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SCIENTIFIC LETTER

Hyperosmolar Hyperglycemic State: A Rare Presentation of Neonatal Diabetes Mellitus

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To the Editor: A 7-wk-old boy was admitted with fever, loose stools, vomiting, and seizures. His father and maternal grandmother were diagnosed with young-onset diabetes mellitus. Examination and investigations revealed acidotic breathing, normal hemodynamics and systemic examination, blood glucose 991 mg/dL, metabolic acidosis (pH 7.18, PaO_2 38 mmHg, $PaCO_2$ 52 mmHg, bicarbonate 17 mmol/L), urea 111 mg/dL, creatinine 0.7 mg/dL, sodium 183 mmol/L, potassium 5.9 mmol/L, negative blood ketones, and calculated effective osmolarity 421 mOsm/kg. Diagnosis of Neonatal Diabetes Mellitus (NDM) with Hyperosmolar Hyperglycemic State (HHS) was made. Treatment included fluid resuscitation (N/4 5% dextrose for 72 h), insulin infusion (0.02 U/kg/h), antiseizure medications, and mechanical ventilation. Over next 48 h, there was improvement in clinical and metabolic parameters when intravenous fluid was stopped, subcutaneous insulin was started, and he was extubated. As he remained euglycemic, subcutaneous insulin was discontinued after 7 d. Further investigations revealed HbA1c 7.7% (normal <6.5%), C-peptide levels 0.52 ng/mL (normal 1.1–4.4 ng/mL), normal ultrasonography of pancreas, and negative next-generation sequencing for 36 genes associated with monogenic diabetes. Follow up till 13-mo of age shows normal development, normal blood sugar values, and HbA1c of 6%.

NDM is a rare disorder present in infants <6 mo and caused by mutations in genes that affect pancreatic beta-cell function (ATP-sensitive potassium channel genes KCNJ11 and ABCC8) [1]. Elevated HbA1c and low C-peptide suggest NDM. Some cases of NDM may respond to oral sulfonylureas, making genetic testing more crucial. HHS is a rare complication of NDM and it is characterized by very high blood sugar, mild acidosis, low ketones, and high osmolarity. The management includes fluid resuscitation and insulin [2]. Pathogenic mutations may be positive in around 50–60% of cases. NDM is transient in half of the cases with remission within 1–18 mo followed by relapse in childhood or adulthood [2–4].

Declarations

Conflict of Interest None.

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