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Submission date: 17-Jan-2023 02:03PM (UTC+0800)
Submission ID: 1993980251
File name: Pediatric_LGS_associated_Encephalomalacia_Cyst_Rwanda.pdf (549.87K)
Word count: 2043
Character count: 11804

Rwanda Medical Journal RMJ

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Hemispherectomy with corpus callosotomy in pediatric Lennox Gastaut Syndrome associated encephalomalacia cyst: The first case in Indonesia

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ABSTRACT

Lennox-Gastaut syndrome (LGS) is a form of severe epileptic encephalopathy in children. LGS with encephalomalacia cysts is rare in children. We report a six-year-old mentally retarded boy who was referred for an intractable seizure. Seizures were tonic, atonic, and dialeptic in frequency. EEG showed generalized SSW discharges of 1.5-2 Hz, polyspikes, and burst suppression typical for LGS. Head MRI showed an encephalomalacia cyst in the right subcortical temporoparietal lobes with hemiatrophy in the right cerebral hemisphere. He was already treated with three antiepileptic drugs, but the seizures persisted. The patient was then performed right hemispherectomy and corpus callosotomy. It resulted in a good response. A combination of hemispherectomy and corpus callosotomy could be promising in this form of epilepsy disease. Seizure reduction was achieved and showed cognitive improvement and hemiparesis.

Keywords: Lennox-Gastaut syndrome, Disease, Encephalomalacia Cyst, Hemispherectomy

INTRODUCTION

Lennox-Gastaut syndrome (LGS) is a form of severe epileptic encephalopathy in children. The onset of LGS arises between 3 and 5 years of age. The prevalence rate is 1–10% of all childhood epilepsies [1]. LGS has been identified by various types of intractable seizures. There is emotional, mental, and intellectual impairment related to loss of ability and behavioral obstacles (depression, aggression, and hyperactivity). Sometimes it is difficult to recognize LGS, because of limited specific biochemical markers, many etiologies, and varied clinical manifestations. The specific electroencephalogram (EEG) feature in LGS shows generalized 1.5–2.5 Hz slow spike-wave (SSW) complexes [1,2]. Treatment of LGS is always difficult, and the possibility of complete seizure control remains grim. The objective is to report a rare case of LGS with an encephalomalacia cyst in an Indonesian child successfully treated with oral antiepileptic drug (OAE) and surgery.

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Received: 02nd March 2022; **Initial decision given:** 29th March 2022; **Revised manuscript received:** 17th April 2022; **Accepted:** 15th June 2022.

 Publisher: Rwanda Biomedical Centre (RBC)/Rwanda Health Communication Center, P. O. Box 4586, Kigali. ISSN: 2079-097X (print); 2410-8626 (online)

Citation for this article: P. I. Gunawan; W. Suryaningtyas. Hemispherectomy with corpus callosotomy in pediatric Lennox Gastaut Syndrome associated encephalomalacia cyst: The first case in Indonesia. Rwanda Medical Journal, Vol. 79, no. 3, pp. 5-8, 2022. https://dx.doi.org/10.4314/rmj.v79i3.1

Case: A six-year-old mentally retarded boy was referred to the Pediatric Neurology clinic at Dr. Soetomo Hospital, Surabaya, Indonesia, with an intractable seizure in 2019. Seizures were tonic, atonic and dialeptic.Each seizure is usually less than 1 minute. The seizure started at 2 months old and became more frequent, tonic, with rapid trunk and limb musculature contraction that moderately reclined over 3-6 seconds. He also suffered from left extremity weakness.

He was born spontaneously, not cried immediately, and delivered by a midwife with a birth weight of 2050 grams. He was born of a non-consanguineous marriage and there was no family history of congenital birth defects. The milestone was delayed. Physical examination revealed an alert child with stable vital signs. The left physiological reflexes were increased with positive pathological reflexes. There was paresis on a left extremity and scalp EEG showed moderate cortical dysfunction with generalized SSW discharges 1.5-2 Hz, polyspikes and burst suppression (Figure 1).

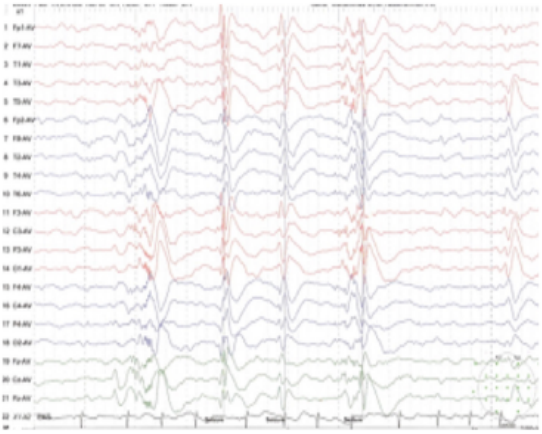

Figure 1: EEG shown moderate cortical dysfunction with generalized SSW discharges 1.5-2 Hz and polyspikes

Head MRI showed encephalomalacial cyst with gliosis in right subcortical temporo-parietal lobes with hemiatrophy at right cerebral hemisphere (Figure 2).

Based on the history, physical examination, EEG, and head MRI, the diagnosis of LGS with a right hemisphere encephalomalacia cyst was considered. He was already treated with three OAE drugs (valproic acid, phenytoin, and clobazam in a maximum dose) since 18 months old, but the seizures still persisted. The patient was then performed right hemispherectomy, and corpus callosotomy (CC).

After the surgery, the condition of the patient improved. No seizure was observed. Otherwise, the problem of speech and hemiparesis had been more severe.

