Neonatal Bartter Syndrome in a Case of Severe Polyhydramnios

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Abstract

We report a case neonatal Bartter syndrome in a 27 old pregnant woman with polyhydramnios first detected at 29th gestational weeks requiring multiple amnioreduction. Ultrasonographic evaluation of fetus was normal and neonatal Bartter syndrome was detected in the neonatal period. As a result neonatal Bartter syndrome should be suspected in early onset severe polyhydramnios cases.

Case Reports

Case 1

A 23-years-old primigravid, 33 3/7 week pregnant patient was referred to our clinic with a diagnosis of polyhydramnios. She had regular antenatal visits, polyhydramnios was first detected at 29th gestational weeks, and physical and laboratory examinations were normal except from candida albicans isolated from servico vaginal culture. Obstetric ultrasound revealed a single fetus with a biparietal diameter 85 mm and femur length 60 mm. Amniotic fluid index (AFI) measurement in 4 vertical pockets was 380 mm. Non stress test (NST) was reactive however irregular uterine contractions was present which did not respond to sedation and hydration but ceased with nifedipine tocolysis. NST (4 times daily) and daily AFI measurement was performed. Amnioreduction was first performed 5 days after admission and 2000 cc amniotic fluid was withdrawn. Additional reduction of 9000 cc was carried out with three different procedures two days apart from each other. At 36th gestational week AFI was 340 mm, amnioreduction was repeated and 2000 cc amniotic fluid was collected. Analysis of amniotic fluid revealed that Lecithine/Sphingomyeline (L/S) ratio was 0.038. A final amnioreduction was performed and 1500 cc fluid was withdrawn 2 days later which made the...
total amount 14500 cc. Labor induction was not elected since L/S ratio suggested prematurity. At 37th gestational week labor ensued and cesarean section was performed due to intrapartum severe variable decelerations after careful artificial rupture of membranes.

**Case 2**

A female infant, weighing 2330 g with Apgar scores 2,5,7 for (1,5,10th minutes consecutively) was delivered. The infant was transferred to neonatal intensive care unit where she was followed for 2 days. Her oral feeding and urinary output were noted as normal and did not have any problems except a low Apgar score in the early neonatal period. The infant was hospitalized for vomiting and poor feeding 27 days after delivery. Dehydration and malnutrition was noted. Blood tests showed hypokalemic metabolic alkalosis (pH: 7.65, serum Potassium: 3.2 mEq/L), increased plasma renin activity (135 ng/ml/h), and increased plasma aldosteron concentration (420 ng/dl). Renal ultrasonography revealed bilateral nephrocalcinosis. A diagnosis of neonatal bartter syndrome was made and ibuprofen (15 mg/kg daily) and potassium supplementation was started. The infant had a marked response to therapy and was discharged after 23 days with normal serum electrolyte levels.

**Discussion**

Polycydrannios is a condition associated with significant perinatal morbidity. It is observed in 1-2.8% of all pregnancies. Polycydramnios may be related to maternal or fetal conditions. However the etiology of polycydramnios may not always identified. Pauer et al. reported that 48% of fetuses had severe malformations in polycydramnios cases. Although gastrointestinal tract, central nervous system and the heart are predominantly involved most of the idiopathic cases. Preterm delivery or underlying fetal abnormalities are main reasons for obstetric complications in the presence of polycydramnios.

Overall, worst prognosis is encountered when there is an underlying defect or central nervous system abnormality. Since a variety of reasons may cause polycydramnios determining the onset and severity of polycydramnios is important.

Bartter syndrome is a rare condition which (in the neonatal form) usually complicates pregnancy with polyhydramnios. It is an autosomal recessive condition affecting ion transport channels in the nephron at the loop of Henle. Polyuria is the most striking feature of fetal bartter syndrome which leads to polyhydramnios in a healthy mother and a morphologically normal fetus. Wladimiroff et al. estimated fetal diuresis may reach to 910 ml in 24 hours at 28th week in fetal bartter syndrome while normal values were between 120-200 ml/24 hours. The clinical manifestation is hypokalemic metabolic alkalosis with increased urinary excretion of potassium and secondary hyperaldosteronism.

Madrigal et al. reported 20 cases of fetal Bartter syndrome. Polyhydramnios was present in 90% of patients (18 of 20). Preterm delivery occurred in 90% (18 of 20), mean gestational age at delivery was 32.5 weeks. Proesmeans, in a study of 9 cases with fetal Bartter syndrome, reported the mean onset of polyhydramnios as 25.3 weeks (ranging between 20 to 30) and mean gestational age at delivery was 31.2 weeks (ranging between 27 to 35 weeks). High chloride concentration in the amniotic fluid was also a constant finding in this study median value of chloride being 115 mmol/L which was significantly higher than the normal mean value which was 108 mmol/L.

Despite the presence of severe polyhydramnios labor did not ensue until after 37th week and electrolyte composition of amniotic fluid was not examined in our patient.

**Conclusion**

Polycydramnios is associated with a significant increased risk for perinatal morbidity and mortality. The incidence of fetal abnormalities are higher and the severity of polycydramnios may indicate underlying CNS malformation even in the absence of apparent fetal abnormalities. Fetal Bartter syndrome, although a rare condition, should be suspected in early onset, severe polycy-
dramnios cases without apparent fetal abnormalities. Analysis of amniotic fluid chloride levels may be of some value in prenatal diagnosis of suspected cases.

REFERENCES