

Fetus papyraceus in dichorionic diamniotic twin pregnancy: a case report

Zgon wewnątrzmaciczny jednego z płodów w II trymestrze ciąży bliźniaczej dwukosmówkowej dwuowodniowej. Opis przypadku

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Abstract

Introduction: The actual rate of multiple pregnancies is significantly larger than that observed during labor due to the fact that in the course of pregnancy intrauterine death of one or more fetuses may occur.

Case presentation: A twenty-six old woman (GII, PII) reported to hospital in 26 weeks of DC/DA twin gestation complicated by intrauterine death of one fetus in the second trimester of a spontaneous pregnancy. Pregnancy ended at term with vaginal birth of a single live fetus. After birth, entanglement of the fetal umbilical cord around the leg of the dead fetus was discovered. It was the most probable cause of death.

Conclusions: Conservative management is preferred in case of intrauterine demise of one of the fetuses in DC/DA twin pregnancy. One of the reasons of fetal death may be entanglement of the umbilical cord around fetal small parts. The time of fetal death can be determined on the basis of the length of the thigh bone (Femur Length – FL).

Keywords: fetus papyraceus / intrauterine death / dichorionic diamniotic twins /

Streszczenie

Prezentujemy przypadek ciąży bliźniaczej DK DO z płodem papierowatym, zakończonej porodem żywego współbliźniaka w 37 tygodniu trwania ciąży.

Faktyczny odsetek ciąż wielopłodowych jest znacznie większy niż obserwowany w czasie porodu. W trakcie przebiegu ciąży dojść może bowiem do zgonu wewnątrzmacicznego jednego lub większej liczby płodów.

26-letnia kobieta (GII PII) przybyła do szpitala w 26 tygodniu ciąży bliźniaczej powikłanej zgonem jednego z płodów. Ciąża powstała spontanicznie. Ciężarna była pod opieką lekarza ginekologa od 10 tygodnia ciąży. Dotychczasowy przebieg ciąży – bez powikłań. Wcześniej wykonywane badania usg wykazywały prawidłowy rozwój płodów. W chwili przyjęcia ciężarna zgłaszała nieregularne skurcze macicy oraz plamienie z dróg rodnych.

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Badanie wewnętrzne wykazało obecność uformowanej szyjki macicy oraz zachowane błony płodowe. Badanie USG: płód „A” martwy, położony poprzecznie, FL 39mm = 22 hbd, bez widocznych wad rozwojowych. (Rycina 1 i 2). Największa kieszeń płynu owodniowego 45 mm. Płód „B” żywy, w położeniu podłużnym główkowym, BPD i FL odpowiadają 26 tygodniowi ciąży, bez widocznych zaburzeń rozwojowych. Prawidłowa objętość wód płodowych. Badanie dopplerowskie - prawidłowy przepływ w naczyniach pępowinowych i wewnątrzpłodowych. Łożysko podwójne złączone (ciąża dwukosmówkowa dwuowodniowa), położone w dnie macicy i na ścianie przedniej. Pobrano badania laboratoryjne oraz posiew z pochwy. Wyniki badań laboratoryjnych – prawidłowe. Zastosowano fenoterol we wlewie dożylnym. Wdrożono postępowanie zachowawcze z oceną morfologii, CRP, profilu koagulologicznego i funkcji nerek w odstępach tygodniowych oraz nadzór nad stanem płodu. Ciężarna wymagała 3 krotnie wsparcia psychologa. Okresowo była wypisywana do domu.

W 37 tygodniu ciąży wystąpiła samoistna czynność skurczowa macicy. Wyniki badań laboratoryjnych prawidłowe. Zapis kardiotokograficzny żywego bliźnięcia prawidłowy. Badaniem wewnętrznym stwierdzono część pochwową całkowicie zgładzoną, rozwarcie 4 cm. Pęcherz płodowy zachowany. W dalszym przebiegu porodu doszło do wtórnego osłabienia czynności skurczowej macicy i zahamowania postępu porodu przez przodujący, znajdujący się w położeniu poprzecznym martwy płód. Po przebicciu pęcherza płodowego i wydobyciu martwego płodu za stópki dalszy przebieg porodu był bez powikłań.

