

# Typical semiology in extra-temporal lobe epilepsy



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It is crucial to know the semiology of extra-temporal lobe epilepsy during the diagnosis of epilepsy. Extra-temporal lobe epilepsy can be classified as frontal lobe epilepsy, which is the most common epilepsy other than temporal lobe epilepsy, parietal lobe epilepsy, and occipital lobe epilepsy. In this lecture, various semiology of extra-temporal lobe epilepsy will be introduced in terms of new ILAE seizure classification.

## Introduction

뇌전증의 발작형 (semiology)은 증상유발구역 (symptomatogenic zone)에 대한 정보를 알려준다.(Carreno and Luders 2008) 이는 뇌전증유발병소 (epileptogenic lesion), 자극구역 (irritative zone), 발작시작구역 (ictal onset zone), 기능이 상구역 (functional deficit zone) 등과 함께 뇌전증유발구역 (epileptogenic zone)을 예측할 수 있는 중요한 소견이다. 뇌전증유발구역을 찾는 것은 뇌전증 수술의 성공에 매우 중요한 역할을 하므로 뇌전증의 발작형에 대한 국소화 및 편측화의 의미를 알고 있는 것은 매우 중요하다.(Tufenkjian and Luders 2012) 실제로 편측화를 시사하는 발작형은 뇌전증 수술의 예후와 관련이 많은 것이 알려져 있다.(Boesebeck, Schulz et al, 2002)

하지만, 이런 발작형에 대한 훈련이 제대로 되지 않은 경우에는 발작형에 대한 서술의 정확도가 떨어질 수가 있으므로, (Heo, Kim et al. 2008) 이에 대하여 정확하게 파악하여야 한다. 발작형의 분류는 Hans Lüders가 제안한 방법이 있고 (표1), (Luders, Acharya et al. 1998, Loddenkemper, Kellinghaus et al. 2005) 또한 이에 대한 용어를 2001년에 ILAE에서 정리한

용어를 사용할 수도 있다.(Blume, Luders et al. 2001) 최근 한 연구자는 이 2가지의 발작형 분류를 비교하여 Lüders의 분류가 수술전 평가같은 특별한 경우에 발작유발구역을 확인하는데 더 효과적이라는 결론을 내리기도 하였다.(Hirfanoglu, Serdaroglu et al. 2017)

최근 ILAE에서는 그림1과 같은 체계에 따른 뇌전증 분류를 새로 제안하였다.(Scheffer, Berkovic et al, 2017) 발작에 대한 분류도 그림2, 3 및 표2와 같다. (Fisher, Cross et al. 2017, Fisher, Cross et al. 2017)

본 강의에서는 측두엽외뇌전증에서 나타나는 특징적인 발작 임상양상(semiology)를 소개하고자 한다.

- ETLE review논문(Dash and Tripathi 2014)
- Seizure semiology(Jan and Girvin 2008)

## 전두엽 뇌전증 (Frontal Lobe Epilepsy, FLE)

전두엽은 국소뇌전증 중 측두엽뇌전증 다음으로 흔하며 전체의 20-30% 정도를 차지한다. 하지만, 전두엽 뇌전증은 뇌전증이 아닌 다른 질환과 혼동되는 경우가 많다.

전두엽 뇌전증의 특징은 다음과 같다. (표 3.) (대한뇌전증 학회 2013)

1989년도에 ILAE를 참고하여 분류한 전두엽뇌전증의 일반적인 발작양상은 표 4와 같다.(1989, O'Muircheartaigh and Richardson 2012)

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표 1. Semiological seizure classification(Luders, Acharya et al. 1998)

**Epileptic seizure**

Aura

Somatosensory aura, visual aura, auditory aura, gustatory aura, olfactory aura, autonomic aura, abdominal aura, psychic aura

