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Seizure Semiology in Focal Epilepsies.

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Conflicts of Interest

- Member of UpToDate ´s Latin American advisory board

Learning Objectives

- Use seizure semiology to correctly identify typical focal epileptic seizures.
- Understand and analyze focal seizures based only on seizure semiology.
- Recognize localizing and lateralizing seizure phenomena that are useful in the diagnosis of focal epileptic syndromes.

Key Message

- Systematic and independent analysis of focal seizures is fundamental in the diagnosis of focal epileptic syndromes.

Focal seizures.

- Focal seizures originate within networks limited to one hemisphere. They may propagate to the same or contralateral hemisphere or engage bilaterally ditributed networks such as the thalamo -cortical system, evolving to generalized seizures.

- Most focal seizures can be grouped in the following main categories :
- **Seizures with alteration or loss of consciousness.**
  - Auras, dyscognitive seizures, dialeptic seizures.
- **Autonomic seizures.**
- **Motor seizures.**
  - **Simple motor seizures:** myoclonic seizures, tonic seizures, clonic seizures, versive seizures.
  - **Complex motor seizures :** hypermotor seizures, automotor seizures, gelastic seizures.

- **Special seizures :** «negative» or «inhibitory» motor seizures such as atonic, hypomotor or negative myoclonic seizures.

Seizures with alterations or loss of consciousness

- **Auras .** Illusions and hallucinations that alter the content of consciousness wthout affecting self awareness. The arousal system, postural muscular tone and fine distal movements are preserved. Eyes are usually open.
- **a) Somatosensory auras .**Abnormal somatosensory sensations, usually parestehsias,that are limited to a clearly defined somatosensory region of the body. Less frequently, the aura may consist in a sensation of heat or pain. The symptomatogenic zone of unilateral somatosensory auras is the contralateral primary sensory cortex .
- **b) Visual auras.**Simple visual hallucinations or illusions originated at Brodmann´s area 17 and 18. They manifest as flashing lights or different colors that may blink and move in the visual field. Patient may report amaurosis during or after the seizure.Easy spread to the parieto - temporal region generating complex visual hallucinations that may evolve to psychic auras.
- **c)Auditory auras:** simple auditory hallucinations like a «buzz» or a noise. Symptomatogenic zone : Heschell´s gyrus. They have no lateralizing value.
- **d)Olfactory auras:** they manifest as an unpleasant smell. They are most frequently seen in patients with temporal mesial temporal lobe epilepsy. They have no lateralizing value.

Seizures with alterations or loss of consciousness

- **Auras**
- **e)Gustatory auras.** They consist of an unpleasant taste. Symptomatogenic zone : insula.They have no lateralizing value.

- **f)Autonomic auras .**Subjective sensations suggesting autonomic alterations such as palpitations, sweating or hot flashes.

- **g) Abdominal auras.** Abdominal sensations frequently reported in patients with temporal lobe epilepsies. They are typically a difficult to describe or unspecific sensation that begins at the epigastric region and frequently raises to the chest .It may also involve throat, head or face.

- **h) Psychic auras.** Complex hallucinations and illusions that usually affect different senses. Most typical examples are distortion of familiarity such as «jamais vu» or «dejá vu», elation or autoscopy.Frequently, these sensations may be associated with emotional alterations such as fear. The temporal lobe is usually involved in these phenomena , including the temporal lobe convexity or the junction with the occipital or parietal lobe.

Seizures with alterations or loss of consciousness

- **Dyscognitive seizures .**
- Characterized by a disturbance of high cognitive functions including aphasic seizures, apraxic seizures and amnestic seizures. Aphasic seizures lateralize to the language dominant hemisphere. Apraxic seizures are thought to arise from activation of mesial and inferior frontal cortex.

- **Dialeptic seizures .**
- Unresponsiveness or markedly decreased responsiveness to external stimuli, activated arousal system, preservation of postural tone and fine distal movements, with total amnesia of the event . Eyes remain open.
- These seizures are more frequently observed during the early part of focal epileptic seizures , especially those arising fromthe temporal lobe and in patients with typical «absences» seen with 3 Hz spike and waves.

Autonomic seizures

- **Autonomic Seizures.**

- Seizures that show objective autonomic signs such as ictal taquicardia or pilomotor seizures. The latter have a somatotopic distribution and may show «Jacksonian march».

- There are other autonomic signs that are observed as part of temporal lobe seizures such as :
- a)Ictal vomiting , probably originating in the insula with no definite lateralizing value.
- b) Ictal spitting, with no lateralizing value.
- c) Ictal hypersalivation , which is more frequent in seizures originating at the non dominant hemisphere.

Motor seizures

- **Simple motor seizures.**

- **a)Myoclonic seizures.**Short muscle contractions lasting less than 200 ms more often seen in generalized epilepsies.

