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Clinical Case Open Access

Unilateral Perisylvian Syndrome and Unilateral Nodular Heteropia: 2 Cases of Neuronal Migration Disorders Presenting as Adulthood Partial Complex Seizures

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Clinical Case Presentation

The development of the central nervous system is a complex process, organized in the following steps: primary neurulation (3-4 weeks), prosencephalic development (2-3 months), neuronal proliferation (3-4 months), neuronal migration (3-5 months), organization (5 months of gestation to after birth), and myelination (after birth) [1].

Neuronal migration consists of nerve cells mobilizing from their sites of origin in the ventricular and sub ventricular zones to their final localization. If such process is disrupted, the central nervous system inappropriately develops, manifesting as one of the following five neuronal migration disorders (NMD):

(1) Lissencephaly

(2) Heteropia

(3) Polymicrogyria

(4) Schizencephaly and

(5) Focal cortical dysplasia

The first case is a nineteen-year-old female patient suffering from partial complex seizures, secondary to sub ependymal grey matter heteropia (Figure 1) along the left ventricle. Heteropia, an inappropriate neuronal migration from the ventricles, can be either periventricular or subcortical; the location of the heteropia is often bilateral and placed along the ventricles [2]. 90% of patients have epilepsy as their main problem.

The second case is a 64-year-old man suffering from partial complex seizures secondary to unilateral perisylvian syndrome, a type of polymicrogyria presenting as cortical dysplasia along the left lateral sulcus (Figure 2). Bilateral cases are more common and more severe, presenting with pseudo bulbar palsy, mental retardation and epilepsy. Unilateral cases may be associated with septo-optic dysplasia.

Learning Points

1. Neuronal migration disorders are five: Lissencephaly, heteropia, polymicrogyria, Schizencephaly and focal cortical dysplasia.
2. They are usually associated with psychomotor retardation and intractable seizures. However, adult cases with well controlled, focal seizures can also be the main clinical manifestation.

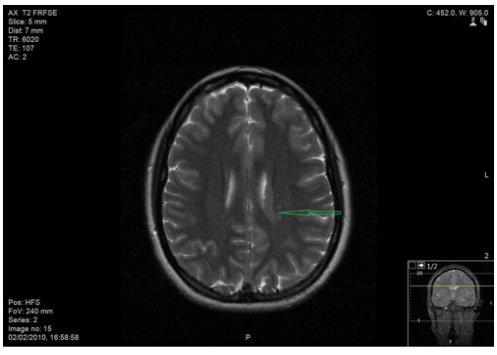

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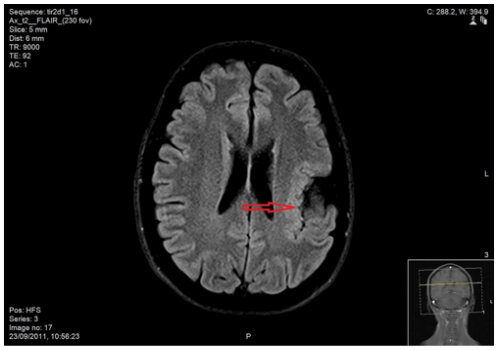
Figure 1: MRI: Sub ependymal grey matter heteropia.



128x90mm (300 x 300 DPI)

Figure 2: MRI: Left unilateral perisylvian syndrome.

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