Neuroimaging of Gray Matter Heterotopias: Study of Eight Patients

I. Chaouche[1]*, A.Akammar[1], N.Bouardi[2], M.Haloua[1], B.Alami[2], Y.Lamrani[2], M.Maaroufi[2], and M.Boubbou[1]

[1]Department of Mother and Child radiology, CHU Hassan II, Sidi Mohammed Ben Abdellah University, Fez, Morroco
[2]Department of Adult radiology, CHU Hassan II, Sidi Mohammed Ben Abdellah University, Fez, Morroco

*Corresponding Author**
I. Chaouche
Department of Mother and Child radiology, CHU Hassan II, sidi Mohammed Ben Abdellah University,Fez,Morroco
E-mail: liamsiismail3998@gmail.com

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Abstract

Heterotopy of the gray matter is a very rare cause of epilepsy in children. It is a disorder of neuronal migration during embryonic life, responsible for the presence of neurons outside the cortex. Gray matter heterotopias are common forms of cortical developmental malformations characterized by the presence of cortical neurons in abnormal positions.
They are caused by the arrest of migration of neurons from the germinal matrix in the wall of the lateral ventricle to the developing cerebral cortex between the 6th and 16th week of gestation. It is usually discovered during the evaluation of children or young adults with refractory epilepsy. In Our study we report a series of 08 patients collected in our hospital within a period of 6 years from 2016 to 2022, diagnosed with heterotopia of the grey matter, however, six patients had benefited from an MRI and two from a CT scan.

Keywords: Epilepsy • Heterotopy • Gray matter • CT-scan • MRI

Introduction

Neuronal heterotopias are rare cerebral malformations defined by the presence of neurons outside the cerebral cortex. This disorder of neuronal migration that affects the neuroblasts during their migration between the ependymal and the cortex occurs between the 10th and 16th week of gestation [1, 2]. It is usually discovered during the evaluation of children or young adults with epilepsy, children with neurodevelopmental abnormalities, or as an incidental finding.
Imaging is the key to positive diagnosis by the CT or MRI [3].

Materials and Methods

Retrospective study of 08 cases investigated in our department of Mother and Child radiology at the Hassan II university hospital of fez, within a period of 6 years from 2016 to 2022.All our patients underwent neurological examinations as well as brain CT angiography and/or MRI.

Results

The age of onset of seizures ranges from four months to five years and age at diagnosis ranges from four years to 20 years with an average age of 9 years, with a slight female predominance with a ratio of 1.66%. Clinically six out of eight patients consulted for refractory epilepsy, one patient consulted for head trauma and another patient for chronic headaches. The epilepsy is the most frequent mode of revelation, it was isolated in four patients, and associated with psychomotor delay in two cases. However the clinical examination revealed the presence of generalized seizures in five patients and partial in one case. The clinical examination was without particularity in the patient who consulted within the context of cranial trauma and the one who consulted within the context of chronic headaches.
Electroencephalogram was performed on a single patient, showing a pathological pattern with diffuse micro-waves, however other patients do not benefit from an electroencephalogram, due to its high cost and difficult access in emergency. The biochemical tests in our study does not show any abnormality.
Our patients had benefited from an imaging by MRI and cerebral CT-scan, six patients had benefited from an MRI and two from a CT scan. The exploration protocol of MRI adopted in our department for epilepsy includes: T2 and T2* axial slices, axial and coronal slices, FLAIR and a three-dimensional T1 acquisition. The brain scan was performed in two patients; the first one was consulted for a head trauma with fortuitous discovery of a heterotopy of the grey matter and the second one was consulted for chronic headaches and the third one for an epileptic seizure disorder.
The types of heterotopias found are the periventricular heterotopy in 5 cases (Figure 1-3).

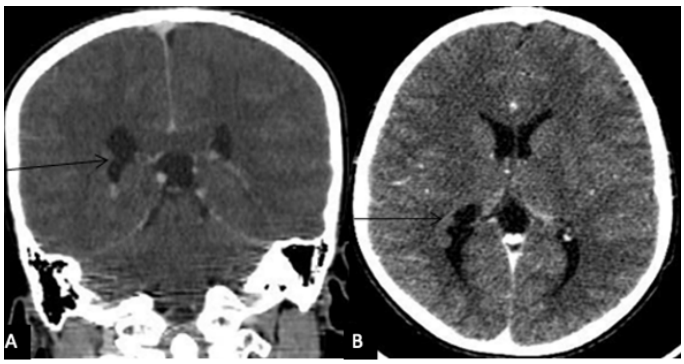

Figure 1. 1A & B)Young child of 05 years old, admitted for chronic headaches, with the presence of periventricular nodular lesions of the same density of the cortex on the CT scan.