- **b) Tonic seizures .**Sustained muscle contractions , usually lasting several seconds that lead to «posturing» . Tonic seizures in patients with focal seizures preferentially affect proximal muscle groups, may be uni or bilateral and tend to be asymmetric.
- Tonic seizures occur most commonly in frontal lobe epilepsy and very rarely in temporal lobe epilepsy.
- Tonic seizures arising from supplementary sensorimotor area show bylateral, usually asymmetric contraction of proximal musculature, sudden onset, short duration (10 – 20 seconds), frequently preserved consciousness and preferential occurrence during sleep. Most often all four limbs are involved with abduction of the upper limbs and asymmetric flexion of the elbows.
- If tonic seizures are clearly unilateral, they have a high lateralizing value pointing to a contralateral seizure onset.

Motor seizures

- **c)Clonic seizures .**Myoclonic contractions that recur regularly at a rate of 0.2 – 5.0 per second.The symptomatogenic zone is usually the primary motor strip.The epileptogenic zone is usaully at , or in close proximity of the primary motor strip when clonic seizures without alteration of consciousness is the first sign of the seizure.

- **d)Versive seizures.** Forced and involuntary turning of the head and eyes in one direction with associated neck extension resulting in a sustained and unnatural position. The frontal eye fields are the symptomatogenic zone.

- Versive seizures can be the first sign or appear earlier in frontal lobe epilepsies opposed to temporal lobe epilepsies.

- Versive seizures have a high lateralizing value to the contralateral hemisphere, especially when they occur within 10 seconds before secondary generalization.

Motor seizures

- **Complex motor seizures.**
- Seizures with complex movements that resemble normal movements or behavior.
- **a) Hypermotor seizures.**Complex movements involving the trunk and proximal segments of the limbs. This results in large movements that may look violent . Movements may be repetitive and stereotyped (e.g pedaling).Consciousness may be preserved during the seizure. They occur mostly during sleep. Most originate from the orbital or mesial frontal region. However, they may also ocurr in temporal lobe and insular epilepsies.Ocasionally, hypermotor seizures resemble sexual activity , like violent writhing, thrusting and rhytmic movements of the pelvis.
- **b)Automotor seizures.**Automatisms involving the distal segments of the hands, feet, mouth and tongue. Automotor seizures are typical of temporal lobe epilepsies ,but can occasionally be seen in frontal lobe epilepsy. Unilateral automatisms have ipsilateral lateralization value when they appear together with contralateral dystonia . Preservation of consciousness may rarely occur in patient whith non dominant mesial temporal epilepsy.
- **c)Gelastic seizures.** The main manifestation is laughing. In about half of the cases an hypothalamic hamartoma is found. Other symptomatogenic zones may be the anterior cingulate cortex and frontal, temporal or parietal lobes.

Additional lateralizing signs.

- **Dystonic posturing.** Sustained (>10 sec) , forced, unnatural positioning of an upper extremity on one side of the body with a clear rotational component. It is a reliable contralateral lateralizing sign in temporal lobe epilepsy. This sign can less frequently occur in extratemporal epilepsies.
- **Ictal speech.**Presence of clearly intelligible speech in a patient that is unresponsive and/or has clear distal automatisms. It has a «soft» lateralizing value to the non dominant hemisphere in patients with temporal lobe epilepsy.
- **Post ictal aphasia.**Is a lateralizing sign to the language dominant hemisphere in temporal lobe epilepsy or in frontal lobe epilepsy with ipsilateral temporal lobe propagation.
- **Todd´s paralysis.**Highly contralateral lateralizing sign.
- **Post ictal nose wipe.** Ipsilateral lateralizing sign in temporal lobe epilepsy.
- **Unilateral eye blinking.** Infrequent, apparently good ipsilateral lateralizing sign.
- **Ictal nystagmus.** Fast phase lateralizes to the contralateral hemisphere .
- **Asymmetric ending .** Ipsilateral latearlizing sign.
- **Peri- ictal urinary urge.** Non dominant hemisphere lateralizing sign.

- **Seizure sequence.**
- Most seizures consist of symptoms that evolve in a sequence due to seizure spreading.
- An accurate semiological description is obtained by taking each of the considered seizures as a component of the sequence and linking them, usually with an arrow . For example : Left visual aura → left hand clonic seizure→ generalized tonic clonic seizures.

- **Using semiology to diagnose a focal epileptic syndrome.**
- Independent analyisis and clasification of seizure semiology.
- Caveats :
  - Seizure semiology obtained by clinical history may be inaccurate or incomplete when compared with video recordings.
  - Seizure semiology reflects activation of the symptomatogenic zone and it can be the result of ictal spread from a more distant, non eloquent epileptogenic zone.
- Adequate epileptic syndrome diagnosis requires also an independent analysis of EEG, structural an functional imaging , clinical data, other molecular or genetic studies , etc. Only then should all these findings be integrated in a syndromatic diagnosis.

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