Symptomatic epileptic syndrome

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Classification (ILAE 1989)

- Limbic epilepsies
  - Temporal lobe epilepsy
- Other types defined by location and etiology
  - Frontal lobe epilepsy
  - Parietal lobe epilepsy
  - Occipital lobe epilepsy

Temporal lobe epilepsy

- Heterogeneous group sharing same topographical S2 onset, often diverse etiology, age onset, prognosis and response to medical and surgical Rx
- Divided into originating from
  - Lateral TLE: more common (2/3 cases)
  - Mesial TLE
- Most common substrate mesial temporal sclerosis, others benign or malignant tumor, viral and other infection and parasitic disease, CVD, developmental malformation, trauma.

Temporal lobe epilepsy

- TLE comprises 30-35% of all epilepsies
- MTS account 65% of cases, mal and female equally affected
- Age onset depend on etiology
- Clinical manifestations
  - Simple partial seizures
  - Complex partial seizures
  - 2 GTCS
  - Focal non-convulsive status epilepticus
  - Secondarily convulsive status epilepticus

Temporal lobe epilepsy

- Ictal clinical manifestations
- Nearly 90% of cases experienced aura
  - Epigastric aura and fear are commonest of MTLE
  - Simple or complex auditory hallucinations mainly characterize LTLE
  - Mental hallucinations and illusions common in both MTLE and LTLE
  - Olfactory and gustatory hallucination less common

Temporal lobe epilepsy

- Subjective ictal symptom of TLE
  - Ascending epigastric aura
  - Experienced (mental or psychic) symptoms
  - Fear and panic
  - Déjà vu or Jamais vu and their variations
  - Auditory hallucinations and illusions
  - Olfactory and gustatory hallucinations
Temporal lobe epilepsy

- **Objective ictal symptom**
  - Usually occur when consciousness is impaired
  - Objective ictal symptom
    - Automatisms
    - Autonomic disturbances
    - Speech disturbances
    - Head and eye deviation as well as dystonic postures
    - Motor arrest with staring
    - Unilateral ictal paresis
    - Unilateral eyelid blinking
    - Ictal vomiting

- **Autonomic disturbance and ictal emeticus**
  - Cardiovascular symptom, mainly tachycardia and arrhythmias, less often bradycardia, asystole or hypertension
  - Brief respiratory arrest, sigh or gasp is common, hypopnea or hyperventilation may occur in late seizure
  - Mydriasis frequent symptom, miosis and hippus pupillae common
  - Feeling of shivering cold somite associated with piloerection, salvation is common
  - Flushing or pallor common encountered
  - Ictal emeticus is exceptional in adlt with TLE

- **Gelastic of TLE**
  - Ictal laughter is rare, attributed mainly to right
  - Age onset variable
  - Galastic seizures had reported produces stimulation of temporo-basal cortex
  - Clinical descriptions are variable
  - Laughter commonly accompanied by a break of contact
  - In 50% of cases has other types of seizures

- **Postictal symptom**
  - Common, often characteristic and may be value in lateralizing TLE
  - Postictal symptom may disproportionately more severe than the ictal symptom and may last for hours

**Diagnostic procedure**

- MRI brain most importance diagnostic test
  - High resolution MRI may detect in 90% of cases
- MRS offer variable insights
- PET and MRR helpful for localization
- EEG
  - Interictal EEG 1/3 has classic spikes or SW by anterior temporal electrode, it yield increased with prolonged EEG
  - In LTLE spike and SW may seen in middle temporal electrode
  - Other 2/3 have normal routine EEG or shows non-specific abnormalities with excess slow in one temporal area
  - Ictal EEG no significant between MTLE and LTLE consist regional ipsilateral rhythmic 4-7 Hz activity

**Differential diagnosis**

- Typical TLE consisting rising epigastric sensation, fear and progression to impairment of consciousness with oro-alimentary automatisms
- DDX from FLE and other extratemporal seizures
  - Although various ictal symptom of TLE, but vast majority of TLE offer no difficulty in DDX if clinical symptom properly analyzed
  - Single ictal symptom may predominance in one or another, but no contribution diagnosis i.e head deviation can found in seizure from any location, but together with elementary visual hallucination suggested DLE, with epigastric aura—TLE, stereotypical and rather violent jerk—FLE
  - Dystonic postures common in FLE or TLE, but FLE-dystonic very early 1st symptom, brief and without impair severe LOC, mainly during sleep
Prognosis
• Prognosis is largely, cause related
• Familial TLE is usually mild
• Hippocampal TLE take a progressive course that can be halted by appropriate neurosurgical procedures
• However, even with same cause, prognosis vary from mild to severe
• In community studies about 10-40% of TLE may go to remission

