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Focal Status Epilepticus Revealing Autoimmune Encephalitis With Anti-Gad Antibodies 65

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Abstract:
Background*: Autoimmune encephalitis with anti-GAD antibodies manifests with varied clinical phenotypes, associated with neurological syndromes: limbic encephalitis, cerebellar ataxia and stiff man syndrome.*
Clinical case report: *We report the case of a 25-year-old female patient with a history of Graves' disease, hospitalized in the emergency room for treatment of recurrent focal status epilepticus.*
Conclusion: *Anti-GAD 65 antibodies should be sought in temporal status epilepticus on the basis of autoimmunity.*
Keywords*: Recurrent focal status epilepticus, Autoimmune encephalitis with anti-GAD antibodies, Anti-GAD antibodies 65.*

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I. Introduction

Autoimmune encephalitis with anti-GAD antibodies manifests with varied clinical phenotypes, associated with neurological syndromes: limbic encephalitis, cerebellar ataxia and stiff man syndrome [1, 2, 3].

Antibodies to GAD 65 (glutamic acid decarboxylase 65) are associated with several neurological syndromes, including stiff person syndrome, cerebellar ataxia, and limbic encephalitis (LE). GAD65 is a crucial enzyme involved in the production of a neurotransmitter called GABA (gamma-aminobutyric acid). GABA is the main inhibitory neurotransmitter in the central nervous system. It is widely distributed in the brain and plays a primary role in reducing central nervous system neuronal excitability. Much remains unknown about the role that GAD65 antibodies play in neuroinflammation and their pathogenic role remains controversial; Furthermore, the T cell-mediated immune response appears to play an important role in the pathogenesis of autoimmune encephalitis with anti-GAD65 antibodies.

It affects women more frequently than men and the median age at which symptoms appear is 30 years (range: 5 to 80 years). Subjects presenting neurological symptoms associated with GAD65 antibodies most often have a personal or family history of autoimmunity, including type 1 diabetes mellitus, thyroiditis, pernicious anemia or vitiligo; which are found in almost half of patients and indicate a genetic predisposition.

Autoimmune encephalitis with anti-GAD65 antibodies is characterized by the subacute onset of convulsions and amnesia. Convulsions are most often refractory to antiepileptic treatment, status epilepticus is rare. Psychiatric manifestations, such as anxiety, depression, apathy and behavioral disorders, are less common than for other antibodies.

II. Clinical Case

We report the case of a 25-year-old female patient with a history of Graves' disease, hospitalized in the emergency room for treatment of recurrent focal status epilepticus: temporal status epilepticus. confirmed on EEG.

Anti-GAD 65 antibodies present positive at serum level 138,4 u/ml (Highly elevated serum levels of anti-GAD antibodies 65 were present). Lactate negative (blood and CSF). Brain MRI reveals a cortical hypersignal with a triangular configuration at the left temporoparietal level.

A bolus of corticosteroids for 5 days and Ig immunoglobulin IV at a dose of 2g/KG/d for 5 days allowed clinical and electrical improvement (Focal status epilepticus).

