Semiology in generalized epilepsy

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연세의대

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Semiology is a keystone of epilepsy diagnosis

- Diagnosis of Epilepsy is based on thorough history
  - ✓ Requires 2 unprovoked seizures.
  - ✓ EEG and MRI are important but only supportive evidences.
  - ✓ Semiology is essential in epilepsy diagnosis, classification, and basement of appropriate treatment.

Semiology?: a branch of linguistics concerned with signs and symptoms.

International Classification of Seizures

ILAE Seizure Classification 1981

I. Partial Seizures
- 1. Simple partial Sz
  - with ▪ motor signs, sensory symptoms
  - ▪ autonomic symptoms & signs, psychic symptoms
- 2. Complex partial Sz
  - simple partial onset f/b impairment of consciousness (IOC)
  - IOC at onset: ▪ IOC only, ▪ IOC and automatisms
- 3. Partial Sz evolving into secondarily GTCS

II. Generalized Seizures
- 1. (clonic) tonic-clonic 2. absence 3. myoclonic
- 4. tonic 5. atonic 6. clonic

III. Unclassified

Semiological Seizure Classification (Lüders et al., 1998)

❖ **Aura**
- somatosensory • visual • auditory
- gustatory • olfactory • autonomic
- abdominal • psychic

❖ **Autonomic Sz (documentation of autonomic dysfunction)**

❖ **Dialeptic Sz (predominant Sx is altered consciousness)**
- typical dialeptic Sz (consistent with typical absence Sz)

❖ **Motor Sz**
- simple motor: - myoclonic, - tonic, - clonic, - spasm - tonic-clonic, - versive
- complex motor: - hypermotor, - gelastic, - automotor

❖ **Special Sz (Szs difficult to classify into one of above 4 types)**
- atonic • astatic • hypomotor
- akinetic • negative • aphasic

Descriptive Terminology for Seizure (Blume, 2001; Fisher, 2017)

Epileptic spasm (formerly infantile spasm)
- sudden flexion, extension, flexion-extension of predominantly prox. muscles (longer than myoclonus, ~1s). Limited forms: grimacing, head nodding... Infantile spasms are the best known form, but spasms can occur at all age

Myoclonic seizure
- sudden, brief (<100 ms) involuntary single or multiple contractions of muscle or muscle group.

Tonic seizure
- sustained muscle contraction lasting a few seconds to min.

Atonic seizure
- sudden loss or diminution of muscle tone without apparent preceding myoclonus or tonic event lasting 1-2 s.

clonic seizure
- Jerking, either symmetric or asymmetric, that is regularly repetitive and involves the same muscle groups

Astatic seizure
- loss of erect posture that results from an atonic, myoclonic, or tonic mechanism (synonym: drop attack).

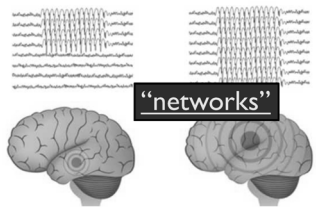
Dystonic
- Sustained contractions of both agonist and antagonist muscles producing athetoid or twisting movements, which may produce abnormal postures

Revised Classification of E. Seizures (ILAE, 2010)

Classification of Seizures

Generalized seizures: Szs originating and rapidly engaging bilaterally distributed networks

- Tonic–clonic (in any combination)
- Absence
  - Typical
  - Atypical
  - Absence with special features
    - Myoclonic absence
    - Eyelid myoclonia
- Myoclonic
  - Myoclonic
  - Myoclonic atonic
  - Myoclonic tonic
- Clonic
- Tonic
- Atonic



