Sub-Ependymal Heterotopia

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A 27 year old female presented in emergency department with simple partial seizures involving right side of face and right upper limb. She had history of recurrent brief episodes of such seizures since last 6 months. Each episode was followed by localized paresis of involved region lasting for few hours and then recovered. She was on antiepileptic drugs as prescribed by local practitioners. On examination she was conscious, oriented with stable vital parameters. There was no audible adventitious sound in heart / carotid arteries. Neurologic examination was unremarkable after few hours of seizure episode. Laboratory investigation revealed no abnormality pertaining to complete blood count, electrolytes, serum glucose and liver and renal function test. No ocular or cutaneous diagnostic hallmarks of tuberous sclerosis were present. Lumber puncture and electroencephalogram was unremarkable. Magnetic Resonance Imaging (MRI) T1 (Figure 1) and T2 (Figure 2) of brain revealed smooth nodular area isointense to grey matter projecting in frontal horn of left lateral ventricle. suggestive of subependymal heterotopia.

Gray matter heterotopia are common malformations of cortical development They are caused by arrested migration of neurons to the cerebral cortex; that is, when neurons which are supposed to form part of the cerebral cortex, fail to climb to the end of their ladder correctly and are permanently situated in the wrong location. Affected patients are generally divided into three groups, depending on the location of the formation: subependymal, subcortical, and band heterotopia. Subependymal heterotopia is by far the most common location for heterotopia. They can exist on either or both sides of the brain at any point along the higher ventricle margins, can be small or large, single or multiple, and can form a small node or a large wavy or curved mass. Patients with isolated subependymal heterotopia usually show female preponderance and present with seizure disorder in the second decade of life. Development and neurologic examinations up to that point are typically normal. Detection of heterotopia generally occurs when a patient receives brain imaging - usually an MRI or CT scan to diagnose seizures that are resistant to medication. Frontal lobe resection provides significant relief from seizures to a minority of patients with periventricular lesions. However our patient refused for surgery and was discharged on antiepileptic drugs.