CASE REPORT OPEN ACCESS

DOI: 10.5281/zenodo.4268384

Behavioral problem, Acute Confusional state and Mutism as Presentation of NMDA Encephalitis

Jawad Ahmad Wasif, Syed Muhammad Hur Abbas*, Gulsam Bashir*

Department of Pediatrics, Letterkenny University Hospital Donegal-Ireland, *Khyber Medial College Peshawar-Pakistan

Review Began 25/09/2020 Review Ended 15/10/2020 Published 28/10/2020

Anti-N-methyl D-aspartate (NMDA) receptor (anti-NMDAR) encephalitis is caused by immunoreactivity against the NR1 subunit of the NMDA receptor. We present the case of an 18-year-old young girl with that presented to us with behavioral problem, acute confusional state and mutism. After a detailed history and a thorough clinical examination the autoimmune encephalitis antibodies profile were sent. This came positive for NMDA receptor and negative for CASPR-2, Type AMPA1/2, LGI-1, DPPX, and $GABA_B$ receptor antibodies. The patient was started on corticosteroids, azathioprine and procyclidine. She was sent home on immune suppressive therapy and called after 4 weeks for follow up. Upon follow up her mutism was gone but the behavioral issue was not significantly improved. Steroids induced psychosis was also observed. She was started on Rituximab with improvement in her behavior. Acute confusional state and behavioral illness preceded the diagnosis of NMDA Encephalitis. The diagnosis of autoimmune encephalitis should be considered a top differential in relapsing patients as it has a very wide range of presentation. Prompt diagnosis and early treatment are the need of the hour as the ramifications of a mis or missed diagnosis could be detrimental

Keywords: Encephalitis; NORSE; NMDA

Citation: Wasif JA, Abbas SMH, Bashir G. Behavioral problem, Acute Confusional state and Mutism as Presentation of NMDA Encephalitis. THE STETHO 2020;1(2):7-9

This is an open access article distributed under the terms of the Creative Commons Attribution License CC-BY 4.0., (http://creativecommons.org/licenses/by/4.0/), which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.

INTRODUCTION

Anti-N-methyl D-aspartate (NMDA) receptor (anti-NMDAR) encephalitis is caused by immunoreactivity against the NR1 subunit of the NMDA receptor. It is beyond doubt one of the most common encephalitis's. The term was first explained and evaluated by Dalmau and his team. They described it as a psychiatric and neurologic condition found in women that had ovarian teratomas. [1] However recent advances and studies suggest that tumour involvement wasn't necessary to be a part of this condition. It can and may present with or without the tumour. Recent literature has shown an overlap of psychiatric and neurological pathology in association of autoimmune encephalitis. [2,3] The disease progresses as Prodromal phase moving into the Psychotic phase followed by a Catatonic stage. [4]

CASE REPORT

We report a case of 18-year-old young girl from Afghanistan presented to us with complaint of behavioral problem, decreased clarity of mind with acute confusion and mutism for 1 month. She was alright back then when she suddenly developed these symptoms. History was negative for fever, autoimmune rheumatic disease and any psychiatric illness. Systemic review was normal for any significant illness. She was treated in Afghanistan by psychiatric doctor but with no response to treatment.

General physical examination showed BP: 110/70mmhg, Pulse: 76/Minute regular and a febrile. GCS: 13/15. No signs of meningeal irritation, anemia, lymphadenopathy, thyromegaly, jaundice edema feet clubbing or cyanosis. Skin and hair normal with good hydration status.

MMF score was 23/31 with no abnormality in neurological examination.

Baseline investigation showed HB: 12 g/dl, TLC: 8000/ mm^3, platelets: 170000/mm^3 and MCV: 84fl ESR: 68/1st hour. Rest of baseline including LFTs, RFTs, Serum electrolytes Urine R/E, serum calcium and uric acid was normal. Thyroids function test normal, ANA & RA factor negative.