Focal seizures: Szs originating within networks limited to one hemisphere

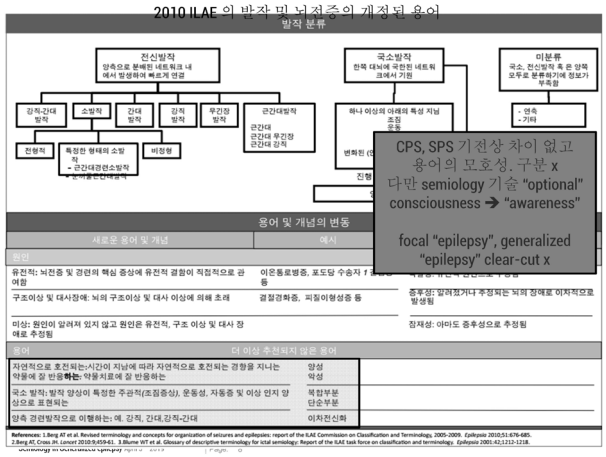
Unknown

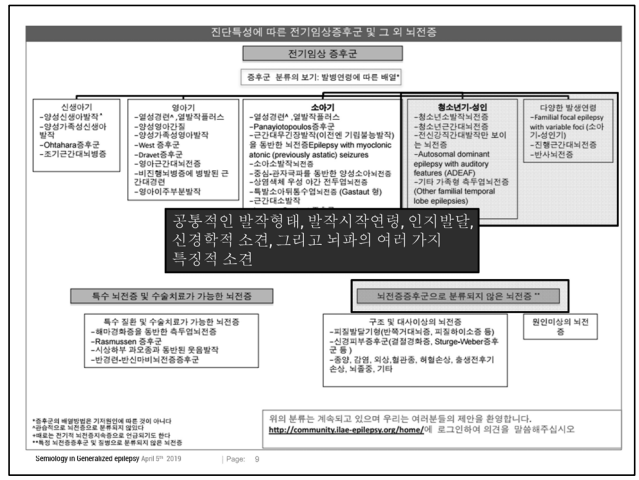
- Epileptic spasms

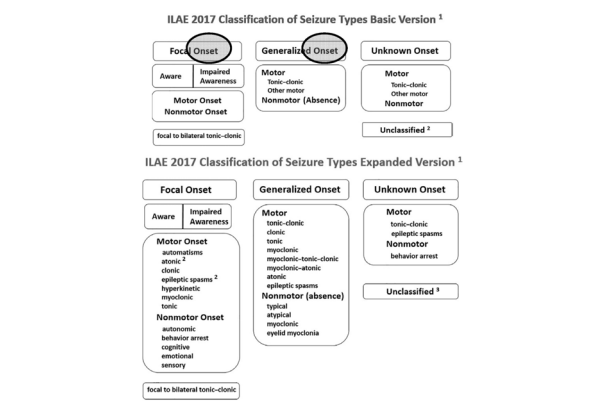
Key points in modelling of brain networks in epilepsy

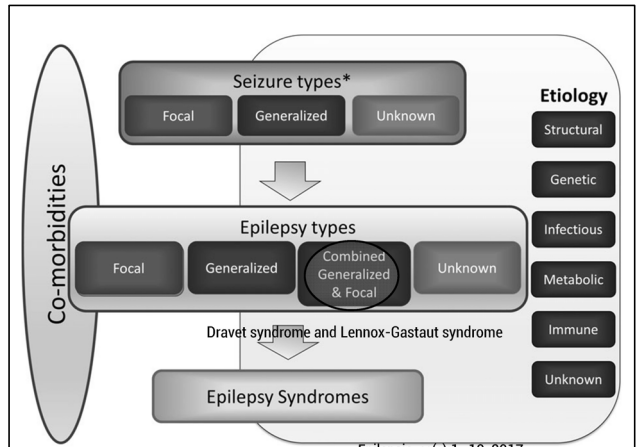
- Seizures emerge rapidly **from normal brain states** and usually self-terminate
  - Neuronal machinery causing seizures is the same machinery present in brain during normal function ("Dynamic" rather than "static")
- **Clinical expression** of seizures necessarily requires the involvement of large scale brain networks to create "symptom"
  - Seizure onset from different brain regions can produce seizures that appear identical
  - "common pathways" necessarily involve large-scale brain networks
- Concept of a **'seizure focus'** is **less clearcut**!
  - Epilepsy **cure** after surgery **is not simple**
  - 22% Pts with "good" postsurgical outcome have SPS despite removal of 'Sz onset zone'; 31% of all op. pts have complex pattern of remission and relapse
    Lancet 2011;378:1388
  - In many pts., there is no single focal region which drives seizure onset, but rather complex networks!

