Ulegyria

Ulegyria refers to a shrunken and flattened cortex due to global hypoxic-ischaemic injury in term infants, centering on the deepest portion of gyri, usually in the parasagittal region. It is here that perfusion is most tenuous and, therefore, most susceptible to ischaemic damage.

see also Bilateral perisylvian ulegyria.

It is one of the leading causes of posterior cortex epilepsy.

Treatment

Presently, there is no well-defined treatment for ulegyria mainly because of the irreversible ischaemic damage done to neurons of an affected area. However, conditions associated with ulegyria, such as epilepsy and cerebral palsy, can be treated using the appropriate treatment. For instance, seizures caused by epilepsy, due to the presence of ulegyria in the occipital lobe, can be controlled using antiepileptic drugs in some patients [1].

In other patients, such as those who suffer from ulegyria in the posterior cortex, drugs are not effective and surgery of the area causing epilepsy is needed [2) 3)]. These treatments treat only the conditions but have no effect on the condition of ulegyria itself.

Case series

Ten patients who underwent surgery for posterior cortex epilepsy with ulegyria and were followed for more than 2 years were included. All patients underwent comprehensive presurgical evaluations. Five patients underwent intracranial electroencephalography (EEG) studies. The posterior cortex including the magnetic resonance imaging (MRI) lesion was resected in all patients. Postoperative follow-up period was 2-12 (mean 6) years.

Nine patients had a history of perinatal distress including asphyxia and prolonged labor. Age at seizure onset was 5-11 years, except one patient. Three patients had visual field defects preoperatively. Ulegyria was unilateral in four patients and bilateral but unilateral-predominant in six patients. In most of the cases, the lesions were in the posterior cerebral artery area or the watershed area between middle cerebral and posterior cerebral arteries. In four of five patients who underwent intracranial EEG, seizure onset zones extended outside the lesions. Postoperative seizure outcome was Engel's class I in seven cases, and class III in three cases. Three of four patients whose seizure onset zones were not completely resected achieved class I outcome. Four of six patients with bilateral lesions achieved class I outcome.

Ulegyria due to perinatal distress is considered to be a major cause of posterior cortex epilepsy. Long-term postoperative seizure outcome is favorable. Resection of MRI lesion is important for seizure relief. Bilateral lesions should not be excluded from surgical indication. The usefulness of intracranial EEG may be limited [4].

Case reports

A study reports, for the first time, a rare case of benign childhood focal epilepsy(BCFE) coexisting with lesional epilepsy secondary to parietooccipital ulegyria. The patient underwent right parietooccipital lobe disconnection plus tailored resection of temporooccipitoparietal junction cortex under electrocorticography (ECoG) monitoring. Post-operatively, there was no impairment of neurological function and the patient only experiences a few breakthrough benign partial seizures during sleep [5].

[1] [2]
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[3] [4]
Usui N, Mihara T, Baba K, Matsuda K, Tottori T, Umeoka S, Nakamura F, Terada K, Usui K, Inoue Y. Posterior cortex epilepsy secondary to ulegyria: is it a surgically remediable syndrome? Epilepsia. 2008 Dec;49(12):1998-2007. doi: 10.1111/j.1528-1167.2008.01697.x. Epub 2008 Jun 13. PubMed PMID: 18557774.

[5]
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