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NEUROCUTANEOUS SYNDROMES

BRAIN MRI GROWTH PATTERNS IN NEUROFIBROMATOSIS 1

Brain growth patterns in 27 children with neurofibromatosis 1 (NF-1) (20 boys, aged 1.0-17.7 years; mean age, 8.8 years) were compared to 43 controls (22 boys, aged 0.1-17.7 years; mean age, 5.9 years) using magnetic resonance imaging (MRI) in a study at Children's Medical Center, Hartford, Connecticut. Patients with NF-1 have a lateral volume expansion of the cerebral hemispheres, accounting for the characteristic megalencephaly, and an acceleration of brainstem growth rate, as measured by brainstem height, with increasing age. Significant morphometric changes compared to controls included a larger bicaudate width, biatrial width, biparietal diameter, iter measures, and descending sigmoid sinus. The regions showing increases in size and growth rate compared to controls are the parenchymal zones that correlate with high-signal T2-weighted lesions on MRI. These data show that patients with NF-1 have dynamic changes in brain morphometry with region-specific parenchymal overgrowth. (DiMario FJ Jr, Ramsby GR, Burleson JA. Brain morphometric analysis in neurofibromatosis 1. Arch Neurol Nov 1999;56:1343-1346). (Rewprints: Francis J DiMario Jr MD, Department of Pediatrics, Connecticut Children's Medical Center, 282 Washington St, Hartford, CT 06106).

COMMENT. Neurofibtomatosis 1 (NF-1), a genetic autosomal dominant disorder, characterized by cafe-au-lait macules, skin freckling, Lisch nodules, and an increased risk of benign and malignant tumors, is frequently associated with learning disabilities and attention deficit disorder. In an editorial (Gutmann DH. Learning disabilities in neurofibromatosis 1. Sizing up the brain. Arch Neurol Nov 1999;56:1322-1323), the relationship of the brain morphometric changes and neurofibromin (NF1 gene protein) to learning disabilities, UBO formation, and brainstem gliomas in NF-1 is discussed. Gutmann DH et al (Ann Neurol Nov 1999;46:777-782) have identified a protein isoform of the NF1 gene, exon 9a neurofibromin, localized in the cytoplasm of forebrain neurons, and first detected after postnatal day 2. The function of this neuron-specific isoform in the developing brain remains to be determined.

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