Subependymal nodular heterotopia
Heather Borders, MD
01/12/2010

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
Disclaimer
This teaching site is partially funded by an educational grant from GE Healthcare and Advanced Radiology Services, PC. The material on this site is independently controlled by Advanced Radiology Services, PC, and GE Healthcare and Spectrum Health have no influence over the content of this site.

Content Download Agreement
The cases and images on this website are owned by Spectrum Health. Permission is granted (for nonprofit educational purposes) to download and print materials to distribute for the purpose of facilitating the education of health professionals. The authors retain all rights to the material and users are requested to acknowledge the source of the material.

Site Disclaimer
This site is developed to reach healthcare professionals and medical students. Nothing this site should be considered medical advice.

Only your own doctor can help you make decisions about your medical care. If you have a specific medical question or are seeking medical care, please contact your physician.

The information in this website is provided for general medical education purposes only and is not meant to substitute for the independent medical judgment of a physician relative to diagnostic and treatment options of a specific medical condition.

The viewpoints expressed in these cases are those of the authors. They do not represent an endorsement. In no event will Advanced Radiology Associates, PC, Spectrum Health Hospitals (Helen Devos Children's Hospital) or GE Healthcare be liable for any decision made or action taken in reliance upon the information provided through this website.