Seizure Semiology

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OBJECTIVES:

1. To understand the definition of seizure semiology
2. To understand the purpose of seizure semiology in the diagnosis of epilepsy
3. To identify parts of a clinical history that are essential to define seizure semiology
4. To identify lateralization and localization of seizures based on clinical features and ability to identify seizure types based on seizure semiology
5. To understand the semiological classification of seizures as defined by Luders, et al in 1998
6. To understand that seizure semiology in infants differs than that seen at older ages
1. Seizure semiology definition
   a. Clinical signs or manifestations before (pre-ictal), during (ictal), or after (post-ictal) a seizure

2. Purpose of seizure semiology
   a. Crucial to obtaining and confirming the proper diagnosis and localization of the seizure focus
   b. EEG, vEEG, MRI, PET CT, etc. are invaluable in helping localize a seizure focus, but if these tests are in discordance with the clinical semiology, this raises suspicion about the accuracy of the localization
   c. Simple and cost effective

3. Clinical history that provides clues to semiology
   a. Pre-Ictal Phase/Prodrome:
      i. Provoking or precipitating factors
         1. Fever, illness, high altitude, lack of sleep, lack of compliance, menstruation, head injury
         2. Headache, behavioral irritability, personality change
         3. Reflex, photosensitivity, hyperventilation
      ii. Environment of occurrence
         1. To exclude syncope or non-epileptic events
      iii. Time of day (i.e. upon awakening or in sleep)
   b. Ictal Onset/Aura:
      i. Brief focal signs or symptoms (the warning)
      ii. Helps to identify the lobe of origin, as well as help with lateralization and localization
   c. Ictal Phase:
      i. Progression
ii. Aphasia vs Ictal speech

iii. Awareness/Consciousness

iv. Duration (Status epilepticus)

d. Post-Ictal Phase:
   i. Deficits of function
   ii. Confusion and/or amnesia
   iii. Weakness (Todd’s paralysis)
   iv. Visual field defect
   v. Dysphagia/Aphasia
   vi. Post ictal speech
   vii. Duration: none vs prolonged

4. Lateralization and localization of seizures based on clinical features

   a. Types
      i. Sensory phenomena
      ii. Psychic Manifestations
      iii. Motor Abnormalities
      iv. Behavioral Manifestations
      v. Automatisms
      vi. Autonomic Manifestations
      vii. Language Abnormalities

   b. Sensory phenomena
      i. Auras
         1. Ictal sensory phenomena occurring at the beginning of a focal seizure and with preserved consciousness
2. Reported by 22-83% of patients

3. The longer the subsequent seizure, the higher the likelihood of forgetting the aura

4. Present more with seizures from the non-dominant hemisphere and during the awake state

5. Patients are generally aware that their aura is a pseudohallucination

6. Most patients with temporal lobe epilepsy report an aura

7. Absence of an aura is more likely with bitemporal than unilateral temporal

8. Several auras?
   a. Patients reporting several different experiences preceding a loss of consciousness may raise concerns about possible non-epileptic events.
   b. A strong predominance of non-dominant hemispheric lateralization was found among these patients, mostly involving the temporal or posterior regions

ii. Somatosensory phenomena
   1. Well localized, discriminatory, and spreads relatively slowly (“Jacksonian March”)-Parietal lobe (primary somatosensory cortex)
   2. Ill-defined, often accompanied by pain, spreads within seconds, contra- or ipsilateral-posterior Insula-Parietal operculum (supplementary somatosensory area)
   3. May also occur from SMA, causing diffuse contralateral or bilateral tingling and a sensation of movement

iii. Cephalic-sensation in the head (nonvertiginous dizziness, light-headedness, or headache)
1. Often a frontal lobe focus
2. Lateralized ictal headache-can be ipsilateral temporal or occipital
3. Post-ictal headache-Non-localizing (except in children with idiopathic focal epilepsies—generally a posterior focus)

iv. Special senses
1. Gustatory Aura (metallic-rubbery taste)-Insular region/Rolandic operculum, parietal
2. Visual Aura (colors, amaurosis)-Contralateral occipital cortex
   a. Ictal blindness-contralateral occipital
3. Elementary auditory
   i. Primary auditory cortex-contralateral to seizure focus
   ii. Auditory aura is a hallmark of an AD form of lateral temporal lobe epilepsy
   iii. Complex auditory-Temporal-Parietal junction
4. Olfactory Aura
   i. Anterior mesiotemporal (“uncinate fits”)
   ii. Always unpleasant, i.e. burning or rotten

