Cystic encephalomalacia and infantile spasm as a complication of transient and mild hyperinsulinemic hypoglycemia

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Introduction: Hyperinsulinemic hypoglycemia (HH) is the most common cause of persistent hypoglycemia in infant posing significant risk of permanent brain damage. Although it is known that hypoglycemic brain injury could cause infantile spasms, the mechanism is not precisely known.

Case Report: A 3140 g, male, full-term infant was admitted with poor sucking and feeding problems on the postnatal second day. The patient was hospitalized, as the venous blood glucose level at admission was 29 mg/dL. There was no consanguinity between the mother and the father. On physical examination, the body weight was 3185 g (25-50p), height was 85 cm (25-50p), and head circumference was 34 cm (50p). The systemic examination was normal. Intravenous bolus glucose was administered to treat hypoglycemia and there was a requirement of glucose infusion at 15 mg/kg/min during follow-up. Phenobarbital was initiated to treat tonic-clonic convulsions. The patient's serum insulin level was 13.7 µg/dL and urine was negative for ketone during hypoglycemia. As the blood glucose level increased with glucagon treatment, the case was diagnosed with HH. Diazoxide treatment was initiated (15 mg/kg/day), and as hypoglycemia improved after the postnatal fifth day, intravenous glucose infusion was terminated, and total enteral feeding was started. The EEG during this period was within normal limits according to the age. As the venous glucose levels were high, diazoxide treatment was terminated at the postnatal 21st day and the patient was discharged on phenobarbital treatment due to the absence of hypoglycemia in the follow-up.

At 2.5 months of age the patient developed infantile spasms with 2-3 clusters per day and were not associated with hypoglycemia (Figure 1). Physical examination revealed microcephaly (head circumference 35 cm) and an EEG was abnormal with epileptiform discharges consistent with hypersencron/modified hypsaritmia (Figure 2). The cranial magnetic resonance imaging (MRI) revealed large cystic encephalomalacia areas in the parieto-occipital area (Figure 3). No diffusion restriction was observed in the diffusion weighted images. The findings were thought to be related to hypoglycemic brain injury. Serum uric acid level, plasma amino acids, carnitine-acyl carnitine profile, ammonium level (91 ug/dL), cerebrospinal fluid analysis, and urine organic acid levels were normal. The infantile spasms did not respond to vigabatrine or adrenocorticotropic hormone (ACTH) therapy and the patient was commenced on valproate and topiramate, which resulted in slightly reduction of seizures.

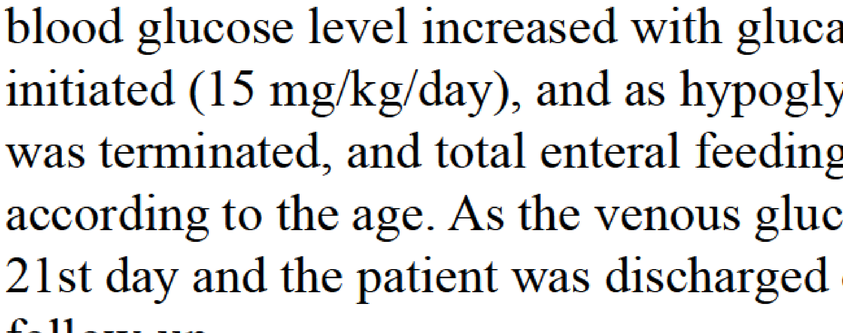

Figure 1. Continous glucose montioring system of the patient showed normoglycemia

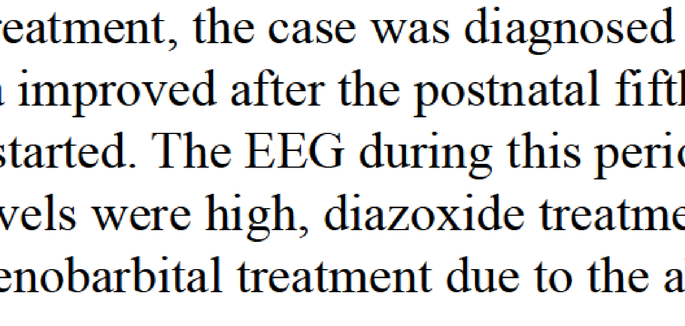

Figure 2. Hypersencron/modified hypsaritmia

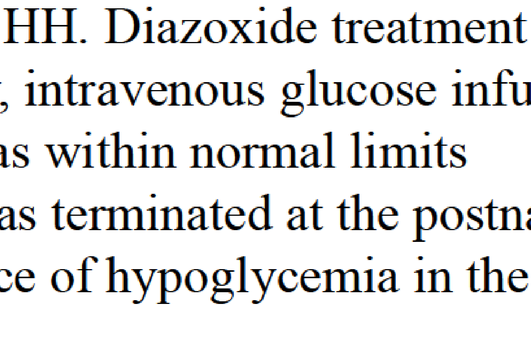

Figure 3. Cranial MRI showing cystic encephalomalacia areas

In conclusion, The present case report emphasized that even HH is short-term and transient; (i) it could cause cerebral damage, (ii) it could present as infantile spasms in the late period, and therefore, (iii) long-term follow-up is important for these patients.

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