Indian Journal of
Neurology

Open Science Publications

Volume 5, Issue 1 - 2024

www.opensciencepublications.com

GABA B Receptor Autoimmune Encephalitis – A Case Report

Case Report

Gupta A*, Bhardwaj S, Munjal J, K Lal S, Nakra R and Lal V

Department of Hematology and Immunopathology, National Reference Lab, Dr Lal Path labs, Rohini , New Delhi, India

***Corresponding author:** Ajay Gupta, Department of Hematology and Immunopathology, National Reference Lab, Dr Lal Path labs, Rohini, New Delhi, India, Email:drajaygupta76@gmail.com

Article Information: **Submission:** 14/10/2024; **Accepted:** 30/10/2024; **Published:** 07/11/2024

Abstract

Background: Although autoimmune encephalitis is a rare disease with an incidence of 0.8/ 100,000, it is associated with cognitive decline, epilepsy, behavioural disturbances, and impairment in the level of consciousness. Therefore, prompt diagnosis and treatment lead to improvement or full recovery in most cases [6]. Encephalitic syndromes are a common medical emergency. The importance of early diagnosis and appropriate treatment is paramount. If initial investigations for various conditions including infectious agents prove negative, other diagnoses must be considered promptly. Autoimmune encephalitis are being increasingly recognized as important (and potentially reversible) non-infectious causes of an encephalitic syndrome [1]. Some encephalitis or seizure disorders once thought idiopathic now seem to be immune mediated [3]. In 2010 the spectrum of known antigens in autoimmune encephalitis has been expanded by GABAB receptors. Until now over 80 patients with GABAB receptor encephalitis have been described [2].

Introduction

Anti-gamma-aminobutyric-acid B receptor (anti-$GABA_BR$) encephalitis, which was first described by Lancaster et al., is clinically characterized by limbic encephalitis (including seizures, cognitive disorders, behavioural changes) and other uncommon clinical syndromes (such as cerebellar ataxia and opsoclonus-myoclonus syndrome) [4]. Approximately 50% of patients are diagnosed with small cell lung cancer (SCLC), and in rare cases, thymoma, malignant melanoma, breast carcinoma, rectal carcinoma, multiple myeloma, esophageal carcinoma, sarcomatoid carcinoma (SC), and gastric adenocarcinoma have also been found [6]. Antibodies against $GABA_B$ receptor are found in serum or CSF [5]. In the majority of patients oligoclonal bands are found in the CSF and 24 hr IgG levels are increased in 92% of patients [7]. Antibody-mediated encephalitis constitutes a group of inflammatory central nervous system disorders that are associated with antibodies against neuronal cell-surfaces proteins, ion channels, or receptors. The most common type of autoimmune encephalitis is autoantibodies against the N-methyl-d-aspartate (NMDA) receptor. Other autoantibodies include those directed against the voltage-gated potassium channel complex (VGKC), the α-amino-3-hydroxy-5-methyl-4-isoxazolepropionic acid (AMPA) receptor, the γ-aminobutyric acid (GABA) receptor, anti-dipeptidyl-peptidase-like protein-6 (DPPX, viz. DPP6), and the glycine receptor [6].

We report a case of a patient who was diagnosed with autoimmune encephalitis due to GABAB receptor autoantibodies who had long history of unexplained fever, diabetes, severe hypertension having normal routine lab and radiological investigations initially and gradually progressing onto having seizures and neurological deficits with deteriorating life threatening health condition.

Case Presentation

A 56 yr old male pt admitted with history of 10 days fever being investigated upon, all routine lab investigations CBC, LFT, KFT ESR,

CRP, Urine R/M, fever profile including MP, Blood C/s, Dengue and X-ray Chest, USG whole abdomen were normal. pt was diabetic, hypertensive with Hba1c 7.0 and BP 142 / 90 mmHg respectively and was on medication for both conditions, pt was put on IV antibiotics for 2 days, fever subsided on 3rd day of admission and discharged on 4th day. After 2 days pt has one episode of seizure, BP 200 / 120 mmHg, MRI brain was normal was put on antihypertensive medication and BP was 140 / 90 mmHg in 2 days . After couple of days pt had repeat episode of seizure , again 2nd MRI was done which was normal.pt also had little delirium. Subsequently he had repeated episodes of seizures with twitching of left side of face and frothing, pt BP was fluctuating and was put on anti-epileptic drugs. looking at the sign / symptoms a provisional diagnosis of Hypertensive encephalopathy was made. After 10 days of being afebrile, pt again had fever and was quite unstable as he was irritable, in continuous delirium, amnesia, even pulling off infusion line. Pt was on antibiotics, anti diabetics, antihypertensives, anti epileptics drugs with BP in normal range. As blood counts were normal, CSF tap revealed TLC 25 cells/ cumm, raised protein, low sugar, CSF c/s was normal and bacterial meningitis was ruled out .pt was put on acyclovir with Anti tubercular therapy and higher antibiotics but didn't respond and a repeat MRI was done after 20 days of 2nd MRI showed T1 hypointense / T2 hyperintense, punctate T2 /FLAIR hyperintensities seen involving the bilateral mesial temporal lobes including amygdala , hippocampi, parahippocampal gyri. Based on the finding of MRI – test for autoantibodies to diagnose Autoimmune encephalitis and test for oligoclonal bands, test for paraneoplastic syndrome antibodies was done. GABA B receptor autoantibodies were positive and oligoclonal bands were positive. Autoantibodies against paraneoplastic syndrome were negative. EEG was done which showed generalized beta activity. PET –CT was negative for any lymph nodal or any distant metastasis, presence of diffuse mild increased FDG uptake also noted in bilateral mesial temporal lobe, bilateral basal ganglia which is suggestive of Autoimmune encephalitis. Finally a Diagnosis of GABA B receptor Autoimmune encephalitis was made after more than one month of onset of 1st seizure. Pt was put on IV immunoglobulins for 5 days, his condition improved. Further pt was advised with follow up and treatment with rituximab (Figures 1 and 2).

