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Propionic Acidemia Presenting as Diabetic Ketoacidosis

Propionic acidemia is a rare, autosomal recessive, inherited inborn error of propionate metabolism presenting as life threatening ketoacidosis progressing rapidly to coma and death [1]. Very few cases presenting with hyperglycemia have been described [2,3].

We report a 11 month old girl with propionic acidemia appearing as diabetic ketoacidosis. This child was referred to us for further care with diagnosis of diabetic ketoacidosis. Before coming to us she was admitted with fever, breathlessness and altered sensorium. There was history of failure to thrive and frequent vomiting. She was drowsy and had acidotic breathing, weak peripheral pulses and delayed capillary refill. Investigations revealed hemoglobin of 8.7 g/dL, normal WBC and differential count and low platelet count of 57000/cmm. The blood glucose was 466 mg/dL and urine ketones. Blood gas revealed severe metabolic acidosis (pH were positive -7.18, bicarbonate -4.4 mmol/l) with increased anion gap and increased serum lactate. Glycosylated Hb was high (16.2%). She received treatment with insulin drip, IV fluids and antibiotics. Acidosis and sensorium improved over the next 24 hours after which she was referred to our institution. She weighed 6 kg (<3rd centile) and measured 72 cm in length. She was given subcutaneous regular insulin of 0.5 u/kg/day with daily monitoring of blood glucose. Insulin had to be gradually decreased and then stopped within 7 days

as she had frequent hypoglycemia. She was re-admitted within 7 days for vomiting, dehydration, tachypnea and drowsiness. Investigations revealed WBC of 3400/cmm with normal platelet count, metabolic acidosis, blood glucose of 349 mg%, urine positive for ketones and high serum ammonia of 202 mcg/dL. She was treated with IV fluids and subcutaneous insulin after which she improved within 24 hours. Urine was analysed with a suspicion of organic acidemia by gas chromatography/mass spectrometry, revealed increased excretion of glycine, 3-hydroxypropionic acid, 3-hydroxybutyric acid, 3-hydroxyvalerate, propionyl glycine, tiglylglycine and methylcitrate, which are the biochemical marker compounds of propionic acidemia. Biotin (10mg) and carnitine was added to the therapy and diet with low proteins (1 g/kg/day) was given.

Our initial diagnosis in this patient was type 1 diabetes mellitus. However, rapid response and recurrent hypoglycemia on insulin therapy, leucopenia, thrombocytopenia and hyperammonemia made us suspect an organic acidemia. Most patients with propionic acidemia present with acute episodes of metabolic acidosis, hypoglycemia, hyperammonemia and characteristic organic aciduria due to the decreased activity of intramitochondrial enzyme propionyl CoA carboxylase before they are a few months old.

In our patient, severe hyperglycemia with ketoacidosis was the first life threatening presentation of propionic acidemia to emphasize that hyperglycemia with ketoacidosis is not always diabetes. One should think of propionic acidemia, especially if there is young age of presentation, hyperammonemia, leukopenia, thrombocytopenia and insulin therapy gets withdrawn rapidly.

Rajesh Joshi and Ankur Phatarpekar

*Department of Pediatrics,
BJ Wadia Hospital for Children,
Acharya Donde Marg,
Parel, Mumbai, Maharashtra, India.
rrj23@rediffmail.com*

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Emergence of Metallo- β -Lactamases and Carbapenem Resistance

We read with interest the recent article by Murki, *et al*. on impact of cephalosporins restriction on incidence of extended spectrum β -lactamases (ESBLs) producing gram negative bacteria [1]. Antibiotics restriction and their cycling are no doubt proven strategies to limit emergence of resistant microbial flora, provided they are employed judiciously. However, while attempting this policy in intensive care units (ICUs), one needs to be careful regarding the inadvertent overuse of carbapenem groups of antibiotics, the most potent weapon in our armamentarium to fight ESBL producing gram-negative organisms. Although ESBLs producing bugs have now become a major threat to the utility of cephalosporins, particularly to the broad spectrum third and fourth generation members of this group, the recent resurgence of another group of beta-lactamases, the metallo- β -lactamases (MBLs) in enterobacteriaceae [2] have far more serious threat to the antimicrobial world. They hydrolyze virtually all beta-lactam antibiotics including extended-spectrum cephalosporins and carbapenems, not inhibited by serine beta-lactamase inhibitors like clavulanic acid, sulbactam, and tazobactam, and more seriously, they

are often plasmid-borne making them readily transferable among various species of bacteria [2]. The most worrying part of carbapenem-resistance is that there is hardly any effective antibiotic to treat these infections. With the detection of a new type of MBL, New Delhi metallo-beta-lactamase-1 (NDM-1) from few Indian hospitals has further compounded the problem [3].

Another worrisome aspect is the fact that these MBLs producing carbapenem-resistant organisms are not only confined to the ICUs of big hospitals in metropolitan cities alone but they have also made deep inroads in to the smaller cities of India too. We share our recent experience of treating similar MBL-producing multi-drug resistant (MDR) gram negative infections emanating from a level-3 neonatal intensive care unit (NICU) at Bijnor, a small city of western Uttar Pradesh. Since April 2009, we have treated 14 such neonates admitted in our NICU where nosocomial sepsis was responsible for emergence of MDR gram negative bacteria. The organisms isolated on automated blood culture (BACTEC 9050) included *Klebsiella pneumoniae* (8, 60%), *Acinetobacter baumani* (2, 10%), and *Pseudomonas aeruginosa* (4, 30%). Modified Hodge test was used to screen for MBLs production. All 14 cases showed distorted carbapenem inhibition zones, indicating production of MBLs. These organisms were resistant to all cephalosporins, aminoglycosides, monobactams, quinolones, piperacillin-tazobactam combination, and even to carbapenems. However,