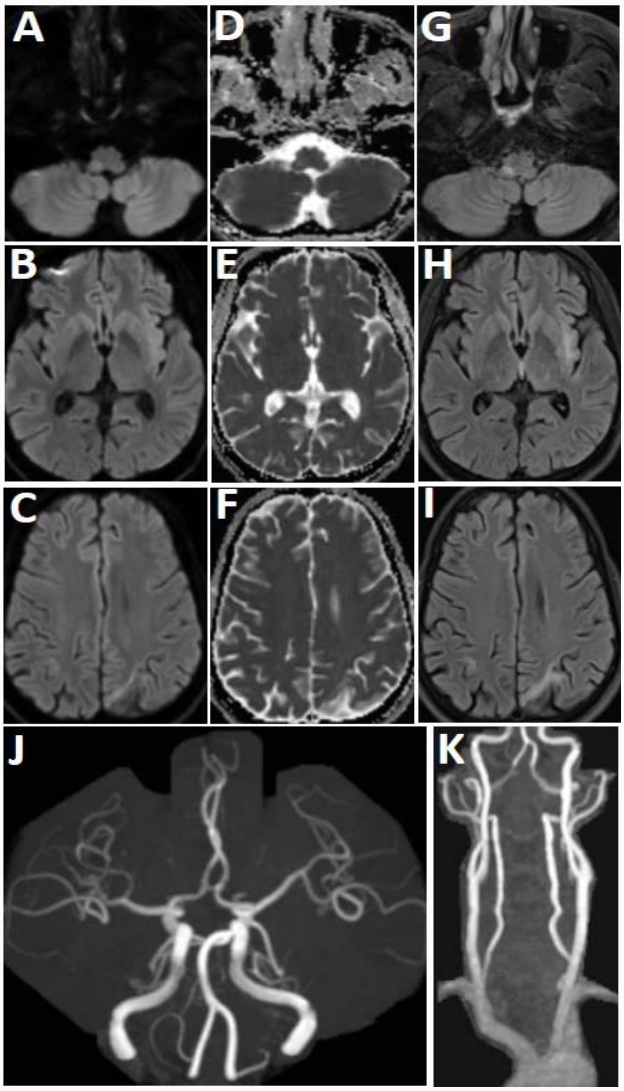

Figure 1: MRI brain axial images (A, B, C- DW sequences; D, E, F- ADC sequences and G, H, I- T2 FLAIR sequences) showing multifocal areas of diffusion restriction and FLAIR hyperintensity involving the right half of the lateral medulla, left frontal and parietal lobes and right parietal lobe. MR Angiogram (J- intracranial vessels and K- neck arteries) was normal.

The characteristic MRI picture and HIV positive status with low CD4 cell count pointed towards possibility of PML. Hence CSF John Cunningham Virus PCR was ordered and it turned out positive. Unfortunately, patient succumbed due to progressive brainstem involvement.

Discussion

PML is a white matter disease of immunocompromised host with predominant fronto-parietal involvement. It has been observed in a variety of immunocompromised states like HIV, SLE; post-chemotherapy, chronic kidney disease etc [1].

In a case report by Finelli et al. [3], misdiagnosis of PML as a stroke mimic was discussed, with case details of two patients who turned out to be HIV positive at a later date. One of these patients had a transient ischemic attack with retrospective evaluation of MRI showing subcortical white matter changes. The other patient had a left thalamic and left parietal lesion that was treated as a chronic infarct, with a diagnosis of a cryptogenic stroke. Both patients were reevaluated at a later date in view of progressive worsening of clinical deficit and had a poor outcome.

Infant et al. [4] have reported a single case of infratentorial PML mimicking an acute ischemic stroke in a patient of chronic lymphocytic leukemia.

Initial presentation of PML as predominant medullary involvement mimicking lateral medullary syndrome was also observed in another case report by Durafort et al. [5] in a patient undergoing hemodialysis. This report highlighted the possibility of PML in an unconventional immunocompromised state such as chronic kidney disease.

Similar to this scenario, our patient did not have a known immunocompromised condition at presentation, delaying the diagnosis of HIV and initiation of ART. The clinical presentation of bulbar weakness and ataxia with gaze evoked nystagmus coupled with a history of COVID pneumonitis in recent past prompted the consideration of a lateral medullary infarct. This clinical suspicion was further strengthened by the diffusion restriction in right lateral medullary lesion. As with other cases described in the literature, progressive worsening of bulbar weakness and sensorium prompted a repeat imaging and search for other etiologies.

In 2012, Liu et al. [6] have published an article from a neuroradiologists' perspective of stroke mimics. They mention Creutzfeldt-Jakob disease CJD, PML and Herpes encephalitis as infections mimicking stroke clinically and radiologically. They noticed that an ADC bright signal in the diffusion restricted lesions should alert the clinician against the diagnosis of infarct.

MRI has a superior sensitivity in the detection and characterization of PML lesions though the diagnosis may be suspected by CT scan [4]. However the diffusion restriction observed in PML lesions can mimic an infarct. On T1W sequence, the lesions are hypointense, due to apoptosis of virus infected oligodendorcytes. There may be an incomplete hyperintense rim on pre-contrast T1W images, representing lipid laden macrophages at the advancing edge of the lesion. T2 W images the lesions appear hyperintense due to demyelination, and typically involve the periventricular and subcortical white matter, having scalloped lateral margin when they involve the subcortical white matter predominantly in the parieto-occipital lobes. Lesions are more conspicuously visualized in the T2 FLAIR sequences, appearing hyperintense against a background of suppressed CSF signal intensity. Mass effect is infrequently described and usually minimal and correlation with shorter survival when seen on initial studies. The pattern may be unilateral, but more often bilateral and asymmetrical with involvement of posterior fossa. PML may also involve the basal ganglia and deep white nuclei as there are white matter fibers in these regions. The lesions typically do not enhance as the blood brain barrier is intact. There have been some reports of peripheral mild or diffuse enhancement suggestive of good immune response and hence improve prognosis. There have been reports with lesions having cystic component suggesting tumors or abscess [4].

