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Case Report

Periventricular nodular heterotopy of the gray matter: A case report☆

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ABSTRACT

Periventricular focal nodular heterotopia is a rare secondary cerebral distortion caused by the interruption of neuronal migration from the periventricular germinal zone to the cortex during the fetal period. Clinically, it may manifest as epilepsy resistant to pharmacological treatments or rarely as mental retardation. We report a case of a six years-old male child who was subject to the intensive care unit for the management of refractory epilepsy. The diagnosis was done by magnetic resonance imaging of the brain, which revealed a nodular periventricular heterotopia of the gray matter. After the management of the status of epilepticus, the child remained spastic, aphasic with no contact with his environment.

Introduction

Periventricular nodular heterotopy of the gray matter is a rare malformation of the cerebral cortex, responsible for epilepsy and mental retardation. Its incidence is still unknown. Positive diagnosis is based on magnetic resonance imaging (MRI) and mapping of ectopic nodules.

Case summary

A six-year old male child was referred to the ICU for refractory epilepsy. The patient was known to be epileptic, treated with sodium valproate since the age of 3; with good psychomotor development.

Upon admission; the patient was unconscious in a post-critical coma, without hemodynamic or respiratory instabil-

Abbreviations: CT, Computed tomography; MRI, Magnetic resonance imaging; ICU, Intensive care unit; PNH, Periventricular nodular heterotopia.

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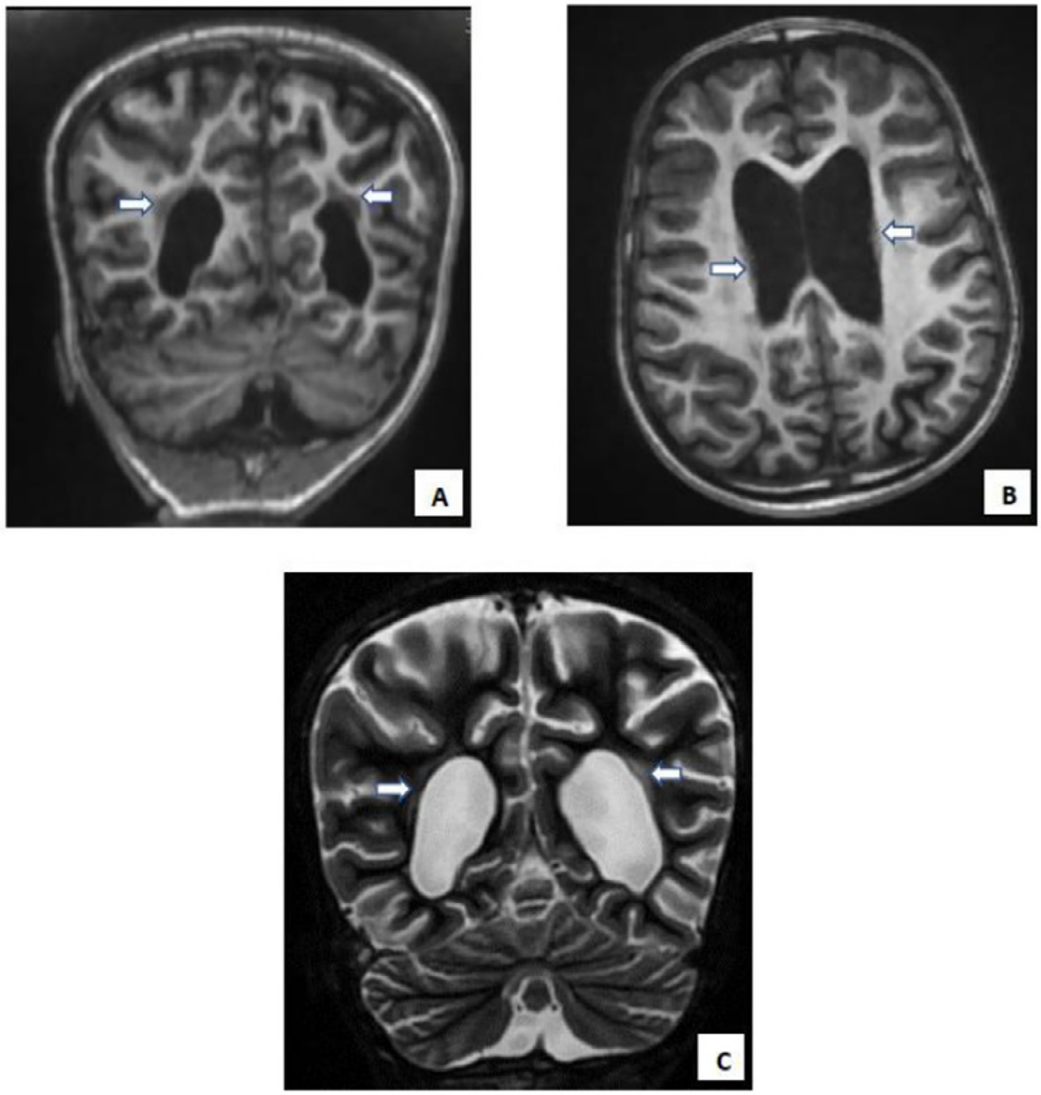