Urodzony płód papierowaty był płci męskiej o ciężarze 200g, grubości zaledwie 1 cm, z łożyskiem ważącym 100g (Ryc. 3). Sznur pępowinowy miał długość 40 cm, zawierał 3 naczynia i był okręcony dookoła podudzia lewego płodu (Ryc. 4). Bliźnię to nie posiadało żadnych nieprawidłowości rozwojowych. Narządy wewnętrzne wykazywały zgodny rozwój z wiekiem ciążowym.

Po godzinie, spontanicznie drogami natury urodziło się bliźnię „B” żywe, płci żeńskiej, o masie 2900g, w stanie ogólnym dobrym (10 punktów w skali Apgar). Śródporodowa utrata krwi wyniosła 250 ml. Badanie makroskopowe popłodu wykazało obecność ciąży DK DO, a badanie histologiczne łożyska obecność wykładników stanu zapalnego. Grupa krwi matki B Rh plus, żywego płodu AB Rh plus.

Matka wraz z dzieckiem zostali wypisani w 3 dobie po porodzie. Do dnia dzisiejszego rozwój dziecka pozostaje prawidłowy.

Słowa kluczowe: **płód papierowaty / śmierć wewnątrzmaciczna / bliźnięta dwukosmówkowe dwuowodniowe/**

Introduction

The actual rate of multiple pregnancies is significantly larger than that observed during labor due to the fact that intrauterine death of one or more fetuses may occur in the course of pregnancy [1]. Depending on the gestation period in which fetal death occurs, there are 3 forms of this complication:

- 1st trimester of pregnancy – ‘vanishing twin’ syndrome,
- 2nd trimester of pregnancy – fetus papyraceus s. compress,
- 3rd trimester of pregnancy – macerated fetus.

The incidence of intrauterine death of one of the fetuses in the second trimester of a twin pregnancy ranges from 1:184 to 1:200 twin pregnancies and 1:17000 – 1:20000 of total births (even higher proportion is observed in triplet pregnancies). During this period, a complete disappearance of the dead fetus, due to a significant degree of development of the tissues, especially bone, becomes impossible. If death occurs between 15 and 20 weeks of gestation, mummification and then pressing by the expanding amniotic sac of the second fetus occurs. A “papyraceus” or a “compressed” fetus is slowly formed [2-4]. Type of twin pregnancy (mono- or dichorionic) and the period in which the death of one of the twins occurred have a significant impact on the prognosis of the surviving fetus and the occurrence of complications in the mother.

We present a case of DC/DA twin pregnancy with a papyraceus fetus, with successful outcome of the second twin in 37 weeks of gestation.

Case Report

A twenty-six year old woman (GII, PII) reported to hospital in 26 weeks of spontaneous twin gestation complicated by the death of one fetus. The patient was under supervision of a gynecologist since 10 weeks of gestation. Previous pregnancy was uneventful. Earlier ultrasound examination showed normal development of both fetuses. On admission, the pregnant woman reported irregular uterine contractions and vaginal spotting. Internal examination revealed a normal sized cervix and amniotic fluid saved. The ultrasound examination found fetus ‘A’ dead, in a transverse lie, FL 39mm = 22 hbd, without visible malformations, (Figure 1 and 2). The largest pocket of amniotic fluid was 45mm. Fetus ‘B’ was alive, in a cephalic longitudinal lie. BPD and FL corresponded to 26 weeks of pregnancy, with no visible developmental disorders. Adequate amniotic fluid. Doppler test revealed normal flow in the umbilical and fetal vessels. Fused double placenta (dichorionic diamniotic gestation), located at the bottom of the uterus and the anterior wall. Laboratory testing and vaginal culture were taken. Laboratory testing results were normal. Partusisten infusion was administered. Conservative treatment was implemented with the assessment of morphology, CRP, coagulological profile (PT, APTT, PDF, platelets and fibrinogen concentration) and renal function in weekly intervals. Fetal surveillance included daily measurement of the number of movements, NST every other day initially, then once a week. Ultrasound performed at weekly intervals showed normal growth of fetus ‘B’, while in twin ‘A’ it revealed a gradual decrease in the volume of amniotic fluid and biparietal dimension (BPD) of the head. Femur length (FL) remained unchanged until the end of pregnancy, allowing to

establish the probable time of fetal death (Figure 2). The patient required professional psychological help three times. Periodically, she was discharged from hospital to home.

