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Case Report
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Anti-NMDAR Encephalitis Presenting with Unilateral Insular Diffusion Restriction on Brain Imaging: Mimicking as a Stroke

*Meghna Pawar, Naveen Kumar, Angshuman Mukherjee, Sulakshana and Surekha D**

Department of Neurology, Pandit Bhagwat Dayal Sharma Post Graduate Institute of Health Sciences, India

Introduction

Anti-N-Methyl-D-Aspartate Receptor Encephalitis (NMDARE) is a rare but most common subtype among autoimmune encephalitis caused by formation of IgG against NMDA receptor (NR1) subunit with internalization of NMDAR receptors and decrease in the receptor- dependent synaptic currents [1]. Herpes Simplex virus encephalitis, or tumor like ovarian teratoma are the most common triggering factors, however in most cases, the exact etiology remains unknown. NMDARE is characterized by varied psychiatric and behavioral symptoms along with combinations of neurological manifestations such as seizures, Movement Disorders (MD), speech disorders, altered consciousness, insomnia, and autonomic dysfunctions. This case highlights the clinical presentations of a patient initially presented like an ischemic stroke but later developed Epilepsia partialis continua and found to be positive for Anti-N-Methyl-D-Aspartate Receptor Encephalitis (NMDAR-E) without any overt etiology.

Case Presentation

A 19-year-old recently married girl without any significant past presented with history of decreased interaction and alteration behavior since last 3 to 4 days.

On neurological examination she seems confused with global aphasia, Cranial Nerve (CN) examination showed Rt sided grade 3 UMN facial palsy and rest of the CN examination was normal. Tone was slightly increased with mild right sided hemiparesis; all the deep tendon reflexes were normal with plantar response was extensor on right side.

All the routine biochemical tests were normal. Her chest X-ray and Echocardiography was normal. Brain Magnetic Resonance Imaging (MRI) showed Diffusion restriction on DWI images and low signal intensity on the ADC in Left insular cortex suggestive of acute infarct (Figure 1). Possibility of young stroke was kept and she was planned for further evaluation. All the viral markers were negative, Vasculitis and APLA profile was also negative.

Two days later she started having frequent clustering of Rt focal motor seizures with impaired awareness then she underwent repeat MRI Brain which showed T2 FLAIR hyperintensity in Medial Temporal Lobe area along with increase in previous DWI restriction in insular area (Figure 2).

She was started on IV Levetiracetam and oral clobazam but her seizure was not controlled. Eventually she develops Epilepsia partialis continua and was put on ventilatory support and started on midazolam infusion but with little improvement in seizure frequency. Her Electroencephalography (EEG) revealed diffuse slowing with absence of sleep markers. Serum sample for autoimmune panel was sent and was strongly positive for NMDAR antibodies, she was started on combination of IV Methylprednisolone and IVIG. Her seizures stopped completely and she started showing improvement in comprehension after 5 days of treatment and eventually extubated. MRI pelvis was performed in search of ovarian teratoma which did not demonstrate any occult tumor. MRI brain repeated on 12th day which showed complete resolution in her previous lesions (Figure 3).

The present case highlights the possibility of Anti NMDAR encephalitis presenting as stroke which is rare in autoimmune encephalitis [2]. Gait disturbances, status epilepticus and insomnia are other uncommon presentation reported till now [3,4].

In a case report by Chandra et al. [5] out of 29 patients with definite NMDAR encephalitis only single patient presented with stroke and in another case series by Gowda et al. [6] out of 24

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Correspondence:
Surekha Dabla, Department of Neurology, Pandit Bhagwat Dayal Sharma Post Graduate Institute of Health Sciences, Rohtak, Haryana, India, Tel: 09416291276

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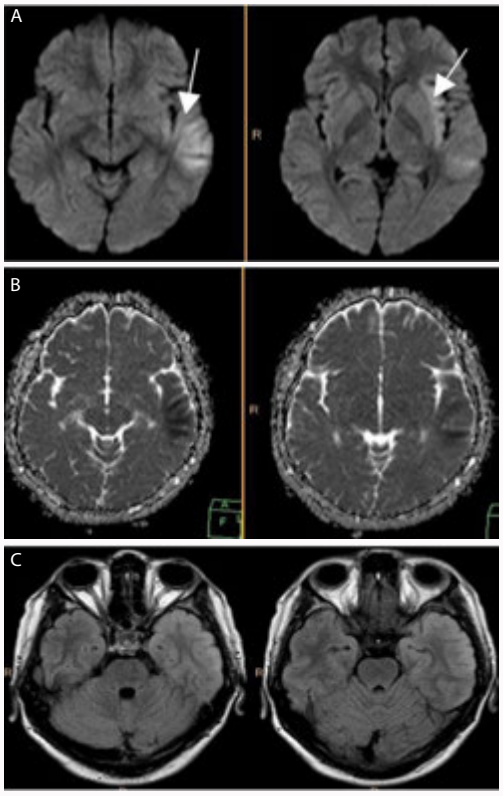

Figure 1: MRI brain showing diffusion restriction in Diffusion Weighed axial Images (DWI) at Left insular cortex (A). ADC map showing low intensity signals at Left insular cortex (B). Normal T2 FLAIR (C).

