Teaching *NeuroImages*: Honeycomb appearance of the brain in a patient with Canavan disease

Sunil Pradhan, MD, DM
Gourav Goyal, MD

Address correspondence and
reprint requests to Dr. Sunil
Pradhan, Department of
Neurology, Sanjay Gandhi
Postgraduate Institute of Medical
Sciences, Raebareli Road,
Lucknow 226014, India
drspradhan@rediffmail.com

Figure 1 Brain MRI

T2-weighted (A, B) and T1-weighted (C, D) MRI of the brain showing severe and diffuse T2 hyperintense and T1 hypointense signal changes in the white matter suggesting Canavan disease. Significant involvement of U fibers is present. Multiple round or oval cystic changes within the white matter give a honeycomb appearance. The radial arrangement of these cystic spaces suggests dilatation of Virchow-Robin spaces due to spongiform degeneration of the white matter.

A 1-year-old girl presented with global developmental delay and generalized tonic seizures for 5 months. She was born of a nonconsanguineous marriage at full term but required resuscitation for 15 minutes during the postdelivery period. Her head circumference was 47 cm (90th percentile). The ocular fundi were normal. She was spastic bilaterally with hyperreflexia and extensor plantar responses. The white

Figure 2 Brain magnetic resonance spectroscopy

Single voxel (8-mL) magnetic resonance spectroscopy of the affected white matter of the brain showing the large N-acetylaspartate peak (arrow).

matter changes on brain MRI¹ (figure 1) and high N-acetylaspartate (NAA) peak on brain magnetic resonance spectroscopy (figure 2) and urine NMR² suggested Canavan disease, an autosomal recessive dysmyelinating disease due to deficiency of the enzyme aspartoacylase that catalyzes breakdown of NAA. Its deficiency results in high NAA levels in serum and urine. The honeycomb appearance of the white matter may be due to leukodystrophy or, possibly, to an additional postnatal ischemic event.

REFERENCES

1. Brismar J, Brismar G, Gascon G, Ozand P. Canavan disease: CT and MR imaging of the brain. *Am J Neuroradiol* 1990;11:805–810.
2. Pinheiro GMS, Basso EA, Fiorin BC. A fast ¹H NMR spectroscopy procedure for quantitative determination of N-acetylaspartate in urine samples. *Clin Chim Acta* 2009; 404:166–168.

From the Department of Neurology, Sanjay Gandhi Postgraduate Institute of Medical Sciences, Lucknow, India.

Disclosure: Dr. Pradhan serves as Associate Editor for *Neurology India*. Dr. Goyal reports no disclosures.

Neurology[®]

Teaching *NeuroImages*: Honeycomb appearance of the brain in a patient with Canavan disease

Sunil Pradhan and Gourav Goyal
Neurology 2011;76:e68
DOI 10.1212/WNL.0b013e318212a887

This information is current as of March 28, 2011

Updated Information & Services	including high resolution figures, can be found at: http://n.neurology.org/content/76/13/e68.full
References	This article cites 2 articles, 1 of which you can access for free at: http://n.neurology.org/content/76/13/e68.full#ref-list-1
Subspecialty Collections	This article, along with others on similar topics, appears in the following collection(s): All Demyelinating disease (CNS) http://n.neurology.org/cgi/collection/all_demyelinating_disease_cns Leukodystrophies http://n.neurology.org/cgi/collection/leukodystrophies Metabolic disease (inherited) http://n.neurology.org/cgi/collection/metabolic_disease_inherited MRI http://n.neurology.org/cgi/collection/mri MRS http://n.neurology.org/cgi/collection/mrs
Permissions & Licensing	Information about reproducing this article in parts (figures, tables) or in its entirety can be found online at: http://www.neurology.org/about/about_the_journal#permissions
Reprints	Information about ordering reprints can be found online: http://n.neurology.org/subscribers/advertise

Neurology® is the official journal of the American Academy of Neurology. Published continuously since 1951, it is now a weekly with 48 issues per year. Copyright Copyright © 2011 by AAN Enterprises, Inc.. All rights reserved. Print ISSN: 0028-3878. Online ISSN: 1526-632X.