

CASE REPORT

Functional Hemispherotomy in a Patient with Rasmussen's Encephalitis: A Case Report

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Abstract

Background: Rasmussen's encephalitis is a progressive disease with recurrent drug resistant epilepsy, unihemispheric brain atrophy, hemiplegia and cognitive decline.

Objectives: This is a disconnection surgery which disconnects the diseased lobe from the normal lobe. This helps in development of the normal cerebral hemisphere and also the child becomes seizure free.

Methods: A five year old boy was diagnosed with Rasmussen's encephalitis. He underwent left sided trans-sylvian functional hemispherectomy. He had uneventful recovery.

Results: Before surgery he had recurrent seizures, frequent drop attacks and right sided hemiplegia and impaired cognition. He used to take three drugs for the control of the seizure. After surgery his mental state had improved, seizures were controlled with only sodium valproate. Patient's mother was happy that he had no drop attacks and was able to sit and speak.

Conclusion: Functional hemispherectomy is a good option for drug resistant seizure in Rasmussen's encephalitis.

Keywords: Rasmussen's encephalitis, Functional Hemispherotomy, Epilepsy, Drug resistance

Introduction

Rasmussen's encephalitis was first described by neurosurgeon Theodore Rasmussen and his colleagues in the late 1950s.¹ Rasmussen's encephalitis is a progressive disease characterised by drug-resistant focal epilepsy, progressive hemiplegia and cognitive decline, with unihemispheric brain atrophy.¹

The characteristic histopathological hallmarks of Rasmussen's encephalitis are cortical inflammation, neuronal loss, and gliosis confined to one cerebral hemisphere. Inflammation is multifocal within the hemisphere and progressive. Microglial and lymphocytic nodules and perivascular cuffing, neuronal death, and neuronophagia are the most common pathological feature. Evidence for an immuno-

pathological basis of Rasmussen's encephalitis is growing. The immunopathological mechanisms recognised to have a role in CNS degeneration can be categorised into three types: antibody-mediated, T-cell cytotoxicity, and microglia-induced degeneration.¹

Surgery still remains the only cure for the seizures caused by Rasmussen's encephalitis. This has functional consequences because the only effective surgery remains complete disconnection of the affected hemisphere (hemidisconnection), either as (functional) hemispherectomy or hemispherotomy.¹

The term hemispherectomy means the removal of a brain hemisphere, usually leaving the basal ganglia block behind.² The indications for functional hemispherectomy or hemispherotomy include damage to one hemisphere accompanied by medically intractable epilepsy or infantile catastrophic epilepsy. Diagnoses include extensive disorders of gyration or widespread cortical dysplasia, Sturge-Weber disease, Rasmussen's encephalitis, hemimegalencephaly, and perinatal infarction.³

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The classic “functional hemispherectomy” approach was described by Rasmussen as a functionally complete but anatomically subtotal hemispherectomy. Several other techniques have since been developed. These include the older technique of peri-insular transcortical deafferentation, the transylvian keyhole, the peri-insular hemispherotomy, the transcortical subinsular hemispherotomy, and the Japanese peri-insular modification.³ Hemispherotomy involves minimal brain removal and completely disconnects the rest of the hemisphere via minimal exposure.⁴ The “peri-insular hemispherotomy” (PIH) is a disconnective procedure included into the lateral approach techniques, and derived from modifications of the functional hemispherectomy. It is composed of three surgical stages called respectively supra-insular window, infra-insular window, and insula resection or disconnection.⁵

For the hemispherotomy four common goals are necessary: disconnection of the cortico-thalamic tract (internal disconnection of the internal capsule and corona radiata), resection of the medial temporal structures, total corpus callosotomy, and disconnection of the orbito-fronto-hypothalamic tract (disruption of the frontal horizontal fibers).⁶

The mechanisms by which hemispherectomy controls seizures are of two types: excision and disconnection. Surgical removal of the neurological tissue responsible for the seizures, the cerebral cortex, can be accomplished by anatomic hemispherectomy or hemidecortication. The mechanical eradication of the epileptogenic tissue should be accompanied by a cessation of the seizures. The same objective may be reached by disconnecting the epileptogenic tissue from the effectors, in this instance, the diseased hemisphere, from the rest of the brain i.e., the contralateral hemisphere and the brain stem. This disconnection can be achieved according to the principles of functional hemispherectomy. In this instance, neurons can still generate epileptic potentials but they have nowhere to go because of the disconnection, so that patients remain seizure-free.⁷ Tan et al had concluded that modified functional hemispherectomy may allow the patients to lead more independent lives by leading to a cessation or reduced frequency of seizures.⁸