Autonomic seizure

Dialeptic seizure

Typical dialeptic seizure

Motor seizure

Simple motor seizure

Myoclonic seizure, epileptic spasm, tonic seizure, clonic seizure, tonic-clonic seizure, versive seizure

Complex motor seizure

Hypermotor seizure, gelastic seizure, automotor seizure

Special seizure

Atonic seizure, astatic seizure, hypomotor seizure, akinetic seizure, negative myoclonic seizure, aphasic seizure

**Paroxysmal event**

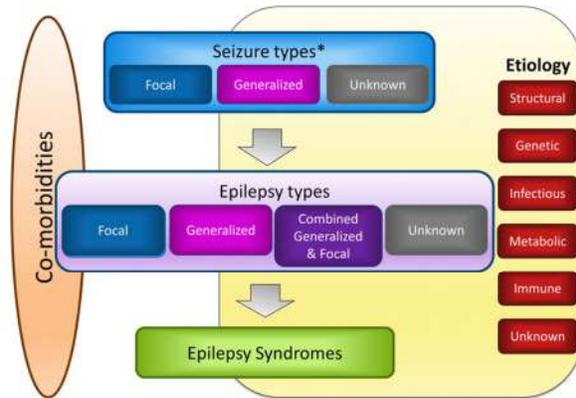


그림 1. 뇌전증 분류를 위한 체계(Scheffer, French et al. 2016)

전두엽뇌전증은 크게 가전두엽, 안와전두엽, 내전두엽 등으로 구분하므로 위의 발작양상에서, 보조운동영역 (supplementary sensorimotor area)와 전대상회 (anterior cingulate gyrus) 등에서 기인한 발작은 내전두엽 부위의 발작증상으로 구분할 수 있다. (Unnwongse, Wehner et al. 2012) 이러한 내전두엽 부위에서의 발작양상은 다음과 같다 (표 5). (Unnwongse, Wehner et al. 2012)

**두정엽뇌전증 (Parietal Lobe Epilepsy, PLE)**

두정엽뇌전증은 국소뇌전증의 5-6%를 차지한다. (대한뇌전증학회 2013) 두정엽 뇌전증은 일반적으로 단순 부분발작 및 2차성 전신발작이 특징적이다. (1989) 가장 흔한 조짐은 감각성 조짐으로 알려져 있고, 그 외에 감정, 현훈, 시각 조짐 등이 있는 것으로 알려져 있다. (Kim, Lee et al. 2004) 최근

**ILAE 2017 Classification of Seizure Types Basic Version 1**

Focal Onset		Generalized Onset	Unknown Onset
Aware	Impaired Awareness	Motor Tonic-clonic Other motor	Motor Tonic-clonic Other motor
Motor Onset Nonmotor Onset		Nonmotor (Absence)	Nonmotor
focal to bilateral tonic-clonic			Unclassified 2

<기본형>

**ILAE 2017 Classification of Seizure Types Expanded Version 1**

Focal Onset		Generalized Onset	Unknown Onset
Aware	Impaired Awareness	Motor tonic-clonic clonic tonic myoclonic myoclonic-tonic-clonic myoclonic-atonic atonic epileptic spasms	Motor tonic-clonic epileptic spasms
Motor Onset automatisms atonic 2 clonic epileptic spasms 2 hyperkinetic myoclonic tonic		Nonmotor (absence) typical atypical myoclonic eyelid myoclonia	Nonmotor behavior arrest
Nonmotor Onset autonomic behavior arrest cognitive emotional sensory			Unclassified 3
focal to bilateral tonic-clonic			

<확장형>

그림 2. IALE 발작 분류

의 한 연구에서는 두정엽 뇌전증의 다음의 조짐 등이 관찰되는 것으로 보고하였다 (표 6). (Francione, Liava et al. 2015)

