Neurological Cortical Dysplasia Associated with Epilepsy in Sudanese Male Twins

A case report and literature Review.

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Epilepsy is the most common neurological disorder affecting young people. The aetiologies are multiple and most cases are sporadic. However, genetic abnormalities have important role. Disturbances of neuronal migration during normal development of the human cortex are recognized as significant causes of mental retardation, symptomatic epilepsy, and congenital neurologic deficits with abnormal neurological development in children. These malformations may be restricted to the brain or may be one component of a generalized malformation syndrome. This review will discuss two male twins with cortical malformation and epileptic seizures failure to thrive, microcephaly and global developmental delay. The classical facies of Miller-Dieker syndrome are not present. The family history suggested a hereditary disorder of XL type.

Key words: epilepsy, lissencephaly, child, cortical migratory disorders, Miller-Dieker syndrome, genetic inheritance twins Sudan.

DISCUSSION:

The development of the cerebral cortex progresses through defined stages including neural proliferation, neuroblast migration and neuronal differentiation. Disruptions in each of these developmental stages can lead to characteristic cerebral cortical malformations.4

Genetic microcephaly and lissencephaly are a significant cause of neurocognitive morbidity in children worldwide, responsible for many cases of mental retardation, cerebral palsy, and epilepsy.6

Recent advances in molecular genetics have led to the identification of several genes involved in the formation of the cerebral cortex and thus accurate molecular diagnosis and appropriate genetic counseling has become available for many patients and their families. Mutations in genes involved in the fine tuning of proliferation and neurogenesis, neuronal migration and differentiation and connectivity, are responsible for neurodevelopmental disorders, such as cortical migratory disorders, which are usually associated with severe retardation and epilepsy.

INTRODUCTION:

1- Professor of Child Health, Head, department of Graduate Medical studies, Faculty of Medicine, University of Gezira, Sudan.
2- Associate Professor of Neurophysiology, Head department of Physiology, Faculty of Medicine, Ribat National University.
3- Associate Professor of radiology, Institute of Nuclear Medicine, Molecular Biology and Oncology-University of Gezira.

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