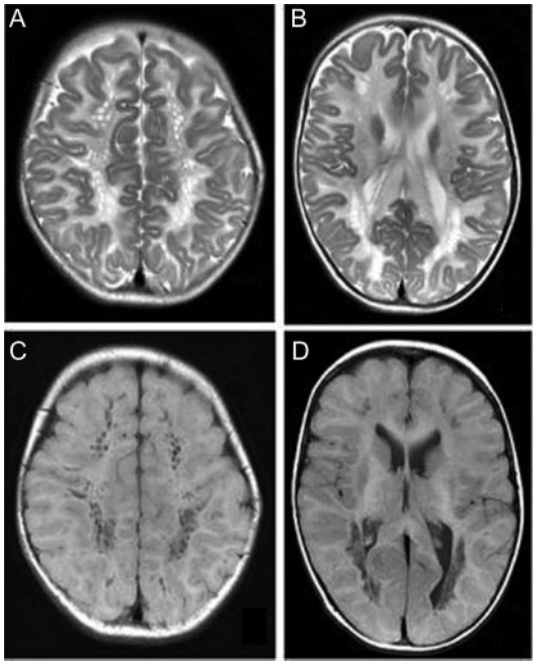
Teaching Neuro*Images*: Honeycomb appearance of the brain in a patient with Canavan disease

Sunil Pradhan, MD, DM
Gourav Goyal, MD

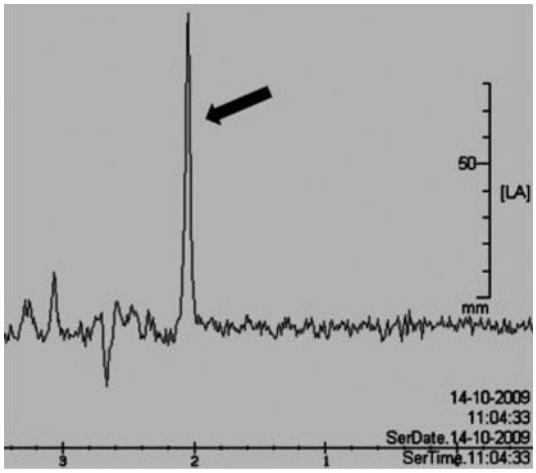
Address correspondence and reprint requests to Dr. Sunil Pradhan, Department of Neurology, Sanjay Gandhi Postgraduate Institute of Medical Sciences, Raebareli Road, Lucknow 226014, India
drspradhan@rediffmail.com

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.

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).

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 matter changes on brain MRI[1] (figure 1) and high *N*-acetylaspartate (NAA) peak on brain magnetic resonance spectroscopy (figure 2) and urine NMR[2] 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 ^{1}H 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.

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Neurology 2011;76;e68
DOI 10.1212/WNL.0b013e318212a887