**표 2.** 국소 발작동안 일어나는 행동에 대한 흔한 기술

Cognitive	Acalculia, aphasia, attention impairment, déjà vu or jamais vu, dissociation, dysphasia, hallucinations, illusions, memory impairment, neglect, forced thinking, responsiveness impairment
Emotional or affect	Agitation, anger, anxiety, crying (dacrystic), fear, laughing (gelastic), paranoia, pleasure
Autonomic	Asystole, bradycardia, erection, flushing, gastrointestinal, hyper/hypoventilation, nausea or vomiting, pallor, palpitations, piloerection, respiratory changes, tachycardia
Automatisms	Aggression, eye-blinking, head-nodding, manual, oral-facial, pedaling, pelvic thrusting, perseveration, running(cursive), sexual, undressing, vocalization/speech, walking
Motor	Dysarthria, dystonic, fencer's posture (figure-of-4), incoordination, jacksonian, paralysis, paresis, versive
Sensory	Auditory, gustatory, hot-cold sensations, olfactory, somatosensory, vestibular, visual
Laterality	Left, right, bilateral

**표 3.** 전두엽뇌전증의 특징

<p>뇌전증 발작의 종류</p> <ul style="list-style-type: none"> <li>• 국소간대운동발작 (focal clonic motor seizure)</li> <li>• 비대칭 강직발작 (asymmetric tonic seizure)</li> <li>• 전두엽운동과다발작 (frontal lobe hyperkinetic seizure)</li> <li>• 전두엽소발작 (frontal lobe absence seizure)</li> <li>• 전두엽덮개발작 (frontal opercular seizure)</li> <li>• 전형적인 내측두엽뇌전증발작과 유사한 전두엽뇌전증발작</li> </ul>
<p>뇌전증 발작의 임상양상</p> <ul style="list-style-type: none"> <li>• 군집성으로 빈번하게 발생하는 뇌전증발작</li> <li>• 짧고 상동적인 뇌전증 발작</li> <li>• 야간뇌전증발작</li> <li>• 갑작스럽게 시작하고 중단되는 뇌전증발작</li> <li>• 정신 조짐을 동반하지 않음</li> <li>• 발작후 혼돈을 동반하지 않음</li> <li>• 발작 초기부터 인지가 빠르게 소실됨</li> <li>• 양쪽 다리의 운동 자동증이 뚜렷함</li> <li>• 발작중자세(ictal posturing)와 강직연축이 뚜렷함</li> <li>• 기괴한 자동증, 종종 비뇌전증발작으로 오진됨</li> <li>• 빈번한 이차전신화</li> <li>• 뇌전증지속상태의 병력이 있음</li> </ul>
<p>뇌파 소견</p> <ul style="list-style-type: none"> <li>• 발작사이와 발작기에 국소적 이상이 관찰되지 않을 수 있음</li> <li>• 양쪽 대뇌반구에서 발작파가 관찰되기도 함</li> <li>• 국소변화가 관찰되기도 하지만, 종종 광범위한 이상소견이 보임</li> </ul>

두정엽은 현훈 및 시각증상도 일어나는 것으로 알려져 있는데, 뇌자극으로 다음과 같은 증상이 발생하는 것으로 알려져 있다 (표 7). (Francione, Liava et al. 2015)

### 후두엽 (Occipital Lobe Epilepsy, OLE)

부분뇌전증의 약 8%에 해당하는 후두엽뇌전증의 국소발작 양상은 시각증상, 눈동자가 움직이는 감각, 안진(nystagmus), 눈뉘떨림(eye flutter), 강제적인 눈감박임, 회전 머리 및 눈 운동 (versive head and eye movement) 등이다. (대한뇌전증학회 2013)

후두엽뇌전증의 발작양상은 다음과 같이 알려져 있다 (표 8). (Blume, Wiebe et al. 2005)

### Conclusion

다양한 발작양상에 대한 숙지는 흔한 측두엽뇌전증과의 구분을 용이하게 해 준다. 발작형의 병력청취와 임상양상에 대한 자세한 관찰로 보다 더 정확하게 진단, 국소화, 편측화를 시행할 수 있다. 이를 통하여, 난치성 국소뇌전증에서 정확한 국소화를 시행하여 수술전 검사 계획을 시행할 수 있다.

