"Frequent Falls and Body Jerks"

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Epilepsy Risk Factors:

Febrile Seizures	No
Age of Onset	67
Head Trauma	No
CNS Infection	No
CNS Neoplasm	No
CVA	No
Family History of Epilepsy	No
Birth History	Unremarkable
Developmental History	Unremarkable
Other	NA

DISCLOSURE

NONE

Case

PMHx: HTN, HLD, CTS

PSx: Appendectomy

Allergies: NKDA

SocHx: Current smoker: cigars 2/day since 2000. No ID use. Drinks 1-2 cans of beer daily. Separated from Wife. Employed as Golf Caddy.

FamHx: Mother-Dementia

Meds: Lisinopril, atorvastatin, aspirin

ROS: bradyphrenia, short-term memory loss, "personality changes", shuffling gait, SOB when bending over, 15 lb weight gain, joint aches from falls, skin tears, dizziness upon standing

Case: HPI

CC: Frequent falls and body jerks

67y/o left handed CM p/w acute onset falls. First episode he fell down the stairs while working as golf caddy. The same day, he unexpectedly fell backwards. This progressed to 5-10x a day where he would stumble with associated spasms. Spasms were described as being right sided. They may be intermittently followed by flexion on LUE. He notes dropping objects. Episodes may include transient confusion where he is unable to complete the task at hand and/or have nonsensical speech. No report GTC events. No precipitating factors. No temporal preference.

Patient was initially treated for vertigo by OSH Neurologist with antivert. Referred to Nsg for consult of falls thought to be due to NPH. Large volume tap did not improve gait complaints. Then seen by Movement disorder clinic who felt patient was suffering from seizures: myoclonus vs psychogenic, referred patient to Epilepsy clinic.

Neurological Exam

- No pertinent findings on Neurologic examination

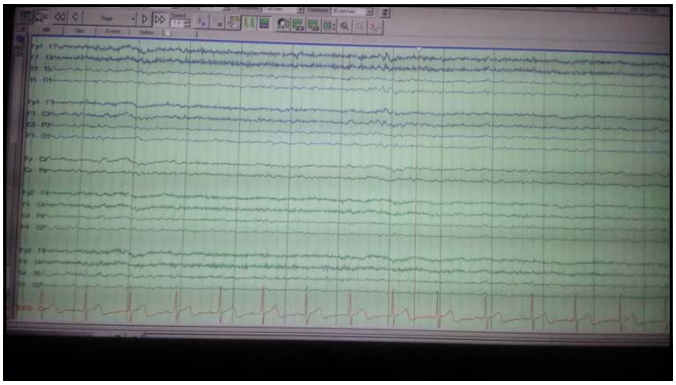
Mental Status	Otherwise cognitively appropriate for age Orientation: oriented to time, place, and person Memory: recent and remote memory intact Attention: attention span and concentration were age appropriate Language: fluent and spontaneous without dysarthric features Knowledge Base: within normal limits
Cranial Nerves	II - visual fields were intact III, IV, & VI - all extraocular movements were intact and no nystagmus noted V - facial sensation was normal and symmetrical VII - eye closure was normal bilaterally and facial contours and strength were symmetrical VIII - non-visualized stimulus response was normal IX & X - uvula midline with normal soft palate movement XI - shoulder shrug strength appeared normal bilaterally XII - tongue protrusion was midline, no fasciculations noted
Motor	Normal bulk and tone. 5/5 strength throughout
Coordination and Gait	Finger-nose-finger, heel-shin, and rapid alternating movements normal; gait deferred
Sensation	Intact to light touch, vibration, and temperature
Reflexes	1+ UEs and patellar; Babinski negative

Case continued

As an outpatient, began on Clonazepam for which he self-discontinued. He was then started on Zonisamide, uptitrated to 400mg qhs.

Pt transferred from OSH due to 2mm parafalcine SDH and large left subcutaneous hematoma on CTH. There was concern that events were increasing following 2 week addition of Levetiracetam. No neurosurgical intervention. Placed on vEEG, no events captured. Levetiracetam discontinued and Clonazepam reinstituted.

