

Umbilical cord stenosis and umbilical cord torsion: a case report

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Abstract

Objective: The aim of this case report is to discuss umbilical cord torsion, which is a rare condition, develops due to cord stenosis and may cause intrauterine fetal death.

Case: Fetal death was detected during the routine examination of a woman who was on 32 weeks of gestation and having her first pregnancy at 26 years old. No problem was encountered during regular antenatal follow-up visits. There was no chronic disease or infection in her medical history. In the postnatal examination of intrauterine dead fetus, it was found that umbilical cord was entangled around the fetus three times and there was a stricture about 1 cm in this torsioned area.

Conclusion: The stricture of umbilical cord may cause fetal mortality. Cord stricture and umbilical cord torsion developing due to cord stricture are rare causes of intrauterine death.

Keywords: Intrauterine fetal death, umbilical stenosis, umbilical cord torsion.

Özet: Umbilikal kord stenozu ve buna bağlı gelişen umbilikal kord torsiyonu: Olgu sunumu

Amaç: Bu olgu sunumunun amacı nadir gözlenen ve intrauterin fetal ölüme neden olabilen kord stenozuna bağlı gelişen umbilikal kord torsiyonunu tartışmaktır.

Olgu: Yirmi altı yaşında ilk gebeliği olan, otuz iki haftalık gebenin rutin kontrolleri sırasında fetal ölüm saptandı. Düzenli olarak takibi gerçekleştirilen hastanın antenatal takiplerinde herhangi bir sorun saptanmamıştı. Gebenin özgeçmişinde herhangi bir kronik hastalık veya enfeksiyon yoktu. İntrauterin ölü fetüsün doğum sonrasında yapılan incelemesinde umbilikal kordun üç kez kendi etrafında dolandığı ve torsiyone olan bu alanda yaklaşık 1 cm striktür varlığı saptandı.

Sonuç: Umbilikal kord striktürü fetal mortaliteye neden olabilir. Kord striktürü ve buna bağlı gelişen umbilikal kord torsiyonu intrauterin ölümün nadir bir sebebidir.

Anahtar sözcükler: İntrauterin fetal ölüm, umbilikal stenoz, umbilikal kord torsiyonu.

Introduction

The rate of intrauterine fetal death is reported as five per thousand.^[1] While infection, detachment and congenital anomalies are considered among the etiological factors of fetal deaths earlier than 28 weeks, the most common causes in pregnancies on 28 weeks and above are growth retardation and ablatio placentae. On the other hand, no factor can be found in 27–50% of intrauterine fetal deaths.^[2]

Although umbilical cord defects are seen relatively less, they are among the severe gestational complications that may lead to high incidence of intrauterine fetal

death. Umbilical cord among umbilical cord anomalies is the most common reason for this case.^[3] In case of the torsion of umbilical cord, blood flow decreases below to the critical level and this may lead fetal hypoxia, oligohydramnios, intrauterine growth retardation (IUGR) and fetal death.^[4] The purpose of this report is to discuss cord stricture, which is seen rare and may lead fetal death, and umbilical torsion case developing after cord stricture.

Case Report

All follow-up procedures of the patient, who had her first pregnancy at 26 years old, were carried out in

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Received: July 27, 2015; **Accepted:** February 22, 2016

Please cite this article as: Akgün N, Köşüş N, Köşüş A, Yardımcı F. Umbilical cord stenosis and umbilical cord torsion developing in parallel: a case report. Perinatal Journal 2016;24(1):47–50.

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Available online at:
www.perinataljournal.com/20160241005
doi:10.2399/prn.16.0241005
QR (Quick Response) Code:



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another center. It was learnt from the investigations performed that the results of double and triple screening tests were normal and no pathology was found in the detailed ultrasonography. The patient referred to our clinic with the complaint that she did not feel the movements of baby for a day, and it was found in the ultrasonographic examination that the fetus with oligohydramnios and consistent with 29 weeks had no fetal heart beat. Later, cervical prostaglandin E1 was applied for labor induction and 1370 g singleton dead male baby was delivered vaginally. After delivery, fetus, placenta and accessories were examined in detail. The fetus had maceration, but no gross fetal anomaly was detected. Placenta was 425 g and in normal appearance. Two arteries and one vein were observed in the umbilical cord, the length of the cord was within normal limits (64 cm) and the number of spirals was normal (0.5 spiral/cm). However, it was seen that the umbilical cord was torsioned three times on its own axis where it enters into anterior wall of fetal abdomen. After the cord detorsioned, umbilical stricture was found on an area of about 0.5 cm beginning from the area where the cord exits anterior abdominal wall, and it was also observed that the cord was edematous and ecchymotic on this area. Also, maceration was seen around the abdomen on an area of about 2 cm (**Fig. 1**). Autopsy could not be performed since the family did not allow. To rule out other causes of fetal death, chromosome analysis, direct radiography, infection panel, autoimmune markers and thrombophilia panel were requested. Direct radiography was done to evaluate malformations in the musculoskeletal system. No pathology was found in the direct radiography (**Fig. 2**). Chromosome analysis could not be performed on the fetus since fetal cells did not reproduce from cell cultures in the genetic examination. Other test results were evaluated as normal. In the pathologic examination of the placenta, perivillous fibrin thrombi and focal chorangiosis were found. Upon the fact that the findings in the first examination indicated a death secondary to the torsion, umbilical cord stricture and torsion were considered as the primary cause of death.

