Case-Based Learning Epilepsy

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Case

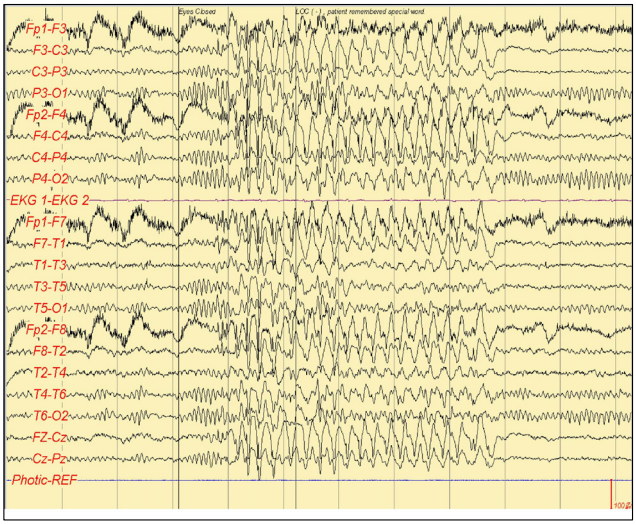
- F/31
- Chief Complaint: easy forgetfulness (치매클리닉)
- Present Illness
  - 2년전부터 slow progressive
  - 건망증이 심하고 어휘력이 떨어지는 것 같다.
  - 간혹 멍해질 때가 있다. LOC: denial
  - 밥이나 약을 먹었는지 기억이 안난다.
  - 1년전부터 일본 생활 (남편이 선교사) 우울증이 있는 것 같다.

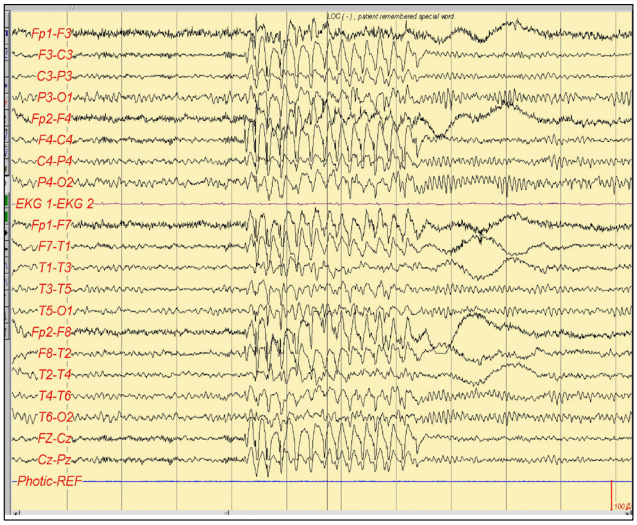
Case

- Past medical history of seizures: denial
- Family history of seizures: denial
- 대학원 졸업, 주부, 자녀 2명, 모유 수유 중
- Impression
  - R/O early onset dementia R/O depression
- Diagnostic plan
  - Neuropsychological test for young adults
  - Simple waking EEG

Results of NPTs

- K-MMSE: 25/30
- 우선 memory면에서 verbal & visual memory의 encoding & storage dysfunctions이 뚜렷한 소견을 보임.
- Frontal/executive functions에서 mental set maintenance & shifting dysfunction, motor programming ability 저하 등의 motor dysfunctions, inhibitory control ability 저하, semantic word fluency 저하 소견을 보임.
- 성격/행동 면에서 약 1년 전부터 irritable한 성향을 보이는 것으로 보고됨. 보호자가 보기에는 다소 우울해 보인다는 보고가 있으나 기분 평가 시 유의미한 depression 소견은 보이지 않음.
- 결론: memory impairment 와 frontal/executive dysfunction 이 뚜렷한 상태로 bilateral frontal & temporal lobe dysfunction 이 의심됨





Diagnosis

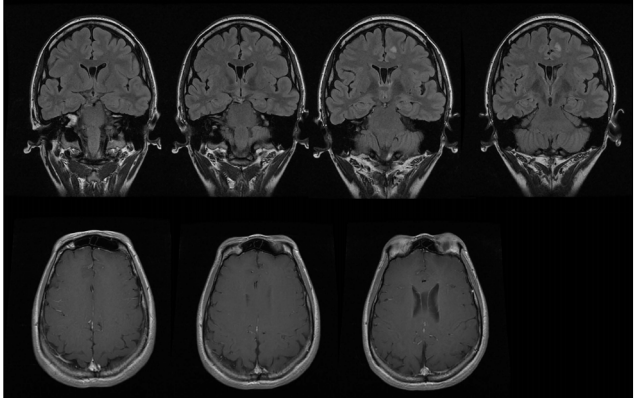
- EEG: brief runs of 4Hz generalized (spike-and) waves
- Diagnosis:
  - R/O Atypical absence epilepsy
- Plan:
  - Levetiracetam 500mg bid
  - Brain MRI?

