

Diagnostic Challenges in Seronegative Autoimmune Limbic Encephalitis Presenting as Faciobrachial Dystonic Seizures: A Case Report

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ABSTRACT

Faciobrachial Dystonic Seizures (FBDS) are sudden brief seizure-like episodes, which are highly characteristic for Autoimmune Limbic Encephalitis (AIE), mostly associated with Voltage-gated Potassium Channel (VGKC), classically Leucine-Rich Glioma Inactivated-1 (LGI1). Timely diagnosis in seronegative patients poses a big challenge, often leading to delay in intervention, causing irreversible neuronal damage. A 66-year-old male with a history of hypertension for seven years on regular medications presented to neurology Outpatient Department (OPD) with one week history of recurrent brief involuntary dystonic movements involving left face and upper limb, along with neuropsychiatric manifestations. Initial assessment revealed mild hyponatraemia. Cerebrospinal Fluid (CSF) analysis, Magnetic Resonance Imaging (MRI) and Electroencephalogram (EEG) were unremarkable. Fluorodeoxyglucose-Positron Emission Tomography Computed Tomography (FDG PET-CT) revealed hypermetabolism in right mesial temporal cortex, bilateral caudate nuclei and putamen. Despite the fact that antibodies were absent in CSF, patient was still started on pulse intravenous corticosteroid therapy and Anti-Seizure Medication (ASM) along with immunotherapy resulting in significant improvement of the patient. The present case highlights the limitations in presently used diagnostic criteria, which rely heavily on MRI and CSF analysis. Advanced imaging techniques like FDG PET-CT are more sensitive, and hence support the diagnosis, when diagnostic criteria are not fulfilled. Timely initiation of immunotherapy based on strong clinical suspicion and advanced imaging is very important to prevent irreversible cognitive impairment.

Keywords: Cognition disorders, Hyponatraemia, Immunotherapy, Positron emission tomography

CASE REPORT

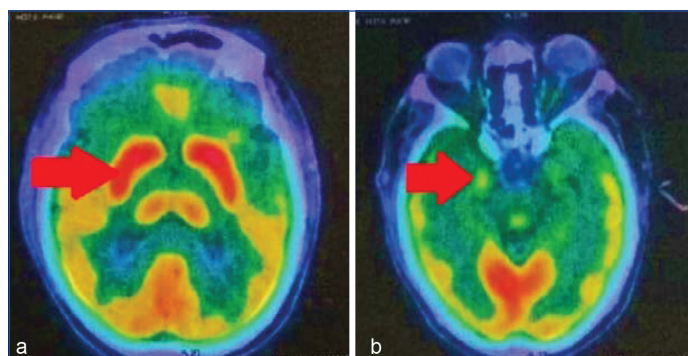
A 66-year-old male presented to neurology OPD with recurrent episodes of abnormal movements for one week. The episodes were characterised by brief involuntary dystonic movements involving the left side of the face and upper limb, without any impairment of awareness, and were associated with progressive behavioural changes including drowsiness, irritability, aggression, reduced social interaction, disturbed sleep, and cognitive decline {Montreal Cognitive Assessment (MoCA) Score - 21/30}. Patient was a known case of hypertension for seven years and was receiving Tablet Amlodipine 5 mg OD on regular medication.

According to family members, he had also been experiencing intermittent behavioural disturbances with delusional ideas for nearly one year prior to presentation. There was no history suggestive of fever, headache, focal limb weakness, loss of consciousness, recent infection, or other systemic illness. At the time of admission, the patient was drowsy but arousable and disoriented to time and place. Neurological examination did not reveal any focal motor deficits, and bilateral plantar responses were flexor.

Laboratory evaluation demonstrated mild hyponatraemia with a serum sodium level of 133 mEq/L. CSF examination was essentially unremarkable, showing a total cell count of 4 cells/mm³, glucose of 62 mg/dL, and protein of 24 mg/dL. Comprehensive autoimmune encephalitis antibody testing performed on both CSF and serum was negative for antibodies against N-methyl-D-aspartate (NMDA) receptor, Alpha-amino-3-hydroxy-5-methyl-4-isoxazolepropionic Acid (AMPA) receptor (GluR1 and GluR2), Gamma-aminobutyric Acid type B (GABA-B) receptor, Leucine-rich Glioma-inactivated 1 (LGI1), and Contactin-associated protein-like 2 (CASPR2). Magnetic resonance imaging of the brain showed no acute infarct,

haemorrhage, or space occupying lesion. However, there were some chronic age-related changes in the form of Grade II Fazekas small vessel ischaemic disease and senile cerebral atrophy. Electroencephalography (EEG) did not demonstrate epileptiform discharges or other significant abnormalities.

Due to clinical semiology, with the strong clinical suspicion for autoimmune encephalitis, FDG PET-CT of the brain was performed, which revealed focal hypermetabolism involving the right mesial temporal cortex along with diffusely increased FDG uptake in the bilateral caudate nuclei and putamen [Table/Fig-1a], and right mesial temporal cortex [Table/Fig-1b]. These findings were highly suggestive of AIE. This hypermetabolism involving the right mesial temporal cortex corresponds to the limbic system involvement, while increased uptake in the bilateral basal ganglia correlated with the patient's FBDSs.



