Title
KRABBES DISEASE - COMPUTED TOMOGRAPHIC AND MAGNETIC-RESONANCE FINDINGS

Journal
ANNALS OF NEUROLOGY, 18(3)

ISSN
0364-5134

Authors
BARAM, TZ
FISHMAN, MA
PERCY, AK

Publication Date
1985

68. Krabbe's Disease—Computed Tomographic and Magnetic Resonance Findings

Tallie Z. Baram, Marvin A. Fishman, and Alan K. Percy, Houston, TX

Krabbe's disease is a rare progressive infantile leukodys-trophy. Clinically, infants display spasticity, irritability, loss of developmental milestones, and blindness. The underlying pathophysiology is dysmyelination secondary to the absence of the enzyme galactocerebroside β-galactosidase. Reported computed tomography (CT) scans have typically revealed cerebral atrophy, although both abnormal lucency and den-sity of the white matter have been described. Progression of the CT appearance from normal through white matter lucen-cies to atrophy has been postulated. To our knowledge, mag-

netic resonance imaging (MRI) studies of patients with Krabbe's disease have not been previously reported. We describe the CT findings in 3 infants with enzyme-proved Krabbe's disease. Aside from atrophy and increased ventricular size, there were dense nonenhancing symmetrical lesions in specific brain areas: the posterior limb of the internal capsule, thalami, periventricular white matter, and quadrigeminal plates. These lesions were most prominent early in the disease. MRI, performed on 2 of these infants, confirmed the abnormalities seen on CT. In addition, white matter lesions., similar to those seen in multiple sclerosis, were seen in the centrum semiovale. Both CT and MRI lesions were progressive and correlated with the clinical stage of the disease present at the time of the study. The symmetrical dense lesions on CT in the deep gray matter, periventricular white matter and brainstem in conjunction with the white matter abnormalities seen by MRI appear to be associated with Krabbe's disease and should raise a clinical suspicion of the disease in the appropriate clinical setting.