PERIVENTRICULAR HETEROTOPIA WITH NEUROPSYCHIATRIC MANIFESTATIONS IN A CHILD

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Abstract

Neuronal heterotopias are collections of normal neurons in an abnormal location, probably secondary to disturbance in radial migration. Various hypothesis include damage to radial glial fibres, premature transformation of radial glial cells into astrocytes. Heterotopias may be found in variety of conditions like neonatal adrenal leukodystrophy, glutaric aciduria type II and GM2 gangliosidosis. Heterotopias often occur as isolated defect that result in epilepsy. Neuropsychiatric manifestations as presentations of periventricular heterotopias is uncommon.

Key-words: Neuronal migration, Periventricular heterotopias, behavioural disturbances

Introduction

Heterotopias are neuronal migration disorders which include periventricular, subcortical and leptomeningeal heterotopias. Periventricular heterotopias are located just beneath ependymal lining of lateral ventricles. They are unilateral or bilateral, located most often adjacent to occipital horns and trigones. Common presentation is seizures in second decade- partial complex, tonic clonic or focal motor(1). Other neuropsychiatric manifestations which include dyslexia, mild intellectual disability, autism, attention deficit hyperactivity disorder, depression, schizophrenia and anxiety are very uncommon(2-5)

Here we report a case of Periventricular heterotopias who presented with behavioural problems.

Case History

7 year old female child, first born of nonconsanguineous marriage with global developmental delay, with birth history of preterm delivery and neonatal admission for 1 week for preterm care presented with behavioural disturbance in the form of temper tantrums, aggressiveness, shouting aloud, beating others, urinating at inappropriate places, speaking obscene words and demanding her mother to always carry on her shoulders for a period of 3 months. Child also complained of recurrent episodes of giddy feeling followed by fall without loss of consciousness for a period of 3 months. Child used to get multiple episodes per day. There was no history of fits. Family history was unremarkable except for isolated speech delay in younger sibling On examination, she was hyperactive with excessive talking. There were no neurocutaneous markers. Head circumference (51cm) was normal. Fundus examination was normal. There were no other neurological abnormalities. On suggestion, we were able to provoke the episode. During the episode, there was no loss of consciousness and child was able to interact with mother with eyes closed. There was no associated tonic clonic movements, urinary incontinence, or tongue biting. The episode was suggestive of nonepileptic attack. Routine blood investigations were within normal limits. Cardiac evaluation and ultrasonogram abdomen were normal. EEG showed generalised slow waves suggestive of mild bilateral dysfunction with no evidence of epileptiform activity. Her IQ was 69 suggesting mild mental retardation. Psychiatrist opined attention deficit hyperactivity associated with conversion reaction. MRI brain showed bilateral periventricular nodular heterotopias. Child was counselled and was started on risperidone. Her symptoms improved partially.

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Discussion

Periventricular heterotopias are neuronal migration disorders in which nodules of grey matter line the lateral ventricles. Neuronal migration occurs between 12th and 26th gestational weeks. The first maturing neurons- Cajal Retzius cells are the major neurons of cortex by day 43 which eventually disappear. The neurons attach and migrate along radial glia (from ventricle to pial surface) in a process called radial migration. In this process, the cortex is formed in an inside out fashion, where the deepest layer of cortex forms before other layers. The six layers of cortex are visible by 27th week of gestation. Mutations in X linked FLNA gene are found in 54% of patients with classical bilateral periventricular heterotopia. Andrew E. Fry et al reported four patients with neuropsychiatric presentations in periventricular heterotropia. Another study done by Chang et al showed reading impairment in 8 out of 10 subjects with periventricular heterotopias. Presentation of periventricular heterotopias with behavioural disturbances is uncommon.

Conclusion

This case is reported for the uncommon association of periventricular heterotopias with behavioural problems. All patients with periventricular heterotopias should also be screened for neuropsychiatric manifestations.

References


Fig.2 - T1 weighted axial image of brain showing periventricular nodular heterotopias