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Perinatal Outcome of a Fetus Presenting with an Isolated Umbilical Cyst

İzole Umbilikal Kist ile Prezente olan bir Fetusun Perinatal Sonuçları

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ABSTRACT
The widespread use of high-resolution obstetric ultrasound has allowed extensive evaluation not only of the fetus, but also of the placenta and umbilical cord. Most of the available data on complication rates and clinical course of fetuses with umbilical cord cysts are limited and is in the form of case reports and small case series. We aimed to present the outcome of a fetus with an isolated umbilical cord cyst on the third trimester. The fetus was delivered by cesarean section because of fetal distress and died three days later following the operation due to perinatal asphyxia. It should be remembered that large umbilical cysts may cause fetal distress by pressure on the cord vessels in case of rapid growth. It is vital to consider the ultrasonographic examination of the umbilical cord as one of the important parts of the examination.
Keywords: Umbilical cyst; fetal anomaly; umbilical cord.

ÖZ
Yüksek çözünürlüklü obstetrik ultrasonun yaygın kullanımı ile sadece fetusun değil, aynı zamanda plasenta ve umbilikal kordun geniş çaplı değerlendirilmesi mümkün olmuştur. Umbilikal kord kistleri olan fetusların klinik seyri hakkındaki mevcut verilerin çoğu sınırlıdır, daha çok komplikasyon oranları ile ilgilidir ve vaka raporları ve küçük vaka serileri biçimindedir. Bu vaka sunumunda; üçüncü trimesterde izole umbilikal kord kisti ile prezente olan bir fetusun sonucunu sunmayı amaçladık. Fetüs, fetal distres nedeniyle sezaryen ile doğurtuldu ve operasyondan üç gün sonra perinatal asfiksi nedeniyle exitus oldu. Büyük umbilikal kistlerin hızlı büyüme durumunda kordon damarları üzerinde baskı ile fetal sıkıntıya neden olabileceği unutulmamalıdır. Umbilikal kordun ultrasonografik incelemesinin, muayenenin önemli kısımlarından biri olarak kabul edilmesi hayati önem taşımaktadır.
Anahtar kelimeler: Umbilikal kist; fetal anomali; umbilikal kord.

INTRODUCTION

The umbilical cord is a vital connection between placenta and fetus. It maintains a stable connection of maternal-fetal interface. Umbilical cord allows the fetal mobility, growth and neuromotor development (1). The widespread use of high-resolution obstetric ultrasound has allowed extensive evaluation not only of the fetus, but also of the placenta and umbilical cord.

The finding of an umbilical cord cyst in a pregnant woman raises a question about the clinical significance of this finding. Since the details on affected pregnancies are based mainly on the findings of case reports, the prognosis and outcome for fetuses with this cord anomaly remain unclear. We herein aimed to present the outcome a fetus with an isolated umbilical cord cyst on the third trimester.

CASE PRESENTATION

A gravida 2, para 1 woman aged 22 years old applied obstetrics and gynecology emergency unit with a complaint of decrease of fetal movements. She was 33w 4d gestational age according to her last menstrual period. Her obstetric history was unremarkable. The patient had not wanted to have first and second trimester screening tests and was not attending her pregnancy follow-ups regularly. Her physical examination and vital signs were uneventful. Current sonographic examination of the fetus revealed a cystic mass with a diameter of 64x54 mm originating from umbilical cord insertion at the umbilicus, containing two arteries and one vein. Polyhydramniosis was also observed.

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The fetal biometric measurements were compatible with 33-34 weeks of gestation and no additional abnormality was observed. In cardiotocography fetal hearth beat revealed bradycardia. Fetal heart rate was between 100 and 110 beats per minute. Cardiotocographic and ultrasonographic appearance of the fetus were shown in Figure 1 and 2. The woman was hospitalized for close fetal monitorization and perinatology consultation. Perinatology consultation confirmed umbilical cyst diagnosis and offered cordocentesis. The patient refused invasive procedure. Cesarean section was performed due to fetal distress on the second day of hospitalization. The newborn died three days later following the operation due to perinatal asphyxia. Postmortem genetic analysis revealed 46, XY karyotype.

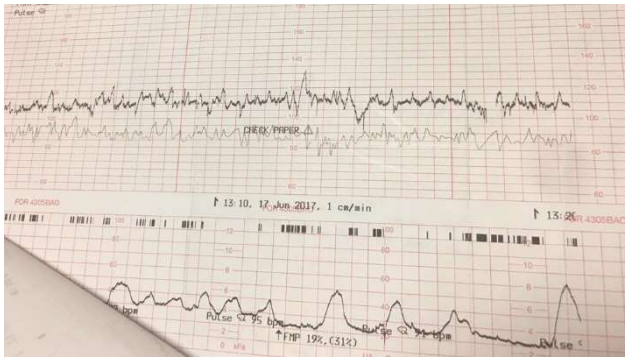

Figure 1. Cardiotocography of the fetus

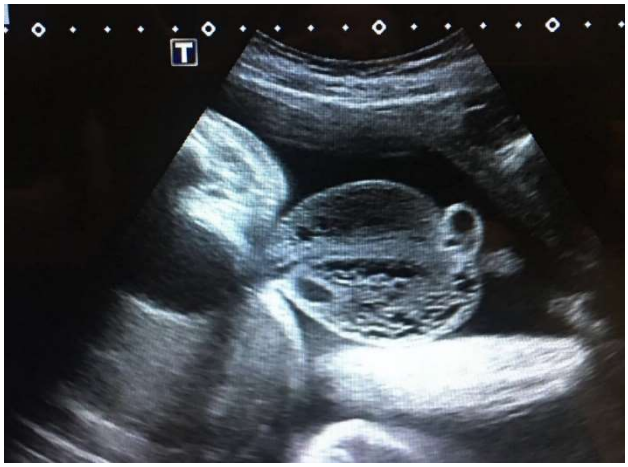

Figure 2. Ultrasonographic appearance of umbilical cord cyst

Figure 3. Umbilical cord cyst of the newborn

DISCUSSION

The etiology of umbilical cord cyst is unknown. Umbilical cord cyst can be easily recognized by ultrasonography at various stages of gestation (2). The prevalence in the first trimester is 0.4% to 3.4%. The second and third trimester prevalence is unknown (3).

