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Polymicrogyria Revealed by Epileptic Seizures in Adults. About Two Cases and Literature Review

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Abstract **Case Report**

Polymicrogyria results from a disruption of the final neuronal migration process leading in small brain convolutions delimited by shallow cortical grooves. We report two cases: The first is a 23-year-old patient, from a poorly followed up pregnancy, with neonatal suffering, delay in motor and mental acquisitions, complaining of recurrent convulsive seizures not improved by medical treatment. The second case is a 39-year-old patient followed up for epileptic seizures of parietal appearance on electroencephalogram, treated by antiepileptic since the 20 years age without improvement. Brain MRI noted bilateral thickening of the cortical band in the parieto-occipital cortical areas in the first patient and in the fronto-parietal areas in the second, with numerous shallow cortical grooves, delimiting shallow convolutions, without anomalies in hippocampal formations. Polymicrogyria is a cortical malformation characterized by the presence of small convolutions with slightly pronounced cortical grooves. Epilepsy is the most common manifestation. It is diagnosed on MRI showing numerous shallow cortical cerebral furrows.

Keywords: Polymicrogyria, epilepsy, malformation.

INTRODUCTION

Polymicrogyria (PMG) results from a disruption of the final process of neuronal migration, that is the organization of the deepest cortical layers. The brain convolutions are small and the cortical furrows shallow.

The goal of our work is to describe the radiological elements in magnetic resonance imaging pointing to a polymicrogyric origin of an epileptic seizure.

CASES PRESENTATION

We report two cases: The first is a 23-year-old patient, resulting from a poorly followed pregnancy, who presented delay in motor and mental acquisitions, complaining of recurrent apyretic convulsive seizures resolving under treatment from an early age. The aggravation of the symptomatology by close crises motivated the consultation and was referred to us for assessment by magnetic resonance imaging (MRI). MRI with gadolinium injection including the epilepsy protocol noted bilateral parieto-occipital cortical thickening with numerous shallow cortical grooves, with no abnormalities in the hippocampal formations or contrast enhancement (Fig 1).

The second case is a 39-year-old woman followed up for epileptic seizures of parietal appearance on electroencephalogram, treated by usual antiepileptic since the age of 20 without improvement. She was referred to us for brain MRI exploration.

Brain MRI including the epilepsy protocol was performed and noted a thickening of the bilateral frontoparietal cortical tape with numerous shallow cortical furrows and noted also agenesis of the pellucidum septum without anomaly of the hippocampal formations noted (Fig 2).

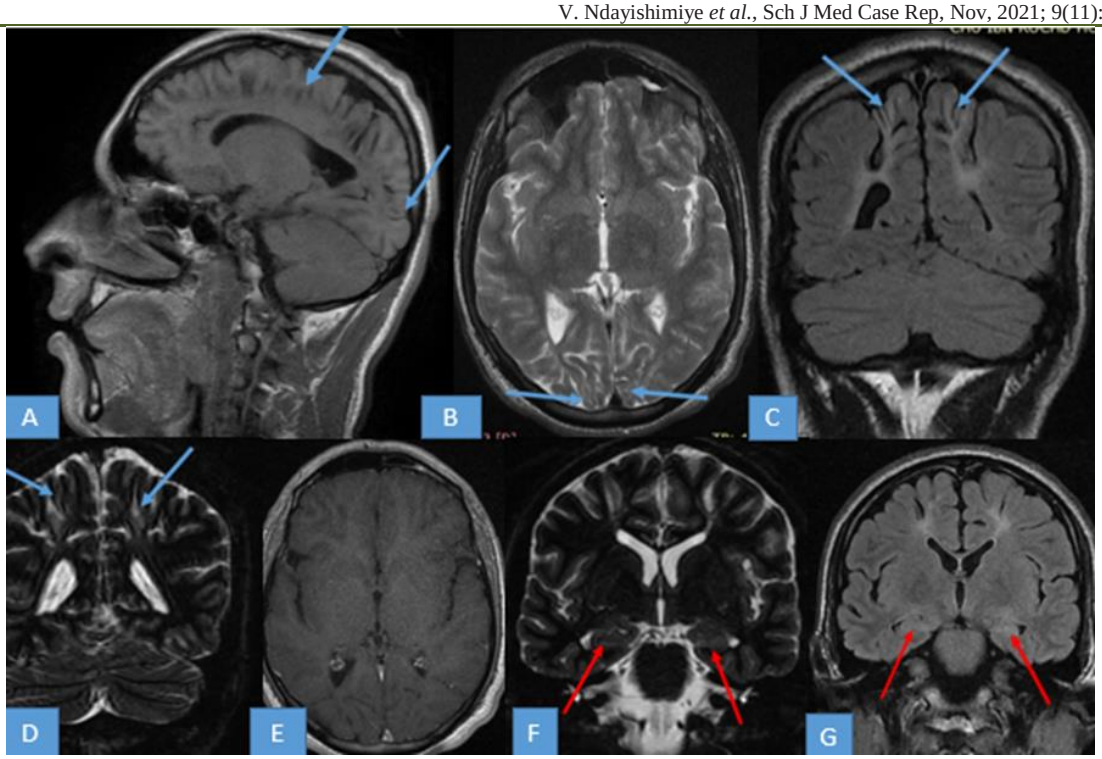

Figure 1: Brain MRI: T1 sagittal (A), T2 axial, FLAIR Coronal (C and G), morphological sequences (D and F) and T1 sequences after injection of gadolinium (E) : bilateral parieto-occipital cortical thickening with numerous and shallow cortical furrows (blue arrows), without anomalies of the hippocampal formations (red arrows) nor of noted contrast enhancement.