2010 ILAE 의 발작 및 뇌전증의 개정된 용어





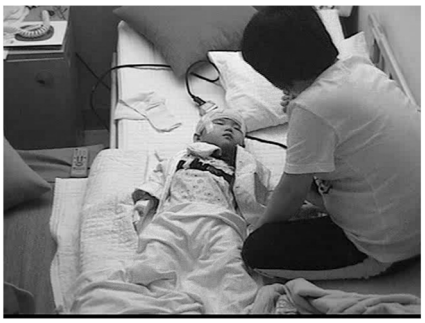




Epilepsia, **(*):1–10, 2017
doi: 10.1111/epi.13709

Cases

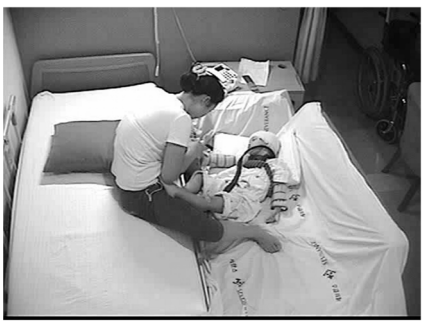
M/9 m



- Epileptic spasm (former infantile spasm):
  sudden flexion, extension, flexion-extension of predominantly prox. muscles (longer than myoclonus, ~1s)
- Epileptic spasm:
  Usually in cluster, flexor spasm is most common.
  Head nodding is common.

West syndrome

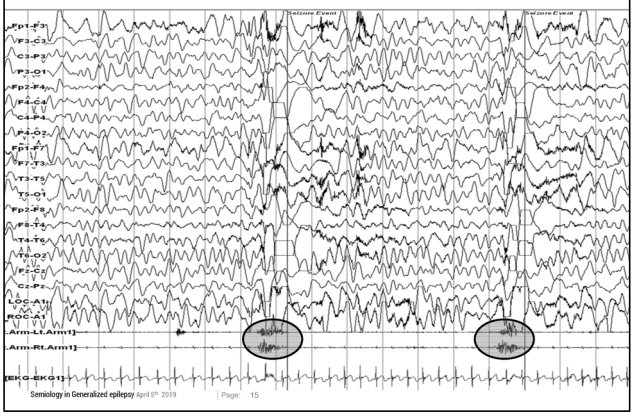
M/2 yrs



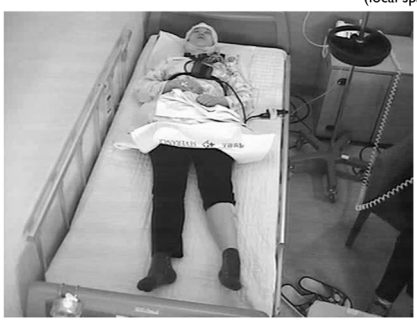
- Epileptic spasm:
  Asymmetric spasm is not infrequent.
  Sometimes it shows complex movements.