[Table/Fig-1]: 18F-FDG PET-CT of brain demonstrating: (a) Axial section- Diffuse hypermetabolism involving bilateral caudate nuclei and putamen (red arrow); and (b) Coronal section - focal hypermetabolism in the right mesial temporal cortex (red arrow), findings supportive of Autoimmune Limbic Encephalitis (AIE).

Based on the characteristic clinical presentation of FBDSs, differential diagnoses considered were focal aware motor seizures, paroxysmal movement disorders, metabolic encephalopathy and AIE. The characteristic FBDS associated with neuropsychiatric manifestations, mild hyponatraemia, and supportive PET-CT findings despite negative antibody testing, a diagnosis of AIE was made. The patient was treated with pulse intravenous corticosteroid therapy of Inj. methylprednisolone 1 g once daily for five days, along with ASM Inj. levetiracetam 1 g i.v. BD. Considering the risk of disease relapse, immunotherapy with rituximab was initiated, following which there was significant improvement in FBDS frequency, behaviour, and sensorium. The patient was subsequently maintained on rituximab (1 g i.v. every 15 days) therapy and remained under regular neurological follow-up. At three month follow-up patient remained seizure free, with significant improvement in behavior, cognition and sensorium. Patient was able to perform activities of daily living independently.

DISCUSSION

The FBDSs are characterised by very brief, frequent, unilateral dystonic contractions involving face and upper limb with preserved awareness. Andrade DM et al., described these episodes mainly as tonic, with occasionally clonic, myoclonic jerks or dystonic posture [1]. These episodes can recur up to hundreds of episodes per day [2]. Unlike focal motor seizures, they usually lack prolonged postictal confusion and frequently demonstrate normal EEG findings. It is plausible to say that there are two disorders: those who have ictal EEG changes and impaired awareness, have seizures, while others have paroxysmal movement disorder [3]. Their close association with AIE, particularly LGI1 antibody-associated disease, makes FBDS highly specific clinical marker that often precedes cognitive impairment [4]. Encephalitis is a clinical syndrome, in which inflammation of brain parenchyma causes neurological dysfunction [5]. If the main characteristic finding in case of parenchymal inflammation is auto-antibodies, which may or may not be associated with the pathophysiology of the condition, it is called autoimmune encephalopathy. If it mainly involves the limbic system, it is AIE. VGKC-antibody-associated limbic encephalitis responds well to immunotherapy and is rarely caused by a tumour [6].

The presentation of FBDS along with neuropsychiatric manifestations, hyponatraemia, PET-CT showing increased FDG uptake in right mesial temporal cortex, and bilateral caudate nuclei and putamen is classically pointing towards the LGI1 (anti-leucine-rich glioma-inactivated) AIE. However, the present case shows a paradoxical situation, where semiology as well as radiology is pointing towards LGI1 AIE, but LGI1 antibodies are not found. This is a rare and diagnostically challenging situation as the widely accepted diagnostic criteria of AIE, as given by Graus F et al., in his different panels are not satisfied [7].

Clinically it fulfils the criteria of epilepsy, but no post ictal confusion, and EEG not showing any epileptiform discharges, are deviating away from the diagnosis of epilepsy.

Here the most striking feature is absence of anti-VGKC antibodies. While the presence of autoantibodies confirms the diagnosis of autoimmune encephalitis, mere absence of these antibodies does not strike it out of our differentials [8]. It is possible that there are many variants of VGKC auto-antibodies which are yet to be

identified, and thus it could not be found during analysis. Delaying the immunotherapy, and waiting for detecting some antibody is not justified as it can cause irreversible cognitive impairment [9].

The 2016 Graus F and Dalmau J criteria rely heavily upon the MRI and CSF findings as its one of the major diagnostic pillars for seronegative cases [7]. In the abovementioned case, FBDS are so specific to autoimmune encephalitis that decision can be made without any imaging or serological testing. It is noteworthy that MRI and CSF were unremarkable and were not pointing towards the diagnosis of autoimmune encephalitis, but PET-CT cleared the picture. PET-CT is often more sensitive than MRI in early or challenging cases [10].

CONCLUSION(S)

The FBDS should immediately raise the suspicion for AIE despite negative serum and CSF autoimmune antibody panels and the absence of definitive MRI or CSF abnormalities. The characteristic clinical presentation, associated neuropsychiatric manifestations, and supportive FDG PET-CT findings strongly suggested an autoimmune aetiology. The patient's favourable response to immunotherapy further reinforced the diagnosis. Earlier diagnosis and initiation of immunotherapy may be possible, thus preventing permanent neuronal and cognitive impairment.

Supplementary materials

Video S1: A clinical video captured during an episode of FBDSs demonstrating brief involuntary dystonic movements involving the left side of the face and upper limb, without any impairment of awareness.

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