Alphabet Soup: Acute Disseminated Encephalomyelitis (ADEM) mimicking Leukodystrophy (LKD)

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Introduction: Acute disseminated encephalomyelitis (ADEM) is an immune mediated demyelinating disease of the central nervous system, usually precipitated by a preceding viral infection. Manifestations include encephalopathy and multifocal neurologic deficits. MRI findings vary widely but most often include bilateral, asymmetric lesions in subcortical and central white matter, cortical gray-white junction, and less commonly in periventricular white matter[1]. The leukodystrophies (LKD) represent chronic conditions with underlying genetic abnormalities that lead to progressive motor and cognitive deficits for which usually no effective treatments exist. MRI in LKD characteristically reveals symmetric lesions located in the periventricular white matter, corpus callosum, internal capsule, and corticospinal tracts[2].

Case Report: A previously healthy 10-year old boy presented to the emergency department with right-sided weakness, right hand tingling and unsteady gait that developed over several hours. No recent illness, travel, or trauma was reported. Initial physical examination was notable for unilateral right-sided lower face, upper and lower extremity weakness with hypoesthesia which initially remitted but then recurred. After admission, his right-sided weakness progressed, and he also developed aphasia and agitation. Brain CT was normal while MRI demonstrated bilateral confluent lesions in the parietal periventricular white matter and splenium of corpus callosum with diffusion restriction, highly suggestive of LKD (Figure 1). CSF obtained by lumbar puncture showed no pleocytosis, normal protein and glucose, negative PCRs for herpes simplex virus and enterovirus, and absence of oligoclonal bands. Inflammatory markers including C reactive protein, procalcitonin, and erythrocyte sedimentation rate were mildly elevated. Viral respiratory PCR panel was positive for rhinovirus/enterovirus. Cortrosyn stimulation test was normal.

Due to the rapid progression of disease suggestive of ADEM the patient was administered a 3-day course of high-dose intravenous methylprednisolone which was associated with swift resolution of his symptoms. He was discharged on a several week prednisone taper. Peroxisomal very long chain fatty acid panel and leukoencephalopathy gene panel both proved to be negative. At his one week follow-up he remained symptom-free. A follow-up brain MRI obtained three months later showed resolution of signal abnormalities (Figure 2).

Discussion: This patient presented with progressive neurologic deficits and MRI findings highly suggestive of LKD. CSF indices were not consistent with an inflammatory or infectious process. However, in the setting of new onset rapidly progressing deficits with encephalopathy and

rhino/enterovirus positivity, intravenous steroids were initiated. The resultant rapid response supported a clinical diagnosis of ADEM.

Conclusion: In this child, MRI signal abnormalities consistent with a genetic leukodystrophy could have led to a delay in diagnosis or even misdiagnosis. Few reported cases exist where the MRI appearance of ADEM suggests a genetic leukodystrophy[2,3,4,5]. This case illustrates the importance of considering the treatable diagnosis of ADEM even if the radiologic and laboratory findings are atypical and suggest another diagnosis.

Conflicts of Interests: No conflicts identified.

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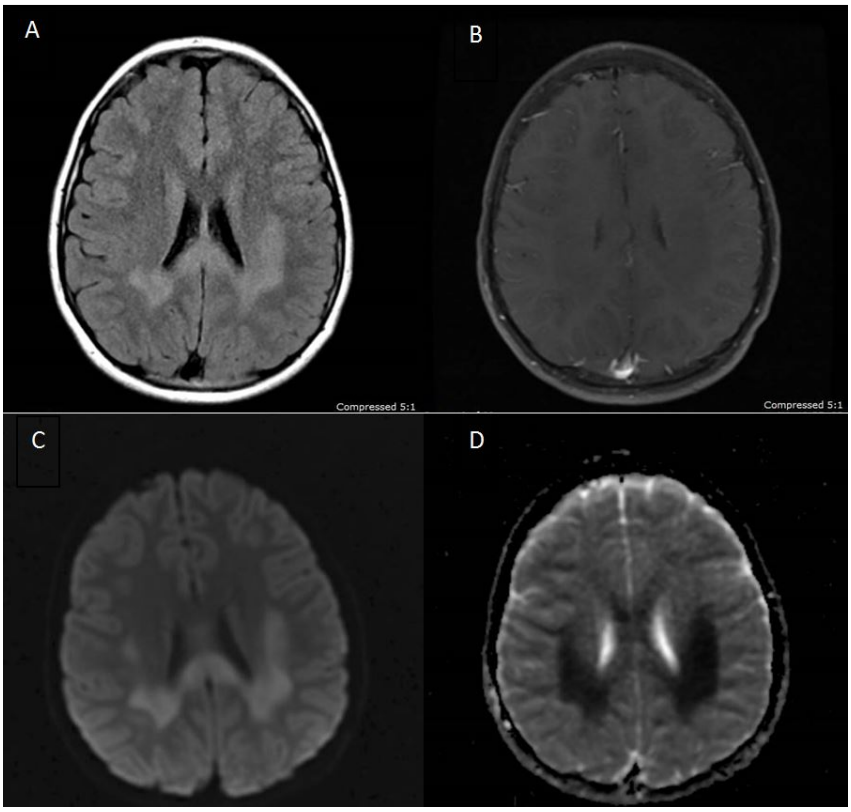

Figure 1. Initial MRI of the brain demonstrates bilateral confluent hyperintense lesions in parietal periventricular white matter and splenium of corpus callosum on axial T2 FLAIR image (A). Post contrast fat suppressed T1 weighted image (B) demonstrates mild hypointensity without enhancement in the lesions. DWI and ADC map (C,D) demonstrate diffusion restriction within the lesions. The findings are suggestive of adrenoleukodystrophy (ALD).

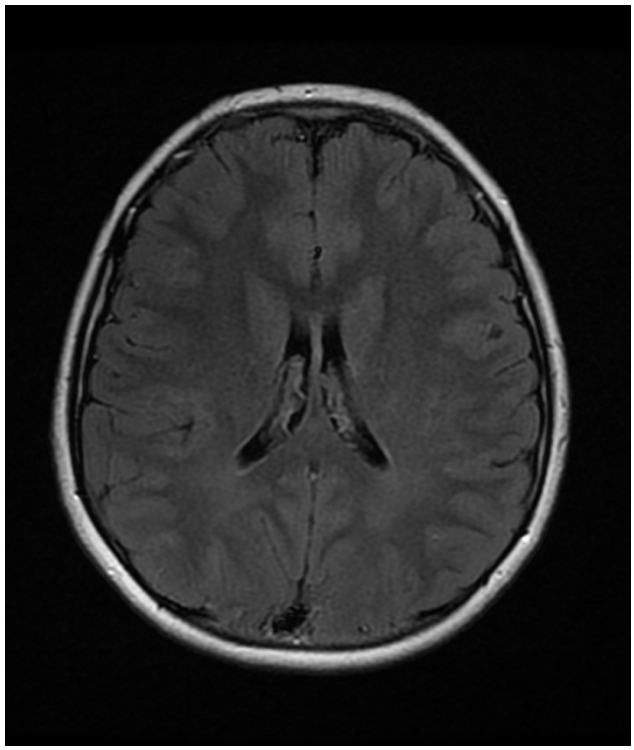

Figure 2. Follow up MRI of the brain demonstrates resolution of lesions in parietal white matter and corpus callosum on the corresponding axial T2 FLAIR image.

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