West syndrome

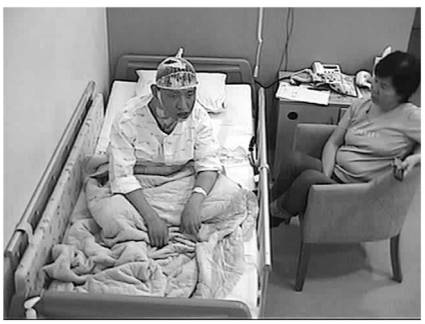
EEG



spasms can occur at all age
(focal spasm in leg)



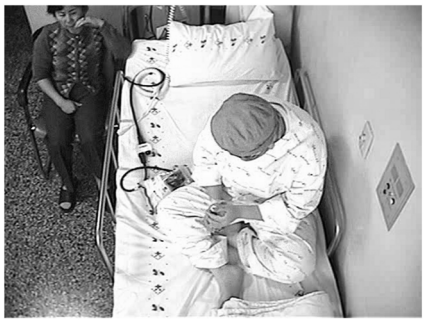
M/19



- Tonic seizure:
  sustained muscle contraction
  lasting a few secs to min.

Lennox-Gastaut syndrome

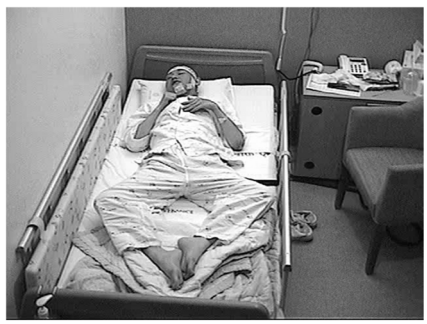
F/29



- Tonic seizure:
  The most frequent seizure types of "drop-attack seizure."

Lennox-Gastaut syndrome

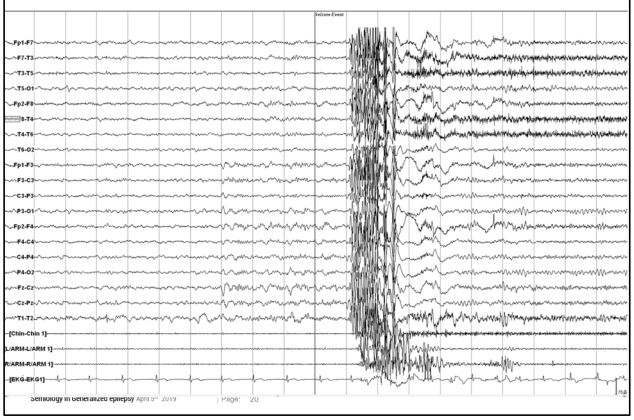
M/19



- Gen Tonic seizure:
  Bilateral symmetric tonic contraction of somatic muscles.

Lennox-Gastaut syndrome

EEG



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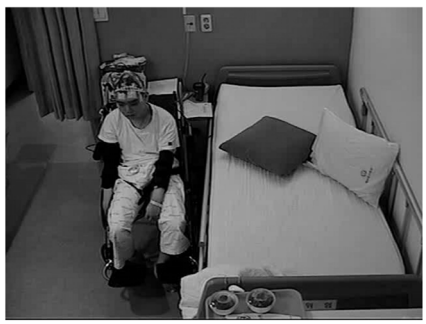


- Gen Tonic-clonic seizure:
  Bilateral symmetric tonic contraction of somatic muscles and then bilateral clonic contraction of muscles.

Lennox-Gastaut syndrome

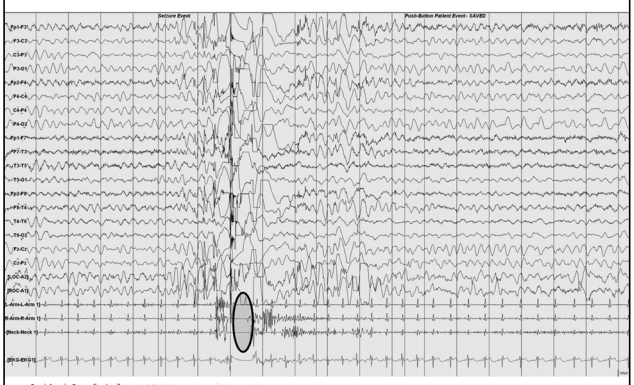
F/14 m

- Atonic seizure:
  Sudden loss or diminution of muscle tone without apparent preceding myoclonus or tonic event lasting 1-2 s.

  unexpected and highly vulnerable to head trauma.

