CASE No 6

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This four-year-old girl, with global developmental delay, was born at term after a pregnancy complicated by gestational diabetes. She was macrocephalic at birth (98th percentile) and myoclonic seizures were noted within 12 hours of birth. Subsequent follow-up by neurology and ophthalmology revealed bilateral optic nerve atrophy. Progressive global cerebral atrophy and delayed myelin maturation were noted on follow-up MRI studies. EEG recordings were suggestive of an emerging hypsarrhythmic pattern. She had recurrent episodes of aspiration pneumonia which exacerbated her partial seizures. On the morning of her death she suffered multiple consecutive seizures and stopped breathing with failed resuscitative efforts.

Material Submitted: Slide transparency, H&E/ LFB stain.