2-044

Atypical Epilepsy in Common Variable Immunodeficiency: A Single Institution Case Series

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Objective

Describe atypical epilepsy presentations in patients with common variable immunodeficiency (CVID) within the University of Utah Healthcare system.

Background

CVID diagnosis requires: hypogammaglobulinemia, poor vaccine response, and onset after the age of four years old, in the absence of an alternative explanation for immunodeficiency.[1]

Patients with CVID are at increased risk of infection, malignancy, and autoimmune disease.[2] These patients may also present with concomitant neurological diseases, most often infective or inflammatory processes.[3]

There is limited data on the coexistence of epilepsy and CVID; clinically, we observed several patients with atypical epilepsies – *association vs coincidence?*

Methods

- Retrospective chart within the University of Utah electronic medical record based on ICD coding for CVID who had at least one encounter in the U of Utah Adult Immunology/Immune Deficiencies clinic, as well as at least one encounter in the U of Utah Neurology Department.
- Patients meeting clinical criteria for CVID by an Immunologist were further examined for co-existing epilepsy.
- Patients were included in the study if they had episodes concerning for seizure, treatment with antiepileptics, and abnormal EEG and/or imaging.

Acknowledgements

This project was supported by the Transverse Myelitis Association and Barbara Gural Steinmetz Family.

Demographics

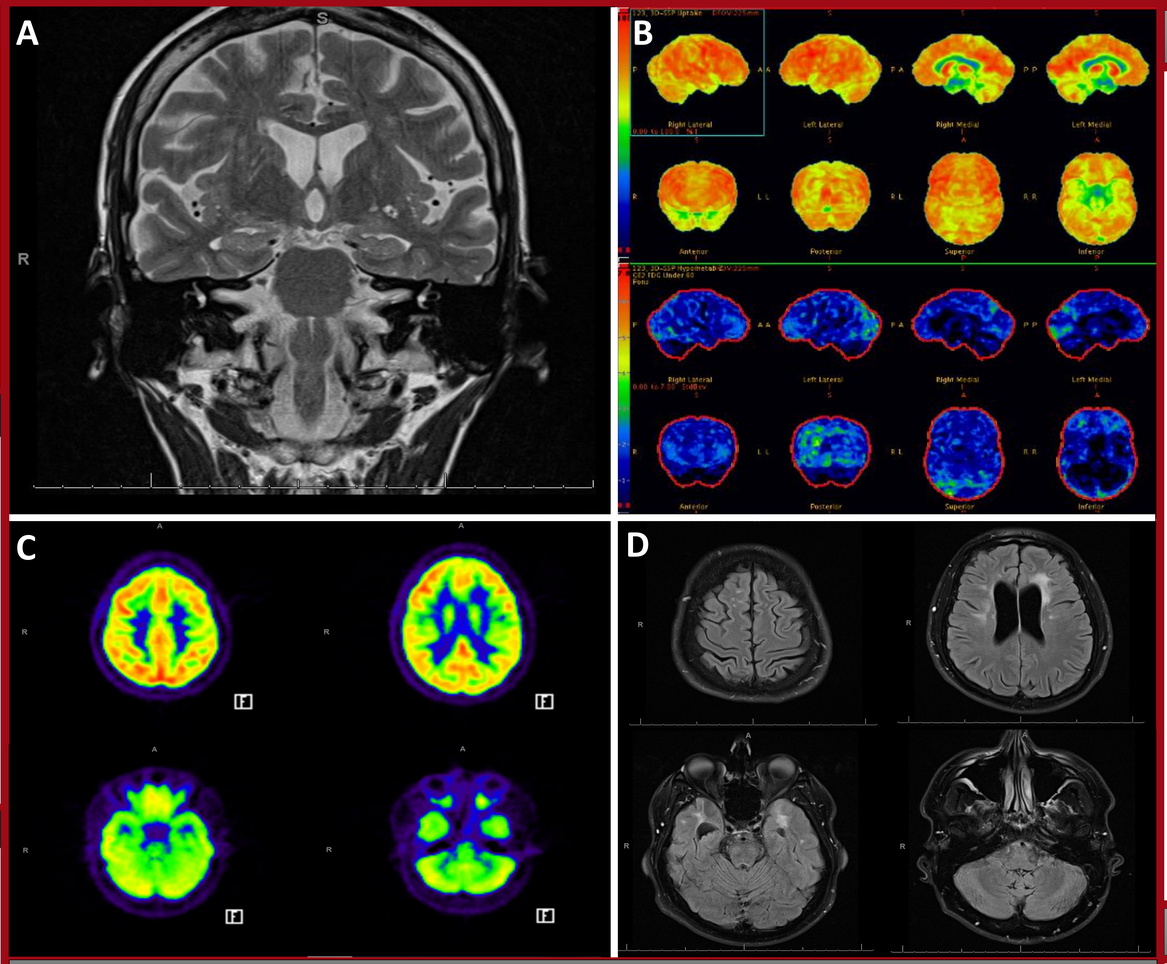
Male	3
Female	2
Alive	4
Deceased	1
Mean Age Seizure Onset (*SD*)	29 (*6.4*)
Mean Age CVID Diagnosis (*SD*)	37 (*22.8*)
Co-Existing Autoimmunity	2
Co-Existing Malignancy	1

