Omer Bashir Abdel Basit, J Clin Case Rep 2016, 6:2 (Suppl)
http://dx.doi.org/10.4172/2165-7920.C1.002

International Meeting on
Clinical Case Reports
April 18-20, 2016 Dubai, UAE

A newborn infant with a reversible cardiomyopathy and non-compaction of the left ventricle

Omer Bashir Abdel Basit
Security Forces Hospital, KSA

A female newborn infant was born after an uneventful pregnancy by an elective cesarean section (CS) because of previous CS. Apgar Score was normal; oxygen saturation 95% and birth weight 2230 grams. At the age one hour the baby started to develop signs of respiratory distress with desaturations. The baby was moved to NICU where she was connected to CPAP. Blood gas showed severe metabolic acidosis and hypoxemia. In view of rising inspired oxygen requirements she was shifted to mechanical ventilation, which was later changed to High Frequency Oscillatory Ventilation (HFOV) plus Nitric Oxide (NO) because of persistent hypoxemia. Clinically there were no abnormal signs in the cardiovascular system and there were no dysmorphic features and the lungs were clear. There was no cardiomegaly on chest X-ray but the ECG revealed low voltage suggesting myocardial dysfunction. An immediate echocardiography revealed cardiomyopathic changes with non-compaction of the left ventricle. At this point the baby was still showing severe hypoxemia, metabolic acidosis and persistent hypotension requiring high doses of dopamine, dobutamine, epinephrine and hydrocortisone. Lactate and ammonia were normal. At the age of 24 hours the baby developed hyponatremia 126 mmol/L, blood glucose of 2.1 mmol/L but BUN, creatinine and potassium were normal. The presence of severe hypotension, metabolic acidosis, hyponatremia and hypoglycemia alerted us to the possibility of adrenal insufficiency and the hydrocortisone dose was increased to physiological dose. This was followed by marked improvement within hours of the blood pressure, sodium and glucose. Maintenance of the mean blood pressure was followed by improvement in oxygenation and mechanical ventilation was gradually phased out. Amino acids and organic acids screening was negative. Thyroid function tests showed very low FT4 going with hypothyroidism but TSH was normal. ACTH and growth hormone were high. 17-hydroxyprogesterone was not elevated. The baby responded well to replacement therapy of hydrocortisone and thyroxin and the cardiomyopathy and the non-compaction of the left ventricle completely resolved on echocardiography. This case represents combined congenital adrenal hypoplasia and hypothyroidism culminating as myocardial failure. The baby is doing well and was discharged home. We are now investigating the hypothalamic-pituitary axis and the possible genetic origin of this case since the parents are cousins.

Biography

Omer Bashir Abdel Basit has received his MBBS degree from University of Khartoum in the year 1971. He has then received his Diploma in Child Health (DCH) from Royal College of Physicians and Royal College of Surgeons of London. He made his Membership at the Royal College of Physicians (MRCP), Ireland (1981) followed by a Fellowship from the same university in the year 1993. Later on he worked as a Consultant Neonatologist at Saudi Council for Health Specialties. His research interests include Perinatal and Neonatal Medicine.

omerabdelbasit@gmail.com

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