In the 30th week of pregnancy, markers of intrauterine infection (body temperature up to 37.8° C; CRP 40 mg/l; WBC 17.2 G/l; analysis of vaginal culture demonstrated the presence of *Enterococcus faecalis*) and disorders in blood coagulation (platelets 120,000/ml; fibrinogen 3.62 g/l; D-dimers 3.12 ug/ml) were recognized. Antibiotherapy (augmentin, metronidazole) and anticoagulant prophylaxis (Clexane 80 mg/day) were administered. Betamethasone was administered 2x12mg. After a week test results returned to normal. Fetus 'B' was in a good condition.

Spontaneous contractile activity of the uterus occurred in the 37th week of pregnancy. The patient was admitted to the hospital again. Results of laboratory tests were normal. Cardiotocographic records of the live twin were normal. Internal examination revealed total effacement of the vaginal portion, dilation 4 cm. Amniotic sac was preserved. In the course of childbirth, secondary decrease in uterine contractile activity and eventually inhibition of the labor progress occurred due to the dead fetus presenting, in a transverse lie. After artificial rupture of the membranes and total breech extraction of the dead fetus, further course of childbirth was uneventful.

The fetus papyraceus born was male, weight 200g, just 1 cm thick, with the placenta weighing 100g (Figure 3). Umbilical cord was 40 cm in length, contained three vessels and was wrapped around the left shank of the fetus (Figure 4). The twin did not have any developmental abnormalities. Internal organs showed development consistent with gestational age.

After an hour, twin 'B' was delivered spontaneously in vaginal birth alive, (female, 2900g, Apgar 10). Five units of oxytocin were administered intravenously immediately, and then a drip with 10 units.

Intranatal blood loss was 250 ml. Macroscopic examination of the afterbirth revealed the presence of DC/DA pregnancy, and histological examination of the placenta revealed the presence of inflammation markers. Maternal blood group was B Rh positive; the blood group of the live fetus was AB Rh positive.

The mother and the child were discharged on the third postpartum day. Up to date the development of the child has been normal.

Discussion

There is a number of factors listed in literature, which may be responsible for the demise of one of the twin fetuses. These include inborn defects, placental insufficiency, IUGR, membranous or velamentous umbilical cord insertion to the placenta, umbilical cord true knot, pinching of the umbilical cord, 'twin to twin transfusion syndrome' between fetuses in monochorionic pregnancy, and in monoamniotic one – kinking and knotting of umbilical cords [5-8]. Also, some injuries in a mother can be the reason [7].

It is believed that in dichorionic pregnancies as well as in monochorionic diamniotic ones, entanglement of the umbilical cord is rarely the direct cause of intrauterine death of one of the twins, but it is the frequent cause in monoamniotic pregnancies. Umbilical cord entanglement can occur as early as in the 9th week of pregnancy i.e. at the moment of the first fetal movements.



Figure 1. Ultrasound image of fetus papyraceus



Figure 2. Fetus papyraceus. FL 39mm = 22.3 hbd.

The described case of a woman in whom this complication occurred in three consecutive pregnancies can suggest its hereditary occurrence.

Entanglement of the fetal umbilical cord may lead both to a partial reduction of blood flow from the placenta to the fetus and chronic hypoxia, with the development of the oligohydramnios and IUGR, as well as the sudden disruption of fetoplacental circulation with subsequent fetal death [10]. The fact that in our case the fetus showed a normal intrauterine growth before death indicates that the fetal demise occurred due to acute umbilical cord entanglement.

The mechanism of umbilical cord becoming entangled and tightened remains unclear. Earlier reports emphasized the importance of the absence of Wharton's jelly [11]. However, this symptom was not present in our case. The length of tightened umbilical cord described in the literature ranged from 34cm to 120 cm (40 cm in our case) [12]. Thus, this feature also does not seem to be significant in the occurrence of this complication.

