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. Elevated CSF protein and mass effect due to fulminating demyelination and inflammation is a possible cause. Familiarity with atypical clinical courses and classic MRI features may avoid unnecessary investigations and delayed diagnosis.

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