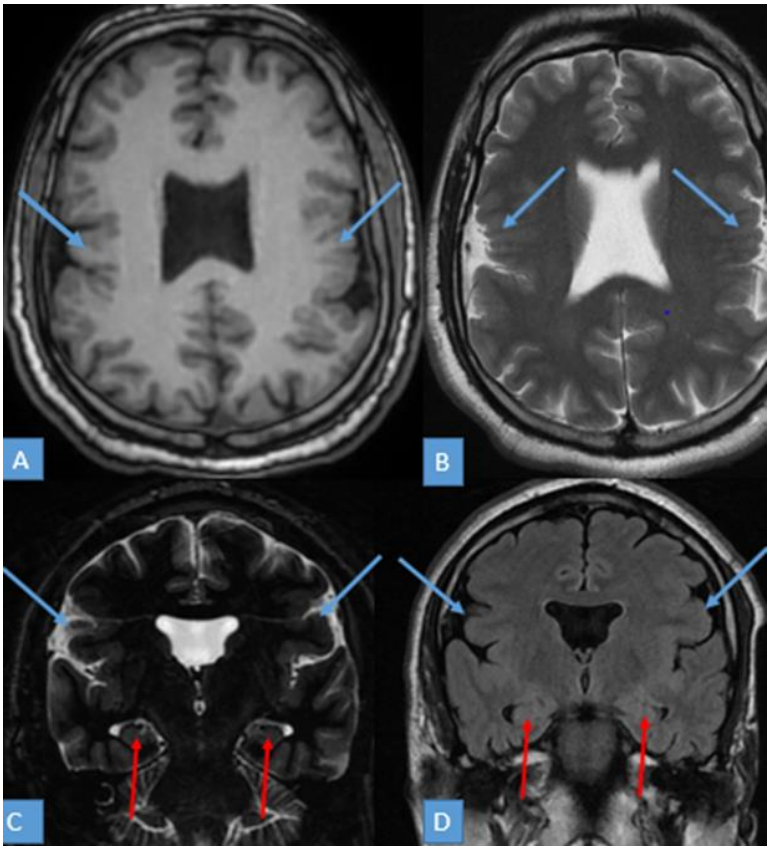

Figure 2: Brain MRI: T1 and T2 axial sequences and (A and B), morphological and FLAIR coronal (C and D): Bilateral frontoparietal cortical thickening associated with numerous and shallow cortical furrows (blue arrows) without anomalies of the hippocampal formations (red arrows). Note an agenesis of the pellucidum septum

DISCUSSION

Polymicrogyria is characterized by the presence of multiple microgyris fused together and separated by shallow grooves [1].

The aetiology of polymicrogyria is unclear. It is currently classified as resulting from abnormalities during late neuronal migration or early cortical organization. Evidence for both genetic and non-genetic aetiologies exists. Polymicrogyria occurs at the periphery of ischaemic insults and in association with congenital infections, particularly cytomegalovirus.

Multiple observations of familial polymicrogyria have been reported, including many pedigrees suggesting X-linked inheritance [2].

Clinical manifestations of patients with PMG have a large spectrum, ranging from isolated selective impairment of cognitive function to severe encephalopathy and intractable epilepsy. The severity of neurologic manifestations and the age at presentation are, in part, influenced by extent and location of the cortical malformation but may also depend on its specific etiology.

Although the incidence of PMG is unknown, it is one of the most common malformations of cortical development and an important contributor to medically refractory epilepsy, as 60– 85% of all patients with the diagnosis of PMG have epilepsy [3, 4].

The posterior perisylvian location is the most common in the frontal cortex, more rarely parietal, temporal or occipital. The cingulate gyrus and the hippocampus are classically unaffected [5, 6].

Polymicrogyria is manifested by the presence of small convolutions with not very pronounced cortical furrows which give a flattened but irregular appearance on the surface of the brain but also at the cortex-subcortical white matter junction; the cerebral cortex appears thinner when the white matter is not myelinated, whereas it is thickened when myelination is complete. The signal of the polymicrogyric cortex is identical to that of the normal cortex in the majority of cases; an hyperintense T2 signal is possible at the cortical level but also at the level of the adjacent white matter. Dilation of the perivascular spaces of Virchow Robin is possible within the underlying white matter. The drainage veins of the polymicrogyric cortex are often dilated [5].

CONCLUSION

Polymicrogyria is a cortical malformation characterized by the presence of small convolutions with slightly pronounced cortical grooves. Epilepsy is the most common manifestation, occurring at an early age but which can manifest in adulthood. It is diagnosed with MRI in the presence of numerous shallow cortical cerebral furrows.

Competing Interests: The authors declare no conflict of interest.

Contributions from authors

All the authors contributed to the conduct of this work. They also state that they have read and approved the final version of the manuscript.

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