MRI in Epilepsy

- The nonacute situation: Ideal practice
  - The ideal practice is to obtain structural neuroimaging with MRI in all patients with epilepsy except in patients with a definite electroclinical diagnosis of IGE such as CAE and JME
  - MRI is particularly indicated in patients with
    - Onset of seizures at any age with evidence of a partial onset on history or EEG
    - Onset of unclassified or apparently generalized seizures in the first year of life or in adulthood
    - Evidence of a focal fixed deficit on neurological or neuropyschological examination
    - Difficulty in obtaining control of seizures with first-line AED treatment
    - Loss of control of seizures with AEDs or a change in the seizure pattern that may imply a progressive underlying lesion.

ILAE, recommendation for Neuroimaging in Epilepsy, 1997 8

MRI



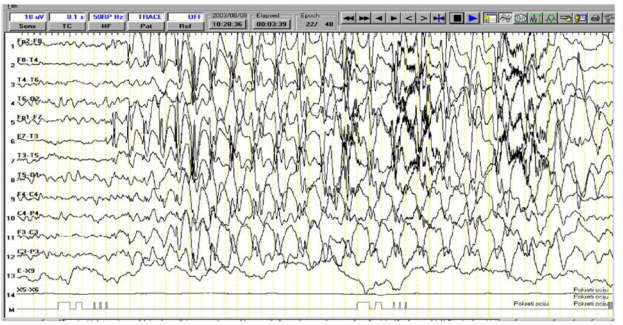
R/O Dysembryoplastic neuroepithelial tumor (DNET)

Secondary Bilateral Synchrony?

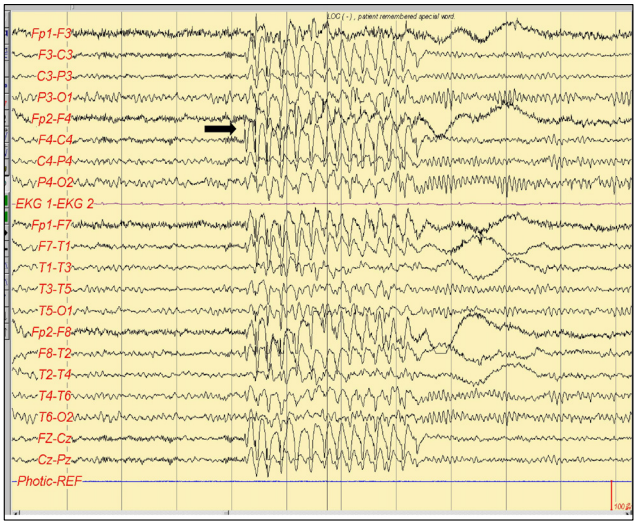
- Generalized interictal epileptiform activity induced by a focal epileptogenic lesion
  - Epileptic focus triggers a mirror image cortical area by transcallosal transmission or through thalamic area that in turn produces a bilaterally synchronous epileptiform discharge
  - Characteristics of SBS
    - They are more often less than 2.5Hz when rhythmic
    - They demonstrates considerable morphological variability from complex to complex
    - They usually contain a single site of phase reversal in transverse bipolar montages
    - They may be consistently asymmetrical
    - Consistently focal epileptiform spikes or sharp waves may be present

From Fisch BJ, EEG Primer 10

6 YO Lt FLE (Frontal Absence)



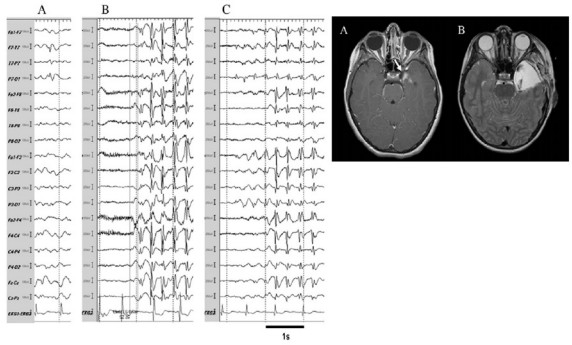
Jocic-Jakubi B et al., Seizure, 2009 11



Clinical Follow-Up

- Clinical course
  - F/U 2 yr
  - 처음에는 많이 좋아진 것 같더니 지금은 조금 좋아진 상태 유지.
  - 약 먹고 졸려하진 않는다.
  - 수술적 치료, further W/U 거부