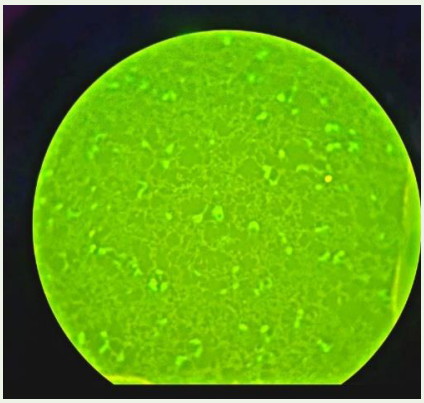

Figure 1: IFA Showing GABA B receptor autoimmune encephalitis on cell based assay kit (Euroimmun, Germany).

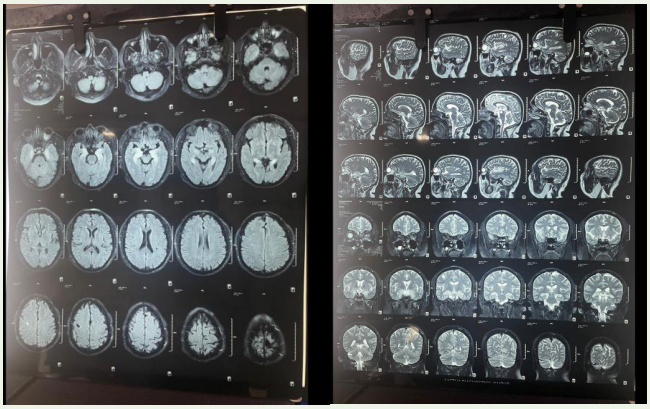

Figure 2: MRI Brain with T2 hyperintense / FLAIR seen involving the bilateral mesial temporal lobes including amygdala, hippocampi, parahippocampal gyri.

Discussion

GABA B receptor autoimmune encephalitis , the diagnosis can be delayed which may be due to the variable presentation of the disease as was seen in this case , the diagnosis was made 1.5 month after initial sign / symptom. This type of encephalitis is often seen in middle aged and older men as was seen in this particular case. Tumour are associated in 50% of cases with more commonly Small cell lung carcinoma however no malignancy was seen in our case. It usually presents with seizures or status epilepticus, ataxia, memory loss as was seen in this case . Some pt might experience fever after which epilepsy, cognitive dysfunction, behaviour abnormalities may be seen and fever may be the first symptom as was seen in this case. MRI is performed early upon pt presentation , however can be negative in large no. of cases [8] as was seen in this case where first 2 MRI were negative. Role of autoantibodies testing in Serum / CSF via cell based assay using IFA technique is of major importance as they tend to be present in early onset when MRI is often negative.

Conclusion

Anti GABA B receptor encephalitis mainly occurs in middle aged men. The onset can be with fever and frequent epileptic seizures, cognitive dysfunctions and abnormal mental behaviour. 50 % cases may be associated with tumour more like lung carcinoma. Comprehensive early testing of Autoantibodies in serum / CSF helps in early diagnosis as MRI may be negative in early stages. A need for early diagnosis and understanding of disease is a good way to prevent misdiagnosis and delayed treatment.

References

1. Wingfield T, McHugh C, Vas A, Richardson A,Wikins E, et al. (2011) Autoimmune encephalitis: a case series and comprehensive review of literature , QJM : International journal of medicine104: 921-933.
2. Schumacher H, Meyer T, Pruss H (2019) GABA B receptor encephalitis in a patient diagnosed with amyotrophic lateral sclerosis, BMC Neurology, article number19: 41.
3. Lancaster E, Lai M ,Peng X, Hughes E, Raizer J,et al. (2010) Antibodies to GABA B receptor in limbic encephalitis with seizures: case series and characterisation of the antigen, The Lancet Neurology 9: 67-76.

Citation: Gupta A, Bhardwaj S, Munjal J, Lal KS, Nakra R, et al. GABA B Receptor Autoimmune Encephalitis – A Case Report. Indian J Neurol. 2024;5(1): 136.

4. Lin J , Li C,Li A, Liu X,Wang R, et al. (2019) Encephalitis with antibodies against the GABA B receptor: high mortality and risk factors, Frontiers in Neurology 10: 1030.

5. Connor KO,Waters P, Komorowski L, Zekeridou A, Guo CY, et al. (2019) GABAA receptor autoimmunityA multicenter experience 6.

6. Santos AB, Hong A, Hong I, Villegas DD (2024) Anti-GABA B receptor autoimmune encephalitis: a report of a rare case in Central America Cureus 16.

7. Zhu F, Shan W, LV R, Li Z, Wang Q (2020) Clinical characteristics of Anti-GABA B receptor encephalitis, Frontiers in neurology11: 403.

8. Abunada M, Nierobisch N, Ludovichetti R, Simmen C,Terziev R, et al. (2024) Autoimmune encephalitis: early and late findings on serial imaging and correlation to treatment time points, European journal of Radiology 12: 100552.

Citation: Gupta A, Bhardwaj S, Munjal J, Lal KS, Nakra R, et al. GABA B Receptor Autoimmune Encephalitis – A Case Report. Indian J Neurol. 2024;5(1): 136.