Lewy Body Dementia Associated with Anti-IgLON 5 Encephalitis Detected on ^{18}F Fluorodeoxyglucose Positron Emission Tomography/Computed Tomography and 99mTc-TRODAT Single-Photon Emission Computed Tomography/Computed Tomography

Abstract

We present a case of Anti-IgLON 5 encephalitis with Lewy body dementia. ^{18}F fluorodeoxyglucose positron emission tomography/computed tomography (18F-FDG PET/CT) scan and 99 mTc-TRODAT-1 SPECT/CT scan were done. 99 mTc-TRODAT-1 scan findings revealed severely reduced concentration of dopamine transporter in bilateral basal ganglia, suggestive of a degenerative parkinsonian disorder. 18F-FDG PET scan findings were suggestive of moderate-to-severe hypometabolism in the bilateral parieto-temporal and bilateral occipital cortices including the primary visual cortices, supporting Lewy body spectrum disease with associated hypermetabolism in the bilateral sensorimotor cortices, bilateral basal ganglia, thalami, brain stem, and bilateral cerebellar hemispheres suggestive of inflammatory pathology.

Keywords: *99 mTc-TRODAT-1, fluorodeoxyglucose positron emission tomography, magnetic resonance imaging*

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we present a case of a 76 year old gentleman male who presented with complains of progressive impairment of memory for 1 year, visual hallucinations with delusions of persecution for 9 months, progressive instability of gait with recurrent falls and urinary incontinence for 6 months, and difficulty in speaking and swallowing of 3 months' duration. He developed loud stridor while breathing for 1 month with disturbed sleep and constant leg movements while lying down along with recurrent episodes of breath-holding with a few episodes of witnessed apneas. He was brought to the hospital with complains of progressively worsening altered sensorium of a few days' duration. On examination, he was found to be drowsy, lethargic, and confused with GCS E2M6V4-12/15, irregular respiration with apneic spells. There was the restriction of vertical eye movements, slowing of saccades and breaking of pursuits with spastic dysarthric speech, drooling of saliva, unstable gait with, increased tone in all four limbs, and bilateral extensor plantar responses; however, there were no cerebellar signs.

Suspecting neurodegenerative and inflammatory pathology, magnetic resonance imaging (MRI) scan was done initially which was unremarkable [Figure 1]. 99 mTc-TRODAT-1 scan was done to rule out Parkinson and Parkinson plus syndromes revealed severely reduced concentration of dopamine transporter suggestive of Parkinsonian syndrome [Figure 2]. ^{18}F fluorodeoxyglucose positron emission tomography/computed tomography (18F-FDG PET/CT) scan was done for further characterization. 18F-FDG PET [Figure 3] revealed moderate-to-severe hypometabolism in the bilateral parieto-temporal and bilateral occipital cortices including the primary visual cortices, suggestive of Lewy body spectrum disease; however, there was associated hypermetabolism in the bilateral sensorimotor cortices, bilateral basal ganglia, thalami, brain stem, and bilateral cerebellar hemispheres which were likely due inflammatory pathology. Statistical parametric mapping (SPM) was done to confirm 18F-FDG PET findings. SPM images were co-registered to a normal axial MRI [Figure 4] showing the areas

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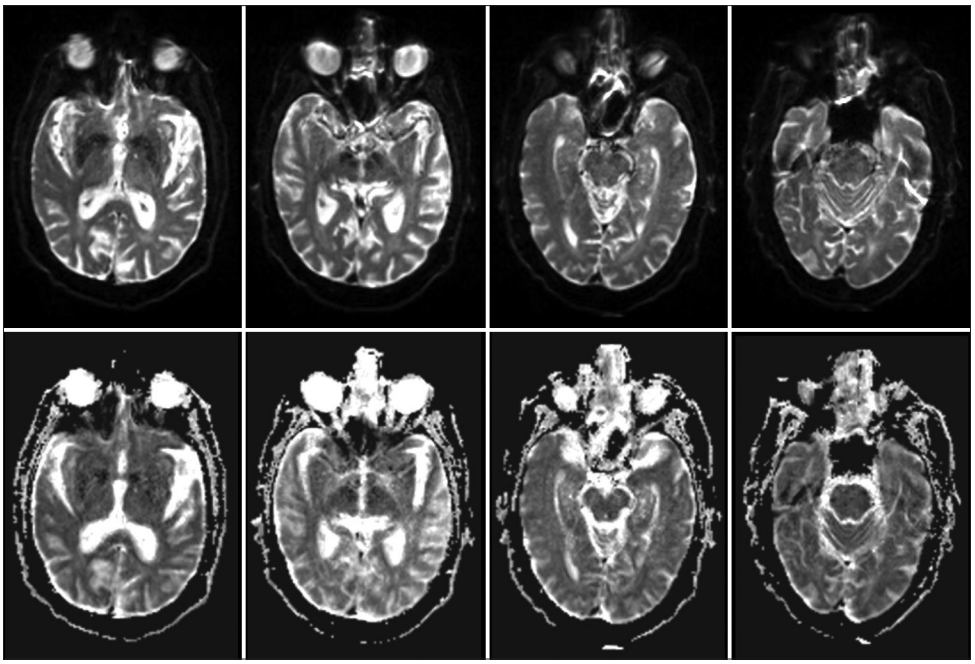