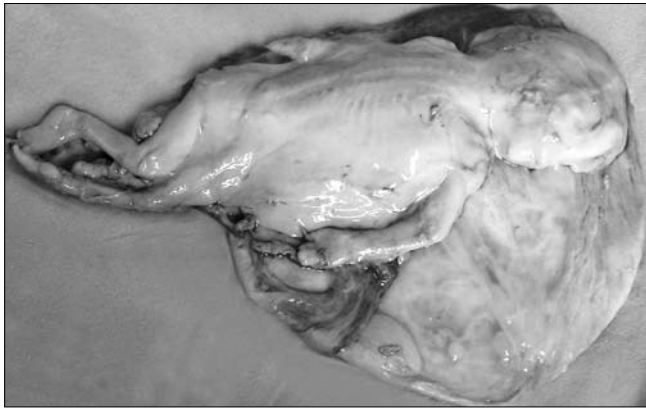
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Figure 3. Fetus papyraceus.

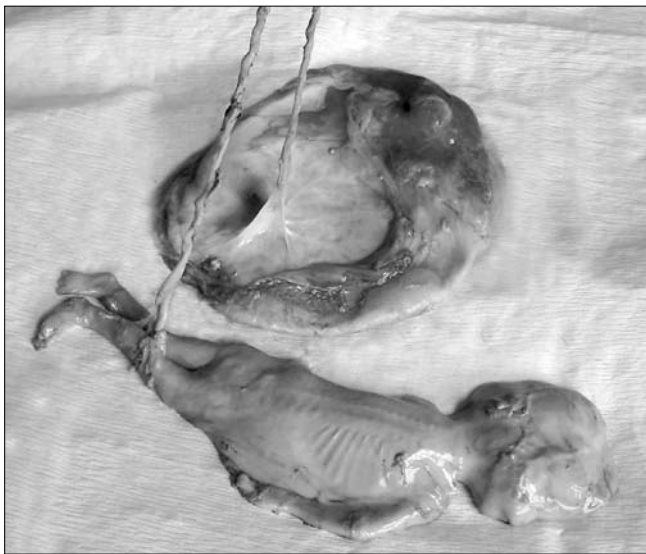


Figure 4. Fetus papyraceus. Umbilical cord wrapped around the left thigh.

Earlier reports suggested that fetus papyraceus occurs more frequently in monochorionic than in dichorionic twin pregnancies. However, according to Benson et al. [14] in DC (12%) and MC (11%) pregnancies the frequency is similar.

Demise of one of the fetuses in the first trimester of pregnancy is a relatively common occurrence. In the DC pregnancies it rarely poses a serious threat to the further course of pregnancy, while in the MC ones it often leads also to death of the second fetus [15]. In contrast, fetal death in the second or third trimester is rare, but it often leads to health and life risks of the surviving fetus and the mother [16]. Complications in mother relate primarily to preterm delivery, intrauterine infection, arrest of labor (in cases where the dead twin is presenting), retained placenta, bleeding during puerperium, and even DIC (extremely rarely) [16, 17]. The prognosis for the surviving fetus to a large extent depends on the type of twin pregnancy. In dichorionic pregnancy chances of survival are much better, and the most frequent complication is prematurity. However, in monochorionic pregnancy the prognosis is poor. Almost 50% twins are born with complications, especially related to the central nervous system [18]. Using Griffiths Mental and Development Scales, Anand et al. [19] found that surviving

co-twins scored significantly lower than singletons. In our case, the twin developed properly.

In case of the death of one of the fetuses in the second trimester of pregnancy, treatment should be conservative – delay delivery until the fetus can survive ex-utero. Due to the possible occurrence of numerous complications, the condition of both the mother and the surviving fetus requires close supervision and specialist obstetric care in the tertiary referral center.

It is necessary to record the intrauterine death of one of the fetuses in the medical documentation, allowing legal protection against the accusation of malpractice and having caused neurological damage to the child during birth.

