Bilateral symmetrical frontal polymicrogyria
Published on 18.07.2012

Clinical History:

The patient has had epilepsy since age 11, with 3-4 seizure episodes per month, refractory to anti-epileptic medications. He has no neurologic deficit. He is suspected to have Ehlers-Danlos syndrome with positive family history in a brother and sister.

Imaging Findings:

There is relatively symmetrical abnormal increase in the cortical infolding and apparent cortical thickening in bilateral anterior and inferior frontal lobes and frontal opercula, with slightly "open" Sylvian fissures (Fig. 1). The subcortical white matter in the involved lobes is reduced with a few tiny T2-hyperintense foci. The mid-frontal and more posterior portions of the brain are intact, and the peri-Sylvian cortex is spared (Fig. 2). No schizencephaly or calcification is seen. The ventricular system is prominent with note of cavum septum pellucidum et vergae.

Mild cerebellar atrophy could be related to chronic antiepileptic drug use (Fig. 3).

Discussion:

Polymicrogyria is a cortical malformation, which results from abnormal post-migrational development [1]. It has been attributed to ischaemic injury or infection during gestation, although genetic contributions from chromosome deletions and X-linked mode of inheritance have been described. Bilateral frontal polymicrogyria has been reported previously in Ehlers-Danlos syndrome [2, 3].

Patients with polymicrogyria typically present with seizure, but may also have microcephaly, developmental delay, and even motor deficits. Distribution of lesions and severity of involvement do not always correlate with clinical manifestation and onset of presentation unless generalized, and it is not uncommon for patients with bilateral involvement to present after the 1st decade [4]. Imaging is important as it allows the diagnosis to be made, obviates biopsy, and may reveal additional non-cortical abnormalities, which could suggest the underlying aetiology in the absence of other congenital abnormalities or inborn errors of metabolism [1]. Imaging findings can also facilitate decision-making for genetic testing [5, 6].

CT is limited and may only reveal an irregular cortical surface. MRI is the most sensitive imaging technique and shows over-folding or thickening of the cortex with "stippling" at the grey-white matter interface. The presence of schizencephaly and calcifications suggests an infectious or vascular cause, while their absence indicates a genetic or disruptive aetiology [1]. In the latter group, further classification is made based on the topographical distribution of
polymicrogyric lesions. In a study of 328 patients, bilateral frontal polymicrogyria was the 4th most common pattern; of these, more than half of patients had lateral ventricle dilatation and significant association with prominent perivascular spaces [4]. The frontal-only subtype is considered a less severe form compared with frontoparietal involvement [6].

Genetic testing is recommended though currently limited to chromosomal analysis in patients with the perisylvian [5] or frontoparietal [6] types of polymicrogyria. Management remains directed towards seizure control, and includes genetic counselling.

**Differential Diagnosis List:** Bilateral symmetrical frontal polymicrogyria, Bilateral frontoparietal polymicrogyria, Bilateral perisylvian polymicrogyria

**Final Diagnosis:** Bilateral symmetrical frontal polymicrogyria

**References:**


Figure 1

Description: Right and left sagittal T1W images demonstrate bilateral anterior inferior frontal involvement with preserved mid-frontal and more posterior portions of the brain, as well as sparing of the perisylvian regions. Origin: Abrigo J, Department of Imaging and Interventional Radiology, The Chinese University of Hong Kong
Figure 2

Description: There is mild cerebellar atrophy which could be related to chronic antiepileptic drug use.

Origin: Abrigo J, Department of Imaging and Interventional Radiology, The Chinese University of Hong Kong
**Figure 3**

**Description:** There is bilateral symmetrical cortical thickening in the anterior frontal lobes, accompanied by decreased white matter with a few small T2 hyperintense signals. **Origin:** Abrigo J, Department of Imaging and Interventional Radiology, The Chinese University of Hong Kong