Figure 1: Axial MRI showing unremarkable DWI and ADC images

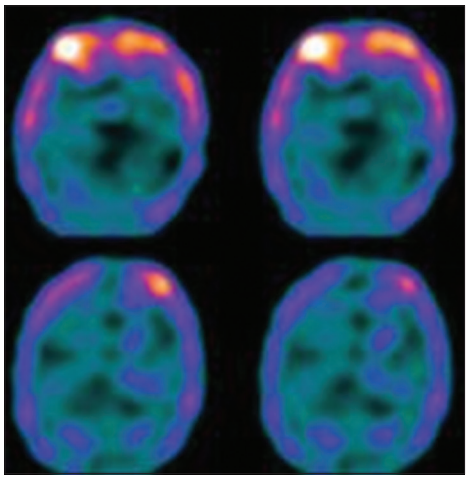

Figure 2: TRODAT scan showing severely reduced TRODAT uptake in bilateral basal ganglia

of hypometabolism (Blue) and hypermetabolism (Red). Antibodies to a neuronal cell adhesion protein IgLON5 were found positive in the cerebrospinal fluid (CSF) and serum. Thus, the diagnosis of IgLON5 encephalitis with Lewy body dementia was established. The patient condition improved after immunoglobulin treatment and the clinical symptoms of the oral discomfort and involuntary movements of mouth were gradually alleviated, and her gait became normal. Anti-IgLON5 disease is a prominent sleep disorder with progressive bulbar dysfunction, gait instability, and abnormal eye movements with cognitive deterioration.[1-3] CSF and serum harbors antibodies against IgLON5 with neuronal-specific tau accumulation.[4,5] The brain metabolic pattern of this patient was similar to the

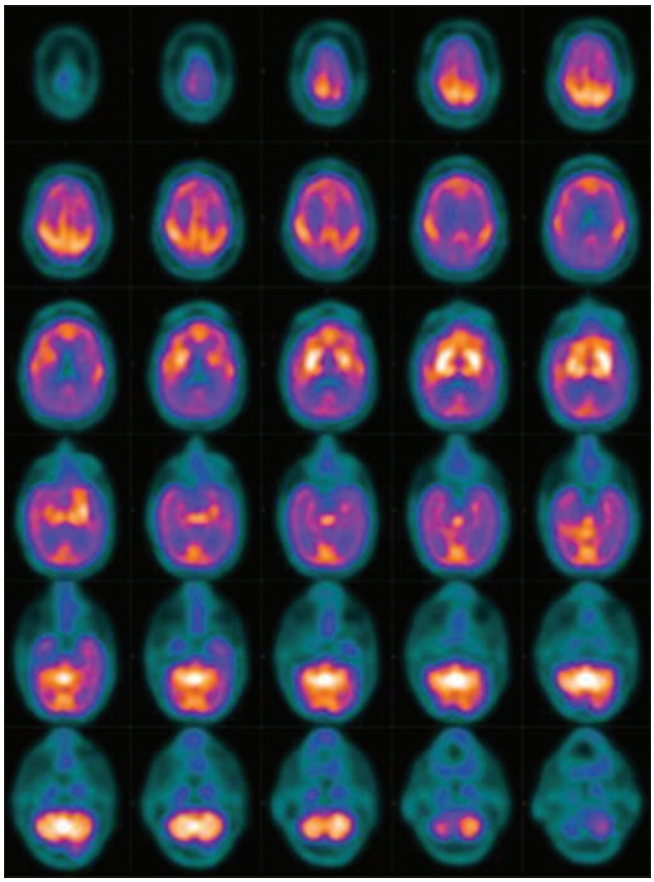

Figure 3: Axial 18F-FDG PET images showing moderate-to-severe hypometabolism in the bilateral parieto-temporal and bilateral occipital cortices including the primary visual cortices with associated hypermetabolism in the bilateral sensorimotor cortices, bilateral basal ganglia, thalami, brain stem, and bilateral cerebellar hemispheres