Management
• Management comprises AEDs and neurosurgical excision of epileptogenic lesion
• Recent practice for TLE and localized neocortical resection conclude
  — Benefit of antero-mesial temporal lobe resection for disabling CPS is greater than AEDs and risks are comparable

The syndrome of mesial TLE
• Strong evidence for progressive behavioral changes especially increasing memory deficit, and increase in contralateral interictal spikes
• Silence period, if present, suggests progression
• Hippocampal atrophy and cell loss may correlate with duration of epilepsy
• All studies are cross-sectional, need longitudinal studies to document progression

Etiology
• Hippocampal sclerosis, pathological substrate
• It consists of hippocampal neuronal loss in a characteristic pattern, predominantly involving the hilar region, CA1 and dentate gyrus, with relative sparing of CA2 (Babb and Brown 1987)
• Cause is unknown, A high incidence of complicated febrile convulsions among patients with hippocampal sclerosis (Falconer 1971; French et al 1993; Maher and McLachlan 1995)

Hippocampal sclerosis
**Etiology**

- Patients without this history often had other cerebral injuries early in life (Mathern et al. 1995; O'Connor et al. 1996)
- Increasing evidence that the epileptogenic process, once begun, is progressive (Beach et al. 1995; Mathern et al. 1995)

**Epidemiology**

- CPS are the most common seizure type, with 40% of epileptic patients reporting such ictal events (Gastaut et al. 1975)
- Not all complex partial seizures, however, are of mesial temporal lobe origin
- Approximately 40% of these seizures originate elsewhere

**Differential diagnosis**

- Partial seizures with impairment of consciousness can originate from any brain area
- Not all mesial temporal onset seizures are due to hippocampal sclerosis (French et al. 1993; Williamson et al. 1993)
- No definitive characteristics distinguish CPS reflecting mesial temporal lobe epilepsy from CPS generated from neocortex or due to mesial temporal lesions

**Differential diagnosis**

- However, mesial temporal lobe epilepsy unlikely when:
  - Seizures; brief and frequent
  - Involve early ictal focal motor
  - Somatosensory
  - Visual or auditory signs and symptoms
  - Drop attacks
  - Often generalize
  - Associated neurologic or cognitive impairments
  - Interictal epileptiform EEG; extratemporal or bilaterally synchronous (Engel 1992; Wieser et al. 1993)
Investigations

• Interictal EEG typically include unilateral or bilaterally-independent anterior temporal spikes, best seen with basal (sphenoidal, earlobe, or true temporal)

• Regional temporal interictal slowing is seen ~ half of patients, and is of lateralizing value (Koutroumanidis et al 1998)

Investigations

• High-resolution MRI often demonstrates hippocampal atrophy on one side

• Ipsilateral hypometabolic temporal lobe on interictal PET (Engel et al 1990; Hajek et al 1993)

• Interictal SPECT unreliable

Investigations

• SPECT is valuable when used to demonstrate a characteristic ictal and postictal pattern (Newton et al 1994)

• Proton magnetic resonance spectroscopy enable to demonstrate decreased N-acetylaspartate in the sclerotic mesial temporal lobe (Hetherington et al 1995; Kuzniecky et al 1998)

Investigations

• Neuropsychological evaluation
  — commonly demonstrates memory dysfunction, which is material-specific to the involved hemisphere
  — related to degree of hippocampal cell loss (O'Rourke et al 1993; Hermann et al 1998)

• Ictal EEG
  — usually reveal a characteristic ictal onset consisting of rhythmic 5- to 7-Hz activity over one temporal lobe, either as the first electrographic change (initial focal onset), or within 30 seconds (delayed focal onset) (Risinger et al 1989)

Prognosis and complications

• Natural history of mesial temporal lobe epilepsy is not known

• When seizures become refractory to medical treatment they are unlikely to remit spontaneously. If the condition continues, increasing memory problems and other behavioral disturbances have been reported (Engel et al 1991)

• Could be a progressive epileptic disorder
Treatment

- Outcome of importance in refractory epilepsy
- Therapeutic alternative and long-term outcome in refractory TLE
  - Medical therapy
  - Surgical therapy

Surgery

- 2% medical, 0% surgical – one year follow up
- Death rate 5 times higher than population
  - Reverts if seizure-free after surgery
- Patient with > 1 seizure in preceding year
  - 81% of those who died
  - 42% of survivors

Surgery

- Recommendation
  - A: patient with partial seizures who failed to 1st line Rx should be referred to epilepsy surgery center
  - B: Patients who meet criteria for antero-mesial resection should be offered surgery
  - U: Insufficient evidence to recommend other neocortical resections

