



Case Report

Autoimmune encephalitis with acute presentation masquerading as viral encephalitis

Ravi Kanth Jakkani^{1,*}, Kiran Kumar Ramineni²¹Dept. of Radiology, Yashoda Hospital, Malakpet, Hyderabad, Telangana, India²Dept. of Neurology, Yashoda Hospital, Malakpet, Hyderabad, Telangana, India

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ABSTRACT

Encephalitic syndromes typically manifest with fever, altered sensorium and seizures. Viral etiology accounts for the significant proportion of cases especially with acute febrile illness. Here we report a middle aged female with anti NMDAR encephalitis with acute presentation mimicking viral encephalitis. She had good clinical and radiological improvement with immunosuppressive therapy.

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1. Introduction

Encephalitis continues to be a significant cause of morbidity and mortality worldwide. Most of the cases of encephalitis are often thought to be mediated by infections, however inspite of battery of investigations exact cause is unknown in good number of cases.¹ Recent advances and understanding in molecular diagnostics and immunological markers led to the better diagnosis and management of immune mediated encephalitis.² Here we report a middle aged female with acute presentation of autoimmune encephalitis.

2. Case Report

A 46-yr-old female patient was brought to the emergency department of our hospital with history of high grade fever, headache, abnormal behaviour and altered sensorium of three days duration. She developed recurrent episodes of focal and generalized tonic clonic seizures, became mute with perioral dyskinetic movements. She was admitted to neuro intensive care unit with the working diagnosis of viral meningoencephalitis. Her vitals included axillary

temperature of 103F, pulse rate of 120 per minute and blood pressure of 110/70 mm of Hg. Seizures continued inspite of adding four antiepileptic drugs. Magnetic resonance imaging (MRI) of brain was done for further evaluation which revealed bilateral symmetrical areas of T2 and fluid attenuation inversion recovery (FLAIR) hyperintensities with diffusion restriction in bilateral medial temporal lobes, hippocampi and external capsules. Figure 1 No significant post contrast enhancement was seen. No evidence of hemorrhage. Complete blood picture, random blood sugar, serum creatinine, serum electrolytes, serum calcium, liver function tests and thyroid profile were within normal limits. Serum Dengue NS I antigen and Immunoglobulin (Ig) M antibody and HIV1, 2 were negative. Antinuclear antibodies (ANA) and thyroid peroxidase (TPO) antibodies were negative. Computed tomography of abdomen and pelvis and chest radiograph were unremarkable. Cerebrospinal fluid (CSF) analysis revealed protein of 40mg/dl, sugar 68mg/dl, cell count 8cells/cumm. CSF infectious encephalitis panel including HSV 1 and 2 PCR was negative. Blood and urine cultures were negative. EEG was abnormal with generalized sharp waves and slowing. In spite of acute presentation based on laboratory investigations and

* Corresponding author.

E-mail address: ravikanthjakkani@gmail.com (R. K. Jakkani).

MRI findings possibility of autoimmune encephalitis was considered. Autoimmune encephalitis panel of serum and CSF was sent. Pending the results she was started on intravenous methyl prednisolone 1gram/day for five days followed by oral steroids. By day 3 of initiation of methylprednisolone seizures subsided completely with significant improvement in sensorium and behavioural changes. By day 7 she was ambulant. CSF autoimmune encephalitis panel revealed anti NMDAR antibody to be positive. By one month she became independent for activities of daily living without further seizures. Oral methylprednisolone was tapered gradually with the addition of Azathioprine. Follow up MRI imaging done after 3 months showed near complete resolution in the diffusion restriction in involved regions Figure 2 with residual FLAIR hyperintensity with mild gliotic changes. She did not have any relapse over the next one year.

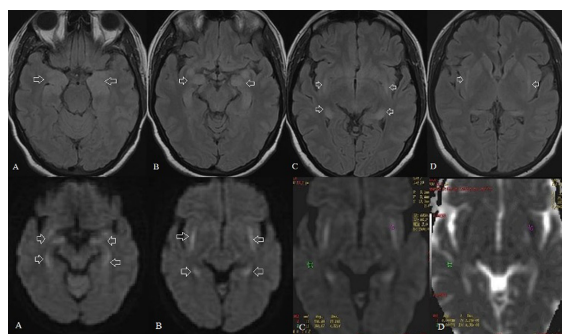


Fig. 1: Top row (A-D) axial FLAIR brain images showing bilateral symmetrical hyperintensities in medial temporal lobes, hippocampi and external capsules (open white arrows). Bottom row (A-D), axial DWI and ADC maps showing corresponding diffusion restriction in bilateral medial temporal lobes, hippocampi and external capsules (open white arrows).

