Question 30

A 20-year-old man presents following a generalised tonic-clonic seizure precipitated by alcohol excess and sleep deprivation. He has an eight-year history of episodes of a brief loss of time without other symptoms. Observers report that these episodes last 10 to 15 seconds during which he stares blankly and is unresponsive to command. His left hand may stiffen and he may fiddle with his buttons using his right hand. He will then look surprised and resume his conversation. Cranial magnetic resonance imagine (MRI) (T1 weighted) is shown below.

The most likely diagnosis is:
A. mesial temporal sclerosis
B. generalised epilepsy with absences
C. hippocampal tumour
D. callosal agenesis
E. temporal lobe neuronal migration abnormality

The key points in the presentation

First seizure with a provoking cause good history for a particular epilepsy syndrome ie absence and arm movements

Management: he needs advice about provoking activities and an EEG within 24 hours then a provoking EEG
He has features of generalised (absence) and localised (arm movement) so he needs work up for a diagnosis

MRI – good for finding structural causes of epilepsy

A. mesial temporal sclerosis
   • MRI detects mesial temporal sclerosis by demonstrating this size asymmetry and abnormal signal within the atrophied hippocampus
B. generalised epilepsy with absences
   • Imaging and physical examination generally normal would need EEG for diagnosis
C. hippocampal tumour
D. callosal agenesis
E. temporal lobe neuronal migration abnormality

Mesial Temporal Sclerosis
The most common cause of complex partial seizures is mesial temporal sclerosis, occurring in 35 to
65 percent of patients who undergo temporal lobe surgery. In mesial temporal sclerosis, the hippocampus is smaller than normal. This usually occurs on one side of the brain, but can occur bilaterally in 10 to 15 percent of cases.

MRI detects mesial temporal sclerosis by demonstrating this size asymmetry and abnormal signal within the atrophied hippocampus (*Figure at left*). Thin-section, high-resolution coronal MR images are best for detecting these abnormalities, which can be subtle. T1-weighted images are best for detecting size asymmetry, and T2-weighted images are most sensitive for detecting signal changes. A special T2-weighted sequence called FLAIR (fluid attenuated inversion recovery) is even more sensitive for detecting signal abnormalities (*Figure at right, below*). With subtle hippocampal atrophy, quantitative three-dimensional volume measurements of each hippocampus can be useful. These volume measurements can be especially helpful when there is bilateral hippocampal atrophy.

http://www.neuro.wustl.edu/epilepsy/pediatric/articleMRIinTLE.html

Table 1. Classification of the epilepsies

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<thead>
<tr>
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<th>Generalized</th>
<th>Localization-related</th>
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<tbody>
<tr>
<td>Idiopathic (genetic)</td>
<td>• Childhood absence epilepsy</td>
<td>• Benign focal epilepsy of childhood (2 types)</td>
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<tr>
<td></td>
<td>• Juvenile absence epilepsy</td>
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<td></td>
<td>• Juvenile myoclonic epilepsy</td>
<td>• ADNFLE</td>
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<tr>
<td></td>
<td>• Epilepsy with grand-mal seizures on awakening</td>
<td>• Primary reading epilepsy</td>
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<td></td>
<td>• Other idiopathic generalized epilepsies</td>
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<td>Symptomatic or cryptogenic</td>
<td>• West's syndrome</td>
<td>• Mesio-temporal lobe epilepsy</td>
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<td>• Lennox-Gastaut syndrome</td>
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<td></td>
<td>• Other symptomatic generalized epilepsies</td>
<td>• Neocortical focal epilepsy</td>
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ADNFLE: autosomal dominant nocturnal frontal lobe epilepsy

Idiopathic Generalized Epilepsies
These syndromes, formerly called primary generalized epilepsies, are the best known group of idiopathic epilepsies. They epitomize the meaning of the term idiopathic: genetic basis, normal neurologic examination, and normal intelligence. EEG shows generalized epileptiform discharges and may show photosensitivity. Seizure types include generalized tonic-clonic, absence, and myoclonic. Within the group of idiopathic generalized epilepsies, distinct entities are distinguished, primarily based on the predominant seizure types and the age of onset. Some syndromes are very well individualized and others have less clear boundaries.

Symptomatic and Cryptogenic Localization-Related Epilepsies

This is by far the most common type of adult-onset epilepsy. These cases are characterized by seizures arising from a localized region of the brain. If the cause is found, these epilepsies are symptomatic; if imaging studies are normal and the cause remains elusive, and they are cryptogenic. The boundary between the two epilepsies is largely dependent on diagnostic and imaging techniques. Causes such as low-grade tumors, hippocampal sclerosis, and subtle cortical dysplasias are identified more often because of advances in neuroimaging. Clinically, seizures may be simple partial or complex partial, with or without secondary generalization. Interictal EEG shows focal spikes or sharp waves, and ictal EEG shows a focal or regional discharge at onset.

Hippocampal tumour

I can’t distinguish this from the temporal sclerosis MRI – hard to find specific information so not a common presentation

D. callosal agenesis
This is a diagnosis made on fetal ultrasound

Agenesis of the corpus callosum can be complete or partial, depending upon the stage of development at which growth was arrested. Normally, the corpus callosum begins to develop anterior to the interventricular foramina of Monroe at about 12 weeks of gestation. It subsequently grows upward and backwards, in a 'C' shape, as the primitive cerebral hemispheres grow laterally and then posteriorly. A normal corpus callosum can be sonographically appreciated by 18 to 20 weeks of gestation.

Total or partial agenesis of the corpus callosum usually occurs (80 percent) in combination with other malformations, although both can occur as isolated anomalies. Most cases are sporadic, but several genetic and chromosomal disorders have been associated with its absence

E. temporal lobe neuronal migration abnormality
Neuronal migration disorders (NMDs) are a group of birth defects caused by the abnormal migration of neurons in the developing brain and nervous system. Symptoms vary according to the abnormality, but often feature poor muscle tone and motor function, seizures, developmental delays, mental retardation, failure to grow and thrive, difficulties with feeding, swelling in the
extremities, and a smaller than normal head. Most infants with an NMD appear normal, but some disorders have characteristic facial or skull features that can be recognized by a neurologist.

So the question

A. mesial temporal sclerosis
   - MRI detects mesial temporal sclerosis by demonstrating this size asymmetry and abnormal signal within the atrophied hippocampus – consistent with presentation

B. generalised epilepsy with absences
   - Imaging and physical examination generally normal would need EEG for diagnosis, consistent with presentation

C. hippocampal tumour
   - MRI good for finding mass – I guess consistent with presentation

D. callosal agenesis – not the answer

E. temporal lobe neuronal migration abnormality – not the answer

So is it A, B or C?
C is the most unlikely from the 3 and so A if you are convinced of a change in the MRI and B if you think the MRI is normal

The answer is A