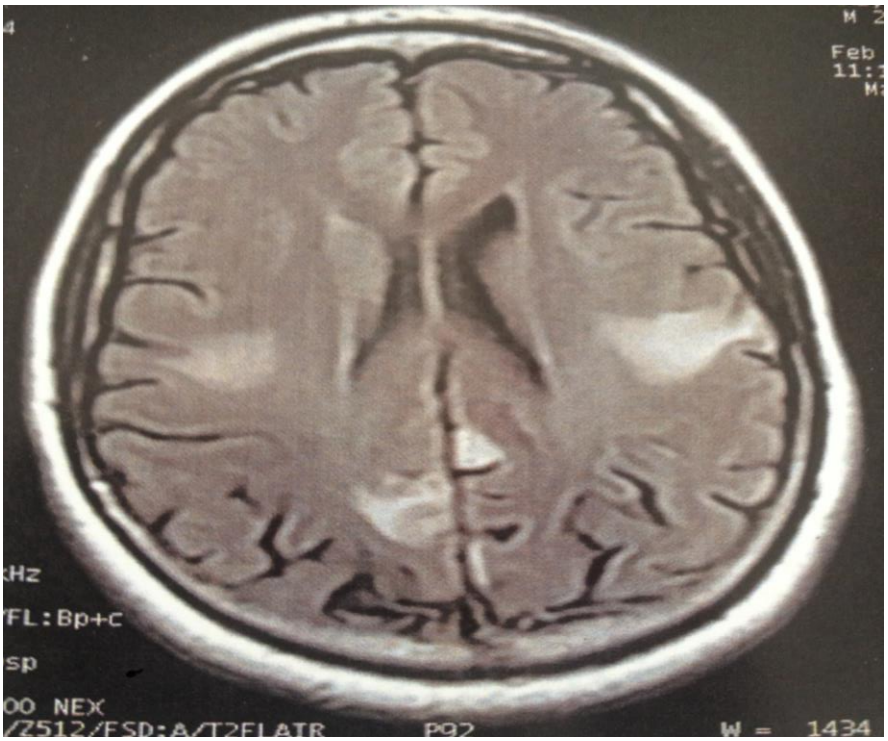

Figure 1. Brain MRI, cortical hypersignal with a triangular configuration at the left temporoparietal level.

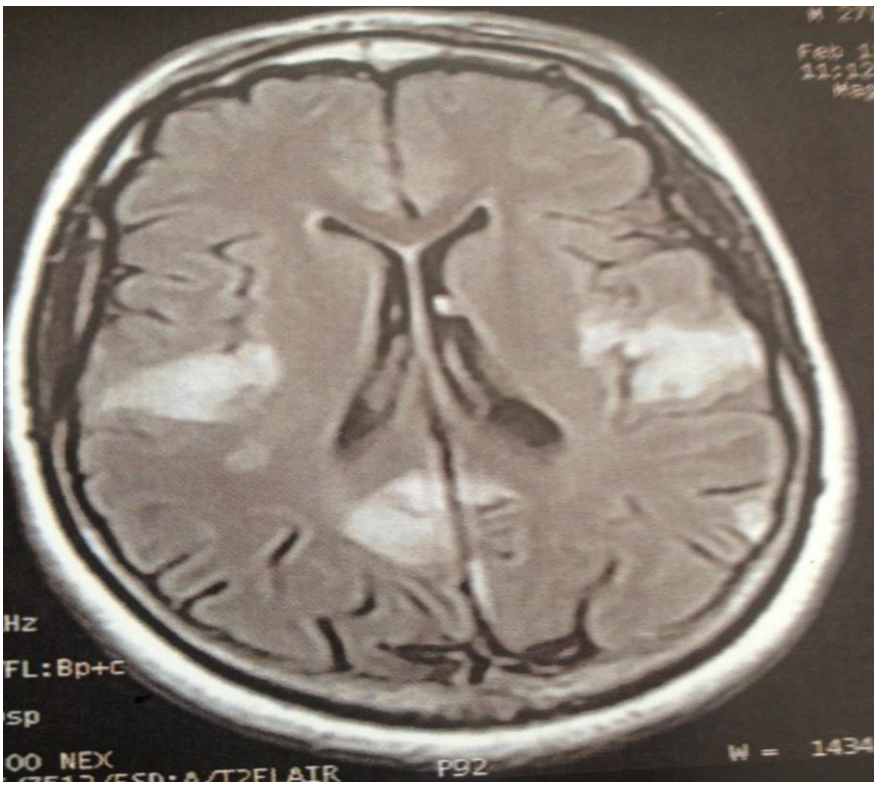

Figure 2. Brain MRI, cortical hypersignal with a triangular configuration.

III. Discussion

In this patient, temporal status epilepticus is the mode of revelation of encephalitis associated with anti-GAD 65 antibodies.

A 5-day course of intravenous methylprednisolone,was effective on focal temporal status epilepticus ,but no effect was observed on seizure frequency, EEG abnormalities, or neurologic symptom. Consequently we used rituximab.

Encephalopathy associated with GAD antibodies may be reversible with immunotherapy. Whereas intravenous immunoglobulin and intravenous and oral corticosteroids had no effect on seizure frequency. (Malter et al,2010)[4] had found 53 patients (aged 17-80 years) with epilepsy and GAD antibodies, treatment with intravenous immunoglobulin, corticosteroids, or cyclophosphamide did not improve seizure control. The use of rituximab may have been effective in our patient and may be of interest in treatment of autoimmune neurologic diseases(Dalakas MC,2008) [5] (Daif A,2018) [6] . This anti-CD20 monoclonal antibody causes selective destruction of B lymphocytes and decreased production of antibodies (Zecca M et al,2003) [7].

IV. Conclusion

Anti-GAD 65 antibodies should be sought in temporal status epilepticus on the basis of autoimmunity.

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