Lennox-Gastaut syndrome

EEG



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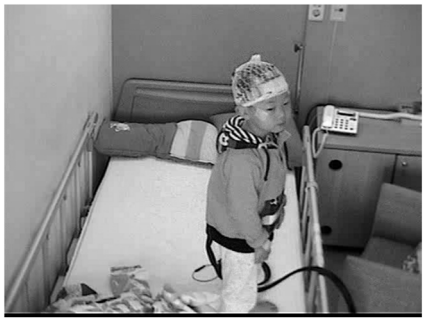

- Astatic seizure:
  Loss of erect posture that results from an atonic, myoclonic, or tonic mechanism
  (synonym: drop attack).

Lennox-Gastaut syndrome

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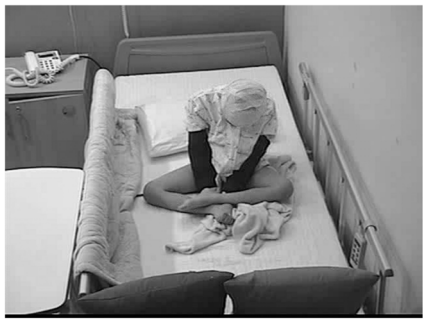
M/11



- Myoclonic-astatic seizure:
  Sudden, brief myoclonus followed by astatic seizure.

Doose syndrome (EMAS)

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F/19

- Myoclonic seizure in JME:
  Have a striking circadian features: occurs within an hour of awakening.
  Sleep-deprivation, photo stimulation, and alcohol is important precipitating factors.

Juvenile Myoclonic Epilepsy

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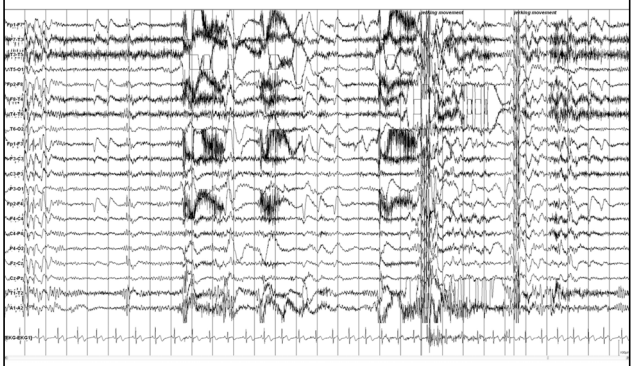
EEG



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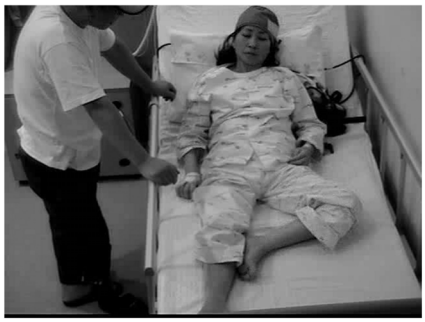

- Lance-Adams syndrome:
  Chronic persistent myoclonus and negative myoclonic seizures, usually in the hypoxic-ischemic encephalopathy.

Lance-Adams syndrome

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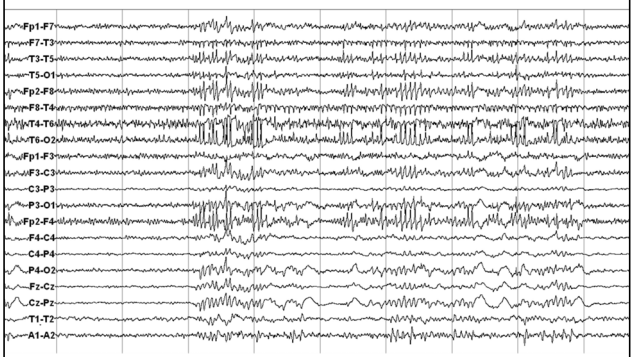

