Infantile Tremor Syndrome (ITS) Presenting as Diabetic Ketoacidosis (DKA)

Kiruthiga Sugumar¹ · Vykuntaraju K. Gowda^{1,2} · Maaz Ahmed¹ · Sanjay K. Shivappa¹

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To the Editor: Infantile tremor syndrome (ITS), is characterized by tremors, delayed development, pallor, skin pigmentation and hair changes [1, 2]. The etiology is commonly attributed to vitamin B12 deficiency [3]. We present a boy who presented initially as diabetic ketoacidosis (DKA) and later was diagnosed to have ITS.

A 1-y-old boy, first born to a non-consanguineously married couple with uneventful perinatal history presented with, delay in attaining age appropriate milestones. The child had not attained head control and social smile. He was exclusively breast fed till 10 mo of age and was started on complementary feeds with inadequate calories and proteins for age. The child was treated in other hospital for cough and tachypnea with ventilator support. The child was found to have persistent metabolic acidosis (ranging from pH - 6.9 to 7.1 and bicarbonate of 3 to 4) requiring intravenous bicarbonate correction, elevated glucose levels (450 mg/dl), positive urinary ketones (+ + +), in view of which he was treated as DKA. At the time of admission to our hospital, child was hemodynamically stable, blood glucose was high, hence treated with insulin infusion and stopped once it was normalized. HbA1C was normal (5.2 mmol/l). His weight was 8 kg; he had pallor, knuckle hyperpigmentation, hypopigmented lusterless scalp hair, hypotonia and areflexia.

Investigations revealed pancytopenia with hemoglobin (Hb) 4 g/dl, mean corpuscular volume (MCV) of 102 fl/red cell. Blood peripheral smear showed macro ovalocytes with hypersegmented neutrophils. Serum vitamin B12 levels were

low (52 ng/ml). Tandem mass spectrometry and Urinary gas chromatography were normal. Child was given packed red cell transfusions and vitamin B12 injections, 1000 mcg/d. Child improved after 3 d, as evidenced by social smile and improvement in appetite. He also developed tremors during hospital stay.

ITS is characterized by pallor, delayed development, knuckle pigmentation and tremors. Our patient had all these features. Investigations revealed pancytopenia, megaloblastic anemia and low vitamin B12 levels. All these findings, supports diagnosis of ITS. The presence of hyperglycemia and ketosis can be explained by stress induced hyperglycemia and starvation ketosis respectively [4]. Acidosis can be due to hemodynamic instability at presentation. As of our knowledge no reports of ITS presenting as DKA are available in the literature. To conclude, ITS, in addition to classical presentation, can present as metabolic acidosis and can mimic DKA and organic aciduria.

Compliance with Ethical Standards

Conflict of Interest None.

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✉ Vykuntaraju K. Gowda
drknvraju@hotmail.com; drknvraju@yahoo.co.in

¹ Department of Pediatric Neurology, Indira Gandhi Institute of Child Health, Bangalore, Karnataka, India

² Bangalore Child Neurology and Rehabilitation Center, No 8/A, First Cross, First Main, Near Adhichunchanagiri Choultry, Vijayanagar, Bangalore, Karnataka 560104, India