TEMPORAL LOBE EPILEPSY AND GESCHWIND SYNDROME

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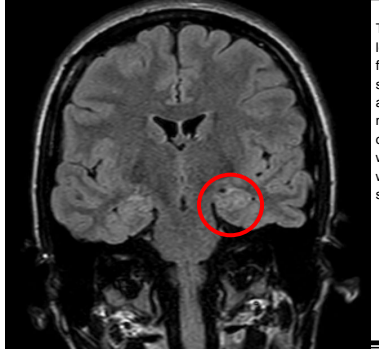
INTRODUCTION

Temporal lobe epilepsy (TLE) is the most common form of localization-related epilepsies. The most common etiology of TLE is mesial temporal sclerosis, thought to be secondary to oxidative stress.[1] TLE typically presents as focal seizures with impairment of consciousness and symptoms such as behavioral arrest, automatisms, or "aura," such as rising epigastric sensation, experiential phenomena (déjà vu, jamais vu, or fear), and gustatory and olfactory hallucinations.[2,3] TLE is also associated with neuropsychiatric symptoms, specifically depression, schizophrenia-like symptoms, and an interictal pattern of behavioral changes known as Geschwind Syndrome. Geschwind Syndrome consists of a triad of hyperreligiosity, hypergraphia, hyposexuality.[4,5] Additional characteristics are bisexuality, schizophrenia-like symptoms, a strong moral compass, heightened emotions, and poor emotional regulation (irritability, impulsivity, egocentric behavior).[5] Hyperconnectivity between the amygdala and temporal lobe has been proposed as the underlying mechanism for development of this behavior change, where significant emotions are attached to seemingly everyday events.[6]

This case describes a patient who presented to the emergency department seeking methamphetamine detoxification. Throughout the hospital course, it became clear that the patient also described symptoms associated with TLE and behavioral changes seen in Geschwind Syndrome. This case underscores the use of Geschwind Syndrome as a diagnostic tool for TLE.

FIGURE 1



T2 hyperintensity in left hippocampal formation with slightly bulbous appearance. Loss of normal granulation of hippocampus which can be seen with mesial temporal sclerosis

CASE

History: A 27-year-old female with a history of substance use disorder presented to our ED for detoxification from methamphetamine. She had been using methamphetamine for the previous 2 months for its 'calming effect' along with suboxone 8mg sublingually to help with sleep, three 12-ounce 7% beers daily, smoking 1 pack of cigarettes per day, and hallucinogens about once a month. Her substance use started six years prior when she was prescribed Vicodin for kidney stones. Regarding hallucinogens, she has religious ideas around its use and described the experience as 'grounding.' She endorsed depression, anxiety, but denied suicidal ideation. She engaged in delicate cutting and burning to distract from internal discomfort. She described hypnagogic and olfactory hallucinations, smells of gas or burning and some 'spacing out' when these occur. She denied seizures but described disorienting experiences of 'deja-vu' and 'jamais-vu' which she first noticed 6-7 years prior to presentation. Past medical history significant for birth via emergency C-section due to concerns for anoxia. She had attentional problems during school that were never addressed but was able to graduate high school. She is an artist and feels compelled to paint and draw; she sells her work online. She identifies as lesbian and endorsed a low sex drive.

Pertinent Labs and Imaging:

CBC, CMP, UA, EtOH: within normal limits
UTox: + amphetamines, + cannabinoids, + buprenorphine
MRI: See Figure 1
EEG: Normal EEG recorded during awake, drowsy, and light sleep stages with no epileptiform abnormalities

MSE: Friendly, cooperative female appearing stated age. Casually dressed with hair dyed red. No psychomotor agitation or retardation. No abnormal movement, good eye contact. Speech normally paced and modulated. Stated she felt depressed but appeared euthymic. Described hypnagogic hallucinations at bedtime and frequent olfactory hallucinations, deja vu, and jamais vu experiences. Acknowledged feelings of micropsia and macropsia akin to Alice in Wonderland. No frank auditory or visual hallucinations. Logical and linear thinking without evidence of delusions. Able to think abstractly and with intact memory. Good insight and judgment.

Assessment/Plan:

The patient is a 27-year-old female with PMH significant for substance use disorder who presented with request for detoxification from methamphetamine. Incidentally, clinical features and MRI point towards probable unrecognized temporal lobe epilepsy with associated Geschwind's Syndrome despite normal EEG. She was referred to neurology for further evaluation and prevention of secondary generalization.

Axis I: Substance use disorder - patient has significant social support at home and has stopped using other substances in the past. Eager to move forward with sobriety without assistance of group therapy
ADHD - previously undiagnosed, was self-medicating with use of stimulant street drugs
Begin methylphenidate 36 mg q day

Axis II: Geschwind Syndrome - personality change associated with temporal lobe epilepsy: hyposexuality, hypergraphia in the form of drawing/art, and hyperreligiosity

Axis III: Mesial Temporal Sclerosis/Seizure activity - despite normal EEG, clinical picture indicates temporal lobe epilepsy without secondary generalization. Begin carbamazepine ER 100 mg bid as seizure prophylaxis

DISCUSSION

There is much controversy as to whether Geschwind Syndrome exists, stemming from the lack of randomized control trials.[7] Seizures related to mesial temporal sclerosis (MTS) are most commonly diagnosed and treated by adolescence, with over 80% of cases of TLE manifesting before age 16. Behavioral changes associated with Geschwind Syndrome are typically chronic and increase over time, making this phenomenon rare.[5,8] A mechanism for development of the syndrome may be hyperconnectivity between the amygdala and temporal lobe; in contrast, Kluver-Bucy syndrome where there is disconnect between these two structures presents with inverse symptoms, a loss of emotionality rather than increased emotions. The constellation of hypergraphia, heightened spirituality, and hyposexuality in conjunction with olfactory hallucinations and 'deja-vu' experiences raised suspicion for Geschwind Syndrome and TLE. Etiology of our patient's MTS was most likely anoxic brain injury during birth. Recognizing potential seizure activity and associated behavior change is imperative to identify epilepsy in patients whose symptoms would otherwise go undetected.

CONCLUSIONS

TLE is the most common localized epilepsy and is most frequently associated with lesions of the mesial temporal lobe. It can present with behavioral arrests, automatisms, or an aura. TLE is also associated with a disputed constellation of behavioral changes known as Geschwind Syndrome, consisting of hyperreligiosity, hypergraphia, and hyposexuality. Geschwind Syndrome develops chronically, and symptoms increase over time. When analyzed in conjunction with other presenting symptoms, Geschwind Syndrome can be helpful to diagnose unrecognized TLE.

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