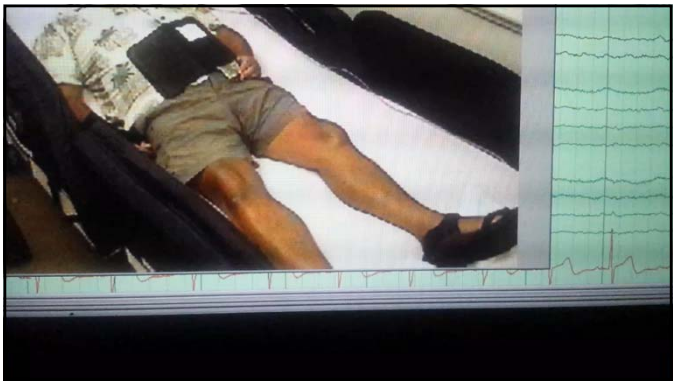
One month later, readmitted again to Neurosurgery for falls with head trauma. CTH showed right holohemispheric SDH of 6mm with extension to the right tentorium and flax. Mild mass effect upon the adjacent brain parenchyma and right lateral ventricle with approximately 1 mm of right-to-left midline shift. Left parietal scalp hematoma without evidence of underlying calvarial fracture. Age-indeterminate comminuted bilateral nasal bone fractures. No intervention. Neurology recommended increasing Clonazepam to 0.5mg tid.



Case

2 days after discharge, readmitted. Ex-Wife frustrated with patient going home then immediately falling, injuring self, and returning back to hospital. No new issues, continues to have episodes. Frequency and characteristics unchanged. No change in patient's medication regimen. Plan to send to Cleveland clinic for nonlesional stereotactic EEG. Discharged with gait belt and helmet.

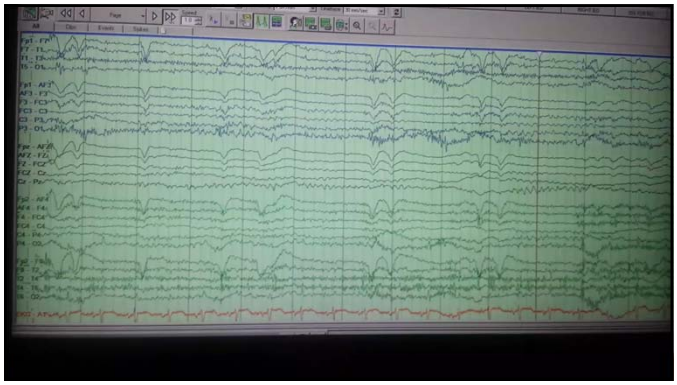
In the meantime, Encephalopathy, autoimmune evaluation, serum panel (ENCES) sent…

