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POLYMICROGYRIA: A CASE OF U/L RIGHT FRONTO-PARIETO-TEMPORAL REGION

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ABSTRACT

Polymicrogyria is a malformation of the cerebral cortex. The word PMG has Greek roots: Poly-many, micro-small, gyri-folds/ convolutions in the surface of brain. In PMG, these folds are unusually small & are many in number. The sign & symptoms of PMG vary from person to person depending on which part of brain is affected. MRI is preferred over CT Scan in diagnosis of PMG. As MRI has submillimeter resolution, it can very well display multiple folds in the cortical area. Treatment is symptomatic & supportive with speech therapy & physiotherapy being the main options in this case.

KEYWORDS

INTRODUCTION:

Polymicrogyria is a malformation of the cerebral cortex secondary to abnormal migration and postmigrational development [1]. Intrauterine infections, ischemic injury to the brain

during intrauterine life or genetic mutation have a causal role in the development of polymicrogyria.[2,3,4] Clinically it presents as developmental delay, and seizure with unilateral polymicrogyria may present with hemiparesis [5-7]. It can affect any part of brain & sign & symptoms vary from person to person.

CASE REPORT

HISTORY:

A 8 year old male patient came to the ENT & Paediatrics OPD , AMC& H, Mohri with complaints of weakness of left upper limb and impaired speech. The child showed delayed milestones & born of a normal vaginal delivery. No history of fever, nausea, vomiting, headache. No history hypertension, diabetes, TB, asthma. No history of seizures. No history of similar complaint in family. There is no history of CMV infection during delivery.

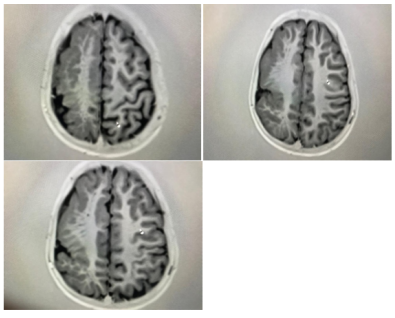
A case of unilateral polymicrogyria with hypotrophic & short left upper limb.

EXAMINATION:

Patient was conscious, cooperative & well oriented to time, place & person. Vital signs were within normal range. Pulse=94 ,BP=100/60, Temperature=98.4, Respiratory rate=24. Weight was 20 Kg which is within normal range for that age. Pallor, icterus, cyanosis, clubbing, & lymphadenopathy were absent. On examination there was weakness of left upper limb along with impaired tongue movements.

MANAGEMENT:

MRI (2D) Brain was done. Child was cooperative, no sedation was required.MRI (2D) Brain showed disorganized cortex/grey matter in the Right fronto-parieto-temporal region & in the Right perisylvian region which appears to have slightly thick cortex with bumpy inner & outer surface & small fine & undulating gyri. Findings are suggestive of PMG in these region. Patient was put on symptomatic & supportive treatment with speech therapy & physiotherapy being the main options.



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DISCUSSION:

The word PMG has Greek roots: Poly-many, micro-small, gyri-folds/ convolutions in the surface of brain. In PMG, these folds are unusually small & are many in number.

It can affect any part of brain or the entire brain. It is unilateral, when it affects one side of brain. It is bilateral when it affects both sides of brain. The sign & symptoms of PMG, vary from person to person depending on which part of brain is affected.

Unilateral Focal PMG is the mildest form as it affects relatively small area on one side of brain. In this form there are minor neurological problems like mild seizures, spastic hemiparesis, mental retardation in variable degree & can be easily controlled with medication. Sometimes patient may not have any symptoms & may be an incidental finding.

Bilateral form has more severe neurological symptoms. In this form sign& symptoms vary from patient to patient & may include recurrent seizure(epilepsy), developmental delay, motor dysfunction including speech & swallowing, crossed eyes, muscle weakness/ paresis. In severe cases there can be intellectual disability, problems in movement & seizures which are not usually controlled with medication.

Most often it occurs as an isolated feature but it can occur as part of several genetic syndromes for e.g. 22q11.2 deletion, Adams-Oliver syndrome, Aicardi syndrome, Joubert syndrome, Zellweger Spectrum disorder, Galloway-Mowat syndrome.

Syndromes from conditions:

Unilateral polymicrogyria

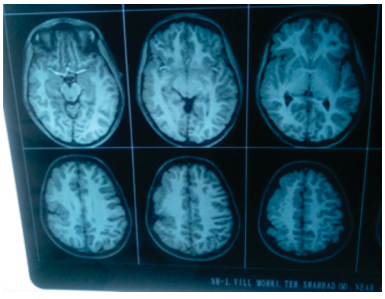
Bilateral frontal polymicrogyria
Bilateral frontoparietal polymicrogyria
Bilateral parasagittal parieto-occipital polymicrogyria
Bilateral perisylvian polymicrogyria
Bilateral generalised polymicrogyria

The gene, SCN3A, when mutated, a language area of Brain known as perisylivian cortex develops abnormal multiple small folds which appear bumpy. Polymicrogyria in this region often lead to impaired oral motor development – including difficulties in swallowing, movements of tongue & articulation of words- especially when both sides of Brain are involved.

DIAGNOSIS:

Advances in technology in past few decades have made possible to study conditions like PMG & syndromes associated with it. This diagnosis on the basis of signs & symptoms is a tedious job as it vary from person to person , on the part of brain affected or PMG may be just part of a syndrome. Multiple syndromes of region specific bilateral symmetrical polymicrogyria have been reported (8).MRI is preferred over CT Scan in diagnosis of PMG. In CT Scan, PMG & Pachygyria appear similar. In pachygyria, there is development of broad & flat regions in the cortical area. As MRI has submillimeter resolution, it can very well display multiple folds in the cortical area. Prenatal diagnosis using fetal ultrasound and MRI may be particularly difficult as the regions of the brain that are involved in this malformation may not have reached the final folding until birth. However there have been studies in which patients with Bilateral Polymicrogyria were identified by prenatal MR imaging and genetic analysis was performed (9).

Neuroimaging shows the characteristics findings of Polymicrogyria as abnormal gyral pattern, increased cortical thickness, and irregularity of the cortical-white matter junction in the form of microgyria [10-12].



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