표 4. 전두엽뇌전증 발작양상 분류

Site of seizure onset	Typical seizure semiology
Primary motor cortex	Contralateral tonic or clonic movements according to somatotopy, speech arrest and swallowing with frequent generalisation. Ipsilateral leg involved in paracentral seizures
SMA	Simple focal tonic seizures with vocalisation, speech arrest, fencing postures, and complex focal motor activity with urinary incontinence
Cingulate	Complex focal motor activity with initial automatisms, sexual features, vegetative signs, changes in mood and affect and urinary incontinence
Frontopolar	Initial loss of contact, aversive and subsequent contraversive movements of head and eyes, axial clonic jerks, falls and autonomic signs with frequent generalised tonic-clonic seizures
Orbitofrontal	Complex focal motor seizures with initial automatisms or olfactory hallucinations, autonomic signs and urinary incontinence
Dorsolateral (premotor)	Simple focal tonic with versive movements and aphasia and complex focal motor activity with initial automatisms
Opercular	Mastication, salivation, swallowing and speech arrest with epigastric aura, fear and autonomic phenomena. Partial clonic facial seizures may be ipsilateral and gustatory hallucination is common

표 5. 최근 보고된 내전두엽 부위의 발작증상

TABLE 1. Summary of Recently Reported Ictal Symptoms Related to the Mesial Frontal Area and Their Supportive Evidence

Ictal Symptoms	Description	Suspected Symptomatogenic Zone	Evidenced by	Reference
Ictal fear	Intense fright with/without facial expression of fear	AC	Ictal scalp EEG and MRI lesion	Alkawadri et al. (2011)
Ictal laughter	Unnatural forceful laughter without mirth	AC and SSMA	Ictal IEEG	Unnwongse et al. (2010)
Cephalic auras	Nonvertiginous sense of head movement	SSMA	Ictal MEG-SAM(g2)	Canuet et al. (2008)
Palilalia	Unsolicited reiteration of utterances, with frequently stereotypic prosody, accelerated rate, elevated pitch, or decreasing volume	SSMA	ES	Cho et al. (2009)
Oral automatisms	Repetitive, stereotyped, semipurposeful movements involving mouth and tongue	SSMA	ES	Unnwongse et al. (2009)
Early ictal face wiping	Stereotyped nonrepetitive gestures of hand to face in a wiping-like motion before complex motor activity	MF	Ictal IEEG and ES	Friedman and Yoshor (2011)
Early ictal body turning	Truncal turning without any tonic element of the extremities for $\geq 90^\circ$ , parallel to the body axis and horizontally within the first 10 seconds of seizure	MF	Ictal IEEG or resection margin	Leung et al. (2008)
Hyperkinetic seizure I	Marked agitation that either included body rocking, kicking, or boxing associated with sitting up or standing up postures	Rostral AC	Ictal IEEG	Rheims et al. (2008)
Hyperkinetic seizure II	Mild agitation primarily characterized by horizontal movements or rotation of the trunk and pelvis	MF, mainly AC	Ictal IEEG	Rheims et al. (2008)
Negative motor seizure	Inability to conduct the voluntary movements or arrest of speech without weakness and decreased tone of the affected limbs	SSMA	Ictal IEEG and ES	Ikeda et al. (2009)
Ictal singing	Diffuse musical elements, being brought together with vocalized in such a way as to produce a recognizable tune	MF	Ictal IEEG	Enatsu et al. (2011)
Pilomotor seizure	Bilateral or unilateral piloerection	MF, mainly AC	Ictal SPECT	Seo et al. (2003)
Paroxysmal arousal	Sudden arousal with variably present dystonic-dyskinetic features, recurring several times during nonrapid eye movement sleep	AC	Ictal SPECT	Vetrugno et al. (2005)

AC, anterior cingulate; ES, electrical stimulation; IEEG, intracranial EEG; MEG-SAM(g2), magnetoencephalography-synthetic aperture magnetometry with excess kurtosis; MF, mesial frontal; SSMA, supplementary sensorimotor area.