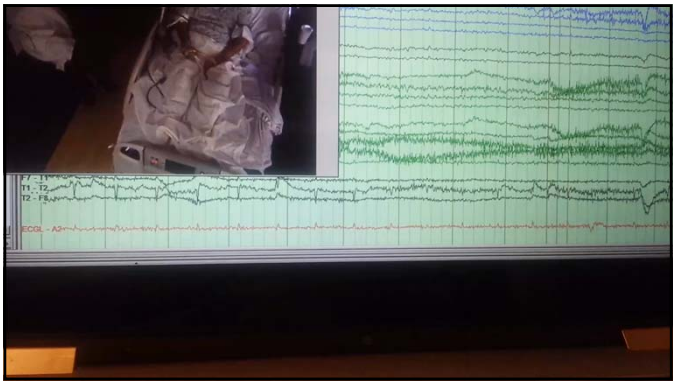


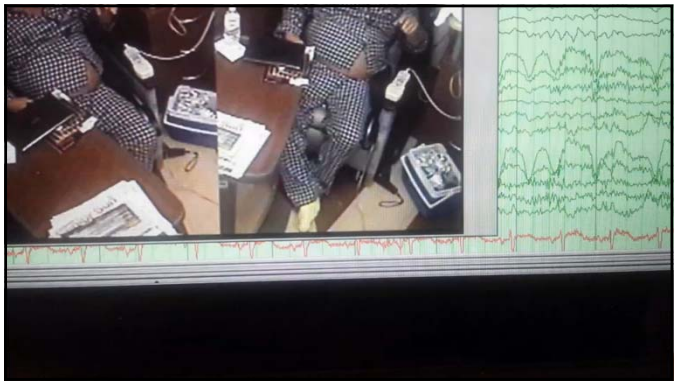
6Hr EEG

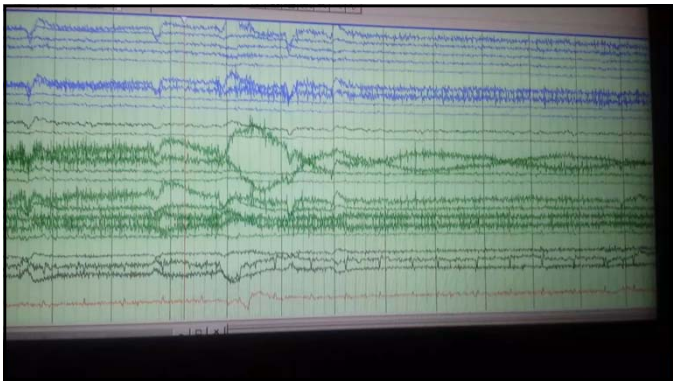
EMU admission:

Semiology-brief, rapid right arm and leg tonic extension
Electrodecrement before, during, and following event
More than 25 events captured during admission
Working diagnosis: Left SMA partial onset seizures









6hr EEG and LTM

Multiple events captured without EEG correlate concerning for epileptic myoclonia vs non-epileptic myoclonia

Work-up

MRI Brain Epilepsy Protocol: No appreciatable seizure focus.

PET Scan: "Images reveal multiple areas of hyper-and hypo-metabolic activity bilaterally, slightly more pronounced on the left. Regions of hypometabolic activity include left superior lateral temporal cortex and left anterior cingulate cortex. Additional foci of hypermetabolic activity are seen in the occipital cortex, likely due to visual activity during the exam. "

Work-up

ENCES: encephalopathy, autoimmune evaluation, serum

- VGKC ab 0.30 (nml<0.02), medium level according to Klein et al
  - LGI1
- Received 5 sessions plasma exchange
- Sx free since Feb 2015

Discussion

- Diagnosis:
  - MRI typically normal or with hyperintensity of mesial TL.
  - CSF normal
  - 20% associated with neoplasm
- Treatment: Immunosuppression

Discussion

LGI 1-leucine rich glioma inactivated 1

- A common autoantigen of voltage gated potassium channel (VGKC)
- Serves as ligand between 2 epilepsy related proteins, ADAM22(A Disintegrin And Metalloprotease Domain) and ADAM23.
- Characterized by seizures, confusion, and memory disturbances. Seizures etiology brief faciobrachial dystonia that are poorly responsive to AEDs and often mistaken for myoclonus.
- When associated with VGKC, yields higher values of VGKC complex positivity ad cortical clinical presentation of disease.
- Unlike other autoimmune diseases, is not typically associated with a type of cancer.

References

Klein et al. Insights From LGI1 and CASPR2 Potassium Channel Complex Autoantibody Subtyping. JAMA Neurol. 2013;70(2):229-234.

Irani et al. Faciobrachial Dystonic Seizures Precede Lgi1 Antibody Limbic Encephalitis. Ann Neurol 2010

Autoimmune and paraneoplastic movement disorders: An update. Baizabal-Carvallo, José Fidel et al. Journal of the Neurological Sciences , Volume 385 , 175 - 184

Discussion

Faciobrachial Dystonic Seizures (FBDS)

- Described as frequent, brief involuntary dystonic movements that are typically preceded by development of limbic encephalitis.
- EEG is typically nondiagnostic
- Seizures are unresponsive to antiepileptic therapy
- Can be associated with dizziness, hyponatremia, and peripheral neuropathy