c. Psychic Manifestations
   i. Déjà vu/Jamais vu-Mesiotemporal (without lateralization)
   ii. Forced thinking-Frontal (more verbal) or mesiotemporal (more emotional) of dominant hemisphere
   iii. Ictal fear and ecstatic auras-Amygdala, cingulate
iv. Orgasmic aura-Non-dominant mesiotemporal or parasagittal regions

v. Ictal autopsopy-Non-dominant parietal lobe

d. Motor Abnormalities

i. Positive motor signs: clonic and/or tonic movement, abnormal posturing, dystonia, head and/or eye deviation

ii. Negative motor signs: muscle weakness, paralysis

iii. Forced and sustained (>5 sec) head and eyes deviation has a PPV of 94% for contralateral localization, mostly temporal and frontal lobes (frontal eye field); rising to 100% if accompanied by neck extension and followed by generalization.

   1. Versive fast head turn-contralateral frontal lobe
   2. Versive slow head turn-contralateral occipital lobe

iv. Mouth deviation also has a high PPV of 92% (contralateral), especially if associated with forced head version

v. Seizures arising from the SMA may have head turn in either direction as compared to the focus.

vi. Early, non-forced head turn is felt to be more likely ipsilateral with involvement of the temporal lobe (mesial) and basal ganglia

vii. Contralateral to seizure focus:

   1. clonic, facial-contralateral, neocortical temporal
   2. dystonic limb posturing-temporal or frontal, putamen
   3. eye deviation-occipital
   4. Fencing posture-frontal lobe, SMA
   5. Figure of 4 sign (extended arm)-usually temporal
   6. focal clonic jerking-peri-rolandic, frontal
   7. Postictal Todd’s paresis
8. RINCH (rhythmic ictal non-clonic hand movement)-temporal
9. tonic limb posturing-temporal, frontal, or SMA
10. unilateral ictal paresis

viii. Y sign-SMA
ix. Tonic seizure (with paroxysmal fast activity on EEG)-frontal lobe (medial-SMA)
x. Ictal smile-non-dominant parietal
xi. Ictal pouting/frown (“chapeaux de gendarme”)-frontal lobe (mesial frontal, anterior cingulate cortex)

xii. Hyperkinetic seizure-frontal lobe, SMA
xiii. Gyratory seizure
   1. contralateral frontotemporal if accompanied with forced head turn
   2. ipsilateral if en bloc version of body
   3. rotation to prone-frontal

e. Behavioral Manifestations
   i. Behavioral arrest-temporal or orbitofrontal
   ii. Bizarre-orbitofrontal

f. Automatisms
   i. Repetitive involuntary purposeless or semi-purposeful movements that are usually inappropriate, but occasionally may simulate relatively normal movements
      1. Oral (lip smacking, sucking, swallowing, chewing)-temporal lobe/inferomesial/hippocampal
      2. Bipedal-frontal lobe
      3. Cough
a. Ictal-dominant hemisphere
b. Post-ictal-non-dominant temporal lobe

4. Eye blinks
   a. unilateral-ipsilateral
   b. Bilateral eye blinks-occipital

5. Gelastic epilepsy-hypothalamic, mesial temporal, or frontal cingulate

6. Genital-ipsilateral

7. Ictal/Postictal drinking-non-dominant temporal lobe

8. Ictal nystagmus-contralateral frontal or occipital lobe

9. Ictal spitting-non-dominant temporal

10. Ictal weeping/crying (dacrystic)-mesiotemporal lobe or mesial frontal, non-dominant, if seen with gelastic seizures are typically hypothalamic

11. Postictal nose wiping-ipsilateral temporal lobe (>90% PPV)

12. Proximal non-manipulative-contralateral, temporal

13. Unilateral limb automatism (manipulative extremity)-ipsilateral

ii. Automatisms and lateralizing signs are more common with increasing age, especially with temporal lobe epilepsy. This likely reflects dynamic brain maturation across different ages.

g. Autonomic
   i. Ictal tachycardia is very common and has no localizing value
   ii. Ictal bradycardia or asystole are very rare phenomena.