The surgical complications were subdivided into the following four subgroups according to the involved anatomical structures: (1) Systemic Complications (2) Skin- and Skull-Covering Complications, (3) Complications Related to Damage to Nervous Structures, (4) Complications Occurring within the Surgical Residual Cavity.⁹

In our patient, we chose transylvian functional hemispherectomy because it is easier to perform, ventricles were larger due to cerebral atrophy, takes less time and blood loss is minimal.

The Case

A five year old boy presented to us on with the complaints of recurrent seizure for two years and drop attack for last one year. His mother informed us that he was relatively seizure free about two years back. Then his seizure returned, and he was repeatedly admitted to hospital. Ultimately, he was given three medicines to control his seizure which included phenytoin, levetiracetum, sodium valproate. Despite on these drugs and with appropriate doses, his seizures persisted. For the last one year he had multiple bouts of drop seizures which resulted in falling down to the ground. He had seizures multiple times a day without any relation to environment. His seizure started from the right side of the body and spread to all four limbs. At this point, he became unresponsive with rolling of the eyeball stiffness of all limbs and drooling of saliva. It was not associated with fever, tongue biting, spontaneous urination or loss of consciousness. After this episode he became agitated and cries a lot. And for last 2.5 years he developed the symptoms repeatedly every 3-4 months and became symptom free after hospitalisation and taking medication. His mother also added that his seizure was getting worse over time. He had no history of headache or vomiting during seizure.

The patient was delivered by normal vaginal delivery. His perinatal period was uneventful, after few weeks the baby stared vacantly and developed stiffness of all four limbs with abnormal crying. But breathing and colour of the body was normal. After 4 days he again developed abnormally staring look and he was admitted in a hospital, there he was hospitalised for two weeks and released at home after relief of symptoms. Then for 2.5 years he was almost free from seizures.

In CT scan there was hypodensity in the left fronto-parietal and temporal region, dilatation of the left lateral ventricle. There was global cortical atrophy on left side. MRI of brain showed global atrophy of left cerebral hemisphere with exvacuo dilatation of left lateral ventricle which was suggestive of Rasmussen encephalitis. In his tractography of brain there was severe reduction in white matter fibres tracts in subcortical U fibres, corona radiata and association fibres in left fronto-parietal lobes (figure 1). EEG showed abnormality in left frontal and temporal regions. CT angiogram showed less vascularity of the left side.

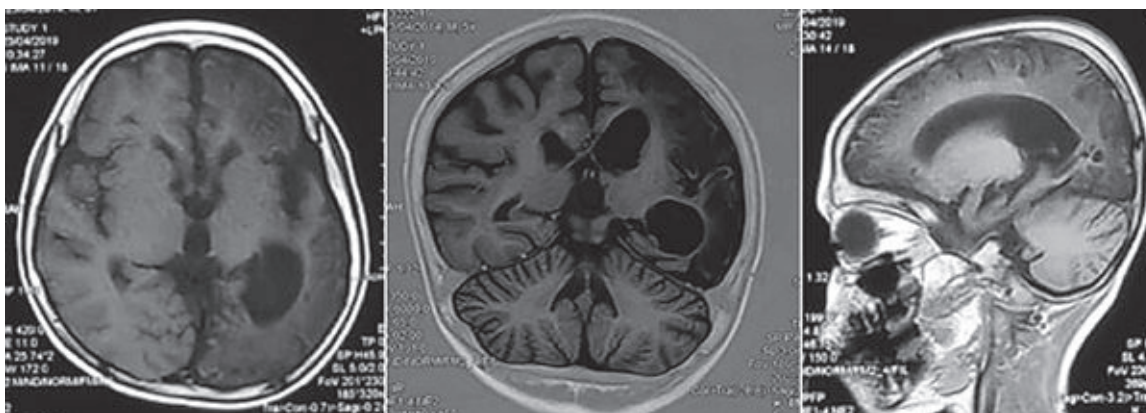


Figure 1: Pre-operative MRI of the patient of RE

He underwent left sided transylvan functional hemispherectomy. During surgery, we had incised the corpus callosum, the descending fibres, and the longitudinal fasciculus. Then the insula was decorticated, keeping the basal ganglia intact. At last, amygdalo-hippocampectomy was done.

