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Case Report

Focal Electrical Status in a Rare Case of Unilateral Perisylvian Polymicrogyria

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Abstract

Centroparietotemporal (CPT) spikes in EEG are commonly associated with childhood epilepsies such as Benign Epilepsy with Centrotemporal spikes (BECTS). This EEG finding can rarely be associated with symptomatic peri-rolandic epilepsies due to pathologies such as perisylvian polymicrogyria (PMG). We report a rare case of left perisylvian polymicrogyria presenting with refractory seizures and focal electrical status in EEG. We highlight the electroclinical features that warrant an MRI brain to rule out a possibility of perisylvian PMG in children presenting with seizures and CPT spikes in EEG.

Keywords: Centroparietotemporal Spikes; BECTS; Perisylvian; Polymicrogyria

Abbreviations

BECTS: Benign Epilepsy with Centro-Temporal Spikes; CPT: Centroparietotemporal; MCD: Malformation of Cortical Development; PMG: Polymicrogyria

Introduction

Centroparietotemporal (CPT) spikes in EEG are a hallmark feature of Benign epilepsy with centro-temporal spikes (BECTS). BECTS has two distinct presentations: i) Classical BECTS: characterized by infrequent focal sensorimotor seizures involving the perioral region during sleep [1] and ii) Atypical BECTS: spectrum comprising of atypical benign childhood focal epilepsy (ABCFE), status epilepticus of BECTS, Landau-Kleffner syndrome and epileptic encephalopathy with electrical status epilepticus during sleep [2,3]. Rarely CPT spikes can be associated with symptomatic localisation related epilepsies secondary to structural lesions involving the perisylvian region. We describe a rare case of left perisylvian polymicrogyria (PMG) present with refractory seizures and focal electrical status involving the left CPT region.

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12 year old girl, presented with history of refractory seizures from 3 years of age. She predominantly had diurnal seizures characterised by facial twitching to right, drooling from mouth and clonic jerks involving the right upper and lower limb, followed by post-ictal right sided Todd's palsy. Apart from seizures she had delayed language milestones and learning disability. She had habitual left handedness.

Her EEG showed left CPT spikes with tangential dipole (fronto-central positivity and temporo-parietal negativity) occupying more than 80% of the EEG record amounting to focal electrical status (Figure 1).

MRI brain (Figure 2) showed reduced gyration in the left frontal lobe with thickening of the grey matter involving the insular cortex.

Figure 1: EEG showing frequent spike and wave discharges over the left centroparietotemporal region with tangential dipole as described.

There was irregularity seen in the grey white matter of the posterior part of the left high frontal lobe suggestive of polymicrogyria. MRI features were are suggestive of pachygyria – polymicrogyria spectrum involving the left frontal perisylvian region.

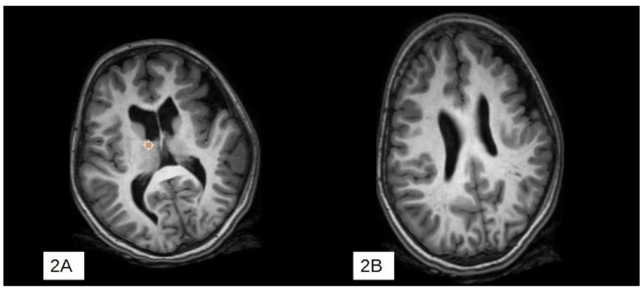

Figure 2A and 2B: MRI brain shows pachygyria - polymicrogyria spectrum involving the left frontal perisylvian region with underlying cortical atrophy.

Discussion

Malformation of cortical development (MCD) are a group of neurological disorders caused due to disruption of the major steps of the cerebral cortical development i.e. cell proliferation/apoptosis, neuronal migration and cortical organization [4-7]. With the advent of modern neuroimaging techniques these disorders have been increasingly recognized as a major cause of medically refractory epilepsy [6]. MCD can be classified into three basic groups [5]. Group I (due to abnormal neuronal and glial proliferation or apoptosis) includes microcephaly, megalencephaly and cortical dysgenesis with abnormal cellular proliferation. Group II (due abnormal neuronal migration) includes heterotopia, lissencephaly, subcortical heterotopia, sublobar dysplasia and cobblestone malformations. Group III (due to abnormal postmigrational development) includes polymicrogyria, schizencephaly, focal cortical dysplasia and postmigrational microcephaly.

The term "polymicrogyria" defines a condition where all or part of the brain surface is divided into an excessive number of abnormally small gyri leading to an irregular cortical surface [8-10]. Various PMG syndromes have been described based on the extent of lobar involvement [9]: bilateral perisylvian, bilateral parasagittal parietooccipital, bilateral frontal, bilateral frontoparietal, unilateral perisylvian, PMG associated with periventricular nodular heterotopia and multilobar polymicrogyria. There are various theories into the pathogenesis of polymicrogyria such as [8-10]: i) hypoxic-ischemic insult ii) congenital cytomegalovirus infection iii) genetic causes including both contiguous-gene and single-gene disorders.

Tangential dipole in EEG indicates deep seated seizure focus from the rolandic sulcus and as described earlier is commonly associated with childhood epilepsies such as classical and atypical BECTS [1-3]. Our case highlights that this EEG finding can rarely be associated with symptomatic peri-rolandic epilepsies due to pathologies such as perisylvian PMG. The red-flags which indicate a possibility of symptomatic rather than benign rolandic epilepsy as seen in our case include: i) clinical features such as language developmental delay and odd semiological features such as Todd's palsy ii) EEG findings of focal CPT spikes.

Conclusion

Perisylvian PMG should be considered as a differential diagnosis in epilepsy syndromes in children having CPT spikes in EEG. MRI brain to rule out a possibility of perisylvian PMG should be recommended in cases of BECTS having atypical electroclinical features as highlighted in our case.

Conflict of Interest

The authors report no conflicts of interest.

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