Conclusions

In case of intrauterine demise of one of the fetuses in DC/DA twin pregnancy, conservative management is preferred. One of the reasons of fetal demise may be entanglement of the umbilical cord around fetal small parts. The time of fetal death can be determined on the basis of length of the thigh bone [Femur Length – FL].

References

1. Landy H, Keith L. The vanishing twin: a review. *Hum Reprod Update*. 1998, 4, 177-183.
2. Bush M, Pernoll M. Multiple pregnancy. In: Current obstetric and gynecologic diagnosis and treatment. Ed. De Cherney A, Nathan L. New York: McGraw-Hill, 2003, 315-325.
3. Leppert P, Wartel L, Lowman R. Fetus papyraceus causing dystocia: inability to detect blighted twin antenatally. *Obstet Gynecol*. 1997, 54, 381-383.
4. Woo H, Sin S, Tang L. Single fetal death in twin pregnancy review of the maternal and neonatal outcomes and management. *Hong Kong Med J*. 2000, 6, 293-300.
5. Akbar M, Ikram M, Talib W, [et al.]. Foetus papyraceus: Demise of one twin in second trimester with successful outcome in second twin at term. *Professional Med J*. 2005, 12, 351-353.
6. Lloveras E, Lecumberri J, Pérez C, [et al.]. A female infant with a 46, XX/48,XY, +8, +10 karyotype in prenatal diagnosis: a 'vanishing twin' phenomenon? *Prenat Diagn*. 2001, 21, 896-897.
7. Peleg D, Ferber A, Orvieto R, [et al.]. Single Intrauterine fetal death (Foetus papyraceus) due to uterine trauma in a twin pregnancy. *Eur J Obstet Gynecol Reprod Biol*. 1998, 80, 175-176-A-16.
8. Reddy K, Petersen M, Antonarakis S, Blakemore K. The vanishing twin: an explanation for discordance between chorionic villus karyotype and fetal phenotype. *Prenat Diagn*. 1991, 11, 679-684.
9. Bakotic B, Boyd T, Poppiti R, Pflueger S. Recurrent umbilical cord torsion leading to fetal death in 3 subsequent pregnancies: a case report and review of the literature. *Arch Pathol Lab Med*. 2000, 124, 1352-1355.
10. Ben-Arie A, Weissman A, Steinberg Y, [et al.]. Oligohydramnios, intrauterine growth retardation and fetal death due to umbilical cord torsion. *Arch Gynecol Obstet*. 1995, 256, 159-161.
11. Hallak M, Pryde P, Qureshi F, [et al.]. Constriction of the umbilical cord leading to fetal death. A report of three cases. *J Reprod Med*. 1994, 39, 561-565.
12. Herman A, Zabow P, Segal M, [et al.]. Extremely large number of twists of the umbilical cord causing torsion and intrauterine fetal death. *Int J Gynecol Obstet*. 1991, 35, 165-167.
13. Szymonowicz W, Preston H, Yu V. The surviving monozygotic twin. *Arch Dis Child*. 1998, 61, 454-458.
14. Benson C, Doublet P, Laks M. Outcome of twin gestations following sonographic demonstration of two heart beats in the first trimester. *Ultrasound Obstet Gynecol*. 1993, 3, 343-353.
15. Malinowski W, Koktyusz R, Stawski P. The case of monochorionic twin gestation complicated by intrauterine demise of one fetus in the first trimester. *Twin Res Hum Genet*. 2005, 8, 262-266.
16. Sahin Z, Sahin H, Surucu R, [et al.]. Intrauterine Death of One Twin: Case Report. *J Turkish German Gynecol Assoc*. 2003, 4, 63-65.
17. Yoshida K, Soma H. Outcome of the surviving cotwin of a foetus papyraceus or of a dead foetus. *Acta Genet Med Gemellol (Roma)*. 1986, 35, 91-98.
18. Pharoah P. Prevalence and pathogenesis of congenital anomalies in cerebral palsy. *Arch Dis Child Fetal Neonatal Ed*. 2007, 92, 489-493.
19. Anand D, Platt M, Pharoah P. Comparative development of surviving co-twins of vanishing twin conceptions, twins and singletons. *Twin Res Hum Genet*. 2007, 10, 210-216.