Teaching Neuro*Images*: Adrenoleukodystrophy presenting as raised intracranial pressure

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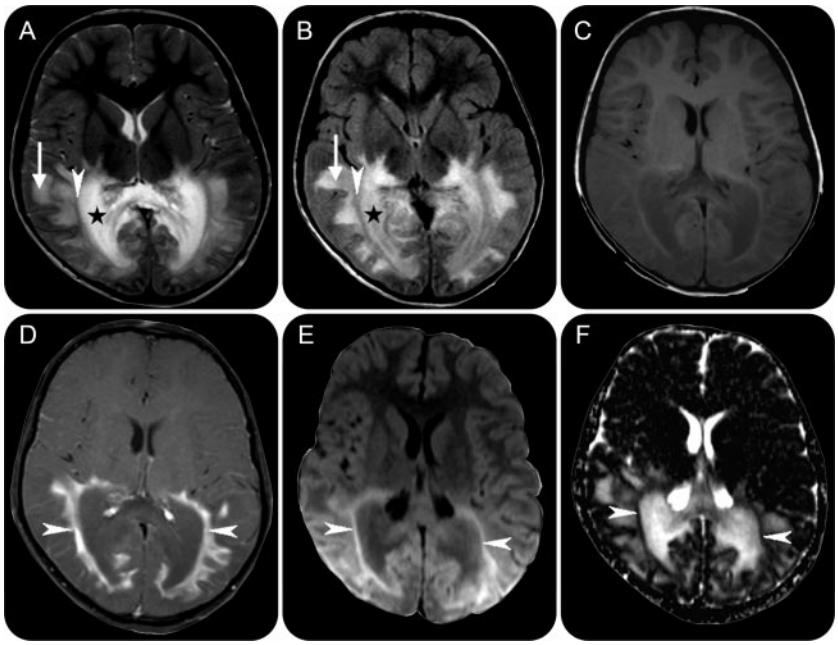
Figure Brain MRI (1.5 T)



Axial images showing symmetric bilateral parieto-occipital white matter changes with local mass effect. (A, B) T2-weighted and fluid-attenuated inversion recovery images show regions corresponding to the 3 classic histopathologic zones of Schaumberg et al.[3]: peripheral hyperintensity (arrow, zone 1); central hypointensity (arrowhead, zone 2); and inner hyperintense zone (asterisk, zone 3). Areas with active inflammation (zone 2 and adjacent part of zone 1) show contrast enhancement (C, D; arrowhead) and diffusion restriction (E, F; arrowhead).

A 7-year-old boy presented with acute onset headache, vomiting, and visual deterioration 2 weeks previously without any cognitive or behavioral changes. Optic fundi showed papilledema, and there were no pyramidal signs or gait disturbances. The remainder of the neurologic examination was normal. CSF protein was 174 mg/dL. Although MRI features (figure) were consistent with adrenoleukodystrophy (ALD), the atypical presentation prompted consideration of alternate diagnoses and therapies. Subsequent progressive vision and hearing loss, bilateral pyramidal signs, and elevated plasma very-long-chain fatty acids confirmed ALD.

Childhood cerebral ALD can present acutely, but rarely with raised intracranial pressure.[1] Elevated CSF protein and mass effect due to fulminant demyelination and inflammation is a possible cause. Familiarity with atypical clinical courses[2] and classic MRI features may avoid unnecessary investigations and delayed diagnosis.

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