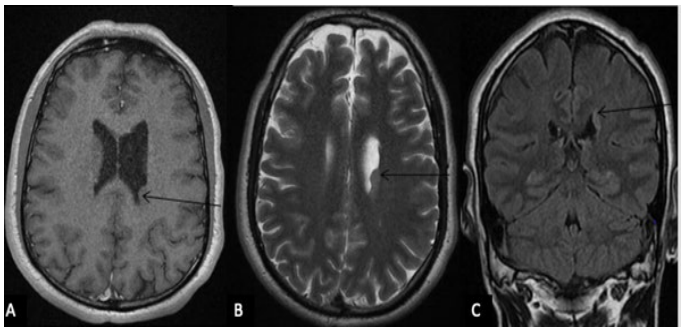

Figure 2. 16-year-old girl, partial phramco-resitant epilepsy with MRI on T1 2A) T2 2B) and FLAIR 2C) sequences, periventricular nodular form of gray matter heterotopia.

infertility.34.9% of respondents that the PCOS would have a detrimental impact on self- confidence. 41. 1% of students agreed that PCOS will make them feel anxious or unhappy. Statement that receiving diagnosis of PCOS need emotional support is disagreed by 41. 6% of respondents. 32

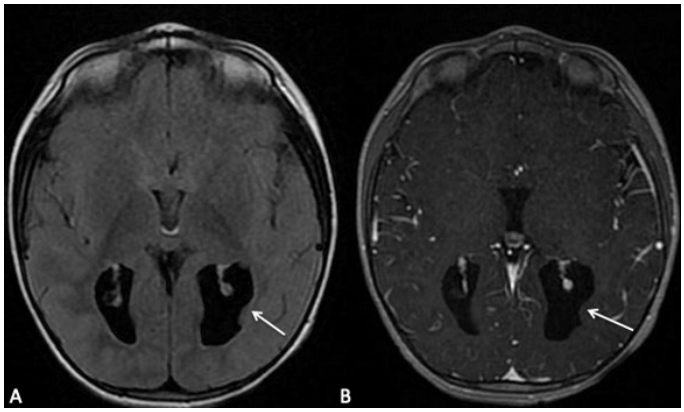

Figure 3. 3 A&B) 4 years old child admitted for a generalized epileptic seizur.

Heterotopy in a band with double cortex in two patients and subcortical nodular heterotopy in one case However, in our series we found an association with other anomalies in two cases: the first one is agenesis of the corpus callosum and the second one with a cavernoma (Figure 4).

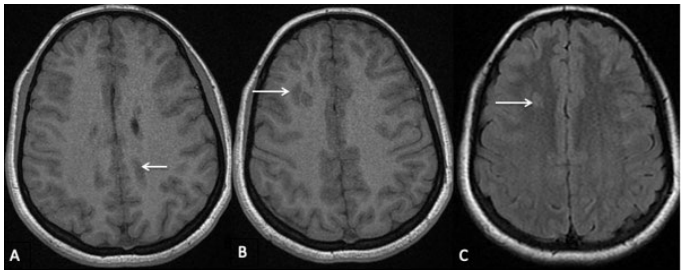

Figure 4. Young adolescent girl, 18 years old, consulted for drug-resistant generalized epilepsy 4A) MRI with T1 SEQUENCES 4B) FLAIR sequence 4C) Presence of nodular lesions, isointense to the SG at the different sequences, in the periventricular area.

The management normally consists of a surgical removal of the epileptogenic focus, however in our series, the initial management was to put all patients under antiepileptic treatment, except the one who had consulted for traumatic context in which we did not indicate antiepileptic treatment since he was asymptomatic. Six patients remained stable on treatment and one patient was lost to follow-up.

Discussion

Neuronal heterotopias are rare cerebral malformations defined by the presence of neurons outside the cerebral cortex. This disorder of neuronal migration that affects the neuroblasts during their migration between the ependymal and the cortex occurs between the 10^{th} and 16^{th} week of gestation, The development of medical imaging means, especially MRI, making the diagnosis of cortical malformations easier.

Gray matter heterotopias are common forms of cortical developmental malformations characterized by the presence of cortical neurons in abnormal positions [4]. They are caused by the arrest of migration of neurons from the germinal matrix in the wall of the lateral ventricle to the developing cerebral cortex between the 6th and 16th week of gestation [5].

It is usually discovered during the evaluation of children or young adults with refractory epilepsy. This malformative pathology affects girls more than boys [6].The pathogenesis is a subject of discussion in the literature, heterotopia is due to an anomaly of neuronal migration following a genetic mutation of the genes coding the neuronal migration or infectious by cytomegalovirus.

They are revealed by an epilepsy associated with or not psychomotor delay, sometimes of incidental findings. Imaging is the key to diagnosis, by revealing abnormally positioned gray matter. RI is the reference examination, more precise than CT scan. Is manifested by nodules in T1/T2 is signal without enhancement after contrast [7]. Imaging shows a regional or focal parenchymal abnormality that follows gray matter on all MRI pulse sequences but is abnormal in location, but usually within the white matter, along glial migration pathways . This can be nodular or mass-like, focal or multifocal, or band-like [8].In our series, the periventricular nodular form is the most frequent, which is consistent with the data in the literature. Few series were published in this sense, which indicates the rarity of this pathology.

Nodular sub ependymal heterotopias pose the differential diagnosis with sub ependymal hamartomas. The differential diagnosis with sub ependymal hamartomas whose signal is different from that of the gray matter. The white matter, basal ganglia and cortex are normal. The differential diagnosis is also made with phacomatoses, in particular the tuberous sclerosis of Bourneville.

This pathology is often drug resistant and requires focal therapeutic intervention. An invasive electroencephalogram study is usually necessary to delineate the epileptogenic zone. The prognosis depends on location and extent of heterotopy, associated malformations and severity of seizures.

Conclusion

Heterotopias of the gray matter may be diffuse cortical or sub ependymal. They are revealed by an epilepsy associated or not with psychomotor delay. MRI is an element of the workup, it allows to make a positive diagnosis, to appreciate the thickness of the heterotopic ribbon and to look for associated malformations.

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