The CSF biochemistry and cytology is normal. Our patient showed raised proteins with normal sugar and no cellularity. In CSF, detection of JC virus by PCR is diagnostic. The sensitivity is 70% and specificity is 90% to 100%. If the PCR for JC virus is negative in a scenario with high degree of suspicion, a repeat CSF examination or brain biopsy is advocated which confirms the diagnosis [7].

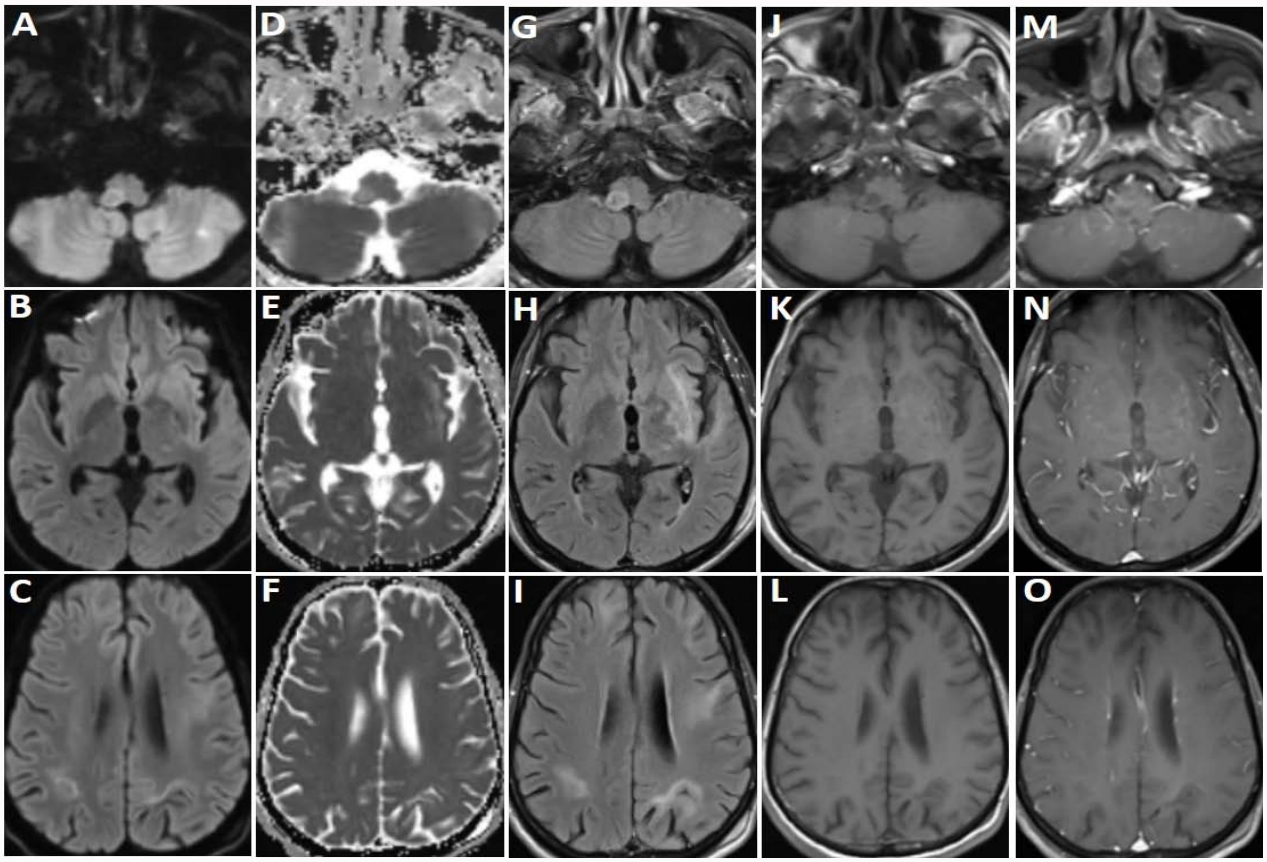

Figure 2: MRI brain during second admission. Axial images (A, B, C- DWI; D, E, F- ADC; G, H, I- T2 FLAIR; J, K, L- T1W pre-contrast and M, N, O- T1W post-contrast sequences) showed expansion of right medullary lesion with peripheral diffusion restriction and FLAIR hyperintensity with new frontal white matter hyperintensities, involving subcortical U fibers with a sharply demarcated peripheral border and an ill-defined inner border. None of the lesions showed any contrast enhancement.

Infratentorial involvement in PML is rare, till now 5 to 6 cases have been documented with brainstem/cerebellar involvement as a presenting feature of PML [8]. Typically, middle cerebellar peduncle, pons and cerebellum is involved clinically and radiologically. Pontine lesions may extend to the midbrain and medulla. Isolated medullary lesions are uncommon (5% to 7%). There is a single case report with bilateral medullary lesion by Matthew et al. [9]. Our patient had a unilateral lateral medullary involvement, which gradually progressed to involve medial structures engulfing the cardiac and respiratory centers.

The delay in diagnosis of PML due to its presentation like a stroke mimic has been discussed by Finelli et al. [3], advising screening of all suspected stroke patients to be screened for HIV.

Conclusion

PML is an important differential for a stroke mimic, both clinically and radiologically. Characteristic MRI features help distinguish between these two clinical entities with separate treatment approaches. Infratentorial involvement in PML is a rare entity with rapidly progressive fatal course.

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