Lennox Gastaut Syndrome with Bilateral Temporo-Parietal Encephalomalacia Cyst in a Child: The First Indonesian Report

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Abstract

A an 11-years-old boy was referred with chief complaint of intractable seizures since 6 years-old. The typical of seizures were varies. The patient was mentally retarded and the EEG showed background slow activity, polyspikes general and slow spike wave complexes bilateral temporal that appropriate wih Lennox Gastaut syndrome (LGS). The Head MRI revealed enchephalomacia cyst in bilateral temporo-parietal that was rare finding in a child with LGS. The patient was treated with triple anti epileptic drug (AED) and ketogenic diet. The treatment reduced the seizures frequencies.

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Introduction

Lennox-Gastaut Syndrome (LGS) is a rare and severe type of epilepsy [1,2]. The prevalence has been estimated at 1-4 percent of all childhood epilepsy, although figures as high as 10 percent have been reported [3,4]. LGS can occur as secondary result of an insult to the brain either during the prenatal, perinatal, or neonatal periods, or can occur in anotherwise previously healthy child.[1] LGS is defined by a triad of symptoms of polymorphic intractable seizures that are mainly tonic, atonic and atypical absence seizures, cognitive and behavioural abnormalities. EEG with paroxysms of fast activity and slow (less than 2.5 Hz) generalised spike-wave discharges (GSWD) [1-4].

The optimum treatment for LGS has not already established. Pharmacological treatment may consists of multiple antiepileptic drugs (AED). In cases of intractable seizures surgical treatments like vagus nerve stimulation, corpus callostomy and electrical stimulation of the centromedian thalamic nucleus are reported to be effective in few cases. Ketogenic diet is reported as an effective option for treatment.[2,4]

The purpose of this case report is to present a rare case of LGS with bilateral temporoparietal encephalomalacia cyst in an Indonesian child.

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Case Report

An 11 years-old boy was referred to Dr. Soetomo Hospital, Surabaya, Indonesia with the chief complaint of intractable seizures since 6 years-old. The seizures usually lasted less than 1 minute and repeated 6-8 times in a day. Recently patient presented with particularly stiffening and frequent drop attacks. There was history of suddenly fell from the bike and often filmed. The typical of seizures were varies. General tonic, atonic and dialeptic seizures were most frequently seen. He was delivered at term spontaneously by vaccum extraction with birth weight 2900 gram and unknown Apgar score.

Physical growth was normal and the developmental milestone was delayed. She was able to walk 22 months and he was able to pronounce the words at 3 years but the articulation not clear. At 3 years old, he was unfocused and had problem of concentrating and speech articulation. Following seizures, the intellegence was decreased. The IQ dropped from 92 to 58. He started to attend special needs school to overcome behaviour problem.

EEG was abnormal with background slow activity, polyspikes general, slow spike wave complexes bilateral temporal that typical with

Lennox Gastaut syndrome (Figure 1). The Head MRI revealed Enchephalomaceal cyst bilateral temporo-parietal (Figure 2). The patient treated with combination of triple AED (Phenobarbital, Phenytoin and Valproic Acid) until reach maximum otherwise the seizures still persisted. Additional ketogenic diet was then considered and it resulted reduce of seizure frequencies.

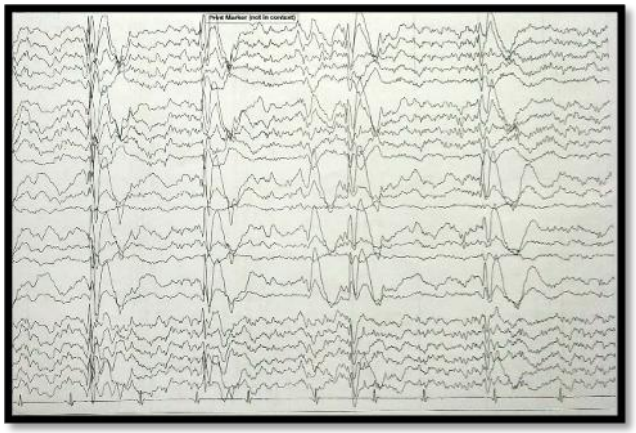

Figure 1. The EEG of the patient that showed slow spike and wave.

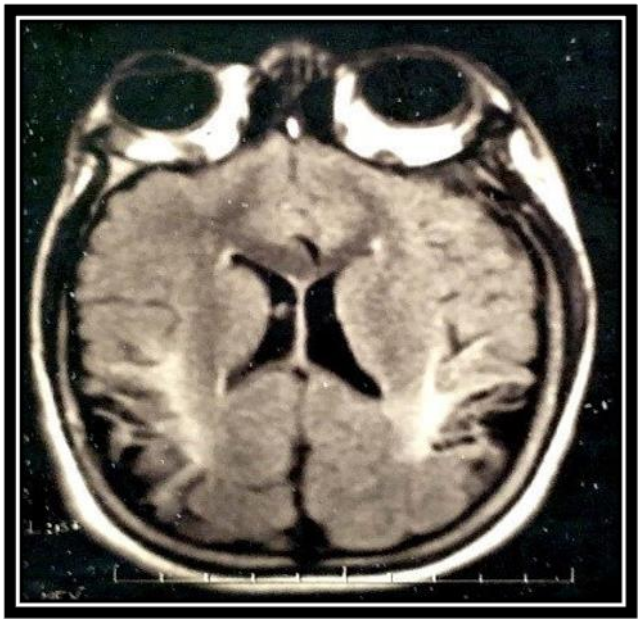

Figure 2. Enchephalomaceal cyst in the bilateral temporo-parietal.

Discussion

Lennox-Gastaut syndrome (LGS) with bilateral temporo-parietal encephalomalacia cyst is uncommon in children bilateral temporo-parietal encephalomalacia cyst is uncommon in children. There were two similar cases that have been reported in adult [5,6]. LGS children with a perinatal hypoxia or other perinatal event will have a higher chance of a preceding West syndrome; early age of onset of LGS; a higher chance to acquire focal neurological deficits and structural brain abnormality on MRI [7-10]. More than 50% of children with LGS had a history of a perinatal event in our study that confirms to previous studies. It is well known that encephalomalacia results from perinatal hypoxia, but it is a polyetiologic condition caused by various damages to immature brain of early infancy and usually results in severe psychomotor retardation [7].

Brain MRI is the most important diagnostic tool to identify the etiology of LGS. The spectrum of brain MRI abnormalities in patients with LGS was similar to a previous study. However, in South Iran study, brain imaging was normal in more than half of the patients that is much higher than the rate observed in that previous study [11].

LGS is usually characterized by a lack of responsiveness to treatment, especially the classic AED. No single treatment regimen could be considered superior to the others, and management depends on the response of the patients [1,2]. In our ccase, treatment with maximum doses of triple AED and ketogenic diet resulted seizure frequencies reduction. Caraballo stated that in LGS treatment with ketogenic diet resulted 15% patients were seizure free, 15% had a 75-99% decrease in seizures, 10% had a 50-74% decrease in seizures [12]. It's difficult to do neurosurgery considering the position of encephalomalacia cyst that were in bilateral region and the risk of surgery, although some epileptic surgery has already performed in pediatric epilepsy in Indonesia [13-16]. The long term outcome is poor in terms of seizure control and intellectual development [3].

Conclusions

The conclusion is LGS with bilateral temporo-parietal encephalomalacia cyst is rare in children. It is usually difficult to treat. Ketogenic diet and epileptic surgery might be an option for seizure control.

Declaration of Interest

The authors report no conflict of interest.

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