Pediatric Neurology Part III: Chapter 164. Lysosomal leukodystrophies: Krabbe disease and metachromatic leukodystrophy (Handbook of Clinical Neurology)

Alfried Kohlschütter

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Genetic deficiencies of lysosomal catabolic pathways lead to storage disorders with multiple organ abnormalities or to degeneration of purely nervous structures. Krabbe disease and metachromatic leukodystrophy are caused by metabolic errors concerning lipids of neural membranes. They are characterized by demyelination of the central nervous system and, variably, the peripheral nerves. Their clinical presentation is a relentlessly progressive motor and mental deterioration starting at any age between infancy and adolescence. MRI demonstrates characteristic lesions of brain white matter. In Krabbe disease, deficient galactocerebroside β-galactosidase activity causes accumulation of lipids in “globoid” macrophages and of psychosine, which is toxic to oligodendrocytes. Diagnosis depends on demonstration of the enzyme deficiency. Experimental treatment is limited to hematopoietic stem cell transplantation, which can favorably alter the course of disease in certain situations. In metachromatic leukodystrophy, deficient activity of arylsulfatase A, or lack of a cofactor, causes accumulation of sulfatide in various tissues and diffuse demyelination. Symptoms are neurological, but gallbladder dysfunction may be present. Diagnosis depends on demonstrating the enzyme deficiency and elevated urinary sulfatide. In a rare variant, multiple sulfatases are deficient. Stem cell transplantation may prevent disease progression in selected cases. Enzyme replacement is being evaluated, and gene therapies are being developed.

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