Ulegyria-The "Mushroom Gyri"

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An eleven years old male child presented with complaints of medically intractable complex partial seizures since three years. Patient had history of delayed cry at birth. Brain MRI was performed at 1.5 Tesla and following sequences were acquired: T1W, T2W, FLAIR, Diffusion weighted and GRE sequences in axial plane with 5mm slice thickness, Inversion recovery (IR) oblique coronal images and oblique coronal T2W images. MRI revealed shrunken groups of gyri with gliosis, especially in the depths of the sulci, but relatively well preserved gyral surfaces in bilaterior posterior watershed territories, which were consistent with the late effects of perinatal hypoxic brain damage (Fig 1 & 2). Bilateral hippocampi were normal in size and signal intensity on oblique coronal T2W and IR images. Epilepsies arising from the occipital lobe or the adjacent portions of posterior temporal and parietal lobe are called posterior cortex epilepsy. Lesions such as tumors, cortical dysplasia, and ulegyria constitute the major cause of posterior cortex epilepsy (1). Ulegyria is caused by perinatal and postnatal injury in the cortical neurons (2). In term neonates, hypoxic-ischemic injury causes lesions predominately in the posterior cerebral artery area or in the arterial border zone between middle cerebral and posterior cerebral arteries and may result in ulegyria (1). Ulegyria tends to occur in a symmetrical fashion and can also involve the perisylvian region (3). Coexistence with hippocampal sclerosis has been reported (4). Clinically, patients present with medically intractable seizures, pseudobulbar palasy (in cases with perisylvian ulegyria) and the age at seizure onset is 5-11 years (1,3). In a newborn, there is greater perfusion in the apices of the gyri than in the cortex at the depth of the sulci. For this reason, tissues at the bottom of the gyri are more susceptible to hypoxic injury (5). Villani et al. (6) examined the radiological characteristics of ulegyria in nine patients and reported that atrophy mainly involved the deep portion of the convolution and spared the apex. The shape of the affected convolution was described as "mushroom gyri" (Fig 2). Ulegyria is diagnosed when a lesion fulfills the following MRI criteria (1): (a) poorly demarcated lesion; (b) atrophy and thinning of the cortex involving mainly the deep portion of the convolution and sparing the apex, resulting in mushroom shaped gyri; and (c) white matter signal abnormalities on T2-weighted and fluid-attenuated inversion recovery (FLAIR) sequences. Polymicrogyria is also found in the watershed between major arterial territories or in the territory of a main artery and should be differentiated from ulegyria. Ulegyria can be distinguished from polymicrogyria by MRI features such as the presence of white matter abnormalities and peculiar mushroom-shaped gyri (1). The usefulness of intracranial EEG may be limited. Resection of MRI lesion is important for seizure relief. Bilateral lesions should not be excluded from surgical indication (1).

References


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