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Bilateral Perisylvian Polymicrogyria in Infant Revealed by Status Epilepticus

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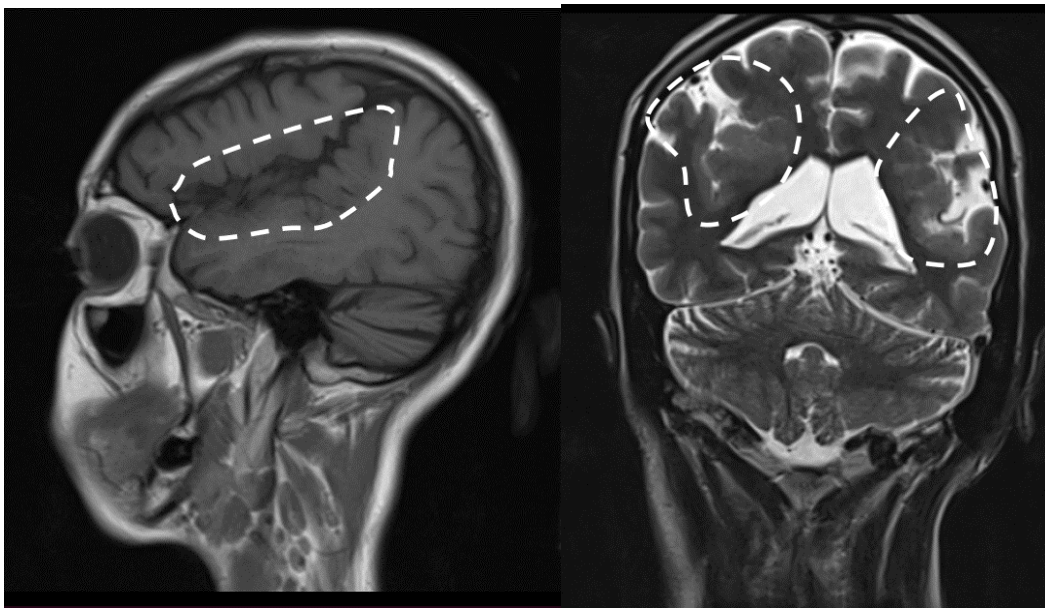
Abstract **Case Report**

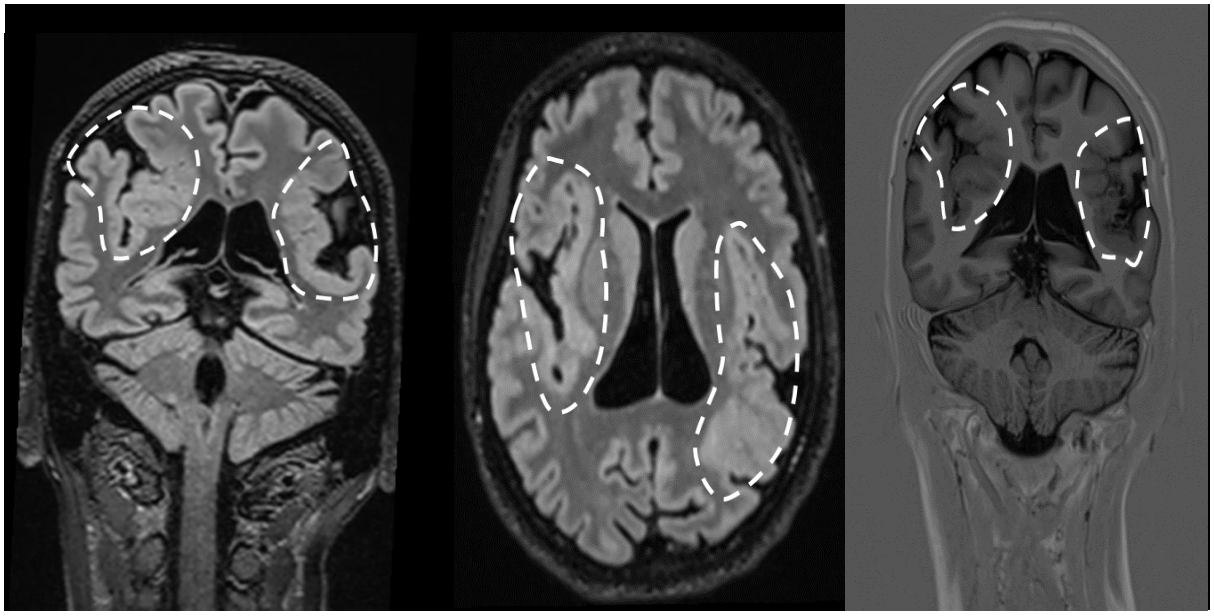
Polymicrogyria is a malformation characterized by an abnormality in late neuronal migration and cortical organization, that causes usually recurrent seizures and or status epilepticus. It results from a combination of genetic and non-genetic factors. We report a case of bilateral perisylvian polymicrogyria in an infant with status epilepticus.