Seizure Semiology

Patient 1	Episodes of disorientation and staring off occurring in 1-2 clusters per month.
Patient 2	Transient epileptic amnesia; staring spells lasting 10-15 seconds with unclear frequency.
Patient 3	Episodes of confusion with speech arrest lasting 10-15 minutes occurring weekly.
Patient 4	Spacing out and panic attack-like episodes lasting seconds occurring daily; also with myoclonic-like spasms occurring several times daily.
Patient 5	Occasional generalized tonic clonic seizures; episodes of alterations of consciousness lasting several minutes with unclear frequency.

EEG Findings

Patient 1	Focal sharp waves in bilateral temporal heads, occurring independently.
Patient 2	Left temporal sharp waves.
Patient 3	Left centro-temporal slowing.
Patient 4	Periods of abnormal sleep architecture with bilateral delta and superimposed alpha activity + occasional sharp waves in the frontal region.
Patient 5	Right focal slowing, along with periodic discharges.



Imaging Findings

Patient 1	MRI brain consistent with right>left temporal lobe asymmetry, consistent with mesial temporal sclerosis (**Figure A**).
Patient 2	MRI brain with right>left temporal lobe atrophy. FDG-PET notable for hypometabolism within the left parietal and occipital lobes (**Figure B**).
Patient 3	MRI brain with non-specific frontal lobe white matter T2 hyperintensities. PET brain with left>right temporal hypometabolism; cerebellar hypometabolism (**Figure C**).
Patient 4	Normal MRI brain.
Patient 5	MRI brain demonstrating frontotemporal periventricular and subcortical white matter hyperintensity and volume loss (**Figure D**). MRI SPECT unrevealing. Focal metabolic activity in right mesial temporal lobe on PET brain.

Discussion and Conclusions

We present a case series of 5 patients within the University of Utah Healthcare system with CVID and co-existing epilepsy.

All patients had atypical seizure semiologies including behavioral arrest, alterations in consciousness, and/or amnestic episodes. Less common were generalized tonic clonic seizures.

A majority of patients had abnormal imaging findings, most commonly temporal lobe asymmetry.

A majority of patients had improvement with antiepileptic therapy. IVIg did not improve epilepsy symptoms; one patient worsened in setting of aseptic meningitis.

Atypical epilepsy and routine EEG should be considered in patients with CVID with abnormal “spells”. In patients with epilepsy, in particular autoimmune epilepsy, baseline immunoglobulin testing should be considered, as some antiepileptics and immunotherapies may lower immunoglobulin levels.[4]

References

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