Recurrent Childhood Stroke-like Episodes

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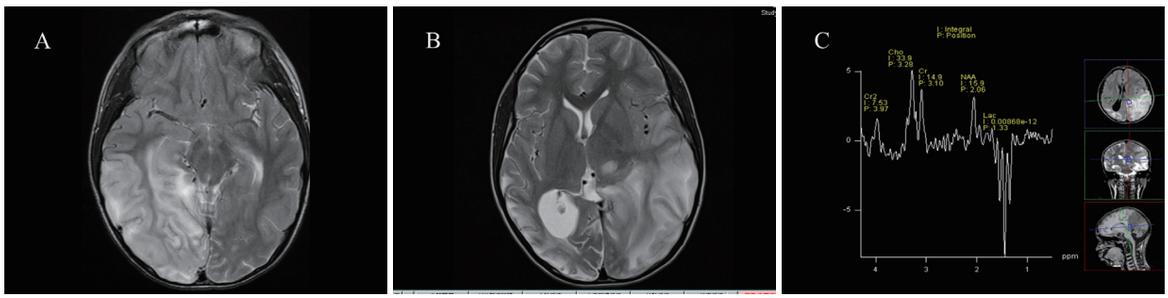
A 9 years old boy suffered from recurrent headache, nausea and blurred vision for 2 months. His medical history revealed failure to thrive, easy constipation and exercise intolerance for more than two years. Initial computed tomography and magnetic resonance images (MRI) of the brain were unremarkable. He presented with ongoing headache and homonymous hemianopia. The follow-up brain MRI revealed high signal intensity on T2-weighted images over affected areas (Figure A). Laboratory investigation revealed an elevated level of lactic acid: 26.5 mg/dL (normal range, <20 mg/dL). Muscle biopsy showed several ragged-red fibers on Gomori-trichrome stain. Subsequently, molecular study demonstrated a typical mitochondrial encephalomyopathy, lactate acidosis and stroke-like episodes (MELAS) syndrome with gene mutation in mtA3243G with 66% heteroplasmy. He suffered from recurrent stroke-like episodes again during the period of follow-up. Repeated MRI 2 years later revealed focal atrophic change with ventriculomegaly over the previous affected areas and new lesions over the right parieto-occipital lobes (Figure B). Proton MR spectroscopy showed the elevated lactate peak (Figure C). Childhood stroke is a rare entity. The characteristic radiological features of infarction-like lesions over the parieto-occipital or parieto-temporal areas can help distinguish between MELAS and ischemic stroke in children.



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REFERENCES

1. Chen JC, Tsai TC, Liu CS, Lu CT. Acute hearing loss in a patient with mitochondrial myopathy, encephalopathy, lactic acidosis and stroke-like episodes (MELAS). Acta Neurol Taiwan. 2007;16:168-172.
2. Tsai J, Tsai HJ, Yang S, Sheu J. Subacute necrotizing encephalomyelopathy (Leigh syndrome) in pediatric patients: a retrospective study. Int J Res Med Sci. 2015;3:3015-3020.
3. El-Hattab AW, Adesina AM, Jones J, Scaglia F. MELAS syndrome: Clinical manifestations, pathogenesis, and treatment options. Mol Genet Metab. 2015;116:4-12.
4. Chi CS. Diagnostic approach in infants and children with mitochondrial diseases. Pediatr Neonatol. 2015; 56:7-18.

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