A 42-year-old man with Addison disease presented to the emergency department with unsteady gait for 1 year. A half-brother had epilepsy and difficulty walking, and died at 6 years of age. Another half-brother has a longstanding gait disorder. Over 3 to 4 months, his gait has become worse with marked stiffness, ataxia, and dysarthria. Neurologic examination demonstrated saccadic pursuit, ocular flutter, severe spasticity throughout, peripheral neuropathy, hyperreflexia, clonus, and bilateral extensor plantar reflexes. MRI is shown in figures 1 and 2. The plasma concentration of very-long-chain fatty acids, which is a general test for peroxisomal disorders, was elevated, confirming the diagnosis of adrenoleukodystrophy (ALD). Very-long-chain fatty acids were also elevated in the surviving brother’s plasma.

Adult cerebral ALD is extremely rare. It is an X-linked peroxisomal disorder caused by mutation in the ABCD1 gene located at Xq28 and involves the nervous system, adrenal cortex, and Leydig cells in the testes.\textsuperscript{1,2}

**Figure 1** Axial fluid-attenuated inversion recovery of brain MRI

![Image of MRI scans showing hyperintensity in various brain regions](image-url)
Gadolinium enhancement of the cerebellum, middle cerebellar peduncles, pons, cerebral peduncle, internal capsule, and genu of corpus callosum.
Teaching NeuroImages: Cerebral adrenoleukodystrophy: A rare adult form
Rania A. Elenein, Sunil Naik, Stephanie Kim, et al.
Neurology 2013;80:e69-e70
DOI 10.1212/WNL.0b013e3182815416

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