A two-year-old girl was admitted to Marmara University Hospital, Department of Pediatrics, Division of Pediatric Neurology, with severe psychomotor retardation and seizure symptoms. EEG revealed diffuse organisation disorder and hypersynchrony in the fronto-parietal region. An MRI examination was within normal limits. Two months later she was unable to walk.

Electron microscopical examination of the skin indicated the presence of specific ultrastructural profiles within the cytoplasm of the eccrine gland cells. A characteristic accumulation of osmiophilic granules in membrane-bounded cytosomes which were globular and lamellar in appearance was observed.

What is the most likely diagnosis of this patient?