Disorders of cortical formation: MR imaging features

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CHU BAB EL OUED
ALGER
Disorders of cortical formation

Heterogenous group:
abnormal structure of cerebral cortex

MCD may be due to:
genetic factors
prenatal injury: trauma, infection or teratogens.

Important cause of epilepsy and developmental delay.
<table>
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<tr>
<th>Development of Cerebral cortex</th>
<th>Embryogenesis From the 2nd Month of gestation Maturation to 24 month after birth</th>
<th>3 main stages</th>
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Embryology

Development of Cerebral cortex

Embryogenesis From the 2nd Month of gestation

Maturatio n to 24 month after birth

3 main stages

- Proliferation
- Migration
- Organization:
  Laminal aggregates/
  vertical columns

Proliferation of neurons and glial cells

Migration from periventricular zone to cortex

Physical biology of human brain development

Frontiers in Cellular Neuroscience
Prenatal detection *(rare)*

Characteristic basic US findings:

- Microcephaly
- Macrocephaly
- Ventriculomegaly
- Asymmetry of brain structures

*Raise suspicion*
Classification of disorders of cortical formations
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- The classification scheme of DCF has tremendously evolved in the past years on the basis of neurogenetics.

- This classification was updated by Barkovich et al in 2001, 2005 and in 2012, based on the causative gene rather than by the clinical phenotype.

- In 1996, Barkovich proposed a classification scheme for DCF, based on the first step at which the developmental process was disturbed. (PMO)
Classification of disorders of cortical formations

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Classification :

malformations liées à des anomalies de la prolifération ou de l’apoptose neuronale ou gliale.

• Microcephałies congenitales (-prolif ou + apoptose)
• Megalencephalie /hemimegalencephalie.
• Dysplasie focale (anlies de prolif type taylor)
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I- Anomaly of prolifération
I-1. Microcephaly

decreased cell production or increased apoptosis in the germinal zone of the cerebral cortex.
I-1. Microcephaly

The cerebral cortex may be of normal thickness or decreased but the enlargement of the SAS is constant.
I-1. Microcephaly

The pattern of Gyration is mostly normal but may be simplified
I-1. *Microcephaly*

Possible associations:
CC atrophy or dysgenesis,  
PMG or heterotopia
I-2. Megalencephaly & Hemimegalencephaly

ch. by enlarged and dysplastic hamartomatous overgrowth of both, one or part of a cerebral hemisphere.

It results from: + ++ neuromas or - apoptosis.

It may be an isolated or associated with syndromes such as:
  - NF type I,
  - Klippel-Trenaunay syndrome, and
  - Tuberous Sclerosis
Hemimegalencephaly

The cerebral cortex may be normal or dysplastic (PMG, lissencephaly, or heterotopia).
Hemimegalencephaly

Blurred GM–WM junction, with variable degree of abnormal T1 and T2 white matter signal intensity (due to heterotopia and astrocytosis).
Hemimegalencephaly

The lateral ventricles are enlarged with a characteristic shape of the frontal horns (appears straight and pointed anteriorly).
I- 3. Focal cortical dysplasia (Taylor type II)

FCD is a heterogeneous group of lesions characterized by the presence of abnormal neurons & glial cells within a localized cortical region.

Clinically: Intractable seizures.

Mild forms to severe forms (with marked cortical dyslamination)
FCD appears as a localized area of **cortical thickening** with a **blurred GM-WM junction**.
I-3. Focal cortical dysplasia

There is macrogyria: an abnormal gyral pattern (widened or deep sulci)
There is a subcortical funnel-shaped focus of abnormal signal intensity extending from the GM-WM junction to the superolateral margin of the lateral ventricle.
I-3. Focal cortical dysplasia

Transmantle sign (FCD)

- Pachygyria
- Cortical thickening
- Blurred GM-WM
- Transmantle sign

Radial band sign TS

- Pachygyria (pellizi 1,2)
- No cortical thickening
- Intact GM-WM
- Subcortical triangular WM, HT2 tubers (ca ++)
# Classification of disorders of cortical formations