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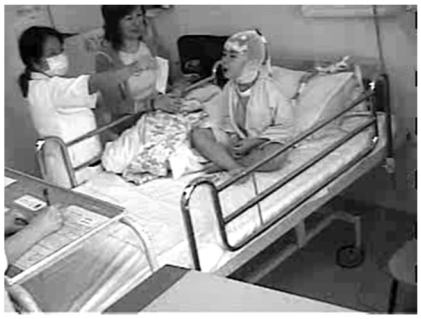
M/6



- Dialeptic seizure:
  Sudden loss of ongoing activity, brief loss of consciousness.
  It can be easily precipitated by hyperventilation (~95%) in CAE.

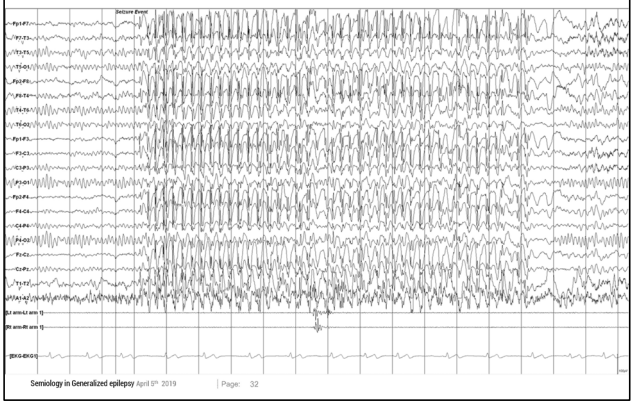
Childhood Absence Epilepsy

F/9

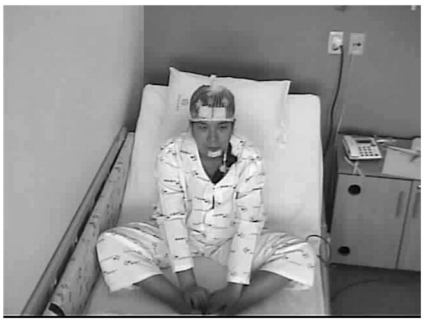
- Dialeptic seizure:
  Subtle automatism (e.g. lip smacking) and incomplete LOC can be easily found in CAE.
  Age 4-8 is the most common.

Childhood Absence Epilepsy

EEG



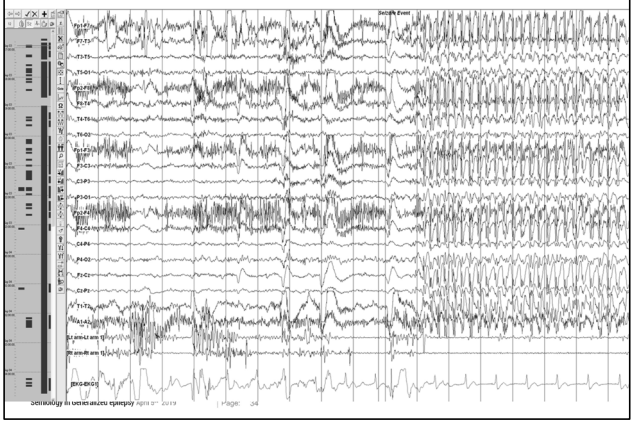
F/25



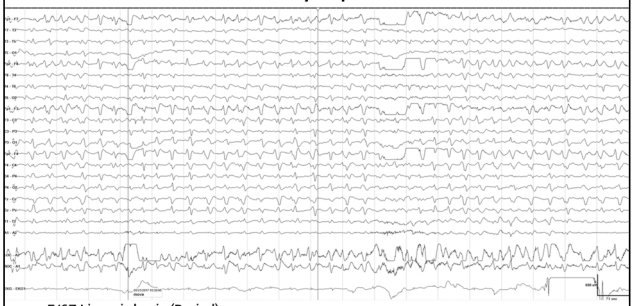
- Dialeptic seizure:
  Longer duration, more common automatism, and more frequent incomplete LOC are typical features of JAE compared to CAE.

