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 \(N\)-acetylaspertate (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


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

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Teaching NeuroImages: Honeycomb appearance of the brain in a patient with Canavan disease
Sunil Pradhan and Gourav Goyal
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