Discussion

Tests such as maternal blood, toxoplasma, rubella, cytomegalovirus, herpes simplex, antinuclear antibody, antiphospholipid antibodies and lupus anticoagulant tests, and thrombophilia panel, fetal autopsy, placental pathological examination and chromosome analyses



Fig. 1. Torsion of umbilical cord and complete periumbilical ecchymosis and skin maceration.

performed to determine the cause of intrauterine death have a significant role for clarifying the etiology of still births with unknown reason.^[5] Yet, the reason cannot be determined in most of the cases. The rate of stillbirths with unknown reason within all stillbirth cases varies between 12 and 50%.^[6]

While the reasons of umbilical cord stricture are not fully known, folding of the cord during the movements when especially umbilical cord is long and fetal movements are intense and the shortage of Wharton gel may lead to umbilical stricture.^[7] The length of umbilical



Fig. 2. Direct radiography of fetus.

cord is determined genetically as well as by fetal movements. While more movements provide the development of longer cord, less fetal movements cause the development of shorter cord.^[7] Normal cord length is 35–70 cm, and umbilical cord is protected against pressure, stress and entanglement issues thanks to the spiral structure of cord.^[7] Cord stenosis is seen more when umbilical cord is long and has high number of spirals and umbilical coiling index is above 0.6.^[8] Some studies indicate that there is an underlying cord stenosis and torsion in cases such as especially fetal cardiac arrhythmia, cardiac failure, oligohydramnios, non-immune hydrops and IUGR.^[9] However, in our case, the length and coiling index rate of umbilical cord were normal and no excessive fetal movements were found in the anamnesis. Also, fetal movements increase with the increase of amniotic increase, and it was reported by some studies that umbilical stenosis is observed more in case of polyhydramnios.^[8] We did not perform the regular follow-ups of the patient; however, no pathology was seen in the anamnesis for her routine follow controls. Oligohydramnios was not found in her previous routine controls. There was oligohydramnios in the ultrasonographic examination during her referral. As known, amniotic fluid amount is determined by the fluid amount flowing into amniotic cavity through fetal hemodynamics, fetal urination, fetal respiration and transmembraneous way. Chronic hypoxia and placental failure are among the most significant reasons of oligohydramnios. In this patient, the reason for detecting oligohydramnios during terminal period may be related with chronic hypoxia increasing due to the exacerbation of umbilical artery stenosis and/or torsion and disruption in fetal circulation and insufficient fetal urination as a result. Abnormalities in Wharton gel protecting umbilical veins on the cord area with umbilical stenosis can be considered among the factors leading to umbilical stenosis and torsion.^[10] Wharton gel consists of mucopolysaccharides containing mainly hyaluronic acid and chondroitin sulfate^[10] and its main role is to replace adventitia in cord veins which do not have adventitia layer. In this way, it protects cord veins by providing elasticity and partial rigidity. Without Wharton gel, vasoconstriction and stenosis-related fetal complications may develop.

Although the disease is sporadic, there are also studies showing that it may present aggregation and recurrence.^[11] Also, the studies state that the risk increases in

twin pregnancies conceived by assisted reproductive techniques after invasive procedures such as amniocentesis.^[12] However, this was not the case in our patient.

Umbilical cord ultrasonography and the benefit of Doppler ultrasonography carried out to establish the diagnosis of umbilical cord anomalies antenatally have still been controversial.^[13] Ultrasonographically measuring the diameter and area of umbilical cord at over 20 weeks of gestation is known to be beneficial to determine risky fetuses for IUGR and intrapartum complication.^[14] The number of the umbilical cord spiral is a significant predictive value in ultrasonographic examination. Umbilical coiling index being >0.6 or >90 percentile is defined as hypercoiling, and IUGR, fetal arrhythmia and decelerations, fetal death and cord stenosis are seen more commonly in this case.^[8,15]

It was shown that the umbilical cord found to be thin ultrasonographically had less Wharton gel compared to normal cords. On the other hand, local cord strictures may develop on any area, but it is seen mostly on the area where umbilical cord enters into fetal abdomen.^[16] In general, umbilical cord complications may manifest themselves with the decrease of fetal movements due to chronic intrauterine hypoxia, decrease in fetal development and oligohydramnios.^[4] However, different clinical results may occur; the patient may also refer to hospital with fetal death without any sonographic indication or intrauterine growth retardation.^[4] It is important to examine umbilical cord in the prenatal evaluation. However, sudden changes may occur in Doppler ultrasonography, development of fetal detorsion, fetal activity and physiological indications.^[17] Therefore, preventing the complication is almost impossible in some patients. Besides, the families who have intrauterine fetal death due to umbilical cord torsion should be informed in detail that there is a recurrence risk, and the family should be recommended a strict fetal monitoring in the follow-up visits of next pregnancy.^[7]

Conclusion

Umbilical cord abnormalities are among the pathologies which are still not clarified for the factors in their etiologies, do not have sufficient data on their diagnosis and treatment but may lead to intrauterine fetal death.

Conflicts of Interest: No conflicts declared.

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