A: Definite, B: Probable, C: Possible, U: Unknown

Surgery

- Seizure freedom is achieved in:
  - Less than 10% of medically treated patients
  - Approximately 60% of surgically treated patients
- In all long-term follow-up studies, the benefits of surgery are sustained
  - Seizures, quality of life, psychosocial aspects

Frontal lobe epilepsies

- FLE manifest with seizure originating from primary epileptic focus within frontal lobe
- Clinical vary and depend on origin and spread of epileptogenic focus
- Frontal lobe occupies 40% of cerebral cortex
- Frontal lobe subdivided into
  - Primary motor cortex
  - Pre-motor cortex
  - Prefrontal cortex
  - Limbic and paralimbic cortices

Frontal lobe epilepsies

- Complex and varied patterns in seizure spread explain variability in clinical and EEG manifestations
- Seizure arising from primary motor cortex and supplementary motor area (SMA) have been relatively well defined, but seizure generate in other region less well specified
Frontal lobe epilepsies

- Demographic data
- FLE may start at any age, accounting 1-2% of all epilepsies, though they are second in prevalence after TLE
- In prospective community-based study, prevalence FLE 22.5% (TLE 27%)

Clinical manifestations

- Various seizure patterns have been recognized
- Motor manifestations are more common and account about 90% of seizures
- Most common FLE
  - Seizure from motor cortex
  - Seizure from supplementary sensori-motor area (SMA)
  - Seizure from other frontal lobe region

Seizure from motor cortex

- Mainly are simple partial seizures, symptom depend on topography of the area involved
- In the lower pre-Rolandic area they may speech arrest, vocalization or dysphasia, tonic-clonic movements of contralateral face or swallowing
- In paracentral lobule: tonic movement of foot may unilateral or contralateral
- Seizure often progress to 2 GTCS, postictal Todd's paralysis frequent

Seizure from SMA

- Distinct and characteristic clustering of symptoms and stereotyped
- Hypermotor seizure of bizarre bilateral, asymmetric tonic posturing
- Characteristic hypermotor seizure of SMA: sudden and explosive bilateral and asymmetrical tonic posturing of limb often with contraversion of eyes and head, vocalization or speech arrest
- Fencing posture best known descriptive term for SMA, although may not be common, one arm is raised and semi-extended above head, while the other limb semi-flex at elbow

Other FLE of particular clinical interest

- Frontal absence
- Unusual force thinking and forced acts
- Gelasic seizure
- Negative motor seizure

Focal status epilepticus of frontal lobe origin

- Undetermined prevalence
- Manifest with prolonged impairment of consciousness and inappropriate behavior
- Symptom fluctuate in intensity and severity over time
- Concurrent turning of head and focal jerking may occur
- Commonly end with 2 GTCS
- Ictal EEG show repetitive fronto-polar, fronto-central and fronto-temporal epileptiform discharges with unilateral emphasis
Frontal lobe epilepsy

- **Etiology**
  - 2/3 are symptomatic with cortical malformation 57.4% tumor 16.4% and trauma and other lesion 26.2%

- **Diagnosis**
  - MRI brain (high resolution) detect abnormalities 67% (79% in TLE)
  - EEG interictal and ictal surface EEG often unhelpful, often normal 50-60% particularly when seizure originate from medial frontal region
  - Prolonged VEM increase EEG yield

Epilepsia partialis continue of Kozhevnikov

- **Type of seizures with various heterogeneous condition**
- **Onset occur at any age, prevalence is extremely small, < 1/1 million**

- **Clinical**
  - Spontaneous regular or irregular clonic muscle twitching of cerebral cortical origin, sometime aggravated by action or sensory stimulus confined to one part of the body and continuing for a period of time

Parietal lobe epilepsy

- **Clinical seizure**
  - Somatosensory
  - Somatic illusions (disturbance of body image)
  - Vertiginous
  - Visual illusion or complex formed visual hallucinations
  - Receptive conductive linguistic disturbances

Frontal lobe epilepsy

- **DDx**
  - Frontal lobe hypermotor seizure should be differentiated from
  - Non-epileptic paroxysmal movement disorders such as
    - Psychogenic movement disorder
    - Familial paroxysmal dystonic choreoathetosis
    - Paroxysmal kinesigenic choreoathetosis
    - Episodic ataxia type 1
  - Sleep disorder
  - TLE
  - Symptomatic frontal absence may similar to absence

Epilepsia partialis continue of Kozhevnikov

- **Cause**
  - Chronic or acute encephalitis
  - Mitochondrial disorder
  - CJD
  - Metabolic disturbances
  - Lesional diseases of the brain
  - Drug

Parietal lobe epilepsy

- **Clinical usually related to epileptogenic location anterior or posterior, dominant or non-dominant hemisphere**
  - Onset with sensorimotor symptom usually associated ant parietal lobe
  - Post parietal: complex symptomatology
  - 50% of cases experienced > one type of seizure
Parietal lobe epilepsy

