

NEURONAL MIGRATION DISORDER

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ABSTRACT

A 3-year-old female child admitted for assessment of developmental delay. No family history of developmental problem. History of third degree consanguinity. Anthropometry:-height -90cms (15th centile), weight - 12.5 kg (<15th centile), Head circumference-44cms (< 3rd centile). On examination shows hypertonia of lower limb extensors, DTR exaggerated, power 4/5 of distal muscles of lower limb and bilateral plantar extensor response. Developmental age is 1 year by using Denver scale. Developmental Quotient for the Child is 33. MRI study showed Grey matter heterotopia of subcortical region in posterior lobe.

KEYWORDS

Neuronal Migration Disorder, Band Heterotopia.

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INTRODUCTION

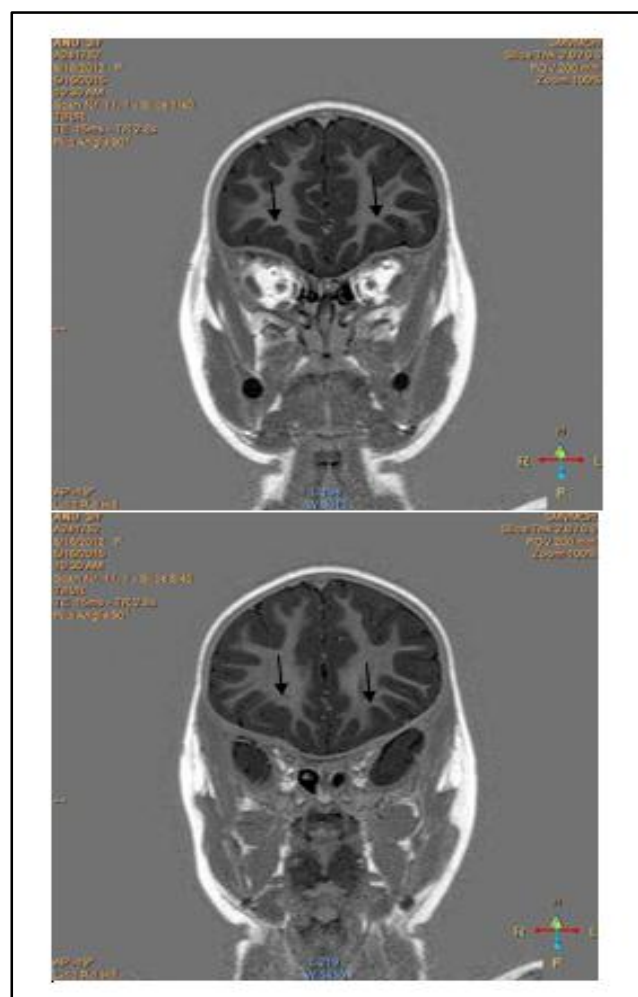
Development of central nervous system is a highly complicated process and is organized in the following steps: primary neural tube formation starts at 3–4 weeks of gestation, followed by prosencephalic development occurs at 2–3 months of gestation, neuronal proliferation starts at 3–4 months of gestation and neuronal migration at the age of 3–5 months followed by organization starts from 5 months of gestation to birth and myelination after the birth will occur. We are presenting a case of neuronal migration disorders.

CASE REPORT

A 3-year-old female child admitted for assessment of developmental delay. This is the second child in the family, elder sibling is a male child and is developmentally normal, no family history of developmental problem. History of third degree consanguinity. Antenatal history was uneventful. Normal vaginal delivery, neonatal period was uneventful. Anthropometry:-height -90cms (15th centile), weight - 12.5 kg (<15th centile), Head circumference- 44 cms (<3rd centile). The child is conscious and cranial nerve examination revealed convergent squint and motor system examination shows hypertonia of lower limb extensors. DTR exaggerated with diminished power 4/5 of distal muscles of lower limb. Bilateral plantar extensor response. Developmental age is 1 year by using Denver scale. Developmental Quotient for the Child is 33. MRI is done.



MRI



MRI: Study showed Grey matter heterotopia of subcortical region in posterior lobe

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DISCUSSION

The development of cerebral cortex results from complex sequential process of cell proliferation, cell migration, cortical organization and formation of neuronal cortex. Grey matter heterotopia is a group of neuronal disorder characterized by ectopic position of neurons. Affected patient are generally classified into two groups, depending on location of formation.

1. Periventricular heterotopia: a) Periventricular Nodular Heterotopia (PNH), b) Periventricular laminar heterotopia, c) Ribbon like heterotopia.

2. Subcortical heterotopia: Subcortical Band Heterotopia falls under lissencephaly spectrum of heterotopia.^[1,2,3] Periventricular heterotopia - mutation in FLNA gene on Xq28 were found on 100% of families with X linked bilateral periventricular nodular heterotopia and 26% of sporadic cases with coagulopathy and cardiovascular malformation may be associated.^[4] Mutation in the gene locus ARFGEF2 (20q13.13) present with PNH with microcephaly. Mutation in gene (C6orf70 6q27, FAT4 (4q-28.1), DCHS1 (11p15.4) are other genes associated with PNH.

In subcortical heterotopia (other than band heterotopia or cortical infolding), no cause has been identified. They can present as, 1. Curvilinear transformed heterotopia with thinning of overlying cortex with or without basal ganglia anomalies. 2. Multinodular heterotopia with thin overlying cortex. 3. Transmantle columnar heterotopic with or without PNH. Heterotopia are most commonly isolated anomalies, but may be part of number of syndromes including chromosomal anomalies and fetal exposure to toxins include alcohol.^[4]

Subcortical heterotopia may present with epilepsy in both men and women. The more extreme heterotopia, the greater the CNS deficit. Bilateral heterotopia is associated with severely developmental delay or mental retardation. Epilepsy associated with grey matter heterotopia is frequently resistant to medication. Epileptogenic foci has not been identified in these patients. It is proposed that reactive changes in perieptopic areas are instrumental. It precludes surgery.^[4]

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