



Heterotopic Gray Matter

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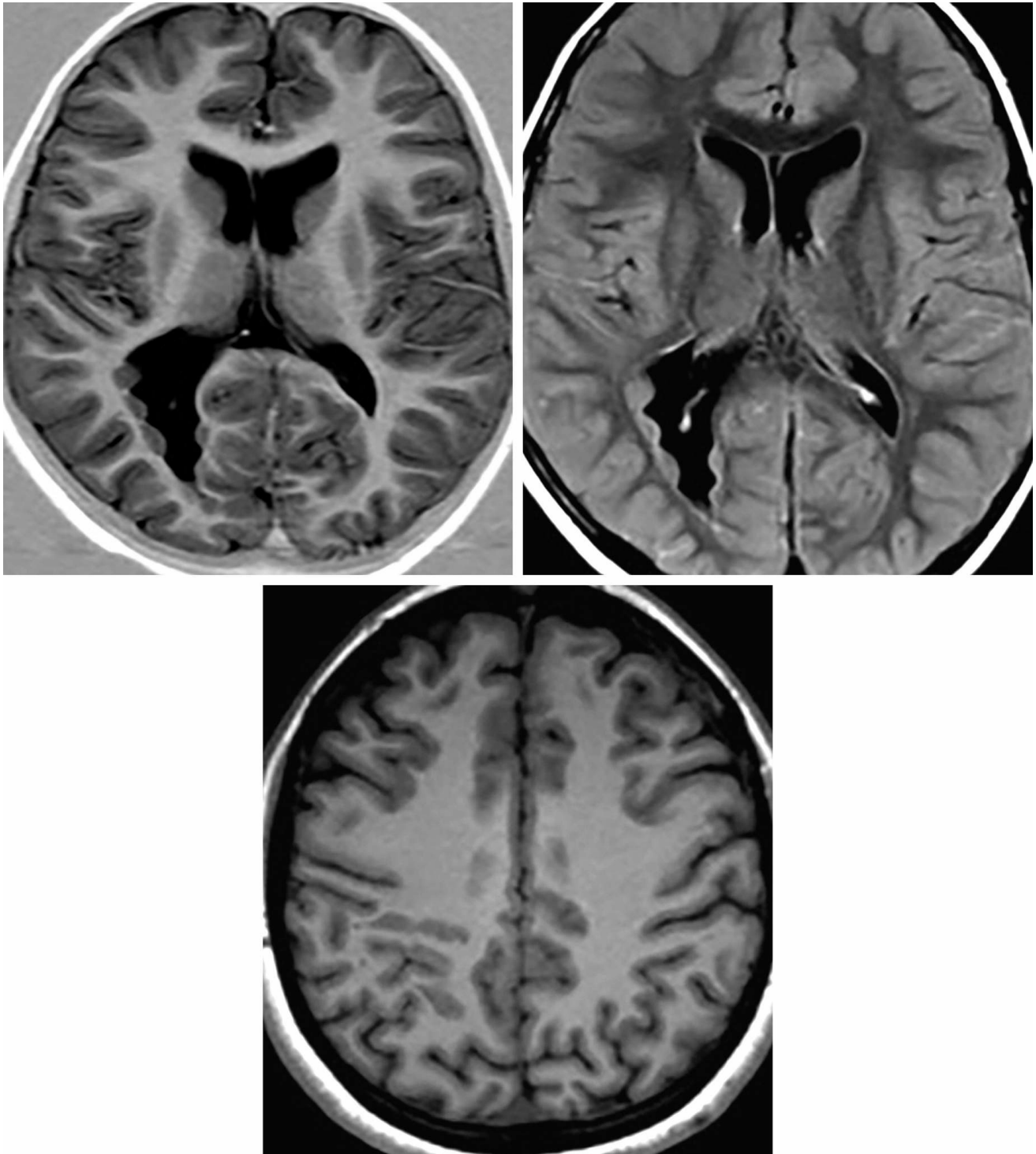


Figure 1: Phase-sensitive (T1-weighted) inversion recovery (top left) and FLAIR (top right) sequences demonstrate a cluster of nodules along the ependymal surface of the posterior horn of the right lateral ventricle.

They have signal characteristics matching gray matter. (Bottom) A T1-weighted image of this patient also demonstrates a band of heterotopic gray matter within the right perirolandic white matter.

Description

- Disrupted migration of neurons from periventricular germinal zone to the cortex

Pathology

- Periventricular nodular type
 - *FLNA* gene commonly involved on chromosome Xq28
- Band-like heterotopia/lissencephaly
 - Deletion of *LIS1* on chromosome 17p13.3 or *DCX* on chromosome Xq22.3-q23

Clinical Features

- Symptoms
 - Young child with variable developmental delays and seizures
- Age
 - Severe cases present earlier in life
 - Typically present by the third decade of life
- Gender
 - Male > female
- Males have worse outcomes

Imaging

- General
 - Abnormal gray matter nodules or ribbons within the white matter anywhere from ventricles to cortex
- Modality specific
 - CT and MRI
 - Nonenhancing masses that follow the density or intensity of gray matter on all images

- Imaging recommendations
 - MRI with contrast
- Mimic
 - [Low-grade gliomas](#) can have a similar appearance but usually do not match gray matter so closely on all MR sequences.

For more information, please see the corresponding chapter in [Radiopaedia](#).

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