

WHAT'S YOUR DIAGNOSIS?

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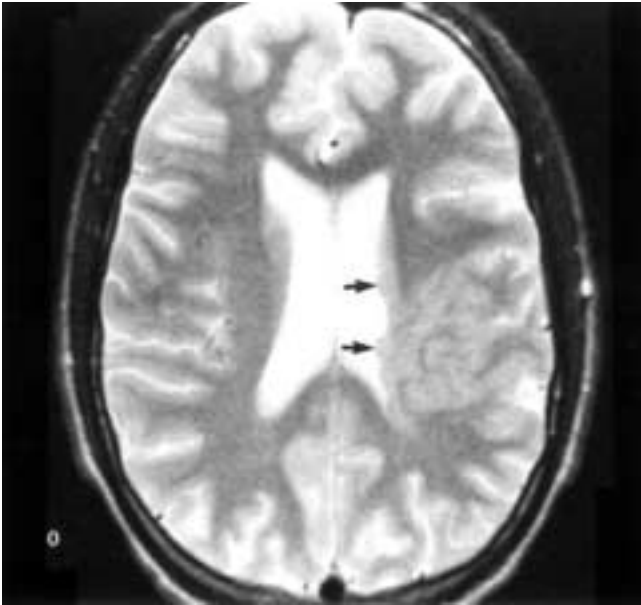


FIGURE 1. An axial T₂-weighted image (TR/TE 400/90 msec) at the level of the lateral ventricles.

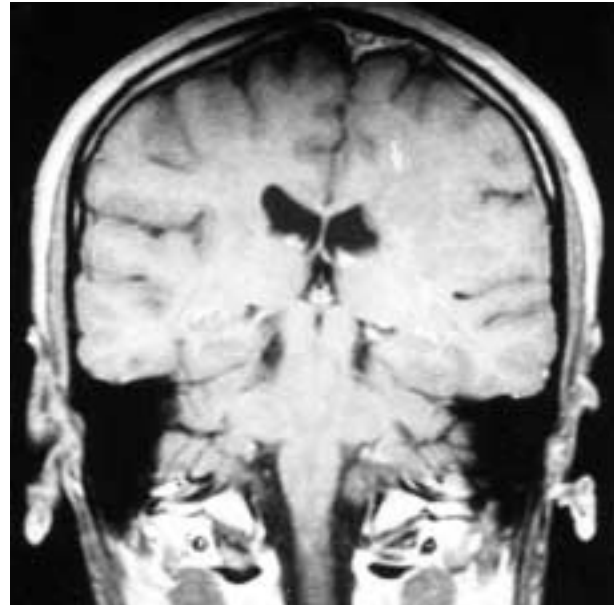


FIGURE 2. A coronal T₁-weighted image (TR/TE 560/16 msec) following intravenous gadolinium-DTPA administration at the level of the third ventricle.

History

A 16-year-old male student presented at the hospital having awoken from sleep with generalized jerking movements of all four limbs. The episode lasted only a few minutes and afterwards the patient experienced a mild headache and tiredness. There was no evidence of urinary or fecal incontinence, although the patient found he had bitten his lip. He was otherwise well with no previous medical, alcohol or drug history of note. Clinical examination and routine laboratory investigations were unremarkable.

Electroencephalogram showed abnormality, demonstrating irregular slow waves associated with sharp waves or spikes arising in the left temporoparietal region. A magnetic resonance imaging scan (Figures 1 and 2) was done. Following the scan, the patient was discharged on phenytoin 300 mg daily and has been well, with no further convulsive episodes during eighteen months of follow-up.

1. What abnormalities are demonstrated in Figures 1 and 2?
2. What's your diagnosis?

ANSWER TO WHAT'S YOUR DIAGNOSIS? (PREVIOUS PAGE)



FIGURE 1. An apparent mass lesion in the left centrum semiovale which indents the body of the left lateral ventricle can be seen. The lesion is of the same signal intensity as cortical gray matter and is consistent with heterotopic gray matter. There is no associated edema. Nodules of subependymal gray matter are also seen lining the left lateral ventricle (arrows).

Diagnosis: Heterotopic gray matter (HGM) in the left centrum semiovale with associated pachygyria presenting with a late-onset seizure.

Discussion: HGM is due to abnormal deposition of collections of neurons during their migration from the germinal matrix to the cortex. HGM most often appears as nodular masses which may range in size from a few millimeters to several centimeters.¹ Heterotopia is associated with a number of conditions, including fetal alcohol syndrome and trisomy 13, and may also be observed in familial members.² Cortical dysplasia is frequently observed in patients with HGM, in particular abnormalities of gyration, most commonly macrogyria and polymicrogyria.³ It is also associated with the absence of the corpus callosum.⁴ Several subtypes of HGM have been described:⁵ 1) laminar, or band heterotopia in the centrum semiovale; 2) nodular, subependymal heterotopia; and 3) subpial heterotopia at the outer margin of the brain.

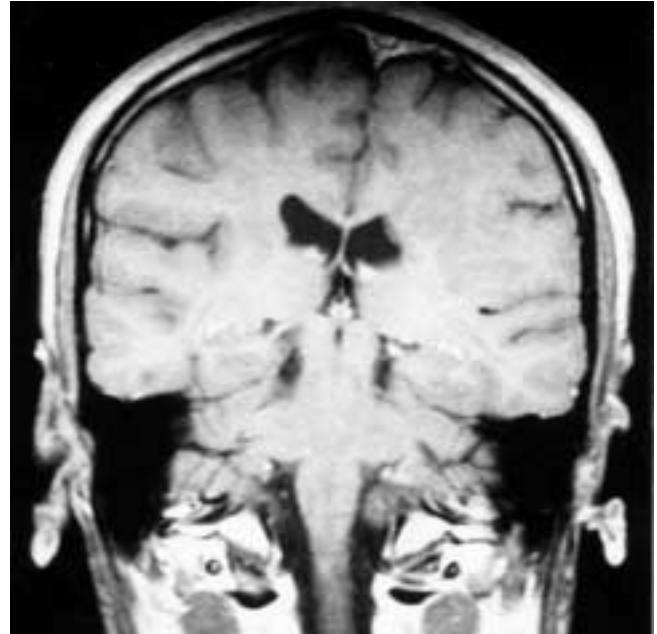


FIGURE 2. The lesion is less clearly visualized (arrows), but is isointense with gray matter, with no evidence of enhancement. There is pachygyria of the overlying cortex.

MRI is the imaging modality of choice for the detection and delineation of HGM (Figures 1 and 2), and will usually allow differentiation from cerebral tumor and avoidance of biopsy.⁶ Heterotopias do not cause disturbance of the blood-brain barrier, and typically have the same density on computerized tomography and the same signal on MRI as cortical gray matter. Abnormal patterns have been identified with MRI, including heterogeneity of signal within the masses and high signal within the surrounding white matter.⁷

HGM may be associated with epilepsy, although the clinical presentation does encompass a broad spectrum of severity. Patients may have normal intellect and motor function, sometimes with apparently extensive cerebral abnormality. Epilepsy may, however, be severe and intractable, and in some of these cases surgery may provide clinical benefits.⁶

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Acknowledgements

The authors would like to thank Mrs. Helen Perry for preparation of the manuscript and Mr. Nick Taylor for his help with production of the illustrations.

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