- Most common type 2/3 of cases
- Various type paraesthesia, dysesthetic and painful sensations such as tingling, tigh, crawling, numbness, thermal, burning, tickling, prickling, creeping electric
- Tingling most characteristic symptom 76%
- Location
  - The face mainly lip and tongue, hand and arm
  - 40% remain static, but somatosensory march may occur

Parietal lobe epilepsy

- Disturbance of body image and somatic illusion
  - 2nd most common ictal symptom
  - Dislocated posture, limb position of movement, feeling that extremity or body part is alien or absent, disassociation and misperception of location and body part identity
  - Most patient have paresthesia associated with illusion
  - Somatosensory inability to recognize the affected body
  - Illusion of movement
  - Other symptom
    - Tongue retractor (5% in PLE)
    - Visual illusions and complex formed visual hallucinations (12% in PLE), image may be larger or smaller, moving
    - Complex disproportions affect dominant PLE
    - Temporary motor seizures and hemiplegia

Parietal lobe epilepsy

- Seizure spreading to extra-parietal regions
  - Simple focal seizure often spread to extraparietal region producing unilateral focal clonic convolution (57%), head and eye deviation (41%) tonic posturing (38%) and automatism (21%)
- Duration
  - Few seconds to 1-2 minutes
- Post-ictal are usually short. Todd’s paralysis (22%) and dysplasia (7%) may be common

Diagnostic procedures

- Interictal EEG may be normal or misleading, 2nd bilateral synchrony common (32%), interictal spikes if present need interpreted cautiously
- Ictal EEG normal in 80% in simple focal sensory seizures
- Localized parietal seizure: rare (11%)
- Postictal EEG may sometime localizing
- Ictal SPECT is very helpful, need rapid injected of the radiotracer

Occipital lobe epilepsy

- May start at any age, OLE account 5-10% of all epilepsies
- In neurosurgical prevalence is 5%
- Clinical manifestation
  - Cardinal symptoms include
    - Elementary and less often complex visual hallucination
    - Blindness
    - Visual illusion
    - Pallinopsia
    - Sensory hallucinations of ocular movement
  - Ictal objective oculomotor symptom are
    - Temporal deviation of the eye
    - Escape from movement of oculomotor
    - Repetitive ocular fixation or eyeblinking

Occipital lobe epilepsy

- Elementary visual hallucination
  - Most common, mainly colored and circular, developed rapidly within seconds
  - Appear in periphery of temporal field become larger and multiplying in course seizure
Occipital lobe epilepsy

- Complex visual hallucination
  - May take the form of persons, animals, objects, figures and scenes
  - May be familiar or unfamiliar
  - Small or larger in hemifield or center, or whole field
- Visual illusions
  - Misinterpretation, false perceptions in size, dimension, shape, position, etc.
- Palinopsia
  - Persistent or recurrent visual image after the exciting stimulus has been removed
- Ictal blindness
  - May follow visual hallucination and progress to other epileptic symptom

Occipital lobe epilepsy

- Tonic deviation of eyes, ocular seizure and epileptic nystagmus
  - Tonic deviation of eyes often, followed by ipsilateral turning of head
  - Ictal nystagmus mainly horizontal, quick phase opposite to the epileptic focus
- Repetitive eyelid closure, eyelid fluttering and eyelid blinking
  - Usually occur at phase of visual hallucination when consciousness impaired
  - However, it may occur alone
- Consciousness is not impaired during the elementary hallucination

Occipital lobe epilepsy

- Ictal or postictal headache
  - Frequent associated with OLE, severe, unilateral throbbing headache 1-2 minutes later, usually associated with vomiting
- Seizure spreading
  - Seizure may spread to other more anterior regions generated symptom from temporal, parietal and frontal lobe seizure, secondarily hemi- or generalized convulsion
  - Infra-calcarine occipital foci will propagate to temporal lobe causing CPS, while supra-calcarine foci propagate to parietal and frontal area giving rise to motor seizure

Occipital lobe epilepsy

- Etiology
  - Lesion may be congenital, residual or progressive resulting from vascular, neoplastic, metabolic, hereditary, congenital, parasite, systemic disease and infection
  - Malformations of cortical development are a common cause
  - Coeliac disease and OLE

Occipital lobe epilepsy

- EEG is essential but certain limitation
  - Interictal EEG usually abnormal with posterior slow wave
  - Unilateral spikes or fast multiple spikes occipital paroxysmal occur
  - May be photosensitivity
  - Ictal EEG
    - Often in symptomatic OLE, ictal discharge is more widespread rather than precise occipital localization
    - No postictal localized slow activity
    - 30% of cases ictal EEG does not show any change
  - Ictal SPECT is very useful