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Pachygyria: A Rare Neuronal Migration Defect

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Abstract: Pacchygyria is a rare embryological defect with serious consequences for the normal child development. In the state of Jammu and Kashmir few cases of the said condition have been reported. The article tends to present a single case reported and a brief review of the matter. The clinical entity is associated with multiple syndromes and a thorough lookout for them should be considered.
Keywords: Cortex, Gyri, Neuronal, Seizures

INTRODUCTION

The Cerebral Cortex has a well defined grey matter and white matter in a balanced proportion. Imbalance in the amount of grey matter with thick cortex and large but relatively few gyri is Pacchygyria. This is a milder form of Lissencephaly and can be associated with many syndromes.

CASE REPORT

A three week old newborn presented with recurrent seizures. The child was borne of normal parents with non consangious marriage. There was no history of any ailment of mother during pregnancy and she was not on any drugs. She was Euthyroid, Euglycemic and Normotensive. The baby was borne by normal delivery and The CT Scan of the head showed Paccygyria especially in the frontal lobes. The blood investigations were normal as was metabolic profile.

DISCUSSION

The Cortical development is a mutiprocess event involving the steps of stem cell differentiation, migration into the cortex and organization into cortical layers. This whole process is cordinated by multiple pathways. Error in any step can lead to defective cortical formation. Mostly the defect arises because of bnormal neuronal Migration. The cells eventually involved in formation of cerebral cortex are deroived from spaces around the ventricles of the brain. There are multiple waves of migration which occur. Migrating neurons are guided by Programmed events as well as certain neuromodulators and chemicals [1]. Any change in signal levels of these substaces can lead to gyral malformation.In severe cases there may be agyria [2, 3]. The brain tends to be smooth and pachygyria can also be termed as partial lissencephaly. There may be related problems like microcephaly, developmental delays, recurrent seizures, feeding problems, swallowing problems, poor muscle tone. The symptomatology of defects differs and depends on the extent of disease and the cortical areas involved. There are mutataions in alpha tubulin gene [4]. Other etiological factors are vascular compromise during embryogenesis and viral infections. It may be associated with Walker Warburg Syndrome, Apert Syndrome, Lowe Syndrome, Gorlin syndrome, Aicardi Syndrome, Shapiros Syndrome or may be inherited as an Autosomal triat as well [5-6].

CT/ MRI are both of value but MRI because of its specificity to delinate white matter and grey Matter is a better technique for Investigation. The ultimate treatment is a multimodality approach.

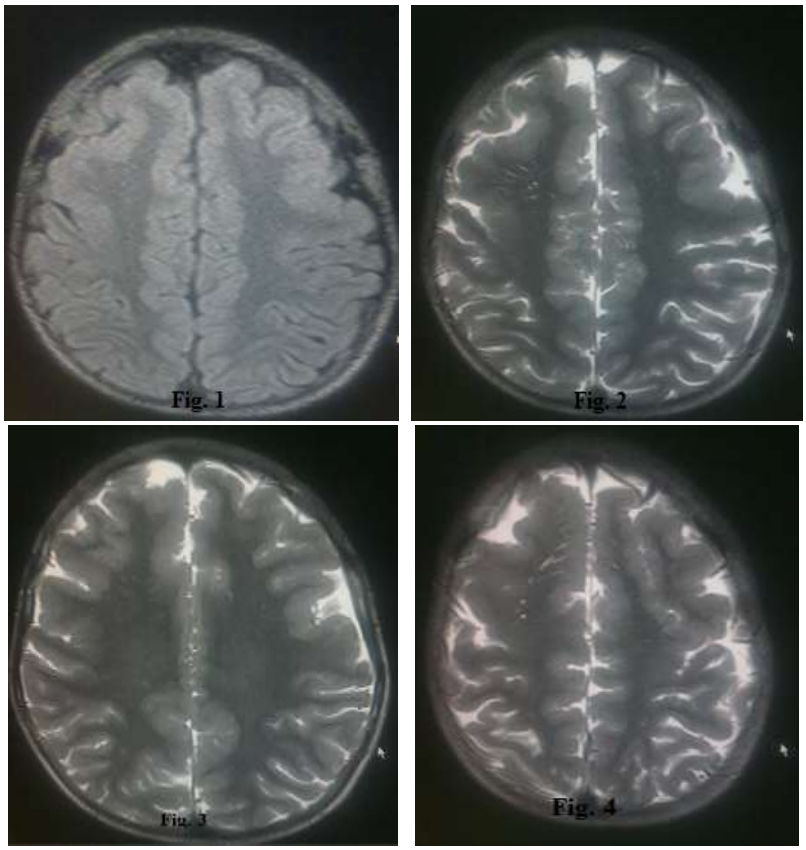

Fig.1-4: Showing Bifrontal Pacchygyria

CONCLUSION

Though Pacchygyria is not a common event but the subsequent development of child with such defects are sources of multiple problems to the child in the form of non development, Mental Retardation, Recurrent seizures with the added fact that mostly they are associated with other syndromes. As such they are a source of potential concern for the Child as well as Parents who find huge difficulties in raising such children should they survive.

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