NEUROCUTANEOUS SYNDROMES

PET FINDINGS IN TUBEROUS SCLEROSIS

The clinical role of positron emission tomography (PET) was evaluated in 23 children with tuberous sclerosis complex examined at the Children's Hospital of Michigan, Wayne State University, Detroit, MI. Focal or multifocal cortical hypometabolism was found in 21 children, some without corresponding CT or MRI abnormalities. PET can provide additional localizing information to CT and MRI when surgery is indicated, but is otherwise of limited value in clinical practice. (Rintahaka PJ, Chugani HT. Clinical role of positron emission tomography in children with tuberous sclerosis complex. J Child Neurol Jan 1997;12:42-52). (Respond: Dr Harry T Chugani, Division of Pediatric Neurology and PET CEnter, Children's Hospital of Michigan, 3901 Beaubien Blvd, Detroit, MI 48201).

COMMENT. In selected patients, PET may provide additional information to that obtained by EEG, CT and MRI, and it can be useful in the pre-surgical evaluation of children with tuberous sclerosis and refractory seizures.

NEUROIMAGING IN NEUROCUTANEOUS SYNDROMES

The radiographic manifestations of neurocutaneous syndromes are reviewed from the Albert Einstein College of Medicine/Montefiore Medical Center, Bronx, NY; and the University of New Jersey, Robert Wood Johnson Medical School, New Brunswick, NJ. *Tuberous sclerosis* is characterized by bilateral calcified subependymal nodules on CT, and areas of hyperintensity on long TR-MRI images, representing dysplastic white matter. *Neurofibromatosis Type 1* abnormalities include optic nerve glioma, hyperintense hamartomas on MR, neural foramina enlargement of the spine with or without dumbbell neurofibromas, scalloping of the posterior vertebral bodies, and kyphoscoliosis. *Neurofibromatosis Type 2* has gadolinium-enhanced bilateral acoustic neuromas, schwannomas of other cranial nerves, especially trigeminal, and meningiomas; also, spinal lesions including meningiomas, schwannomas and intramedullary ependymomas and astrocytomas. *Sturge-Weber syndrome* diagnostic signs include CT gyriform calcification, especially parieto-occipital, ipsilateral cortical atrophy with thickening of the calvarium, prominent paranasal sinuses, elevation of petrous ridge, enlarged choroid plexus, and MRI post-gadolinium pial angiomatosis. *Von Hippel-Lindau disease* lesions on MRI include cerebellar hemangioblastoma (30%), mostly cystic with an enhancing mural nodule, and spinal cord hemangioblastoma (5%). *Ataxia-telangiectasia* is manifested on CT and MRI by progressive cerebellar atrophy, particularly the anterior vermis. (Wolf SM, Mitnick RJ, Traeger EC, Moshe SL. Radiologic review of the neurocutaneous syndromes. Acta Neuropediatrica 1996;2:160-175). (Reprints: Steven M Wolf MD, 111 E 210th St, Montefiore Medical Center, NW7-EEG Lab, Bronx, NY 10467).

COMMENT. This article is replete with excellent illustrations of neuroimages of neurocutaneous syndromes, essential in diagnosis. For reviews of the significance of UBOs on MRI in neurofibromatosis 1, see Progress in Pediatric Neurology III, PNB Publ, Chicago, 1997.