



Pachygyria associated with microcephaly in a newborn

Yenidoğanda pakigiri ile ilişkili mikrosefali

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Dear Editor,

Cerebral malformations due to neuronal migration disorders are studied in four groups: Agyria/pachygyria, heterotropy, polymicrogyria, and cortical dysplasia (1). In this letter, we aim to share our ideas on the possibility that newborns may experience resistant epileptic seizures in the neonatal period and neuronal migration defect may arise in neonates who are born with microcephaly.

We learnt that the patient referred to our hospital with a pre-diagnosis of neonatal convulsion was the third living child of a 27-year-old mother after her third pregnancy. The patient was born by normal spontaneous vaginal way and was delivered as 3500 grams without the need of resuscitation. Having evaluated the leaping movements of the bilateral upper extremities as convulsions, the patient was referred to our center on the first postnatal da.

The pathological findings on physical examination were as follows: head circumference: 31cm (<3p); lifeless newborn reflexes; generalized hypotonia; and there was hypoactivity in its deep tendon reflexes. Our patient did not have any dysmorphic features except for microcephaly. The results of the following laboratory tests were normal: complete blood count, glucose and blood biochemistry, blood gas values. C-reactive protein (CRP) and IL-6 values (6.34 pg/ml) were within normal limits. The screening for TORCH infections did not show any pathological factors.

The cranial MRI displayed images suggesting pachygyria bilaterally in the fronto-temporo-parietal regions (Figure 1).

The electroencephalogram (EEG) examination revealed recurrent rising slow-wave and sharp slow-wave activity along the trace in the bilateral parieto-occipital, bilateral fronto-temporal, and fronto-central regions. The patient started to receive phenobarbital and phenytoin therapy due to seizures.

We found out that convulsions were not under control in the follow-ups and therefore added intravenous midazolam infusion to the phenobarbital and phenytoin therapy. As the seizures stopped after the midazolam therapy, we added topiramate to the treatment. In the follow-up conducted in its third postnatal month, it was observed that the seizures were under control with phenobarbital and topiramate therapy.

The normal development of the cerebral cortex is carried out by the proliferation, differentiation, and, the eventual migration of the precursors in the germinal matrix to the cortex in the sixth week of gestation. Disruption of the neurone migration for any given reason in any of the stages leads to a group of congenital malformations that contain defects in the regulation of cortical development such as lissencephaly, pachygyria, and schizencephaly (2, 3).

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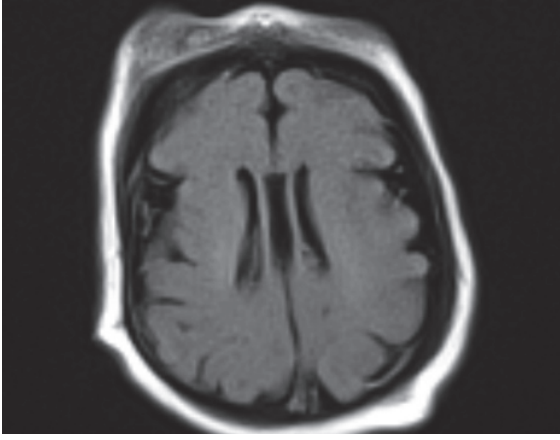


Figure 1. Cranial MRI displayed images suggesting pachygyria in the bilateral fronto-temporo-parietal region.

While agyria refers to the absence of cortical gyri, pachygyria defines the reduction in the gyrus number with a flat and shallow view of the cerebral cortex separated into sulci. Clinically speaking, pachygyria leads to seizures, developmental delay, and motor deficits.

Patients affected by pachygyria may show resistant epileptic seizures as well as mental retardation (4).

We, therefore, aim to share our opinion that pachygyria should be considered as one of the neuronal migration defects while evaluating microcephalic newborns with refractory epilepsy seizures and such patients should undergo cranial magnetic resonance imaging (MRI). Regards,

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