Generalized Spike-and-Wave Complexes from Focal Epilepsy



Kakisaka et al., Epilepsy & Behavior, 2010

Generalized Spike-and-Wave Complexes from Focal Epilepsy

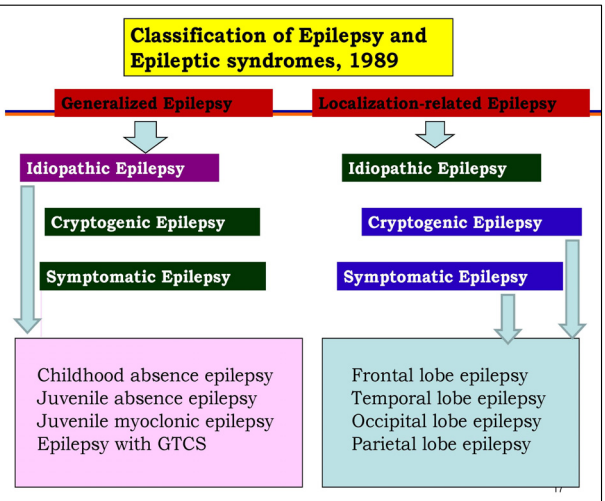
- Successful surgery for epilepsy due to early brain lesions despite generalized EEG findings
  - Wyllie et al. Neurology, 2007
  - 50 pediatric patients, 30-100% epileptiform discharge generalized, contralateral maximum, or contralaterally localized
  - 90% congenital, perinatal, or acquired during early infancy
  - 72% seizure-free
  - The generalized and contralateral epileptiform discharges may be a manifestation of potentially reversible secondary epileptogenesis resulting from an interaction between the early lesion and the developing brain

Classification of Seizure 2010

Generalized seizures
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
Unknown
Epileptic spasms

16

Classification of Epilepsy and Epileptic syndromes, 1989



1989 Epilepsy Syndrome Classification

- Idiopathic: Age-related onset, clinical, and electrographic characteristics, and presumed a genetic etiology
- Symptomatic: A known or suspected disorder of CNS
- Cryptogenic: Presumed to be symptomatic the etiology is not known

18

2010 Classification of Epilepsies

- Dichotomy of focal versus generalized has been abandoned: To separate the manifestation from the underlying pathology
- **Causes**

Idiopathic => Genetic

Symptomatic => Structural/metabolic

Cryptogenic => Unknown

as yet unknown

19

Advocates of 2010 Classification

- Reflecting scientific breakthroughs in genomic, neuroimaging, and neurophysiology
- More transparent language: Abandoning terms such as “idiopathic”, “cryptogenic” and “symptomatic”
- Aligning “idiopathic” with “benign” was discarded: ex) Dravet syndrome
- Distinction between CPS and SPS was discarded:
  - Wild guess in childhood epilepsy
  - Driving issue should be decided on individual basis
  - No contribution in drug trial
- Keeping it as simple as possible but not simpler
- Artificial grouping of generalized epilepsies was abandoned
- Churchill “It may not be a great system, but we don’t have better one”
- It is not a closure

Berg AT, 2011

20

Cons of 2010 Classification

- Genetics are never simple: A case with gene and trauma, genes with less strong association with epilepsy
- No subtypes of focal epilepsy: Genes, clinical features, EEG findings distinguish focal seizures arising in frontal lobes with motor automatism and focal seizures arising in temporal lobe with auditory symptoms
- Preserving “focal” and generalized seizures, while abandoning “focal” and “generalized epilepsies
- No explanation about replacement of “partial” by “focal”: Focal conjures the image of a cortical dot
- Abandoning “CPS”:
  - Most parents can tell if the consciousness is preserved in their children
  - Does 5% overlap justify elimination of terms?
  - Dyscognitive cannot be used to illustrate impaired consciousness

Devinsky O, 2011

21

Take Home Message

- MRI should be taken in most of epilepsy patients
- EEG patterns of generalized seizures can be observed in patients with focal epileptogenic lesion as secondary bilateral synchrony
- In new epilepsy classification, dichotomy of focal versus generalized seizures has been abondomed.

22