CASE REPORT

CONGENITAL BILATERAL PERISYLVIAN SYNDROME: A CASE REPORT

Abdullah Bin Imaad, Nayab Khan, Rustam Alam Shah, Imaadur Rehman
Department of Radiology, Islamabad Diagnostic Centre, Islamabad, Pakistan.

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ABSTRACT

Congenital Bilateral Perisylvian Syndrome (CBPS) is a rare brain structural abnormality. The specific anomaly is called polymicrogyria, which is a type of abnormal brain development. In polymicrogyria, the normal process of cortical development is disrupted during the late stages of neuronal cellular migration. This leads to the formation of many small folds on the surface of the brain and abnormal layers of cells on cortical surface.[1,2,3] In CBPS, there are excessive small folds and unusual layers of brain cells in the Sylvian fissures (a specific area of the brain) and the surrounding cortex.[1]

CBPS can vary in terms of where it occurs in the brain, its symmetry, what causes it, and the specific symptoms it causes.[1] The symptoms experienced by individuals with BPP depend on which part of the brain is affected. In this case, we are reporting the findings of an MRI scan of a 9-year-old boy who has congenital bilateral perisylvian syndrome (CBPS).

Keywords: Epilepsy, Congenital bilateral perisylvian syndrome, Polymicrogyria.

Introduction

Polymicrogyria is a common type of brain malformation that affects the development of the cortex, which is the brain s outer layer. It can cause various clinical problems such as epilepsy (seizures), intellectual disability, difficulties with movement and speech problems.

Congenital bilateral perisylvian syndrome (CBPS) is an extremely rare disorder of the brain that occurs later in development. It is characterized by a combination of epilepsy, bilateral perisylvian polymicrogyria (abnormal folding of the brain in specific areas), and pseudobulbar palsy (a condition affecting the muscles involved in speech and swallowing).[4,5]

This syndrome was first described by a medical professional named Graff Radford, who observed and documented the specific features and symptoms of this condition.[4]

Correspondence : Dr. Imaad ur Rehman
Department of Radiology,
Islamabad Diagnostic Centre,
Islamabad, Pakistan.
Email: imaadur@gmail.com

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The patient, who is 9 years old, had a history of delayed development milestones and occasional seizures with paralysis of bilateral lower limbs for 10-20 min whenever seizures happened. Patient had speech difficulties as a toddler, however, speech problems subsided with time. The patient also experienced intermittent episodes of dizziness (vertigo). The patient's birth history was normal, and there were no incidents of trauma. To investigate further, an MRI scan of the brain was performed.

The MRI results revealed thickening of the gray matter around the sylvian fissure on both sides, with an irregular cortex and widening of the fissures (depicted in (Fig.1,2 & 3). Bilateral sylvian fissures were displaced and extended up to the perirolandic region. Additionally, inverted appearance of the lateral ventricles also seen, which is a characteristic feature

of congenital bilateral perisylvian syndrome (CBPS). After getting the report of MRI, the patient was started on antiepileptic medication (Epival) and patient responded well to it. His seizures have subsided completely in a period of 3-4 months. Now he has been off epival for about 5 months and is symptom free currently.

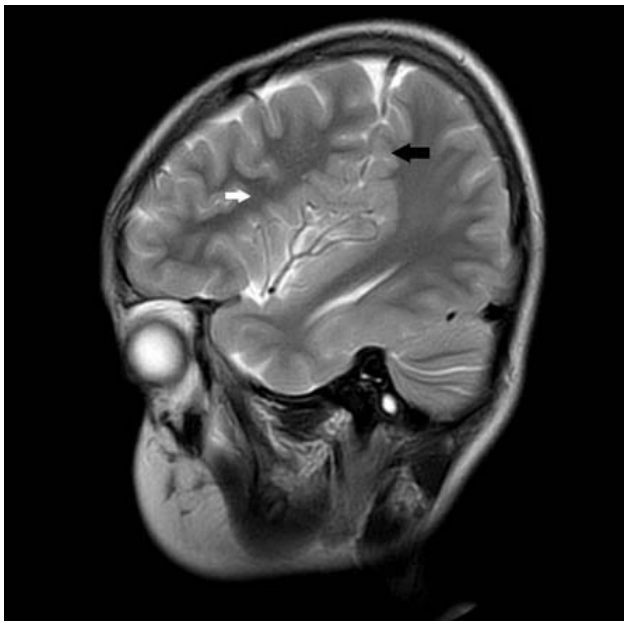

Figure 1: Sagittal T2 weighted image reveals the presence of polymicrogyria, which is depicted as cobblestone-like patterns. These patterns can be observed throughout the perisylvian cortex (indicated by white arrow) and extend into the parieto-occipital area (indicated by black arrow).

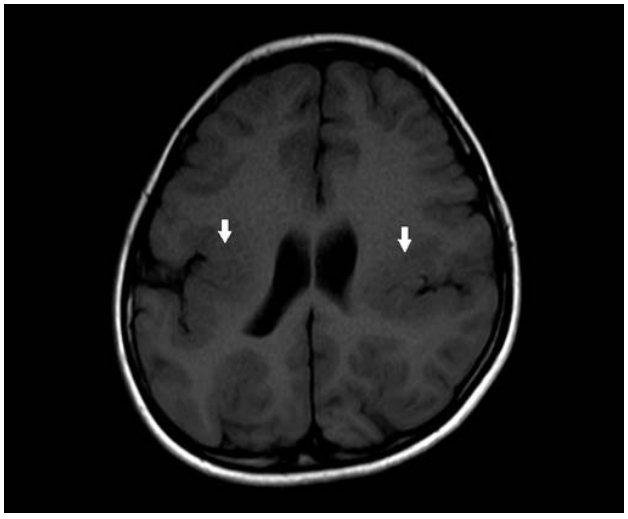

Figure 2: Axial T1 weighted image displays the thickening of the gray matter around the sylvian fissure on both sides of the brain. The cortex, the outer layer of the brain, appears irregular (indicated by arrows).

