



Cerebral polymicrogyria: case report

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Abstract

We describe a rare case of unusual development of the cerebral cortex in a 2-year-old boy, who died in status epilepticus. The brain regions, corresponding to the normal superior and middle frontal gyri, were covered with numerous unusually small gyri, a condition consisting with a case of polymicrogyria.

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Key words [cerebral cortex] [developmental anomaly] [polymicrogyria] [epilepsy]

Introduction

Disruption of the major steps of the cerebral cortical development – cell proliferation/apoptosis, neuronal migration and cortical organization, result in a group of disorders called malformations of cortical development [1–4]. They are frequently associated with the presence of grossly unusual cortical gyri. With the improvement of modern imaging techniques this malformations have been increasingly recognized as a major cause of seizure disorders [3]. We herewith describe such a rare autopsy case.

Case Report

During routine forensic autopsy of the body of a 2-year-old boy, who died in status epilepticus, an unusual cortical formation was observed. On the superolateral surface of both hemispheres the size and location of the gyri differed considerably. In the anterior part of the cerebral area that includes the both left and right frontal lobes, there were numerous unusually small gyri. Careful examination of the brain surface, however, revealed that not the whole frontal lobes were affected, but the unusual gyri actually covered some regions corresponding to the normal superior and middle frontal gyri. On both sides the Sylvian sulcus was well developed. The central sulcus was well seen as a border between the unusual and normal cerebral surface. The gyri of

the parietal and occipital lobes behind the central sulcus were relatively of normal size. The postcentral sulcus was better expressed than the precentral sulcus, which was possible to be recognized only in its lower part. The other parts of the cerebral cortex seemed normally developed.

Discussion

Based on the supposed embryonic developmental stage at which the disruption is purported to cause the anomaly, the malformations of cortical development can be classified into three basic groups [2]. Group I (abnormal neuronal and glial proliferation or apoptosis) comprises microcephalies, megalencephalies, cortical dysgenesis with abnormal cell proliferation. Group II (abnormal neuronal migration) includes heterotopia, lissencephaly, subcortical heterotopia and sublobar dysplasia, cobblestone malformations. Group III (abnormal postmigrational development) includes polymicrogyria and schizencephaly, focal cortical dysplasia and postmigrational microcephaly.

The reported autopsy findings are consistent with a case of cerebral polymicrogyria. According to Friede the term “polymicrogyria” defines an excessive number of abnormally small gyri leading to an irregular cortical surface [5]. This condition can be localized to a single gyrus, may involve a part of the hemisphere, may be bilateral and

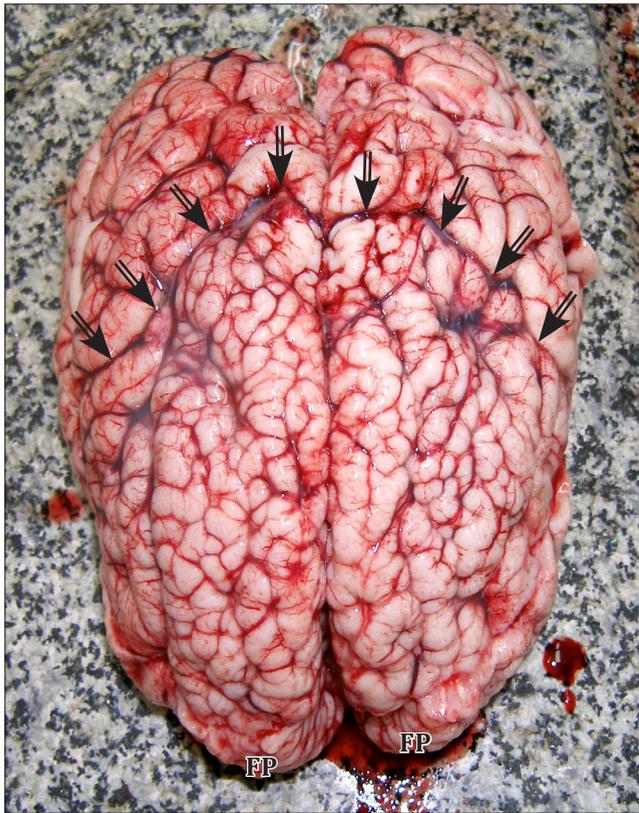


Figure 1. Antero-superior view of the fresh brain at autopsy, showing areas covered by unusually small gyri. **Arrows** indicate the central sulci on both sides. (**FP:** frontal pole)

asymmetrical, bilateral and symmetrical (as in our case) or diffuse throughout the cerebral cortex. According to the lobar topography established, various polymicrogyria syndromes have been described: bilateral perisylvian [6], bilateral parasagittal parietooccipital [7], bilateral frontal [8], bilateral frontoparietal [9] and unilateral perisylvian or multilobar polymicrogyria [10]. The main type is bilateral perisylvian polymicrogyria clinically characterized by a combination of pseudobulbar palsy, spastic tetraparesis, learning difficulties and epilepsy [1].

Polymicrogyria is associated with a wide number of patterns and syndromes and with mutation of several genes [2, 4]. Its pathogenesis is still unknown. The brain pathology comprises at least two major histological types [1]: the most common four-layered cortex with simplified organization and the unlayered cortex. Because the most changes occur in the middle cerebral artery territory, probably in some cases hypoxic-ischemic insult may be related to the genesis of polymicrogyria. A congenital cytomegalovirus infection may also have an important role [1].

The described here case of frontal bilateral polymicrogyria is undoubtedly connected with the status epilepticus [8].

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