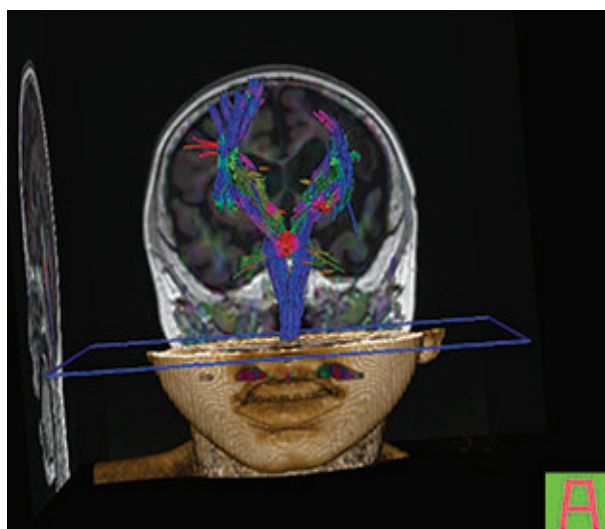


Figure 2: MRT showing loss of the U fibres.

After surgery his recovery was well. He could communicate. His hemiparesis of right side was as before. He developed aseptic meningitis on the first post-operative day, which improved after two days. His seizures were well controlled with only sodium valproate. His mother also said that his apatite had also improved. His stitches were removed on the 7th post-operative day. He could sit without support and made eye to eye contact. He also could call his mother and father.

