Personality Changes in Bilateral Superior Frontal and Parafalcine Frontoparietal Polymicrogyria: A Rare Case Report

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ABSTRACT
Polymicrogyria is a neurodevelopmental abnormality which results in the formation of excessive, small, abnormal, partially fused gyri with superficially located sulci replacing the normal gyral pattern. Intellectual disability, global developmental delay, epilepsy, language deficits, and motor deficits are commonly reported in patients with polymicrogyria. We present here the case of a young male with a rare pattern of bilateral superior frontal and parafalcine frontoparietal polymicrogyria, who had a mild intellectual disability, intractable seizures along with personality changes. This case report also highlights the relevance of neuroimaging in such cases, possible explanations of personality change in polymicrogyria and relevant management issues with a review of the literature.

Key words: Neurodevelopmental abnormality, neuro-imaging, personality changes, polymicrogyria

INTRODUCTION
Polymicrogyria is the most common developmental malformation of cerebral cortex resulting from disordered neuronal organization where morphologically, excessive, small, abnormal, partially fused gyri with superficially located sulci replace the normal gyral pattern.[1-3] Intrauterine infections, ischemic injury to the brain during intrauterine life or genetic mutation have a causal role in the development of polymicrogyria.[1,3] Patients with polymicrogyria, commonly present with intellectual disability and global developmental delay.[1,3,4] However, intractable epilepsy, oromotor disorder, and quadripareisis are also reported.[1,4] Distribution of polymicrogyria may be focal or generalized, unilateral or bilateral, and symmetrical or asymmetrical. Perisylvian area is the most common site of distribution.[1,3] Polymicrogyria may exist alone or in association with other developmental malformations of the brain.[1] We hereby report a case of polymicrogyria with difficult to treat seizures and dissociative personality disorder.
CASE REPORT

A 19-year-old male belonging to a middle-class family presented with complaints of episodes of abnormal body movements associated with shouting and running around. He also had complaints of headache, which was dull-aching and persistent with fluctuating severity for 1 year. The patient had suffered multiple similar episodes, and during one such episode, he had injured his left shoulder and had a shoulder dislocation. A typical episode, as described by patient’s father, occurred when patient woke from his sleep, started shouting in fear, and running around. For this patient consulted a General Practitioner who prescribed him phenytoin 300 mg/day and risperidone 2 mg/day. There was no improvement with the treatment. Due to nonresponse, he consulted different physicians, and was prescribed sodium valproate 1000 mg/day, topiramate 100 mg/day, carbamazepine 800 mg/day, clobazam 10 mg/day, olanzapine 7.5 mg/day, and clonazepam 1.5 mg/day in divided doses in different combinations for adequate period (6–8 weeks). Electroencephalograms done at multiple occasions were reportedly normal. After a combined treatment with anti-epileptics and antipsychotic (olanzapine), the patient used to be asymptomatic for about 4–6 days but only to relapse despite being compliant.

Before the psychiatric consultation, he had frequent, brief episodes (3–4 min, 15–20 episodes/day) of shouting and abnormal behavior, with or without tonic body movements followed by loss of consciousness. Patient’s family members also report about fearfulness, visual hallucinations (seeing snakes), and assaultive behavior associated with the seizure episodes. There was no obvious psychosocial stressor, which might be temporally correlated with the onset of patient’s symptoms. There was no history of substance use. Patient’s sleep, appetite, and self-care were adequate.

His past history was not contributory. Family members did not report about any delay in developmental milestones. There was history of the bipolar affective disorder in first-degree biological relative (elder brother).

Premorbidly, he was highly sensitive to criticism with low frustration tolerance and used to avoid responsibilities. There was a history of nonaggressive stealing from home and indulgence in activities like gambling. He was manipulative and had a callous emotional concern. Patient’s maladaptive personality changes used to further exacerbate during seizure worsening were significantly impairing and were a major concern of the family. His general physical and systemic examinations including neurological examination were found to be within normal limits. Routine hemogram, liver function test, renal function test, and serum electrolytes were within normal limits. IQ assessment using Raven’s progressive matrices was suggestive of mild intellectual disability. Magnetic resonance imaging (MRI) of the brain showed diffuse polymicrogyria in bilateral superior frontal and parafalcine frontoparietal regions [Figure 1a and b].

On the basis of available clinical information, he was diagnosed to be suffering from dissociative personality disorder with mild mental retardation (as per International Classification of Diseases, Tenth Edition) with complex partial seizures.

He was initiated on oxcarbazepine 300 mg/day, which was gradually increased to 900 mg/day over 3 weeks, along with calcium supplementation. He responded well to the above treatment and did not have further episodes of seizure. He was followed over 6 months and found to be maintained well on same treatment.

DISCUSSION

Patients with polymicrogyria, frequently present with treatment-refractory epilepsy. Our patient had complex partial seizures which were well controlled with oxcarbazepine 900 mg/day. His dysfunctional behavior was also improved with this medication.

Studies reveal that neurodevelopmental disorders like autism are associated with polymicrogyria. After exhaustive literature search in PubMed/MEDLINE, Google Scholar using keywords polymicrogyria, conduct disorder, antisocial/dissocial personality disorder, we were unable to find any description regarding association of conduct disorder or antisocial personality disorder in patients with polymicrogyria.

Mavili et al., in their study explored the correlation of neuro-imaging (MRI-brain) findings with clinical test, and serum electrolytes were within normal limits. IQ assessment using Raven’s progressive matrices was suggestive of mild intellectual disability. Magnetic resonance imaging (MRI) of the brain showed diffuse polymicrogyria in bilateral superior frontal and parafalcine frontoparietal regions [Figure 1a and b].

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manifestations. As per this study, patients with unilateral polymicrogyria usually have milder symptoms, which develop late, whereas those with bilateral polymicrogyria often have gross developmental delay (motor and linguistic) with early onset. The patient had milder symptoms (mild intellectual disability and epilepsy without any neurological deficits), even though there was bilateral involvement. Even after the involvement of the parafalcine (parasaggital) frontoparietal areas, there was no suggestion of spastic paraparesis, bladder dysfunction, or any other motor deficit.

Leventer et al., in their study classified polymicrogyria and described different patterns (common and rare) of polymicrogyria. The pattern of distribution of polymicrogyria in this index patient was bilateral superior frontal and parafalcine (parasaggital) frontoparietal, which seems to be an extremely rare pattern.

Bilateral frontoparietal polymicrogyria may be associated with novel GPR56 mutation as found in a recent study. The association of personality changes with polymicrogyria may be explained by following mechanisms:

- Neurodevelopmental abnormality of frontal cortex (which is the determinant of personality, executive functions, and behavior) may attribute to the personality change
- Long standing epileptic process
- Associated intellectual disability
- Chance association.

In this case, the personality changes seem to be of neurobiological origin and possibly indicate toward a rare syndromal association. Neuroimaging (MRI-brain) is an important investigation modality for identification of such conditions, hence should be used to rule out any organic cause of behavioral changes.

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Conflicts of interest
There are no conflicts of interest.

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