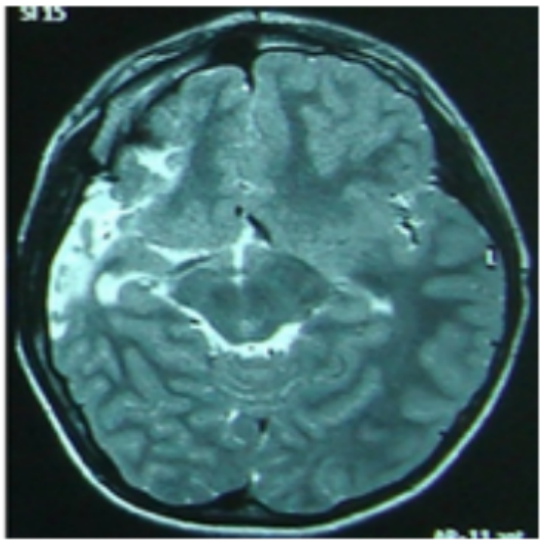

Figure 2: Head MRI showed encephalomalacial cyst with gliosis in right subcortical temporo-parietal lobes with hemiatrophy at right cerebral hemisphere

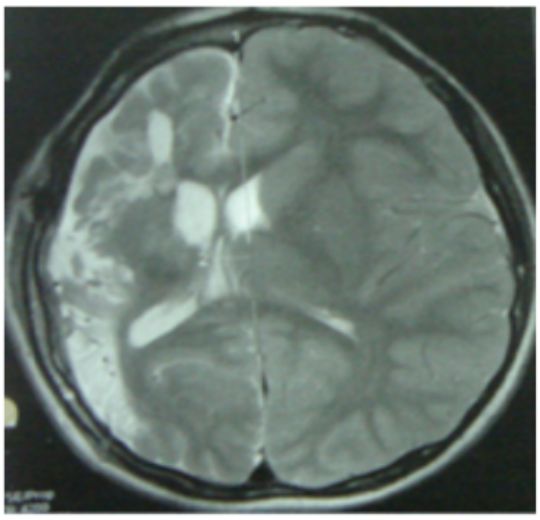

Figure 3: The head MRI evaluation showed encephalomalacia cyst in right cortical to subcortical fronto-temporo-parietal and right cerebral hemiatrophy.

The patient had received topiramate 25 mg twice a day. The head MRI evaluation after two months

of follow-up showed an encephalomalacia cyst in the right cortical to subcortical fronto-temporo-parietal and right cerebral hemiatrophy (Figure 3). EEG evaluation described mild general cortical dysfunction. No burst suppression and SSW were found.

DISCUSSION

The varied clinical manifestation and progression of LGS make diagnosing this disease difficult [2]. In our case, it is a rare and unique case that the LGS was associated with an encephalomalacia cyst in a child. There was a case of LGS in an adult male with multicystic encephalomalacia that had been reported; otherwise, the neurological problem was not severe [3]. It is well known that encephalomalacia may result in neurological sequelae and psychomotor problems.

Reducing seizures in LGS remains problematic. It's estimated that over 90% of children with LGS suffer from OAE resistance. Surgical procedure for individuals with OAE resistance remains the most expected option for long-term seizure management [4]. Hemispherectomy is a surgical method which comprises partial or total evacuation of the affected cerebral hemisphere or disconnecting of the afflicted cerebral hemisphere from the unafflicted side. Caraballo et al. considered some patients with medically intractable epilepsy, including LGS, who experienced hemispherectomy. The outcome is good for syndromes from a hemispheric lesion correlated with hemiplegia [5]. In focal lesions, resective surgery may be beneficial. Ostendorf stated that resective surgery indicated convulsion freedom and improvement in another 15% seizure reduction with a mean follow-up of almost 3 years [4].

CC is a palliative surgical method that requires achieving craniotomy, and surgically dissecting the corpus callosum to avoid seizure driving between hemispheres. Some part or entire of the corpus callosum may be performed. You et al. stated that some patients who experienced CC mostly had a higher than 50% reduction in seizure frequency, and about 35.7% had a higher than 75% reduction [6]. Lee et al. have experienced CC in LGS, otherwise, the result was unsatisfactory. The patient was then undergoing staged total callosotomy, and the patient certainly accomplished a seizure-free state with EEG evaluation return to normal [7]. Nonetheless, Mamelak et al. described that anterior 1/2-2/3 callosotomy as an expanded total callosotomy for compressing generalized tonic-clonic seizures, drop attacks or both [8]. Ding et al. reported a study combining respective surgery with CC for 68 children with non-focal lesional LGS. A combination of resective surgery with corpus callosotomy could provide supportive seizure management and distinct improvements in quality of life in pediatric LGS [9]. Combination CC and hemispherectomy had also been performed in severe epileptic encephalopathy "proteus syndrome" and resulted in significantly reduced seizures [10].

Disconnection syndrome (DS) is a postsurgical obstacle that prompts acute or long-term side effects following surgery [4]. Speech problems, including difficulty initiating speech and hemiparesis, are frequent symptoms following acute DS.

It is fascinating that the EEG following 2 months of the surgical process described mild background slow activity with no epileptiform waves. Data support the assumption that the corpus callosum plays a part in provoking epileptiform activities. Matsuo et al. stated that the corpus callosum not only conducts seizure discharges but has a reciprocally supporting effect that induces epileptogenic activities in both hemispheres. Nonetheless, the actual aspect of the corpus callosum in epileptogenesis continues to be regulated [11,12].

CONCLUSION

A combination of hemispherectomy and corpus callosotomy could be promising in a selected patient with OAE-resistant LGS-associated encephalomalacia cyst supported with EEG discharges. Seizure reduction is achieved and showed modest cognitive improvement and hemiparesis.

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