Double cortex syndrome - A case report

Published on 06.08.2013

DOI: 10.1594/EURORAD/CASE.11143
ISSN: 1563-4086
Section: Neuroradiology
Area of Interest: Neuroradiology brain
Procedure: Diagnostic procedure
Imaging Technique: MR
Special Focus: Congenital Case Type: Clinical Cases
Authors: Chethan Belgur, Yugandhara Shah, Ranganath K
Patient: 9 years, female

Clinical History:

A female child aged 9 years presented with history of seizures and developmental delay.

Imaging Findings:

On T2 weighted axial and coronal sequence note the presence of diffuse laminar band of fairly thick grey matter deep to cortex and almost completely paralleling the cortex giving a “cortex within cortex” or “double cortex” appearance. Between two layers of the grey matter there is a stripe of white matter, which appears hypointense on T2 weighted sequences. The inner grey matter is completely differentiated from the lateral ventricles with an intervening stripe of white matter, which appears hypointense on T2 weighted sequences. Note accentuated perivascular spaces in this intervening white matter. The bilateral basal ganglia appear normal. The overlying cortex shows shallow sulci across bilateral posterior parietal regions, more so across the right side resulting in shallow gyri with smooth outer surface - mild lissencephaly. The bilateral lateral ventricles appear dilated.

Discussion:

Subcortical band heterotopia also called as “Double cortex syndrome” [2] is one of the disorder resulting due to the disturbances in the neuroblast migration and is the prominent cause of epilepsy and abnormal neurological development [1]. It is now classified in the agryia-pachygyria-band spectrum [1]. It occurs very rarely in males. It usually occurs due to mutations in the double cortin gene, (DCX) (Xq22.3-q23) most commonly, during which the band heterotopia occurrence predilection is more towards anterior aspects of brain and much less frequently in the LIS1 (17p13.3) gene, during which the band heterotopia occurrence predilection is more towards posterior aspects of brain [2]. The subcortical heterotopia is further grouped depending on the thickness and extension of the heterotopic cortex as follows: (1) thin partial frontal SBH with no involvement of the posterior regions; (2) thin partial posterior SBH with no involvement of the frontal regions; (3) medium or thick intermediate SBH that is always more prominent posteriorly; (4) diffuse thin SBH; (5) diffuse medium or thick SBH; and (6) anterior pachygyria that merges into posterior SBH [2]. The patient most commonly present with epilepsy and is mostly intractable in about 65% of cases [1].

CT features:
CT shows an abnormal band like hypoattenuating area in the region of white matter [3].

MRI features:
MRI is the diagnostic modality of choice due to its excellent capacity of grey and white matter differentiation. MRI in
cases of complete subcortical band heterotopia shows a four layered appearance of cerebral parenchyma which usually extends from the frontal to the occipital region. The four layers from outside to inside are normal cortex, intervening white matter, heterotopic band of grey matter and periventricular white matter. The periventricular white matter shows an abnormally smooth interface with a thick diffuse layer of heterotopic grey matter [3]. The normal cerebral cortex and bilateral symmetrical circumferential subcortical layer of band heterotopias being separated from each other by a thin white matter band also shows a characteristic 3 layer cake appearance. [4]

**Differential Diagnosis List:** Subcortical band heterotopia, Pachygyria, Subependymal nodular heterotopias

**Final Diagnosis:** Subcortical band heterotopia

**References:**


Description: T2 weighted axial image shows diffuse laminar band of fairly thick grey matter deep to cortex extending from the frontal to occipital lobe and paralleling the cortex showing a “double cortex” appearance. Origin: Ragavs diagnostic and research centre
Description: Axial T2 w image shows dialated lateral ventricles. Origin: Ragavs diagnostic and research centre
Description: T2 w coronal image shows band of grey matter deep to cortex with a intervening white matter in between and periventricular white matter shows a four-layered appearance of cerebral parenchyma. Origin: Ragavs diagnostic and research centre
**Description:** T2 weighted coronal image shows shallow sulci across bilateral posterior parietal region, more so across right side resulting in poorly formed gyri with smooth outer surface - mild lissencephaly.

**Origin:** Ragavs Diagnostic And Research Centre