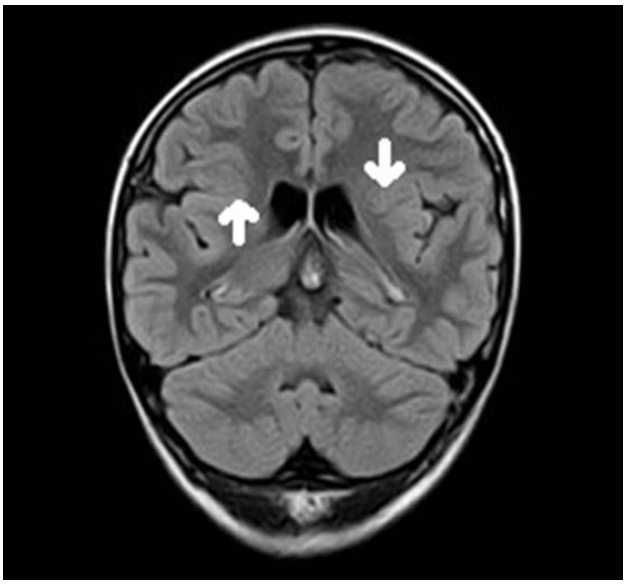

Figure 3: Coronal fluid-attenuated inversion-recovery (FLAIR) image reveals the thickening of the gray matter around the sylvian fissure on both sides of the brain. The cortex, which is the outer layer of the brain, appears irregular (indicated by arrows).

Discussion

Polymicrogyria is a complex disorder with a wide range of features and presentations. It can have different causes, distribution patterns, clinical and pathological manifestations, and imaging features.[5] On the basis of their distribution, many syndromes of bilateral symmetrical polymicrogyria are described that should be recognized by neuroimagers.[1] One of them is called bilateral perisylvian polymicrogyria (BPP), also known as congenital bilateral perisylvian syndrome(CBPS).[3] This syndrome is associated with various clinical and imaging features. The exact cause of polymicrogyria is unclear, but both genetic and non-genetic factors are believed to contribute.[3,5] Genetic causes can be sporadic or familial, with different inheritance patterns observed in affected families including X linked, automsomal dominant and autosomal recessive patterns.[3,5,6]

CBPS can also be caused by issues with blood circulation in the brain or congenital cytomegalovirus infection during fetal development.[3,5,6] The timing of the anomaly usually occurs during the fifth or sixth month of pregnancy, resulting from a disruption in brain development after cell migration.[7]

The clinical manifestations of this syndrome include

immobility of the tongue and throat, epilepsy, and mild cognitive impairment.[6,7] In sporadic cases, more severe neurological symptoms are present, such as developmental pseudobulbar palsy (difficulty with oropharyngeal function and speech), epilepsy, mental retardation, and even arthrogryposis (joint contractures).[1,3,7] Other patients may exhibit delayed development, palatal dysfunction, hypotonia, or motor deficits.[3]

Familial cases of CBPS tend to have a lower incidence of clinical manifestations, as individuals with milder symptoms are more likely to be identified and evaluated. A deveopmental reading difficulty without any physical and mental retardation may be the reason of seeking medical assistance is said group.[3,5] Seizures are common in this syndrome and can present in various forms.[3] Seizures usually present between 4-12 years of age and are usually poorly contolled. Seizures can be of various forms such as generalized tonic-clonic, infantile spasms, typical and atypical absence seizures and drop attacks eventually progressing to Lennoxx-Gastaut syndrome.[4]

Antenal and prenatal diagnosis using ultrasound is difficult as the affected regions of the brain may not have the final folding.[4] CT scan may not identify the presence of polymicrogyria.[4] Hence, MRI remains investigation of choie for identification of CBPS and for grading its severity (Grade 1 being the most severe form and Grade 4 being the mildest form): Grade 1, perisylvian polymicrogyria reaching upto the frontal or occipital pole; Grade 2, polymicrogyria extending beyond the perisylvian region but not reaching either pole; Grade 3, polymicrogyria restricted to perisylvian region only; Grade 4, polymicrogyria confined to posterior perisylvian regions.[3,7,8] Our case was classified as Grade 3 CBPS .

Prognosis for epilepsy in perisylvian syndrome is unpredictable, and seizures often prove difficult to treat, often being resistant to antiepileptic medications.[4] Fortunately, in our case patient responded well to antiepileptic medication and his seizures subsided completely in a period of 3-4 months. Now he has been off epival for about 5 months and is symptom free currently.

There is currently no cure for the syndrome, but a multidisciplinary approach involving speech therapy, oral motor skill training, swallowing therapy, and tongue muscle exercises can be beneficial for patients. In severe cases of feeding impairment, a percutaneous gastrostomy tube may be necessary.[8] Callosotomy (surgical separation of the brain's hemispheres) has been successful in treating intractable drop attacks, and if unsuccessful, a vagus nerve stimulator can be considered as an alternative treatment option.[7]

Conclusion

Congenital Bilateral Perisylvian Syndrome (CBPS) is a rare brain disorder that occurs due to issues during the migration process of brain cells. It can be identified through brain MRI and should be considered as a potential diagnosis when infants or children present with oro-motor dysfunction (problems with mouth and throat movements), signs of pseudobulbar palsy (difficulty with controlling facial muscles), developmental delay, and seizures.

Conflict of Interest: None

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