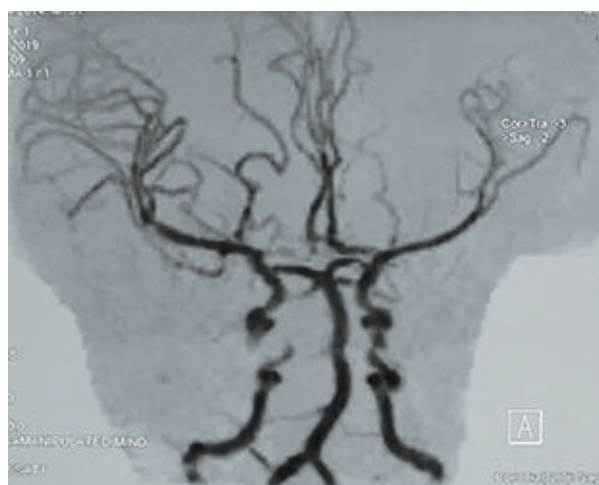


Figure 3: MRA showing marked decrease in blood vessels in left side

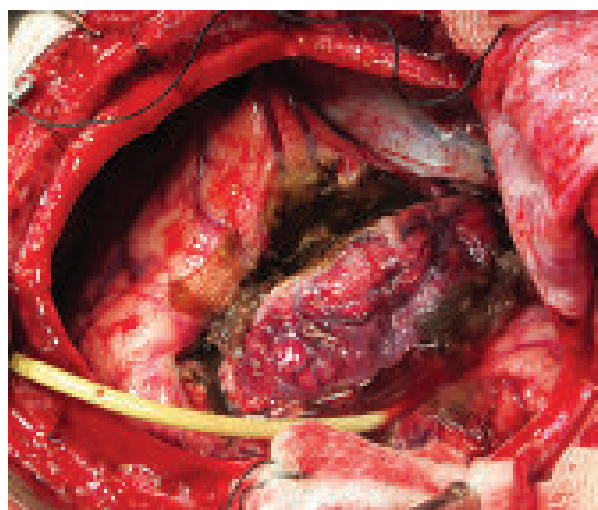


Figure 4: Intraoperative photograph after Hemispherectomy

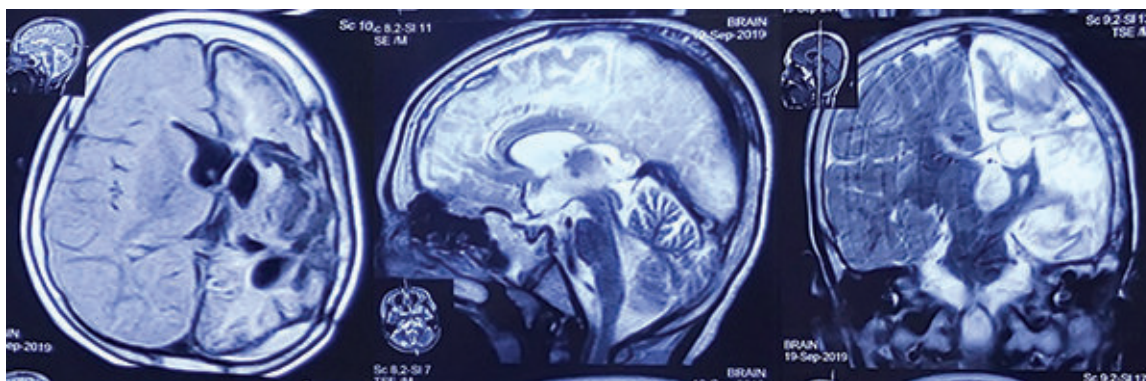


Figure 5: Post operative MRI axial coronal and sagittal section showing the intact basal ganglia after surgery.

He was discharged on the 10th post-operative day with the advice for follow up after 1 month. After one month his wound had healed well and he could sit with support though he could not walk. He had been feeding well and he had no attack of seizure. He was in follow up after discharge. After six months he was better and with only single drug having no seizure. After one year he was seizure free and we had decreased the dose of the medication. But his right side was weaker especially the upper limb.

Discussion

In a study on 115 patients of hemispherectomy or hemispherotomy, Jonas et al found that after post hemispherectomy, 78.6% of patients were seizure free at 6 months (n=112), 6.3% at 1 year (n=97), 70.4% at 2 years (n=88), and 58.0% at 5 years (n=50). And post hemispherectomy, 11.2% of patients were no longer taking AEDs at 6 months, 25.0% at 1 year, 42.9% at 2 years, and 38.2% at 5 years.¹⁰ Our patient also was on only sodium valproate and was seizure free.

Hemispherectomy and hemispherotomy may be the only very effective therapy for achieving freedom from seizure in RE patients, with a seizure-free rate of 80%.¹¹ Danielpour et al reported of two patients. Both patients were able to walk, had improved speech, and were free from their incapacitating seizures, although minor seizures persisted in 1 patient.¹² Our patient was not able to walk but tried to walk and had better postural stability after surgery. His tendency to fall was also absent.

In a series, by Pinto et al, the patients were divided into two groups. In first group four patients had developmental improvements and in second group three patients had developmental improvements. The

mean interval between assessments was 2.3 years.¹³ Our patient was assessed. He showed better communication with mother and had eye to eye contact after surgery. His speech and attention had also improved.

Hemispherectomy was not without any complication. Some patients need VP shunt placement, some had shunt related infection and some had aseptic meningitis.⁹ In our patient there was sign of aseptic meningitis. Though it is too early to comment on whether the patient will need reoperation in the near future.

The lateral hemispherectomy approach provides better corridor, decreased operative time and also relatively little blood loss and also less reoperation rate. Therefore, we chose the lateral transylvian hemispherectomy in our patient. Cook et al had compared anatomical (37 cases) and Rasmussen functional hemispherectomy (32 cases) with a new modified lateral hemispherotomy (46 cases). Of these, number of Rasmussen encephalitis was 21 cases.¹⁴

After surgery the patient had movement of the right upper and lower limbs the muscle power was 3/5. There was no facial deviation. This may be because in the absence of useful finger movements and when foot tapping was not possible, hemispheric deafferentation was not result in increased motor deficit. This has been ascribed to the role of the ipsilateral connections coming from the contralateral unaffected motor cortex.¹⁵

Conclusion

In children with Rasmussen's encephalitis, drug resistant epilepsy is a problem. This not only interferes with development, but also a state of great anxiety for

the parents. Hemispherectomy or hemispherotomy is an important surgical treatment which gives the patient freedom from intractable epilepsy. As we gain more experience, we will be able to provide more benefit to our patients.

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