There is a difference in the clinical significance and the prognosis between first and second-third trimester umbilical cord cysts. Many first trimester cysts disappear by the second trimester (4). Since the presence of umbilical cord cysts on second and third trimester sonography has been associated with chromosomal abnormalities including trisomy 13 and 18 as well as structural defects such as vertebral defects, omphalocele, tracheo-esophageal fistula, imperforate anus, and angiomyxoma of the cord (5-9); cases of umbilical cord cysts should be followed up by detailed investigation for additional malformations.

Both Smith et al. (10) and Sepulveda et al. (11) concluded that there were additional fetal anomalies in 80-85% of the cases. Ross et al. (12) reported 100% correlation between fetal anomalies and persistent second trimester cysts. Shipp et al. (13) reported fetal anomalies in 38% of the cases. In the study of Zangen et al. (14), the umbilical cord cyst was the only abnormal finding in the seven of the 10 cases. In our case; although the patient had not wanted to have first and second trimester screening tests and was not attending her pregnancy follow-ups regularly, her last ultrasonographic evaluation did not reveal any additional fetal anomaly.

Umbilical cord cysts are categorized into true and pseudocysts. Pseudocysts are more common and not surrounded by epithelium. They are formed by liquefaction of Wharthon's gelly and localized edema (15). True cysts are composed of allantois and omphalomesenteric canal residues. It is not usually possible to differentiate the true and pseudocysts by ultrasonography (14). In our case; although umbilical cyst could not be categorized into true or pseudocyst in prenatal ultrasonography, the result of pathologic examination was pseudocyst.

Some studies have shown that there is a relationship between morphological structures of the cord cysts and chromosomal anomalies. It has been suggested that fetuses with small and numerous cord cysts are more often associated with chromosomal anomalies than fetuses with single large cysts (16). Sepulveda et al. (11) showed a correlation between small multiple cysts and aneuploidy. Ross et al. (12) suggested that those close to fetal or placental insertion sites are correlated with fetal aneuploidy and also concluded that smaller cysts had a better prognosis than larger cysts. Ghezzi et al (9) found that multiple umbilical cord cysts were more frequently associated with an increased risk of aneuploidy and miscarriage. In the presented case; the fetus had a single large umbilical cyst with a diameter of 64x54 mm originating from umbilical cord insertion at the umbilicus. Cordocentesis was advised for prenatal genetic diagnosis, but the patient denied invasive procedure. Postmortem genetic analysis of the newborn revealed 46, XY karyotype.

The outcome for fetuses with cord anomaly remain unclear. However, there are a few series reporting the outcome of pregnancies with umbilical cord cysts. In a study of Shipp et al. (13) presented a series of 13 cases of umbilical cord cyst that were detected during the second and third trimesters. They stated that overall, 12 of the 13 newborns survived and the vast majority had a favorable outcome. Sepulveda et al. (11) reported the outcome of 13 fetuses with umbilical cord cysts in the second and third trimesters of pregnancy. Eleven of the cases had additional sonographic findings. Prenatal karyotype testing was carried out in 10 of these fetuses. Aneuploidy was noted in seven cases. In the 3 cases with normal karyotype, isolated omphalocele in one and multiple anomalies were found in two cases. Two chromosomally normal fetuses with associated multiple structural defects and all chromosomally abnormal fetuses died in utero or during the neonatal period. Smith et al. (10) reported the outcome of 3 cases with umbilical cord cysts. One, in which

a transient cyst was detected at the end of the first trimester. This case had a normal outcome. Two other cases in which the cyst was detected at 23 and 39 weeks of gestation, were diagnosed as having trisomy 18. Hannaford et al (17) compared patients with umbilical cord cysts to those with normal umbilical cords and they did not find an association with umbilical cord cysts in the first trimester and pregnancy complications. They expressed umbilical cord cysts in the first trimester may in fact be physiologic findings in the development of a normal umbilical cord. They suggested that first-trimester umbilical cord cysts should not be considered markers of poor pregnancy outcomes if they resolve by the start of the second trimester. Ruiz Campo et al. (18) revealed that the prognosis of this finding seems to be favorable when isolated and there is no relation between prognosis and gestation weeks at diagnosis. In the presented case, the umbilical cyst was detected in the third trimester. Patient was presented with fetal bradycardia. Any accompanying fetal anomaly was encountered. The cyst was single, located near the umblical cord insertion, with a diameter of 64x54 mm and containing two arteries and one vein (Figure 2 and 3). Cesarean section was performed due to fetal distress. Fetal bradycardia and fetal distress seemed to be as a result of compression of umbilical vessels. The physical examination and appearance of the newborn was compatible with perinatal asphyxia. There was no syndromic stigmats. The newborn died three days later following the operation due to asphyxia. Postmortem genetic analysis of the newborn revealed 46 XY.

We herein presented a case with umblical cord cyst. Most of the available data on complication rates and clinical course are limited and is in the form of case reports and small case series. We believe that it is vital to consider the ultrasonographic examination of the umbilical cord as one of the important parts of the examination. It should also be remembered that large umbilical cysts may cause fetal distress by pressure on the cord vessels in case of rapid growth.

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