Keywords: Polymicrogyria, bilateral, perisylvian, infant, status epilepticus.

CASE REPORT

20 yo female patient with dysarthria since childhood, with recurrent generalized tonic-clonic seizures beginning at the age of 16 controlled with drugs then lost of view, presenting currently status epilepticus, laboratory tests were normal and the electroencephalogram demonstrated temporary irritative activity. The Magnetic resonance imaging (MRI) was performed showing multiple abnormal bilateral perisylvian microsulci without author malformation





MRI images on sagittal T1, coronal T2 coronal and axial T2 Flair, and coronal TIR demonstrate thin white matter digitations within multiple small gyri localized in both parietal lobes and on perisylvian having a cauliflower-like aspect with a pseudo fusion of microsulci

DISCUSSION

Neurons reach the cortex but distribute abnormally, forming multiple small undulating gyri hence the name "polymicrogyria", it can be focal or diffuse and bilateral. It accounts for approximately 20% of all malformations of cortical development [1].

The signs and symptoms associated depend on the surface of the brain affected; The affected patients experience minor neurological symptoms such as mild seizures that can be easily controlled with medication. To severe neurological problems such as recurrent seizures; delayed development; diplopia; speech and swallowing troubles; and muscle weakness or paralysis for patients with bilateral polymicrogyria. It can result from both genetic and environmental causes [2].

Bilateral perisylvian polymicrogyria also called a congenital bilateral perisylvian syndrome, is a irare neurological disorder that affects the cerebral cortex

Most cases of bilateral perisylvian polymicrogyria occur sporadically in people with no family history of bilateral perisylvian polymicrogyria and the pattern of inheritance depends on the cause. Genetic causes may include mutations in single genes and [3] reported in association with twin pregnancy complications including twin–twin therapies aim transfusion syndrome and intrauterine death of a co-twin. It has additionally been reported in association with amniotic band syndrome.

A diagnosis of bilateral perisylvian polymicrogyria is typically based on a thorough physical examination, a detailed medical history, and a complete neurological evaluation, which may include tests such as:

- Magnetic resonance imaging (MRI) – a diagnosis of polymicrogyria is usually made by MRI because MRI can detect the brain's small folds more effectively than other imaging techniques.
- Electroencephalography (EEG)
- Computed tomography (CT) scanning
- Genetic testing different types of genetic tests may be performed to determine if there is a genetic cause for the bilateral perisylvian polymicrogyria

The diagnosis of polymicrogyria is made by imaging brain structures using MRI since computed tomography (CT) and other imaging methods do not have a high enough resolution or adequate contrast to identify the small folds that define polymicrogyria. It is important to choose optimal imaging techniques (including thin slices) and age-specific protocols to provide the best contrast differentiation between grey and white matter with good spatial resolution and adequate signal noise ratio Typical findings include regions of the cerebral cortex with a complex set of small gyri that appear fused and separated by shallow sulci. These findings may be isolated to one side of the brain (unilateral) or may involve both sides of the brain (bilateral).

With high-quality MRI, microgyri and microsulci may be appreciated and it is usually an isolated finding, but also it may be seen in association with other brain malformations including grey matter heterotopia and ventriculomegaly, as well as abnormalities of the white matter, corpus callosum, brain stem, and cerebellum.

In 5% of cases, polymicrogyria is detected on prenatal ultrasound examination by the presence of abnormalities, usually microcephaly and/or associated brain malformations [4].

The treatment of bilateral perisylvian polymicrogyria may require the coordinated efforts of a team of specialists. Paediatricians, neurologists, surgeons, physical therapists, and others because it is so difficult to correct it, but some treatments can improve some symptoms. Such as anti-seizure medications to control seizures. People with bilateral perisylvian polymicrogyria may also benefit from physical therapy and/or speech therapy [5].

Early intervention is important in ensuring that children with polymicrogyria reach their potential. Special services that may be beneficial to affected children may include physical therapy, special remedial education, speech therapy, and other medical, social, and/or vocational services.

CONCLUSION

Bilateral perisylvian polymicrogyria is rare affection, that can be diagnosed nowadays with the new techniques of imaging "MRI", but it is still difficult to control and its management required the participation and coordination of various disciplines, however, the prognosis is still unpredictable and depend to each individual.

Above all of that, genetic counselling can be of benefit to prevent other future affected children.

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