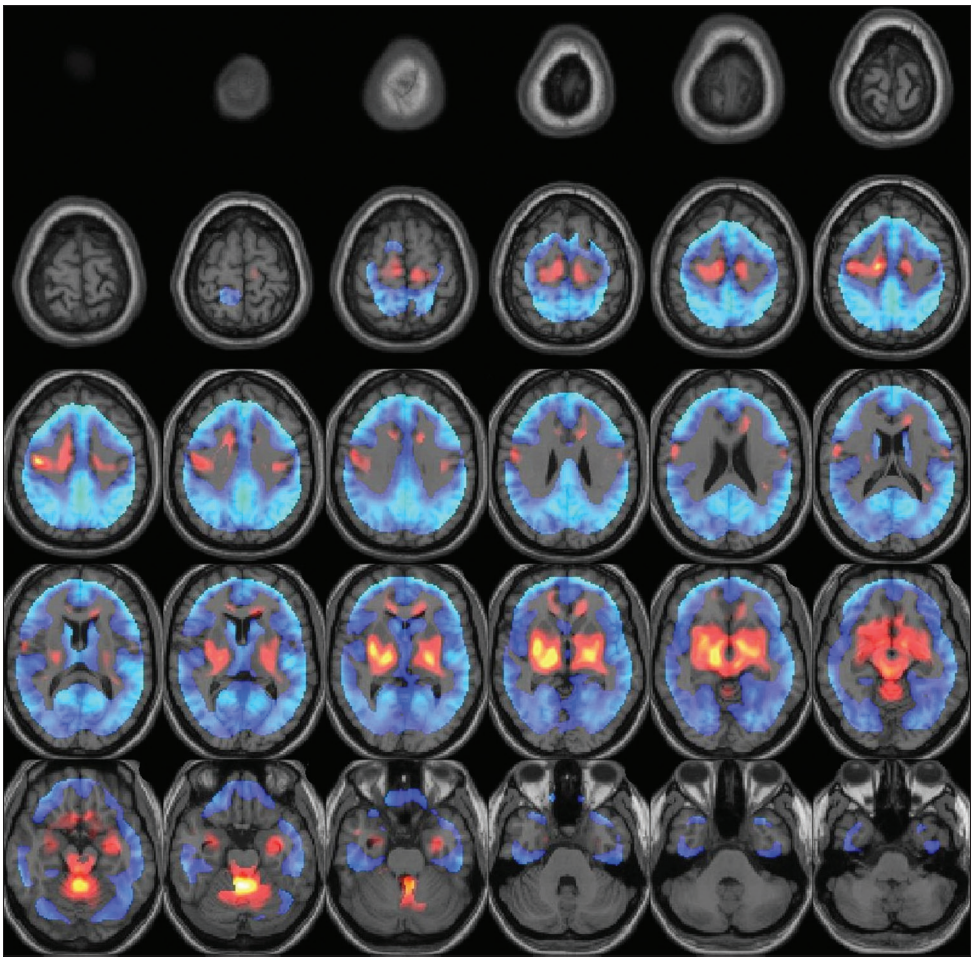

Figure 4: SPM images co-registered to a normal axial MRI showing the areas of hypometabolism (Blue) and hypermetabolism (Red)

case reported by Zhang *et al*.;[6] however, in our case, overlap was seen between degenerative parkinsonian disorder and anti-IgLON5 antibody-mediated encephalitis. Very few overlap syndromes with IgLON 5 encephalitis have been reported in the literature.[7,8] The 18F-FDG PET/CT with 99 mTc-TRODAT-1 scan may have a potential role in establishing the diagnosis of overlapping syndromes of degenerative Parkinsonian disorders with encephalitis and help to monitor response to treatment.[9,10]

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Conflicts of interest

There are no conflicts of interest.

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