Case Report

A case of hypokalemic metabolic alkalosis

M. Harsha, Shafath Ahamed*, Sundari

ABSTRACT

Bartter syndrome presents with hypokalemia; metabolic alkalosis; increased urinary excretion of sodium, potassium, and chloride; and normal blood pressure. It has a good prognosis after treating with indomethacin. Incidence is rare in neonatal period.

KEY WORDS: Hypokalemic metabolic alkalosis, Indomethacin, Neonatal Bartter syndrome

INTRODUCTION

Bartter syndrome is an inherited renal tubular disorder characterized by hypokalemia, hypochloremic metabolic alkalosis, normal blood pressure with hyperreninemia, and increased urinary loss of sodium, potassium, and chloride. The neonatal form of Bartter syndrome is rare and clinically manifests with failure to thrive, polyuria, and episodes of dehydration. There is improvement with administration of indomethacin and potassium supplements.

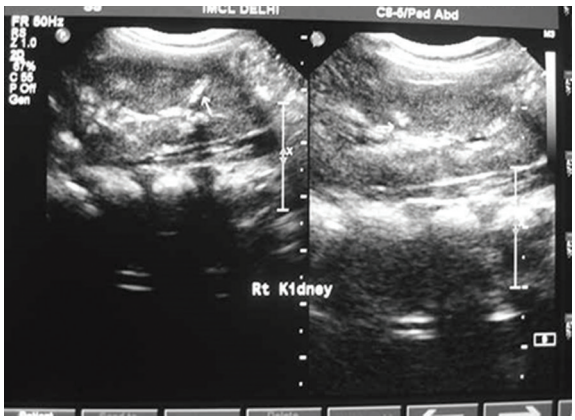
CASE REPORT

A 15-month-old male child presented with vomiting 2–3 episodes per day, poor oral intake, and passing urine frequently with failure to thrive since birth. Antenatally polyhydramnios was detected in 5th month of pregnancy. The male baby born to a non-consanguineous married couple with birth weight of 1.8 kg and no history of birth asphyxia. He was exclusively breastfed for the first 3 months of life after which cow milk was given, and by 5th month of life, complementary feeds were introduced.

At the age of 14 months, he developed fever with respiratory distress, needs hospital admission for 10 days, and was treated with intravenous antibiotics. He was diagnosed with metabolic alkalosis, and there was no history of loose stools, cyanotic spells, seizures, altered sensorium, and focal neurological deficits.

Examination showed weight 4 kg (expected 11.21 kg), length 60 cm (expected 79.4 cm), and head circumference 42 cm (<3rd centile). He had no dysmorphic features. The blood pressure was normal. Systemic examination was within normal limits, and neurological examination did not disclose any localizing signs.

Laboratory investigations: Hypochloremic metabolic alkalosis, hypokalemia, increased urinary losses of K^+, Ca^{++}, and raised aldosterone level [Table 1], confirming the diagnosis of Bartter syndrome. Ultrasound abdomen revealed bilateral medullary nephrocalcinosis.



Ultrasound abdomen revealed medullary nephrocalcinosis.

The child was managed with indomethacin 2 mg/kg, potassium supplementation, and supportive treatment along with dietary advice. On discharge, there was clinical and biochemical improvement with serum potassium of 4.2 mEq/L. On regular follow-up visit, up to present age of 5 years, he has showed a consistent gain in weight.

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Department of Pediatrics, Sree Balaji Medical College and Hospital, Chennai, Tamil Nadu, India

***Corresponding author:** Shafath Ahamed, Department of Pediatrics, Sree Balaji Medical College and Hospital, Chromepet, Chennai, Tamil Nadu, India. E-mail: paedsbmch@gmail.com

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Table 1: Laboratory investigations of the case

Investigations	Case
Serum potassium	2.2 mEq/L
Serum calcium	11 mg/dL
Serum magnesium	3.1 mg/dL
Serum chloride	96 mEq/L
Serum aldosterone	1,0897.4 IU/L
PH	7.8
Bicarbonate	40.4 mEq/L
Urine sodium	38 mEq/L
Urine potassium	8.8 mEq/L
Urine calcium	3.2 mEq/L
Urine creatinine	10.2 mg/dL
Urine calcium:creatinine ratio	0.22
Urine chloride	38 mEq/L

DISCUSSION

Bartter syndrome is an inherited renal tubulopathy, which may manifest during the neonatal period, infancy, or childhood.[1] The sodium potassium 2 chloride cotransporter or the luminal potassium channel ROMK causes neonatal Bartter syndrome, where tubular losses of sodium, potassium, chloride, and water cause secondary hyperaldosteronism.

Gitelman syndrome, however, is a phenotypically related channelopathy instead of being a variant of Bartter's syndrome, affecting sodium chloride cotransporter. Na/Cl cotransporter (NCCT) is present in the distal convoluted tubule. Linkage analysis and mutational studies have revealed defects in the gene encoding sodium chloride cotransporter NCCT.[2] The neonatal variant is particularly uncommon.

Antenatal features include polyhydramnios and premature delivery.[3] Amniotic fluid shows consistently elevated chloride levels, Antenatally polyhydramnios was detected in 5th month of pregnancy.[4-7] After birth, rapid weight loss may occur. Lethargy and poor feeding often develop.[8] Special facial features such as triangular face, prominent forehead, large eyes, strabismus, protruding ears, drooping mouth exist as also sensorineural deafness, convulsion,s and increased susceptibility to infections.

Metabolic alkalosis with hypokalemia occurs in the 1st week of life. Urine has low-specific gravity with very high sodium, chloride, and calcium levels, whereas potassium is normal.[8] However, after 1–3 weeks, the level of potassium in the urine rises with relatively less level of sodium than in the 1st week of life. Prostaglandin levels are high, both in blood and in urine.[8,9] Hyperprostaglandin E_2 is a secondary phenomenon due to fluid and electrolyte loss and is suppressed by appropriate fluid and electrolyte replacement over a period of time. Serum renin and aldosterone levels are also very high and are important in establishing the diagnosis. Untreated infants fail to thrive and may die in a few days as a result of dehydration, poor feeding, or severe electrolyte disturbance. Mild mental retardation is linked to delay in diagnosis and treatment.[8]

Therapeutic efforts should be directed to correct dehydration and electrolytic imbalance.[10] The treatment of neonatal barter syndrome consists of supplementation of potassium (1–3 mEq/kg/day) and after 6–12 weeks of life, administration of indomethacin (2–3 mg/kg/day).[1,3] Ibuprofen (30 mg/kg/day) has similar effect, but indomethacin has been more widely used. The treatment results in striking clinical improvement although serum potassium levels may not increase to above 3.5 mEq/L. Potassium-sparing diuretics and angiotensin-converting enzyme inhibitors have also been used.[1] Prenatal diagnosis can be made by the high-chloride content of the amniotic fluid and mutational analysis of genomic DNA extracted from cultured amniocytes.[11] Bartter syndrome and other renal tubulopathies should be considered in an infant with no obvious cause of failure to thrive and unexplained polyuria. Early investigation and treatment should begin to prevent long-term side effects such as growth failure, nephrocalcinosis, and renal failure.

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