Diabetes ketoacidosis and Recurrent Childhood Stroke-like Episodes

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A 13-year and 4-month-old girl was brought to the emergency department due to fever, dizziness, vomiting, and blurred vision. Laboratory data revealed hyperglycemia (350 mg/dL) with an HbA1C of 7.3%, ketonuria, and lactic acidosis (98.4 mg/dL). The initial impression was diabetic ketoacidosis. During admission, recurrent focal impaired awareness seizures were noted, and magnetic resonance imaging (MRI) of the brain revealed multiple brain infarctions in the bilateral cerebrum (Figure 1 A). Mitochondrial gene report showed A3243 G with 64% heteroplasmy (Figure 1 B), and mitochondrial encephalomyopathy, lactic acidosis, and stroke-like episodes (MELAS) was diagnosed. At 16 years and 7 months old, recurrence of vomiting and onset of right hemianopia and mild right limb weakness were observed and follow-up T2 images showed massive edema in her left parieto-occipital region (Figure 2 A). At 16 years and 10 months old, she developed clonus in her left hand associated with an unsteady gait and blurred vision. MRI of the brain revealed recurrent brain infarction, and T2 images showed massive edema of the right parieto-occipital region (Figure 2B).

MELAS is a rare disease entity and occasionally comorbid with mitochondrial diabetes in childhood [1]. Characteristic radiological features of MELAS include infarction-like lesions over the parieto-occipital or parieto-temporal areas, which help distinguish MELAS from childhood ischemic stroke[2, 3].

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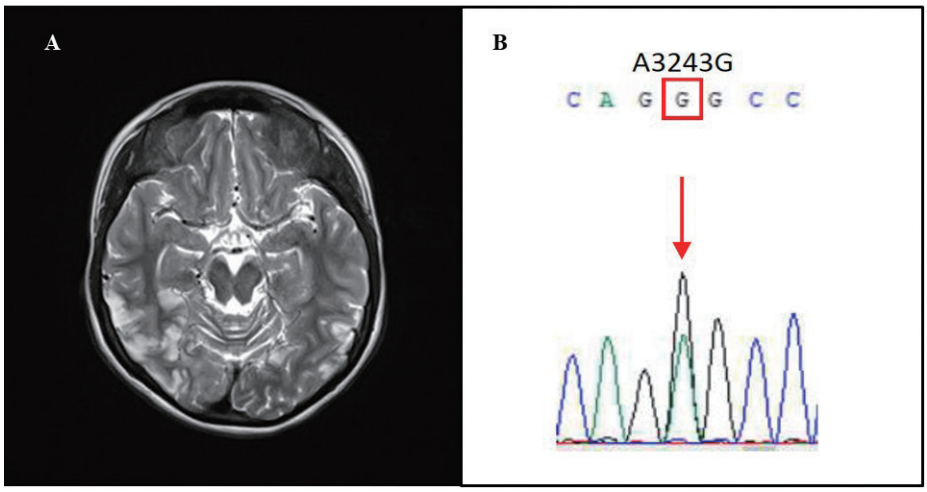

Fig. 1. MRI of brain showed of multiple ill-defined long T2 areas in cortical / subcortical regions of bilateral temporo-parieto-occipital lobes, more on the right side (A). Mitochondrial gene showed mutation in mtA3243G with 64% heteroplasmy. (B)

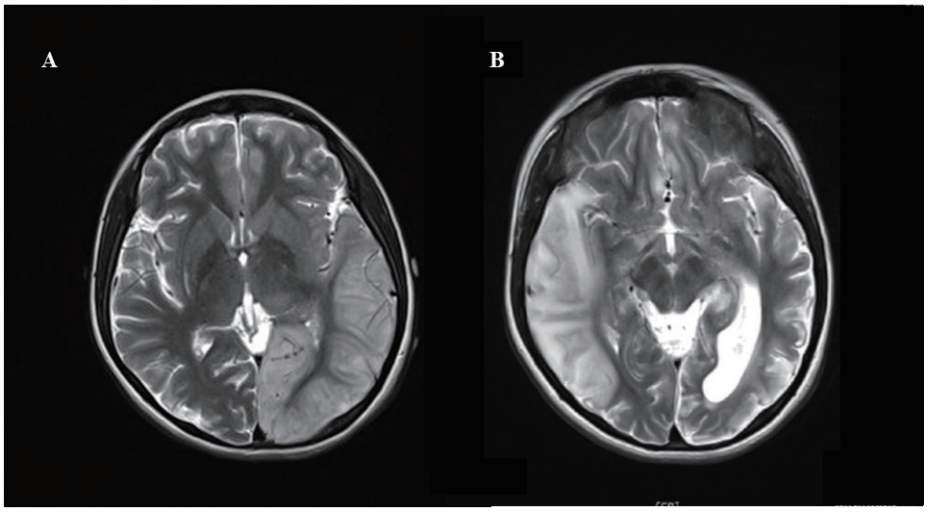

Fig. 2. At 16- years- 7- month old, MRI of brain showed long T2 edema in left parieto-occipital lobe (A). Presence of a large area long T2 edema in right temporo-parieto-occipital lobe, and dilatation of the occipital horn of left lateral ventricle at 16- years- 10- month old.

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