Fig. 1 – Brain MRI in 3DT1 axial and coronal sequence (A, B), T2 coronal sequence (C): presence of nodular lesions (large white arrows) in the periventricular area in T2 isosignal and T1 hyposignal, signal similar to that of the grey matter. Festooned aspect of the ventricular wall with triventricular hydrocephalus and cerebral atrophy.

ity. Somatic examination showed an apyretic, eutrophic child without dysmorphia.

Cerebral computed tomography (CT) scan with contrast showed diffuse cerebral edema, and cytobacteriological and biochemical study of cerebrospinal fluid was normal.

Given the persistence of focal tonic-clonic seizures, under phenobarbital alone, then in combination with sodium valproate and clobazam, a brain MRI was performed which showed nodular periventricular heterotopia, diffuse cerebral atrophy, especially in the hippocampus, associated with unobstructed quadriventricular hydrocephalus (Fig. 1).

A combination of clobazam and levetiracetam at maximum dose stopped the clinical seizures. Moreover, an electroencephalogram was requested in front of tachycardia, arterial hypertension and polypnea associated with desaturation. It revealed a poorly organized and slowed down tracing testifying to a diffuse suffering, with paroxysmal discharges in the bilateral temporal area, predominantly right.

The evolution was as such: absence of return to the previous state with rupture of contact, aphasia, diffuse muscle spasticity as well as abolition of swallowing reflexes.

Discussion

Gray matter heterotopia is a congenital malformation during cortical development characterized by the reunification of normal neurons outside the cortex. This migration disorder affects neuroblasts during their migration from the germinal zone between the 10th and 16th week of gestation [1-3].

Heterotopic cells can take a laminar or nodular form. According to the level of migration, 3 categories are dis-

tinguished: periventricular (subependymal), subcortical, and banded or double cortical heterotopic [3].

Periventricular nodular heterotopia is the most frequent type. These are neurons that have never begun migration, remain adjacent to the lateral ventricles with macroscopically normal cerebral cortex, and retain their function. There is evidence that these heterotopic nodules can form white matter connections between themselves and the overlying cortex [2,4].

Abnormalities in neuronal migration have been reported in association with genetic abnormalities such as point mutations in the filamin A gene (FLNA), vascular abnormalities, environmental abnormalities such as methylmercury poisoning and exposure to ionizing radiation. Other associations, such as fetal alcohol syndrome, neurofibromatosis type I and trisomy 13; were also noted [5,6].

Periventricular nodular heterotopia is most often revealed by seizures, often partial and drug-resistant, beginning between the ages of 2 and 14 years with a mean age of 14 years. Cognitive disorders and psychomotor delays are less frequent in PNH but remain one of the revealing signs of the pathology [7]. Despite an often normal IQ, patients with PNH often present reading disorders (dyslexia), including widely distributed heterotopic reading [4,8]. Spasticity was described in a single patient at age 2 years with generalized seizures (a study of 33 patients) [7].

When studying the electroencephalogram of patients with PNH, the abnormalities found are often unilateral or bilateral temporal epileptic discharges [7]. Magnetic resonance imaging (MRI) shows isointense heterotopic clusters in the cortex, without enhancement after gadolinium injection. Other abnormalities may be associated such as hydrocephalus and cerebral atrophy [9] which was observed in our patient.

Surgical treatment is necessary in case of drug-resistant seizures to usual treatments and consists of excision by stereotactic techniques of small heterotopic nodules [10].

Conclusion

Periventricular nodular heterotopia is a defect in the migration of the gray matter. It is a rare cause of epilepsy resistant to the usual pharmacological treatments, as well as other manifestations such as dyslexia. The diagnosis is made by MRI and the response to treatment depends on the location, number, and distribution of the nodules.

Patient consent statement

I confirm in my own words that the legal representer of the patient whose case is reported in this article, gave his legal consent and declare that she is informed of all the written information related to her son's medical case, and accept it to be published.

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