      1. Temporal lobe, no lateralizing value>>frontal
   iii. Dyspnea is linked to the insula
iv. Ictus emesis (early ictal vomiting)
   1. Non-dominant temporal seizures, anterior insula
   2. Benign occipital epilepsies (Panayiotopoulos syndrome)

v. Ictal urinary urge-non-dominant temporal seizures

vi. Piloerection-Ipsilateral temporal seizures (dominant hemisphere)

vii. Vertigo-Insular-temporal-parietal junction

viii. Epigastric rising-temporal>orbitofrontal>cingulate

ix. Mydriasis, flushing, urinary incontinence-no localizing value

h. Language Abnormalities

i. Ictal speech arrest, ictal jargon-Temporal lobe seizures, dominant hemisphere (speech areas: inferior rolandic and supplementary motor)

ii. Ictal speech preservation-Temporal lobe seizures, non-dominant hemisphere

iii. Postictal dysphagia/aphasia-Dominant hemisphere involvement/dominant temporal

iv. Guttural vocalizations (moaning, grunting, screaming)-Frontal lobe seizures (orbitofrontal or parasagittal)

v. Rarely, ictal humming or singing may occur (reported in about 1% of patients admitted to an epilepsy unit)-Temporal lobe and prefrontal cortex (no clear lateralization)

i. Generalized Epilepsy semiology

i. Absence

   1. Typical: <15 seconds suspension of awareness and behavioral arrest, no post-ictal state, may have eyelid movements, eye opening, or automatisms, often provoked by hyperventilation
2. Atypical: slower LOC and recovery, longer duration, hypertonic or atonic component, less likely provoked by hyperventilation

ii. Myoclonic
   1. Fraction of a second, vary in severity from a twitch to a fall, no LOC
   2. Must distinguish from non-epileptic myoclonus

iii. Clonic-rhythmic bilateral body jerks

iv. Tonic
   1. Sudden LOC with generalized tonic posturing, may be asymmetric (can be associated with drop attacks)
   2. Common pattern: flexion of the trunk and extension of the extremities with abduction of the shoulders, +/-vocalization

v. Tonic-Clonic

vi. Atonic-Can vary from drooping of the head to massive loss of tone with falling, associated with drop attacks

vii. Focal phenomena can occur in idiopathic generalized epilepsy (IGE) and can be potentially misleading.

viii. With a prevalence up to 70%, patients with IGE have also had auras prior to their seizure (similar to that of patients with partial epilepsy)

   1. The aura tends to be more midline and difficult to localize. Visual auras are also described.

   a. Ictal symptoms are produced by epileptic interference of one of the four “spheres”: sensorial sphere, consciousness sphere, autonomic sphere, motor sphere
i. Auras-ictal manifestations having sensory, psychosensory, and experiential symptoms

ii. Autonomic seizures-main ictal manifestations are objectively documented autonomic alterations

iii. Dialeptic seizure-the main ictal manifestation is altered consciousness that is independent of ictal EEG manifestations

iv. Motor seizure-mainly motor symptoms
   1. Simple-simple, unnatural movements elicited by electrical stimulation of the primary or supplementary motor area (myoclonic, tonic, clonic, tonic-clonic)
   2. Complex-complex motor movement occurring in inappropriate setting, i.e. automatisms, hypermotor seizures, automotor seizures, gelastic seizures

v. Special seizures-“negative” features (atonic, astatic, hypomotor, akinetic, negative myoclonic seizures, and aphasic)
   b. Seizure evolution is indicated by consider each semiological component of a seizure, listed in order of appearance and linked by arrows. Limit is 4 components.
      i. Ex. L visual aura → L hand clonic seizure → GTC seizure

6. Seizure semiology in infants
   a. Different from adult seizures due to brain immaturity, different brain metabolism, different causes for the seizures
   b. More likely due to disorders of cortical formation, intrauterine pathology, channelopathies, or inborn errors of metabolism
   c. Semiology of infantile focal seizures:
      i. Aura-often absent
      ii. Clonus, limb-present
iii. Cyanosis, perioral-prominent (particularly with temporal lobe origin), oxygen desaturation (apnea-dominant temporal)

iv. Dystonic posture-absent

v. Hand automatisms-absent

vi. Loss of consciousness-hard to determine

vii. Myoclonus, diffuse-sometimes seen at start or end of focal seizure

viii. Secondary generalization-rare

ix. Spasms-can be concurrent with focal seizures

x. Tonic postures, symmetric-frequent

d. Most common manifestations of infantile focal seizures

i. Spasms, asymmetric or combined

ii. Behavioral arrest with (eye) version (hypomotor-frontal)

iii. Focal clonic

iv. Focal tonic
References