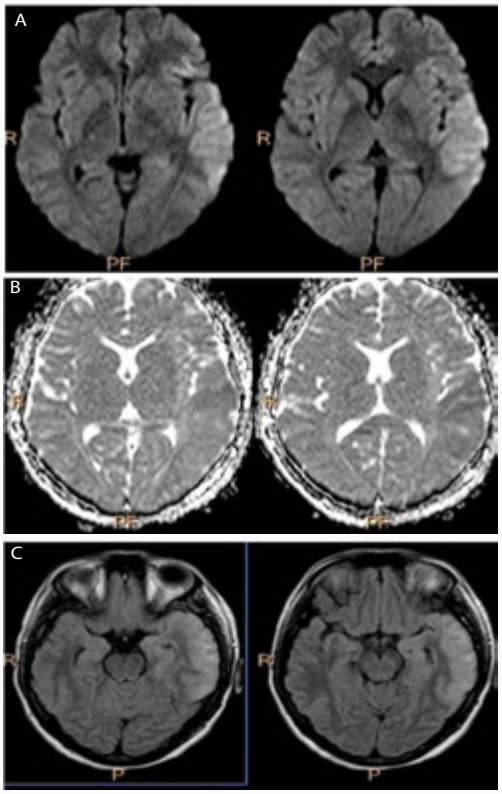

Figure 2: MRI brain showing diffusion restriction in DWI in Left insular cortex (A). ADC map showing low intensity signals at Left insular cortex (B) & T2 W FLAIR hyperintensity in Left medial temporal lobe (C).

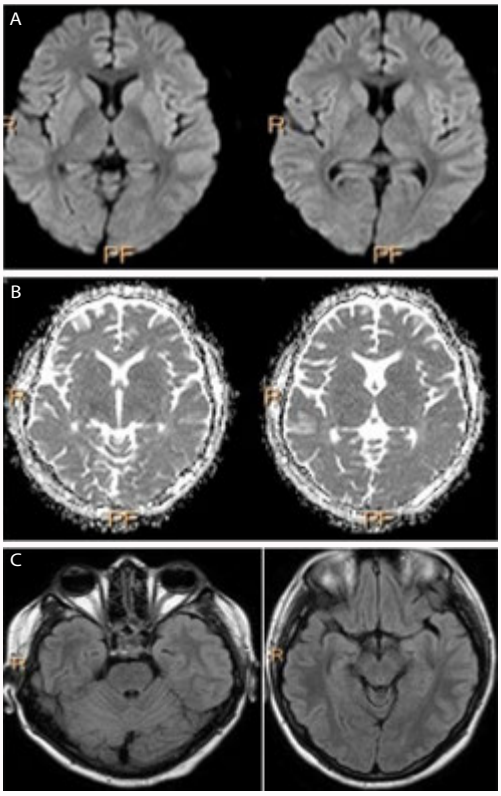

Figure 3: MRI Brain showing complete resolution of lesions (DWI, ADC & FLAIR).

seropositive anti-NMDAR encephalitis only six children presented with hemiparesis/stroke-like episode. A case report by Stavitsky et al. also reported stroke like presentation in patient with NMDARE [7].

Pathophysiology for stroke like presentation in NMDARE is not well established. One of the proposed mechanisms is synthesis of NMDAR antibodies after exposure to triggering factors (HSV infection, Teratoma). This antibodies after binding with NMDAR receptors leads to overactivity of the receptors through excessive presynaptic glutamate release causing excitotoxicity and acute neuronal injury [8].

Elevated levels of pro-inflammatory cytokines like interleukin-6 and interleukin-17A has been reported by Byun et al. [9]. This cytokine causes regional inflammation and leads to arterial thrombosis [10].

In conclusion NMDARE if not diagnosed early may result in permanent neurologic deficits or sometime even mortality. We describe here a girl who presented like a stroke clinically and radiologically but when she develops Epilepsia partialis continua then a second possibility of NMDARE was considered and managed accordingly resultant patient improved remarkably.

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