Neonatal Bartter Syndrome in a Case of Severe Polyhydramnios

AĞIR POLİHİDRAMNİOS OLGUSUNDA GÖRÜLEN NEONATAL BARTTER SENDROMU

A. Filiz AVŞAR, MD,[a] Cantekin İSKENDER, MD,[b] Melike DOĞANAY, MD,[b]
Özlem Başak ALTUNDAĞ, MD,[b] Savaş ERDEM, MD[a]

[a]Department of Obstetrics and Gynecology, Ankara Atatürk Research and Teaching Hospital,
[b]Department of Obstetrics and Gynecology, Zekai Tahir Burak Women's Hospital, ANKARA

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.

Key Words: Neonatal Bartter syndrome, polyhydramnios

Özet

29. gebelik haftasında polihidramnios tanısı alarak çok sayıda amnioredüksiyona ihtiyaç duyan bir gebelikte neonatal Bartter sendromu olgusunu sunduk. Fetusun ultrasonografik değerlendirmesi normaldi ve neonatal dönemde Bartter sendromu tanısı kondu. Neonatal Bartter sendromu erken başlangıçlı ciddi polihidramnios vakalarında ayırıcı tanıda düşünülmelidir.

Anahtar Kelimeler: Neonatal Bartter sendromu, polihidramnios

Turkiye Klinikleri J Gynecol Obst 2007, 17:54-56

Polyhydramnios is an important obstetric problem which complicates pregnancy. A variety of maternal or fetal conditions may lead to polyhydramnios. Therefore determining the onset and severity is important since they may be related to the underlying cause.

Antenatal Bartter syndrome, an autosomal recessive renal tubular disorders, characterized by hypokalemic metabolic alkalosis is a rare cause of polyhydramnios. Bartter syndrome should be considered in polyhydramnios cases without appearent fetal abnormalities.

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 servicovaginal 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

Geliş Tarihi/*Received:* 29.03.2006 **Kabul Tarihi/*Accepted:*** 01.09.2006

Yazışma Adresi/*Correspondence:* Cantekin İSKENDER, MD
Zekai Tahir Burak Women's Hospital,
Department of Obstetrics and Gynecology, ANKARA
c_iskender@yahoo.com

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. Dehydratation 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

Polyhydramnios is a condition associated with significant perinatal morbidity. It is observed in 1-2.8% of all pregnancies.[1] Polyhydramnios may be related to maternal or fetal conditions**.** However the etiology of polyhydramnios may not always identified. Pauer et al. reported that 48% of fetuses had severe malformations in polyhydramnios cases.[2] Although gastrointestinal tract, central nervous system and the heart are predominantly involved most of the idiopathic cases.[3] Preterm delivery or underlying fetal abnormalities are main reasons for obstetric complications in the presence of polyhydramnios.[4]

Overall, worst prognosis is encountered when there is an underlying defect or central nervous system abnormality. Since a variety of reasons may cause polyhydramnios determining the onset and severity of polyhydramnios is important.[5]

Bartter syndrome is a rare condition which (in the neonatal form) usually complicates pregnancy with polyhydramnios.[6,7] 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 polyhdramnios 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.[8] The clinical manifestation is hypokalemic metabolic alkalosis with increased urinary excretion of potassium and secondary hyperaldosteronism.[7-9]

Madrigal et al. reported 20 cases of fetal Bartter syndrome. Polyhydramnios was present in 90% of patients (18 of 20). Preterm delivery occured in 90% (18 of 20), mean gestational age at delivery was 32.5 weeks.[7] 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.[9]

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

Polyhydramnios is associated with a significant increased risk for perinatal morbidity and mortality. The incidence of fetal abnormalities are higher and the severity of polyhydramnios may indicate underlying CNS malformation even in the absence of appearent fetal abnormalities.[5] Fetal Bartter syndrome, although a rare condition, should be suspected in early onset, severe polyhy-

dramnios cases without appearent fetal abnormalities. Analysis of amniotic fluid chloride levels may be of some value in prenatal diagnosis of suspected cases.

REFERENCES

1. Barkin SZ, Pretorius DH, Beckett MK, Manchester DK, Nelson TR, Manco-Johnson ML. Severe polyhydramnios: Incidence of anomalies. AJR Am J Roentgenol 1987;148: 155-9.
2. Pauer HU, Viereck V, Krauss V, Osmers R, Krauss T. Incidence of fetal malformations in pregnancies complicated by oligo- and polyhydramnios. Arch Gynecol Obstet 2003;268:52-6.
3. Lazebnik N, Many A. The severity of polyhydramnios, estimated fetal weight and preterm delivery are independent risk factors for the presence of congenital malformations. Gynecol Obstet Invest 1999;48:28-32.
4. Mazor M, Ghezzi F, Maymon E, et al. Polyhydramnios is an independent risk factor for perinatal mortality and intrapartum morbidity in preterm delivery. Eur J Obstet Gynecol Reprod Biol 1996;70:41-7.
5. Biggio JR, Wenstrom KD, Dubard MB, Cliver SP. Hydramnios prediction of adverse perinatal outcome. Obstet Gynecol 1999;94:773-7.
6. Sieck UV, Ohlsson A. Fetal polyuria and hydramnios associated with Bartter's syndrome. Obstet Gynecol 1984;63(Suppl 3):22S-24S.
7. Madrigal G, Saborio P, Mora F, Rincon G, Guay-Woodford LM. Bartter syndrome in Costa Rica: A description of 20 cases. Pediatr Nephrol 1997;11:296-301.
8. Wladimiroff JW, Barentsen R, Wallenburg HC, Drogendijk AC. Fetal urine production in a case of diabetes associated with polyhdramnios. Obstet Gynecol 1975;46: 101-2.
9. Proesmans W. Bartter syndrome and its neonatal variant. Eur J Pediatr 1997;156:669-79.