Classical Lissencephaly
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History
Newborn male with seizures.

Diagnosis
Classical Lissencephaly

Discussion
The germinal matrix lines the ventricular margins and gives rise to neuroblasts. Around 8 weeks gestation, neuroblasts begin to proliferate and associate with specialized bipolar glial cells. The glial cells extend from the ventricular margin to the pial surface and provide a scaffolding to allow peripheral migration of neuroblasts. Newly formed cells migrate outward through pre-existing cell layers to reach superficial cortex. Neuronal migration in the cortex results in 6 cell layers and is completed by 6–8-months gestation.

Lissencephaly is a devastating brain malformation associated with seizures, profound developmental delay, and often death. Lissencephaly associated with facial dysmorphism is referred to as Miller-Dieker syndrome and is associated with deletions of chromosome 17p13.3 (LIS1 gene). The LIS1 gene is responsible for functional dynein and normal formation of the glial scaffolding.

The lissencephalic brain has total or near complete absence of sulcation. The lack of cerebral convolution results in a vertical sylvian fissure and a "figure of eight" appearance of the cerebral hemispheres. Histologically, the cortex has four instead of the normal six cell layer cortex. Counterintuitively, the two most superficial layers of cortex are formed early by the neurons that migrated normally, whereas the deeper layers are composed of neurons formed later that did not migrate. The third layer contains astrocytes, oligodendroglial cells, and dysplastic neurons. The fourth layer is the densely cellular and radially oriented.

Findings
MR-Axial T2 images show a thin superficial neuronal layer separated from a thick deep neuronal layer by a hyperintense cell-sparse layer. Subependymal nodular heterotopias represent the extreme form of neuronal dysjuction (migratory arrest).

Reference
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