

# CASE REPORT

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## HETEROTOPIA

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**ABSTRACT:** Cortical dysplasias i.e. cortical developmental disorders are emerging as important and common causes of epilepsy with the advent of modern high-resolution MR scanning. Among these, heterotopias are the most common developmental disorders in patients with epilepsy.<sup>1</sup> Heterotopia is the name given to focal collections of ectopic neurons in the cerebral hemispheres, i.e. normal cells present in an abnormal location. It is believed to be the result of prematurely arrested radial neuronal migration. Here we are presenting a rare case of an adult male with epilepsy, found to be having heterotopias on MR scanning.

**CASE REPORT:** A 28 years old male presented with complaint of five episodes of generalized tonic-clonic seizures within last 12 hours. This episode started at night when patient was asleep and occurred without any preceding aura. According to his attendants, it started with a cry and then clenching of teeth followed by convulsions of the whole body which lasted for about 2 minutes. It was followed by post-ictal confusion, headache and generalized weakness. He was incontinent during one of these episodes. Patient regained consciousness in between the attacks. There was no Todd's paralysis following any of these attacks. Patient had a past history of such like seizures for last 8 years. Attacks used to occur mostly during sleep, at an interval of about 6 months to 1 year. Initially he did not take any anticonvulsant drug regularly. Only 6 months back he started taking phenytoin 300 mg/day regularly, but 10 days prior to admission he suffered another seizure in spite of medication and, therefore, he left medicine. He was admitted to our ward 5 hours after the last seizure, fully conscious, well oriented and came walking by himself. His vitals were normal. CNS examination including fundus was non-contributory except for positive babinski response on left side.

Hb, TLC, DLC, ESR, urine, and platelet count were normal. Blood urea, fasting blood sugar, S. creatinine, S. sodium and potassium, S. calcium and phosphorus and ECG were normal. MRI of brain revealed (i) sub ependymal grey matter heterotopia along the occipital horn and atrium of right lateral ventricle. (ii) sub cortical Heterotopia in right temporal lobe. Patient was put on sod. valproate 600 mg per day and is on follow up without seizures.

**DISCUSSION:** Cortical dysplasia (CD) i.e. cortical developmental disorders may be classified as (i) generalized or diffuse, and (ii) focal.

#### DIFFUSE CD

Lissencephaly  
Polymicrogyria  
Pachygyria  
Hemimegalencephaly  
Tuberous sclerosis

#### FOCAL CD

Isolated focal CD  
Schizencephaly  
Microdysplasia  
Heterotopia

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Band Heterotopia  
Periventricular nodular dysplasia

Cortical dysplasias when generalized are usually associated with developmental delay and a static encephalopathy from birth, as well as severe epilepsy. In focal CD, seizures may be the only symptom. Focal dysplasias may account for 15-20% of adult population with intractable epilepsy. Many cortical dysplasias have a genetic basis or they may also be environmental and related to congenital causes, infections, toxins and radiation.

Heterotopia is collection of neurons that are in abnormal locations because of the arrest in their migration from the germinal matrix to the cortex. A wide variety of causes has been associated with heterotopia; these include fetal alcohol syndrome, trisomy 13, exposure to ionizing radiation and methyl mercury poisoning.<sup>2</sup> Patients with heterotopic grey matter almost always present with seizure disorders<sup>1</sup>. Three types of heterotopia are recognized – (i) subependymal or periventricular heterotopia (ii) subcortical heterotopia at the outer margin of the brain and (iii) band or laminar heterotopia in the semicentrum ovale.<sup>3</sup>

Subependymal heterotopia are rests of neurons that lie along the margins of the lateral ventricles and are believed to be the result of premature arrest of radial neuronal migration.<sup>4</sup> These are usually sporadic, however, some cases are familial and demonstrate an X-linked pattern of inheritance. In these a defect in the gene filamin 1 has been identified. Filamin 1 encodes for an actin cross-linking phosphoprotein that is required for normal migration of neuroblasts to the cortex.<sup>5</sup>

Clinically, the patients with subependymal heterotopia generally have normal development and motor skills and they may present with seizures during their 2<sup>nd</sup> or 3<sup>rd</sup> decade of life. MR is far more sensitive than CT in the detection of subependymal heterotopia. On MR scan, they appear as ovoid lesions within the subependymal region and are isointense with grey matter on all imaging sequences without any perilesional edema or contrast enhancement. The lesion may be one or two or may include nodules along the length of the lateral ventricles.

Subcortical Heterotopia is ectopic neuronal rests that may occur at any location from the periventricular white matter to the cortical white matter junction. These are of two types—nodular and curvilinear. MRI in both types shows areas within the subcortical white matter that follow cortical grey matter in signal intensity on all MR pulse sequences. Thinning of the cortex overlying the malformation and a decreased volume white matter within the affected hemisphere are usually evident.<sup>4</sup> Curvilinear Heterotopia are irregular linear regions of ectopic gray matter with multiple sites of contiguity with the overlying cortex. The overlying cortex exhibits deep infoldings toward the malformation and linear signal voids due to vessels along with regions containing fluid isointense to CSF may be seen within the area of heterotopia. Nodular heterotopias are lobulated gray matter foci within the subcortical white matter and the lesion varies from less than a centimeter to several centimeters in size and overlying cortex exhibits deep infoldings towards the malformation.

Patients with band heterotopia often present with “double cortex”.<sup>1</sup> The patient may present at any age although they are usually seen in childhood with developmental delay and variable severity of mixed seizure disorders. Band heterotopia is more commonly seen in females (90%) and rarely seen in males. On imaging, this appears as bands of grey matter situated between the lateral ventricle and cerebral cortex and separated from both by a layer of normal appearing white matter.

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Band heterotopia may be complete, surrounded by simple white matter, or partial. On PET imaging using FDG, band heterotopia has been found to have glucose uptake that is similar or greater than normal cortex.<sup>6</sup>

The present rare case is combination of subependymal and subcortical heterotopias presenting as seizure disorder.

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