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Tigroid pattern: Metachromatic Leukodystrophy

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Clinical image

Metachromatic leukodystrophy is a lysosomal storage disease caused by arylsulfatase deficiency, which results in lipid accumulation leading to impaired myelination. It is the most frequent inherited leukodystrophy with an autosomal recessive mode of inheritance. Brain MRI reveals in T2WI, bilateral and symmetrical high signal intensity abnormalities of the white matter with low signal intensity bands creating a characteristic "Tigroid appearance" (Fig 1A, yellow arrows), initially reaching the periventricular white matter, the posterior limb of the internal capsules and the cerebellum (Fig 1B, white arrows), with sparing of the subcortical U fibers. The corpus callosum may be affected (Fig 1C, red arrows). Cortical atrophy may be observed at an advanced stage of the disease.

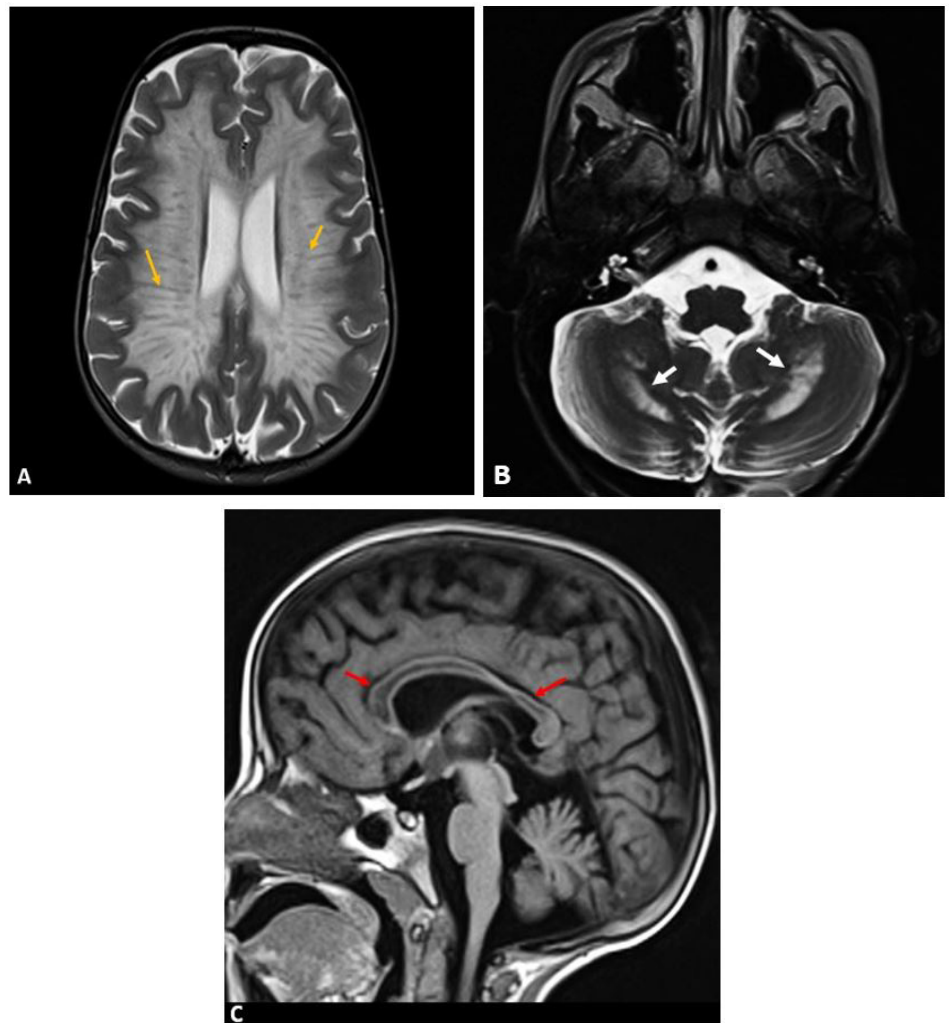


Figure 1: Brain MRI in a 5-year-old girl with psychomotor regression since the age of 3, who presents an extra-pyramidal syndrome, showing "tigroid pattern" of white matter in T2WI (A, yellow arrows), associated to involvement of cerebellar white matter (B, white arrows) and a T1WI low-signal intensity of the corpus callosum (C, red arrows).