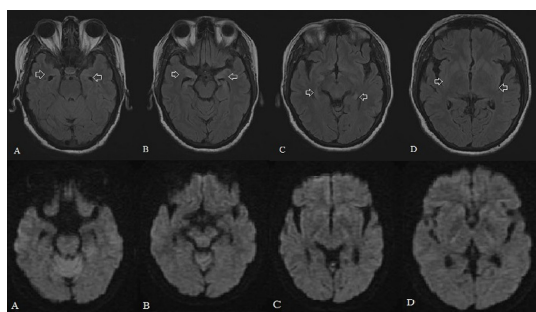


Fig. 2: Top row (A-D) follow up axial FLAIR brain images showing significant resolution of bilateral symmetrical hyperintensities in medial temporal lobes, hippocampi and external capsules with mild gliotic changes (open white arrows). Bottom row (A-D), axial DWI images showing complete resolution of previous hyperintense signal in medial temporal lobes, hippocampi and external capsules (open white arrows).

3. Discussion

Autoimmune encephalitis is a broad group of disorders mediated by neuronal autoantibodies against intracellular, intracellular synapse-related and cell surface antigens. Anti-NMDAR encephalitis is the most common one among this group.³

Anti-NMDAR encephalitis is mediated by Ig G antibodies against Glu N1 subunit of NMDA receptor. It affects all age groups but is more common in young adult females.⁴

Typical presentation of Anti NMDAR encephalitis is sub acute with prodromal symptoms like fever, headache and malaise. It is then followed by early stage of altered sensorium, confusion and behavioural changes. Dyskinetic movements are seen in late stage. Refractory seizures are another clinical presentation. Initial prodrome often mimics viral encephalitis. Anti NMDAR encephalitis is associated with ovarian teratoma in about 40-50% cases and other neoplasms like ovarian, pancreatic, breast cancer or lymphoma. Neoplastic association is absent in some cases as in our case, however neoplastic screening workup needs to be done in all cases.⁴

MRI brain can be unrevealing in Anti NMDAR encephalitis. It has a predilection to affect the limbic system, involving medial temporal lobes, hippocampi, insular region, inferior frontal lobe and gyrus rectus either bilaterally or unilaterally. T2 and FLAIR hyperintensity is seen in affected regions with no post contrast enhancement. Hemorrhages are absent. Diffusion restriction is uncommon, but has been reported.⁵ Imaging findings can closely simulate other viral encephalitis like HSV encephalitis and is often difficult to distinguish. Diffuse and lateral temporal lobe, insular involvement with sparing of basal ganglia is more common in HSV encephalitis and often with asymmetrical involvement. Presence of hemorrhages and enhancement also favour HSV encephalitis.⁶ Predominant psychosis, behavioural disturbances, dyskinetic movements and autonomic instability favour anti NMDAR encephalitis, while acute onset of febrile illness favours HSV encephalitis.^{6,7} Moderate lymphocytosis is seen in CSF with neuronal autoantibodies seen in CSF and serum. EEG shows non specific changes, however in about 30% cases a characteristic delta brush pattern, slow waves with overriding fast beta activity.⁸ Most of the patients show very good clinical response with intravenous steroids, plasma exchange and immunoglobulins by 3-4 weeks. In case of inadequate treatment response second line immunotherapy drugs like cyclophosphamide and rituximab are added.⁹

4. Conclusion

Autoimmune encephalitis workup should be included in otherwise unexplained encephalitic syndromes even with

acute presentation especially in the context of supportive brain imaging findings. Early initiation of appropriate immunosuppressive therapy helps in better outcome.

5. Conflict of Interest

None

6. Source of Funding

None

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Author biography

Ravi Kanth Jakkani Consultant Radiologist

Kiran Kumar Ramineni Consultant Neurologist

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