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Neonatal Diabetes Mellitus and Hypertrophic Pyloric Stenosis: A Case Report

AA Okolo*, JA Omene**, and JC Odita†

Summary

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An unusual case of neonatal diabetes mellitus and hypertrophic pyloric stenosis is presented. The salient features were severe metabolic acidosis, hyperglycaemia, glycosuria, ketonuria and ketonaemia. The clinical course of the disease was charaterised by increased sensitivity to insulin, recurrent episodes of hypoglycaemia and seizures. The prognosis for intact survival was poor.

Introduction

Neonatal diabetes mellitus is uncommon and two types have been described.[1-3] The first type, associated with ketosis and acidosis requires life-long insulin therapy. The other type which occurs within the first six weeks of life, undergoes spontaneous remission.[1] Hyperglycaemia and deydration are the cardinal signs. An unusual case of neonatal diabetes mellitus and hypertrophic pyloric stenosis is presented in an effort to highlight the problems of diagnosis and management.

College of Medical Sciences, University of Benin, Benin City

Department of Child Health
*Lecturer
**Professor

Department of Radiology
+Senior Lecturer

Correspondence to: Professor JA Omene, Department of Child Health, College of Medical Sciences, University of Benin, Benin City.

Case Report

Baby AO, a male infant, was delivered to a 22-year old primigravida by spontaneous vertex delivery after an uneventful pregnancy. Both parents were young, healthy and unrelated. There was no family history of diabetes mellitus. The Apgar score at birth was 7 at 1 minute and 8 at 5 minutes, respectively. The infant was small for gestational age (SGA) with a birthweight of 2.1 kg and an estimated gestational age of 40 weeks. The length was 44cm and the head circumference was 32cm.

Two hours after delivery, he presented with severe respiratory distress. The lungs and cardiovascular system were essentially normal. The blood sugar level was 6mg/dl and packed cell volume (PCV) was 39%. The anaemia was corrected by simple blood transfusion. The symptomatic hypoglycaemia was treated with 2ml per kg of 25% dextrose solution administered intravenously as a bolus and then followed by an intravenous infusion of 10% dextrose solution.

Subsequent serial blood glucose levels were within normal limits. On the 3rd day of life, hyperventilation was observed, The anterior fontanelle was depressed; the weight had dropped to 1.95kg, representing a 7% loss of the total body weight. Further investigations revealed a blood sugar level of 200mg/dl; sodium, 125mEq/L potassium, 6.4mEq/L; pH, 7.23. Base excess was 14.9 and standard bicarbonate was 12.5mEq/L. Ketonuria, glycosuria and mild ketonaemia were present. These metabolic derangements were corrected within 24 hours of insulin (0.5 units of insulin/kg) therapy and rehydration. The total dose did not exceed 3.5 units per day. The patient was rehydrated with 4.3 dextrose in 0.18 saline. He had several episodes of hypoglycaemia on insulin therapy, three of which were symptomatic thus illustrating marked insulin sensitivity.

Persistent vomiting began at the age of 28 days until the age of 39 days. The diagnosis of upper GIT obstruction was considered and confirmed radiologically by barium meal studies at the age of 39 days. The radiological evaluation revealed evidence of hypertrophic pyloric stenosis. On the 40th day of life, he underwent an operation. The post-operative course was uneventful. The insulin requirements varied widely between 1.5 units/kg/day and 2.5 units/kg/day. There was a rapid weight gain when the blood sugar levels were maintained below 220mg/dl with an average insulin requirement of 0.8 units per kg/day. Before discharge against medical advice, at the age of $4\frac{1}{2}$ months, he weighed 4.5 kg and the head circumference was 35.5 cm.

Discussion

Transient diabetes mellitus is uncommon. Gentz and Cornblath[2] reviewed the literature and reported 50 cases in the first six weeks of life. Of these cases, 11 were considered to be permanent diabetes. In nine cases, death occurred shortly after hyperglycaemia was detected.

The exact incidence of this clinical entity is unknown and it is usually sporadic in occurrence. A positive family history of diabetes mellitus was present in 35% of the cases described by Cornblath and Schwartz,[1] while Milner, Ferguson and Naidu[4] reported transient neonatal diabetes in three siblings. A positive family history of diabetes mellitus was absent in the present case.

A number of aetiological factors have been suggested which includes transient hypothalamic imbalance, adrenocortical disturbance, insulin resistance and absolute hypoinsulinism due to hypoplasia of beta-cells.[4-6] Recent studies by Pagliara, Kerl and Kipnis[6] support the concept that a temporary delay in maturation of beta-cell function may occur in these infants. These authors demonstrated no increase in plasma insulin and negligible changes in the concentration of plasma glucose in response to intravenous tolbutamide during the diabetic state. However, the glucose- and tolbutamide-mediated insulin release was normalised on remission of the diabetic state. In contrast, plasma insulin levels are persistently low in permanent diabetes mellitus.[1] Since no plasma insulin levels were measured in the present case, it was difficult to classify it as permanent or transient diabetes mellitus.

Previous reports[14-10] have emphasised the role of infection in the pathogenesis of neonatal diabetes mellitus. Furthermore, a rarer mode of presentation of neonatal diabetes mellitus is an initial manifestation with hypoglycaemia. Such infants are usually small-for-date.[11] It should be noted that our patient was small-for-date (below the tenth percentile) and the initial blood sugar level was 6mg/dl. A more interesting finding in the present case was the presence of ketonaemia. Cornblath and Schwartz[1] have stressed the absence of ketosis in neonatal diabetes mellitus. The four cases reported by Hutchison[12] had neither ketonuria nor ketonaemia. It would appear therefore, that the present case is the first to be reported presenting with ketonaemia.

Acid-base disturbances, specifically, metabolic acidosis are frequently encountered. The present patient had metabolic acidosis. Changes in serum

electrolytes that have been described, include hypernatraemia and very rarely, hyponatraemia and hyperkalaemia.[1] Hyponatraemia and hyperkalaemia were the main electrolyte imbalance in our patient. This apparent hyponatraemia is not to be interpreted as salt depletion and hypotonic dehydration. In fact, the effective osmolarity of the plasma was 305.6 milliosmoles per litre. The elevated osmolarity in the face of hyponatraemia is partly explained by the contribution of glucose to the effective plasma osmolarity.[13]

To our knowledge, the associated hypertrophic pyloric stenosis in an infant with neonatal diabetes mellitus as demonstrated in this case, is the first to be described. The delay in the diagnosis of the cause of vomiting and consequent dehydration was due to the fact that this symptom and the sign are also present in ketoacidosis. Radiological evaluation was helpful in arriving at the correct diagnosis.

The prognosis in this syndrome is usually good in terms of the duration of insulin therapy.[1] The mean duration of therapy was 69 days with a range of 0–540 days in 35 cases reviewed from the literature by Cornblath and Schwartz.[1] Long-term follow up for psychomotor development has revealed evidence of mental subnormality in three of four cases described by Hutchison.[11] Two of their mentally subnormal cases had hypoglycaemia during the early stages of insulin therapy. Our patient had numerous episodes of hypoglycaemia and subsequently developed microcephaly (head circumference, 35·5cm) at $4\frac{1}{2}$ months of age.

Neonatal diabetes mellitus may be underdiagnosed unless regular blood sugar levels are estimated in all high risk neonates, especially the small-for-date and septicaemic infants.

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