

## Landau-Kleffner Syndrome: An Acquired Epileptic Aphasia

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Figure 1. Focal right centroparietal spikes in awake EEG (in different sections)

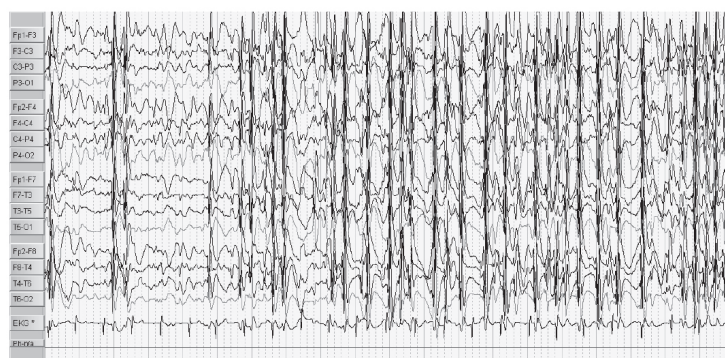


Figure 2. focal right centroparietal spikes and synchronous bilateral frontocentroparietal spike-waves in awake EEG.

A 15-year-old girl, right handed, who has acquired regression of language ability followed by epilepsy. She had age-appropriate development before age 4.5 years. At the time, seizure with limbs twitching was noted once during sleep. At age 6, she had seizures occasionally. Additional to seizures, she was noted to be gradually decreased oral speech with deteriorated intonation, inattention and rage outbursts. The awake EEG showed right focal centroparietal epileptiform discharges with synchronous bilateral frontotemporal spike-waves, (figure 1, 2) while the sleep EEG was normal. However,

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**Figure 3.** Continuous spikes and waves during slow wave sleep (sensitivity: 15uV/min, speed: 20sec/epoch).

5 months later, additional to the temporal spikes, paroxysmal generalized spike-and-wave activity was first noted in the sleep EEG. (figure 3)

Carefully reviewed her history, within 2 years after onset, she showed an auditory inattention to verbal stimulation (even as her name being called). Although she still responded to environmental sounds (such as ringing of the phone), her oral verbal expression deteriorated gradually with only few jargons. Her song-singing ability had disappeared. She communicated mainly through unarticulated sounds or using natural gestures and signing. Apart from the language impairment, the neurologic examination, brainstem auditory evoked potential and brain MRI were negative findings. She had received antiepileptic drugs such as oxcarbazepine, carbamazepine, or topiramate during her course. Electrical status epilepticus during slow wave sleep pattern in the auditory associated cortical areas without clinically concomitant seizures, together with the acquired auditory agnosia and speech deterioration, led to the diagnosis of Landau-Kleffner syndrome at age 7.

She still had twitching of mouth angle or left hand at night or early morning but with decreasing frequency between ages 7 and 10. However, further deterioration of verbal language and intermittent outburst of emotion rage were noted. Seizure free has been noted since 13-year old. The awake and sleep EEG patterns returned to normal. Her behavior and emotional problems also disappeared. However, her oral-auditory language problem did not resolve through the years, even she had received series of speech therapy. Her verbal skills were markedly impaired and the nonverbal abilities found to be below average. She was advised to receive sign language education.

Landau-Kleffner syndrome (LKS) is defined as an acquired aphasia, onset typically after age 3 years, with an epileptiform EEG and/or clinical seizures<sup>(1)</sup>. The EEG pattern of LKS shows frequent temporo-parietal discharges, and is activated by slow-wave sleep. In LKS, as with continuous spike and waves during slow wave sleep (CSWS) syndrome, seizures and EEG abnormalities resolve during adolescence. Most LKS children begin with symptoms of verbal auditory agnosia, and eventually the acquired aphasia. The aphasia is characterized by fluctuating course and mainly phonologic paraphasia. They learn to use visual modality to communicate, and are taught as deaf by using sign language. The language impairment would persist after the resolution of abnormal EEG<sup>(2-4)</sup>. The mood, social and behavior problems are not accompanied by the language regression. Most cases are idiopathic in nature and have no significant focal pathology in brain MRI<sup>(4)</sup>.

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