Juvenile Absence Epilepsy

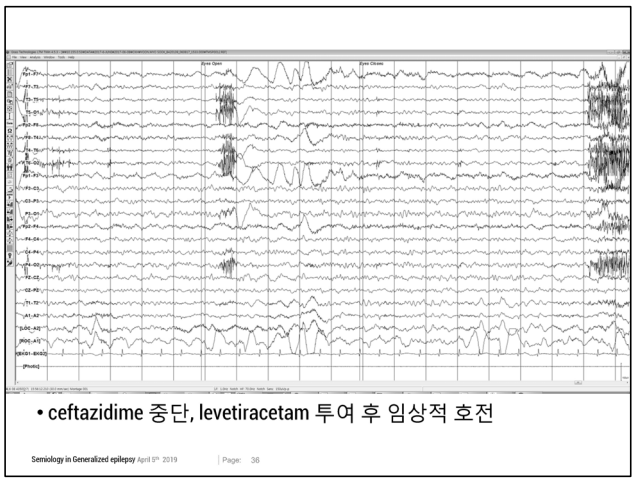
EEG



case 1: Generalized NCSE in critically ill patient



- F/67 Liver cirrhosis (B-viral)
- Spontaneous bacterial peritonitis
- ceftazidime (5 days) meropenem (10 days, stopped 6 days ago)
- **drowsy mental status;** r/o hepatic encephalopathy, Ammonia 교정후에도 의식 호전이 없다 MRI: nonspecific



• ceftazidime 중단, levetiracetam 투여 후 임상적 호전

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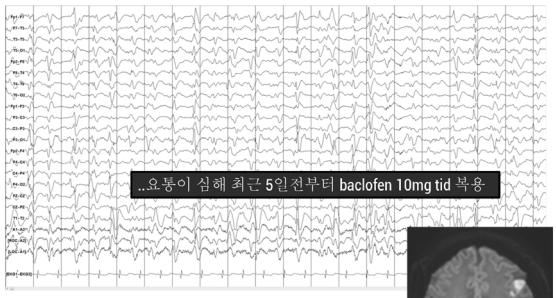
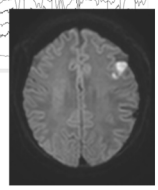
case 2: Generalized NCSE in critically ill patient



..요통이 심해 최근 5일전부터 baclofen 10mg tid 복용



- F/74
- ESRD on HD, s/p cholecystectoimy
- 이틀째 의식 저하 (drowsy)로 내원, stroke 으로 입원
- 3일간 점점 의식 악화되어 **coma**

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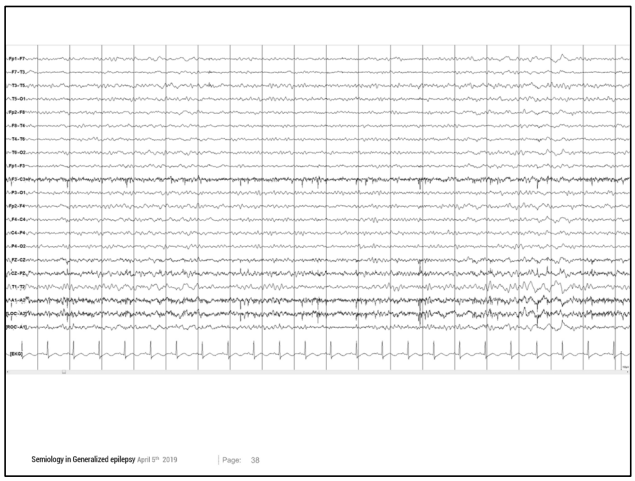

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