표 6. 두정엽뇌전증의 발작양상

A) 조짐		B) 객관적 발작 징후	
Sign	N° of Pts (total 40 pts)	Sign	N° of pts (total 40 pts)
Somatosensory paresthasias	20 (62.5%)	Eye +/- head deviation	22 (55%)
illusions	15	contralateral	13 (32.5%)
disesthesias	4	ipsilateral	9 (22.5%)
	2	Automatisms	17 (42.5%)
Visual	10 (31.2%)	simple motor	3 (7.5%)
visual illusions:	9	gestural	6 (15%)
metamorphopsia	4	oro-alimentary	3 (7.5%)
blurred vision	2	vocalization	3 (7.5%)
macropsia	2	bipedal, ictal automatic locomotion	2 (5%)
micropsia	2	Motor contralateral	35 (87.5%)
amaurosis	2	clonic	18 (45%)
positive elementary visual hallucinations	3	dystonic /hypertonic	14 (35%)
complex visual hallucinations	1	negative myoclonus	3 (7.5%)
Vertiginous	9 (22.5%)	Version	3 (7.5%)
rotatory vertigo contr/ipsil	2-Mar	contralateral	2 (5%)
disequilibrium	2	ipsilateral	1 (2.5%)
sensation of falling	1	Autonomic	3 (7.5%)
dizziness	2	Hypermotor	1 (2.5%)
Psychic	6 (18.7%)	Aphasia	5 (12.5%)
Cephalic sensation	6 (18.7%)	Post-ictal focal paresis	7 (17.5%)
Epigastric/thoracic	5 (15.6%)		
Fear/anxiety	4 (12.5%)		
Auditory	4 (12.5%)		
Gustatory	2 (6.2%)		
Pain	2 (6.2%)		
Autonomic	1 (3.1%)		

표 7. 두정엽 뇌자극시 나타나는 주관적인 전정기관 및 시각 양상

Stimulation site	Vestibular symptoms (N° of patients)	Visual symptoms (N° of patients)
Precuneus	“falling flat” (1) subjective vertigo (2) disequilibrium (1)	blurred vision (3) macropsia + object motion (1) “a moving object” (1)
Intraparietal sulcus	body oscillations “like being on the sea” (2)	double vision (1) blurred vision (1) metamorphopsia (1)
Superior parietal lobule	subjective vertigo (1) cephalic subjective vertigo (1) “pulsation of the head” (1)	blurred vision + object motion (1) object motion (1)
Inferior parietal lobule	“falling into a vortex” (1)	blurred vision (1) object motion (1)
Supramarginal gyrus	“falling out of bed” (1) subjective vertigo (1) body oscillations “like being in a boat”(1)	blurred vision (1) “halo around the image” (1)
Parietal cingulum	“pulsation inside the head” (1)	blurred vision + object- motion (1)

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표 8. 후두엽 뇌전증의 발작양상

	Lateral	Mesial	Both	Total
Number of patients	11	20	10	41
Visual aura	10	15	10	35
Visual unformed	9	13	9	31
Visual formed	5	8	6	19
Never visual	1	5	0	6
Autonomic	3	4	3	10
Abdominal cephalic	1	6	4	11
Unilateral somatosensory	2	2	0	4
Bilateral somatosensory	3	3	0	6
Dyscognitive	9	17	6	32
Never aura	0	2	0	2
Unilateral motor	8	10	5	23
Bilateral motor	10	16	8	34
Any motor	10	17	10	37
Versive contralateral	2	8	3	13
Versive ipsilateral	0	1	0	1

All relationships in this table are statistically non-significant.

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