CASE REPORT

Emergency Surgery for Refractory Status Epilepticus

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Background: Management of refractory status epilepticus in children is extremely challenging. **Case characteristics**: Two children with medically refractory status epilepticus, both of whom had lesional pathology on MRI and concordant data on EEG and PET scan. **Intervention:** Emergency hemispherotomy performed in both patients. A complete, sustained seizure freedom obtained postoperatively. **Message**: Emergency surgery is a treatment option in selected cases of drug refractory status epilepticus with lesional pathology and concordant data.

Keywords: *Management, Outcome, Super-refractory status epilepticus.*

Status epilepticus is a serious medical emergency in pediatric practice with a potential for significant morbidity and mortality. Almost 30-40% of SE is refractory to first and second line treatment and needs coma producing therapy for controlling seizures [1,2]. Subsets of these patients do not respond to coma therapy too and are considered to be super-refractory. Treatment of this super-refractory status epilepticus is extremely challenging with scant literature on effective therapies. In selected cases and at experienced centres, epilepsy surgery could be considered as a therapeutic option once medical management has failed [3-5].

We describe two children with medically refractory status epilepticus (RSE) due to lesional pathology in brain, who responded well to emergency epilepsy surgery.

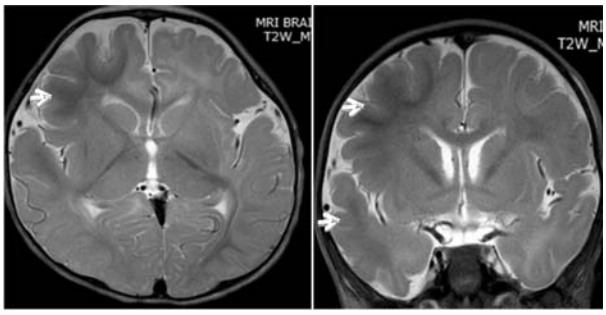
***Case* 1**

A 4.5-month-old boy was referred for neonatal onset drug-resistant epilepsy. Seizures started from 15 days of life in the form of flexor spasms. Initially infrequent, these events later increased to daily jerks which responded transiently to Inj. ACTH and oral topiramate. At this time, his developmental milestones and examination was normal. A brain MRI done elsewhere showed a right hemispheric cortical dysplasia. An EEG showed predominantly right hemispheric epileptiform discharges.

At one year of age, he came back with progressively increasing flexor spasms since 7 months of age. He also had significant left hemiparesis. An option of hemispheric surgery was declined by the parents. He responded to the ketogenic diet and was also continued on multiple anticonvulsants. A month later, he was referred from a local hospital after developing status epilepticus.

The infant had generalized convulsive seizures and was deeply comatose on admission. Seizures persisted despite multiple antiepileptic drugs and midazolam infusion. Video-EEG showed predominantly right sided ictal as well as interictal epileptic discharges and right sided burst-suppression pattern. A repeat cranial MRI confirmed the right hemispheric cortical dysplasia (***Fig* 1**). A FDG-PET scan showed increased metabolism in the right hemisphere suggestive of ongoing ictal activity.

An emergency right hemispherotomy was done after 10 days of uncontrolled status epilepticus. Histopathology confirmed the diagnosis of cortical



(a) *(b)*

Fig.1 *Brain MRI T2 weighted axial (a) and coronal images (b) of case 1 showing the right hemispheric dysplasia seen as fronto-temporal hypermyelination (hypointense white matter)-hallmark of dysplasia in infancy.*

dysplasia. Post-operative course was uneventful. Seizures stopped completely and the child remained seizure-free for next 6 months on three antiepileptic drugs (Topiramate, Sodium Valproate and Leviteracetam). Seizure recurred after 6 months due to presence of incomplete disconnection at the temporal stem as revealed by the tractography; hence, a second surgery was done for complete disconnection. Child remains seizure-free at 24 months post-operative follow up. He is gaining milestones and can stand independently though his left hemiparesis persists.

Case 2

A 7-year-old boy presented with a history of right-sided brief, focal motor seizures from the age of 4 years following an encephalitis-like illness. The seizures occurred once in 8-10 days but frequency had increased recently to daily events associated with transient right sided Todd's paralysis. A poor right hand grip was noticed since last few months. He had failed trials of multiple antiepileptic drugs.

The child was admitted in March 2015 with worsening seizures, which later evolved during hospital stay to a status epilepticus. His video-EEG showed clinical as well as subclinical seizures arising from the left temporal region. Repeat brain MRI showed progressive left cerebral atrophy with thinning of left putamen and ill-defined patchy T2 hyperintensities in left frontal and peri-insular cortex suggestive of Rasmussen's encephalitis (***Fig* 2**). An emergency hemispherotomy was performed after 7 days. Histopathology confirmed the diagnosis of Rasmussen's encephalitis. Seizures stopped completely after surgery and the child remains seizure-free at 12 months follow up. He has started speaking and is ambulant.

DISCUSSION

In both cases, RSE with lesional etiology and concordant data, emergency surgery was done for seizure-control.

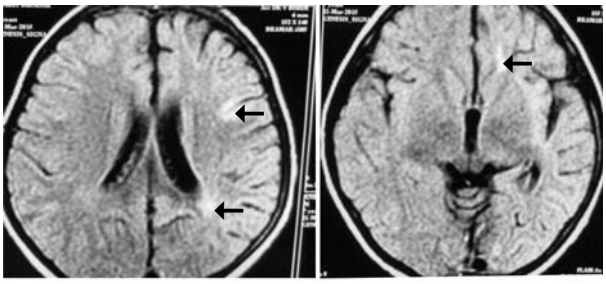


FIG. 2 *Brain MRI showing progressive left cerebral atrophy and ill-defined patchy T2 hyperintensities in left hemisphere in second patient.*

Similar approach has also been used previously as a treatment of RSE [3,4]. The published evidence-base consists of 36 patients reported in 15 small series and case reports, and the surgeries included focal cortical resection, lobar and multi-lobar resection, anatomic and functional hemispherectomy, corpus callosotomy and multiple subpial transactions [4].

Surgery has been carried out as early as 8 days after the onset of RSE but generally considered only after weeks of SE [3]. Hemispherectomy and hemispherotomy are routinely used for intractable hemispheric epilepsies, with good seizure control in 50-85% [6-9]. Seizure-free rate is highest in infantile hemiplegia syndromes and Rasmussen's encephalitis [7].

Over the last decade, newer less invasive disconnection techniques of hemispherotomies appear to achieve postoperative seizure control which is comparable to anatomical and functional hemispherectomies, with a significantly lower rate of complications [9]. This could be potentially life-saving procedure for RSE. In addition there is better postoperative developmental outcome [6]. Our experience with hemispheric surgeries has been previously detailed [10].

In conclusion, emergency epilepsy surgery is a therapeutic option in RSE in selected cases and at experienced centres, once medical management has failed. Hemispherotomy is a surgical procedure of hemispheric disconnection which seems to be safe even in infants as demonstrated in our case.

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