MRI brain with contrast and MRI done with no abnormality (images given below). Lumber puncture done with normal opening pressure, normal biochemistry and cellularity. CSF PCR was negative for virus and Mycobacterium tuberculosis. Patient was started on conservative management with antibiotic and antiviral but no response to treatment.

Autoimmune encephalitis antibodies profile was sent which came out to be positive for NMDA receptor and negative for CASPR2, Type AMPA1/2, LGI1, DPPX, and $GABA_B$ receptor antibodies.

She was started on corticosteroids along with azathioprine and procyclidine. Initial response to corticosteroid was poor. So, she was given IVIGs with some improvement in her condition. she was sent home on immune suppressive therapy and called for follow up after 4 weeks.

During subsequent follow up her mutism was gone but behavioral problem was not significantly improved with some steroids induce psychosis. She was started on Rituximab with improvement in her behavior. She was fully oriented in TPP

DISCUSSION

Antibody testing to confirm the diagnosis should be requested in all suspected cases especially those presenting with psychiatric signs and symptoms. The current methods for testing antibody detection has a sensitivity and specificity of 100%. [5]

It is beyond doubt one of the most common encephalitis's. The term was first explained and evaluated by Dalmau and his team. They described it as a psychiatric and neurologic condition found in women that had ovarian teratomas. [1] However recent advances and studies suggest that tumor involvement wasn't necessary to be a part of this condition. It can and may present with or without the tumor.

We highlight and stress upon the atypical presentations, prompt diagnosis and early treatment of anti-NMDA-receptor encephalitis as the ramifications of a mis or missed diagnosis could be detrimental. the extensive investigations, detailed history and physical exam is ambiguous in giving us a definitive diagnosis early on, especially in case of overlapping neurological and psychiatric presentations having strong associations with the medical condition.

CONCLUSION

The diagnosis of autoimmune encephalitis should be considered a top differential in relapsing patients as it has a very wide range of presentation. Prompt

This is an open access article distributed under the terms of the Creative Commons Attribution License CC-BY 4.0., (http://creativecommons.org/licenses/by/4.0/), which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.

diagnosis and early treatment are the need of the hour as the ramifications of a mis or missed diagnosis could be detrimental.

REFERENCES

1. Dalmau J, Tüzün E, Wu HY, et al. Paraneoplastic anti-N-methyl-D-aspartate receptor encephalitis associated with ovarian teratoma. *Ann Neurol*. 2007;61(1):25–36. https://doi.org/10.1002/ana.21050
2. Tsutsui K, Kanbayashi T, Tanaka K, et al. Anti-NMDA-receptor antibody detected in encephalitis, schizophrenia, and narcolepsy with psychotic features. *BMC Psychiatry*. 2012;12:37. Published 2012 May 8. https://doi.org/10.1186/1471-244X-12-37
3. Mann A, Machado NM, Liu N, et al. A multidisciplinary approach to the treatment of anti-NMDA-receptor antibody encephalitis: a case and review of the literature. *J Neuropsychiatry Clin Neurosci*. 2012;24(2):247–254. https://doi.org/10.1176/appi.neuropsych.11070151
4. Kuppuswamy P, Takala C, Sola, C. Management of psychiatric symptoms in NMDAR encephalitis: a case series, literature review and future directions. *Gen Hosp Psychiatry*. 2014;36(4):388–391. https://doi.org/10.1016/j.genhosppsych.2014.02.010
5. Dalmau J, Gleichman AJ, Hughes EG, et al. Anti-NMDA-receptor encephalitis: case series and analysis of the effects of antibodies. Lancet Neurol 2008;7:1091-98.

Address for Correspondence:

Jawad Ahmad Wasif, Department of Pediatrics, Letterkenny University Hospital Donegal-Ireland
Cell: +92-333-3331666
Email: jawadwasif7@gmail.com

This is an open access article distributed under the terms of the Creative Commons Attribution License CC-BY 4.0., (http://creativecommons.org/licenses/by/4.0/), which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.