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CLINICAL QUIZ

Persistent hypoglycemic attacks during hemodialysis sessions in an infant with congenital nephrotic syndrome: Questions

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Case summary

A 5-month-old female infant was referred to our clinic to maintain her chronic hemodialysis (HD) program. She was born 3100 g at 38th week of gestational age from fifth-degree consanguineous parents. She was diagnosed with congenital nephrotic syndrome (CNS) at the age of 2 months and treated with albumin infusions and diuretics. Biopsy showed mild mesangial cell proliferation in glomeruli, dilatation of tubules, and hyaline degenerations in arterioles. At the age of 3.5 months, the patient was hospitalized for sepsis, seizures, severe pulmonary edema, and kidney failure. Lumbar puncture revealed no signs of meningitis, and cranial MRI findings were normal. She was treated with continuous renal replacement therapy and broad-spectrum antibiotics, as well as phenobarbital. High-dose vasopressor therapy was required due to septic shock and hydrocortisone was empirically started. Thyroid hormone replacement was used for hypothyroidism. After the stabilization of clinical status, chronic peritoneal dialysis was started. After 1 month of peritoneal dialysis, the patient was required to switch to HD due to fungal peritonitis and was then transferred to our clinic.

The answers to these questions can be found at https://doi.org/10.1007/s00467-018-3982-7

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On admission at the age of 5 months, she was anuric; her weight (4750 g), height (61 cm), and head circumference (38 cm) were below the third percentile. Blood pressure was 95/62 mmHg (75th percentile). She was receiving nasogastric tube feeding. Skin examination was normal except a skin tag on the occipital region and peritoneal catheter scar. Physical examination revealed no other abnormality. Erythropoietin, active vitamin D, and phenobarbital treatment were continued. Corticosteroid treatment was gradually tapered and discontinued. Serum immunoglobulin levels were within normal limits for her age (IgM 35 mg/dL, IgA 31 mg/dL, IgG 345 mg/dL), and lymphocyte subgroup analysis revealed no signs of immunodeficiency. Abdominal ultrasonography revealed bilaterally enlarged hyperechogenic kidneys and normal findings for the other abdominal organs. Echocardiographic examination was normal. Panel screening via next-generation sequencing covering well-known genes for CNS, including *NPHS1*, *NPHS2*, *LAMB2*, *PLCE1*, and *WT1*, revealed no mutations.

Intermittent HD was performed three times a week for 4 h using a two-lumen HD catheter (7F) in the right internal jugular vein. The Fresenius 4008 machine and Fx-ped (0.2 m^2) dialyzer were used for HD. System priming with 5% albumin, plasma, or saline was applied for the dialysis sessions with extracorporeal blood pump rates ranging from 30 to 40 mL/h. Anticoagulation was provided by standard heparin infusion. Her continuous tube feeding was stopped to hinder hypotensive attacks during HD sessions, but hypoglycemic attacks and autonomic symptoms (sweating, irritability) were observed during the sessions. Intradialytic parenteral nutrition was started with a glucose perfusion rate of 4 mg/kg/min, and the dialysis solution was changed to a glucose-containing HD solution; however, hypoglycemia persisted. Metabolic screening for hypoglycemia showed no signs of hepatic enzyme deficiencies or fatty acid oxidation defects. Serum electrolytes and insulin levels were within the normal range. Total and free carnitine levels were normal. Repeated

measures of serum cortisol levels were very low (2 mg/dL). ACTH stimulation test showed no appropriate cortisol response and the patient was diagnosed with primary adrenal insufficiency. After the initiation of hydrocortisone treatment, blood glucose levels stabilized. A gastrostomy tube was placed surgically at 9 months of age, but the patient died from severe sepsis 10 days after the surgery.

Questions

1. What is the differential diagnosis for persistent hypoglycemia in HD patients?
2. Which symptoms should warn the clinician for adrenal insufficiency in HD patients?
3. What are the causes of adrenal insufficiency in HD patients?
4. What is the potential cause of CNS and adrenal insufficiency in this patient?

Compliance with ethical standards

Informed consent for genetic testing was obtained from the parents.

Conflict of interest The authors declare that they have no conflicts of interest.