

Neuropathology of Epilepsy

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Case-Based Questions (please see page 3 for answers)

1.	Which activity is typically part of Phase II evaluation for epilepsy?
a.	Ablation of the epileptogenic lesion
b.	Neuropathology evaluation
c.	Placement of intracranial electrodes
d.	Video EEG

2.	Which ILAE type of hippocampal sclerosis is associated with CA4-predominant neuronal loss?
a.	Type 1
b.	Type 2
c.	Type 3
d.	Type 4

3.	Which epilepsy-related disorder is associated with pathogenic variants in GNAQ?
a.	Hippocampal sclerosis
b.	Rasmussen encephalitis
c.	Sturge-Weber syndrome
d.	Tuberous sclerosis

Scroll to Page 3 for answers.

Answers

Question 1 Correct answer and rationale: C) Placement of intracranial electrodes

Phase II includes surgical placement of depth electrodes or grid electrodes. Obtaining a video EEG is a Phase I procedure, surgical ablation or resection is Phase III, and neuropathology evaluation is post-surgical.

Question 2 Correct answer and rationale: C) Type 3

Hippocampal sclerosis ILAE type 3 is also known as “end-folium sclerosis.” Type 1 is associated with more diffuse neuronal loss or with CA1/CA4 neuronal loss; type 2 features CA1-predominant loss of neurons and gliosis. Type 4 refers to lack of neuronal loss (isolated gliosis).

Question 3 Correct answer and rationale: C) Sturge-Weber syndrome

A manifestation of Sturge-Weber syndrome (encephalofacial angiomatosis) is leptomeningeal vascular malformations that can cause epilepsy. Sturge-Weber is most commonly caused by a somatic mosaic *GNAQ* mutation (80-90% of cases). Tuberous sclerosis is caused by alterations in *TSC1* or *TSC2*. Hippocampal sclerosis and Rasmussen encephalitis have no currently known genetic basis.