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<td>Partial : Band heterotopia</td>
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Anomaly of undermigration (Transmantle)

The cortex is abnormally thickened and sulcation largely absent with 2-4 instead of normal 6 layer cortex.
II-A-1. Complete (classic) lissencephaly

Cerebral configuration is oval or hourglass with shallow Sylvian fissures due to lack of or incomplete operculization.
II-A-1. Complete (classic) lissencephaly

The cortex is thick because it encompasses radial columns of the arrested cells.
II-A-1. Complete (classic) lissencephaly

The subcortical WM is thin, with a lack of the normal gray-white matter interdigitation.
II-A-1. Complete (classic) lissencephaly

The subcortical WM is thin, with a lack of the normal gray-white matter interdigitation.
II-A-1. Complete (classic) lissencephaly

There is a circumferential band of high signal intensity on T2-WI, most prominent in the parieto-occipital cortex, corresponding to a sparse cell zone with increased water content.
Lissencephaly variant

Associated with a large spectrum of extra cortical malformations: hypoplastic cerebellum, and a small brain stem with Dandy-Walker syndrome.
II-A-2. Band heterotopia

Or double cortex, Rare malformation
Ch by a band of GM in the middle of the WM, located somewhere between the cortex and the ventricles.
II-B Peri ventricular Heterotopia: Anomaly of the neuro ependymal tissue
II-B-1. Nodular Periventricular Heterotopia

Group of neurons and glial cells located along the ependyma of the ventricles
Subependymal heterotopia / epilepsy, +/- cognitive impairment.

MRI:
Nodules of grey matter lining the ventricular wall, protruding in the ventricle giving it an irregular appearance, Focal, uni or bilateral.
Nodular Periventricular Heterotopia

Heterotopia sub ependymal

Tuberos sclerosis
Heterotopia : péri ventriculaire diffuse bilatérale

Diffuse and bilateral
Affecting only girls (genetic mutation located on the X chromosome)
II-B-3. Subcortical Heterotopia

Less frequent, different morphology, probably of different origin than PVH

À groupe of neurons and glia located in the white matter, more or less spread, can appear multinodular or curvilinear
II-B-3. Subcortical Heterotopia

Clinically: epilepsy, Intellectual & motor deficit depending on the size of the aggregate, and its masse effect on the cortex

À groupe of neurons and glia located in the white matter, more or less étendu, multinodular or courbe lineaires
II-C. Lissencephalies type II or Cobblestone complex

Post migrational or end migrational disease

The cortical surface is pebbled (due to presence of ectopie neuroglia in the meninges), Usually associated with muscular distrophy and eye abnormality
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III-1. Polymicrogyria

PMG are a heterogeneous group of malformations. They are characte‌rized by:

- an irregular cortical surface,
- an excessive number of small partially fused gyri, separated by shallow sulci.
III-1. Polymicrogyria: « focal or diffuse »

4 Subgroups of PMG: “Association”

1. Schizencephalic slits or Calcifications: Infectious origin
2. No schizencephaly & calcifications: Genetic origin
3. Malformative syndrome
Schizencephaly consists of a linear slit, lined with Gray matter substance, crossing the entire hemisphere, from the lateral ventricle to the outer surface of the cortex.
III-2. Schizencephaly: Closed Type 1, Open Type 2
Ill.1. Polymicrogyria

Apart from the Irregular surface (thick cortex + numerous gyri + shallow sulci) in type I of PMG there’s an Enlargement of SAS.
MR imaging is essential to demonstrate the morphology, distribution, and extent of disorders of cortical formation.

Moreover, it can identify the associated congenital anomalies and related syndromes and suggest the genetic abnormalities.
1/ Disorders of Cortical Formation: MR Imaging Features Abdel Razek
AJNR 30 Jan 2009

2/ Apport de l’IRM dans les malformations cérébrales Docteur Bennouna Siham, Fes, Maroc 2015

3/ barkovitch radiopediatry

4/ radiopedia.