History
13 month old infant with seizures.

Diagnosis
Subependymal nodular heterotopia

Discussion
Subependymal heterotopia consist of clusters of disorganized neurons and glial cells that are located in close proximity to the ventricular walls. They may lie in the wall of the ventricle and project into the ventricular lumen or lie within the periventricular white matter. They are frequently associated with seizures and variable intellectual deficits. Subependymal heterotopia are frequently associated with epilepsy; they may be isolated or may occur in conjunction with other malformations such as callosal agenesis, Chiari II malformations, polymicrogyria, and basilar cephaloceles. Disorders of cortical formation are commonly caused by mutation in a specific gene that acts in a dominant or X-linked fashion. Several new genes and new mutations of known genes for disorders of cortical formation have been mapped or cloned. PVH are usually bilateral with predilection for the right cerebral hemisphere due to later migration of the right-sided neuroblasts. On CT scans and MR images, subependymal heterotopia appear as nonenhancing periventricular nodules that are isointense to gray matter on all pulse sequences. They may be diffuse and symmetric, and when diffuse as in this case, may form a contiguous undulating bandlike mass around the lateral ventricles, or may be nodular and noncontiguous. Associated cortical malformations are unusual.

Findings
Numerous confluent nodules of heterotopic gray matter lining the lateral ventricles, right more than left; involvement of the trigones, occipital and temporal horns. Effacement of the posterior right lateral ventricle secondarily. No other malformation was identified.

Reference
American Journal of Neuroradiology 30:4-11, January 2009
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