Conjoined twins in the course of a triplet dichorionic diamniotic pregnancy

Bliźnięta nierozdzielone w ciąży trojaczej dwukosmówkowej, dwuowodniowej

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Słowa kluczowe: bliźnięta nierozdzielone, ciąża trojacza, perinatologia, obumarcie wewnątrzmaciczne.

Abstract

Conjoined twins (CT) is an extremely rare complication in a monoamniotic twin pregnancy, affecting approximately 1 in 80,000 births. In our case report, we present the occurrence of this complication in a patient with a triple dichorionic pregnancy. The complication was diagnosed in the 15th week of pregnancy. The degree of organ connection in CT did not predict the survival of the conjoined foetuses after delivery. The third foetus was morphologically normal and had a separate chorion. The patient did not decide to undergo selective CT foeto-reduction in the second trimester. The natural course resulted in spontaneous intrauterine death of the affected foetuses in the 30th week of pregnancy, followed by spontaneous preterm labour in the 36th week of pregnancy. The description of the case along with photographic documentation allows the diagnosis, procedure, and the spontaneous course of pregnancy with such a complication to be traced. The attached discussion deals with similar descriptions of such rare cases in the literature and proceedings.

Streszczenie

Bliźniaki zrośnięte (CT) to skrajnie rzadkie powikłanie występujące w ciąży bliźniaczej jednoowodniowej, dotykające około 1 na 80 000 ciąż. W niniejszej pracy przedstawiamy przypadek pacjentki w ciąży trojaczej dwukosmówkowej, u której zdiagnozowano to powikłanie w 15. tygodniu ciąży. Stopień połączenia narządów u bliźniaków nierozdzielonych nie rokował ich przeżycia po porodzie. Trzeci płód był morfologicznie prawidłowy i posiadał oddzielny worek owodniowy i kosmówkę. Pacjentka nie zdecydowała się na selektywną terminację płodów nierozdzielonych w drugim trymestrze. Naturalny przebieg skutkował spontaniczną wewnątrzmaciczną śmiercią chorych płodów w 30. tygodniu ciąży, po czym nastąpił spontaniczny poród przedwczesny w 36. tygodniu ciąży. Opis przypadku wraz z dokumentacją fotograficzną pozwala na śledzenie diagnostyki, postępowania i naturalnego przebiegu ciąży z tym rzadkim powikłaniem. W dyskusji omówiono również podobne przypadki opisane w literaturze oraz sposoby postępowania w takiej sytuacji.

Introduction

Conjoined twins (CTs) are an extremely rare complication of a monoamniotic pregnancy. Siamese twins are formed in the situation where the division of a zygote starts after the 13th day of conception. It is estimated that this situation occurs once in every 80,000 births, of which only half results in live birth [1].

The paper presents the case of conjoined foetuses in a triplet pregnancy, which is an extremely rare complication of pregnancy, as well as a narrative review of the literature on the diagnosis, procedure, and natural course of this complication.

Case report

A 23-year-old pregnant woman in her second pregnancy was admitted to the Clinic of Gynaecology and Obstetrics of the Provincial Combined Hospital in Kielce for diagnostic tests in the 15th week of pregnancy due to suspected foetal abnormalities. The first pregnancy proceeded without complications. The patient did not suffer from chronic illnesses, nor was she treated for infertility.

During an ultrasound examination, a triplet dichorionic pregnancy was confirmed, and the presence of 2 CTs with the features of cephalo-thoraco-

omphalopagus in one of the amniotic cavities was detected. Inside the other cavity there was a foetus with normal ultrasound morphology.

The patient was presented with information on the unfavourable prognosis and increased risk of complications that might affect the healthy foetus. The parents were also informed about the possibility of selective CT termination. During the perinatal consultation, the parents did not express an intention to terminate the pregnancy. After performing laboratory tests, the pregnant woman was discharged from hospital in good condition, with the recommendation of undergoing diagnostics in the second trimester of pregnancy.

The patient was again admitted to the hospital at week 23 + 6 of pregnancy to assess the degree of the organ fusion in the foetuses. During the hospitalisation, an magnetic resonance imaging (MRI) examination was performed, which did not yield an unambiguous diagnostic image. An ultrasound examination proved to be more helpful. At week 25 of pregnancy, an ultrasound showed a live, normally developing single foetus, without morphological defects, with estimated foetal weight of 750 g (39.2 percentile) (Figure 1). The placenta was located on the posterior wall. In the other amniotic sac, with an increased volume of amniotic fluid (MVP 86 mm) and a separate placenta located on the lateral and posterior wall, the presence of conjoined foetuses was confirmed.

The foetuses shared cerebral hemispheres and had separate cerebellums. Four lower limbs and four upper limbs were visualised (Figure 2). There were also 2 separate structures corresponding to the hearts, in extreme lateral positions of the fused chest. Both hearts were structurally abnormal. The smaller heart was located anteriorly, probably two-chambered, with one haemodynamically common atrium, one atrioventricular valve, and one haemodynamically active large vessel merging into the arc of the ductus arteriosus. The other, larger heart was located on the opposite side. It had 4 chambers with an abnormal origin of large vessels. Above the defect in the dextroposition, one large vessel merged into the aortic arch. However, the second vessel was not visualised. The lungs, liver, and parts of the intestines also remained fused. The foetuses had separate bladders. It was determined that the degree of fusion did not allow a favourable prognosis of separation and that the defects of the foetuses were lethal.

An interdisciplinary council, consisting of specialists in perinatology, paediatric surgery, and neonatology, decided to implement palliative treatment of the conjoined foetuses after birth. The patient was discharged home.

The patient was hospitalized again at week 30 of pregnancy. During hospitalization, cardiotocographic supervision of the healthy foetus was carried out. On the day after the admission of the pregnant woman, a midwife did not detect audible cardiac activity of the CTs. In the ultrasound examination an intrauterine demise of the CTs was confirmed. Cardiac function and haemodynamic parameters of the non-conjoined foetus remained normal. Obstetric examination revealed an unfavourable cervix.

A decision was made to implement expectant management and observation of the pregnancy, controlling inflammatory parameters, coagulation parameters, and foetal well-being. Nadroparin 40 mg 1 × 1 sc and dexamethasone antenatal steroid treatment in 4 doses of 6 mg were introduced. At week 36 of pregnancy, uterine contractions began. In view of the progress of delivery, and an increase in inflammatory parameters and D-dimers, a decision to complete the pregnancy by caesarean section was made.

Caesarean section was performed in a typical manner. The foetuses were extracted in the following order: female conjoined foetuses with a total weight of 2880 g, in brown amniotic fluid, then the live triplet – live premature son with a body weight of 2275 g, in clear amniotic fluid. The postoperative period proceeded without complications.

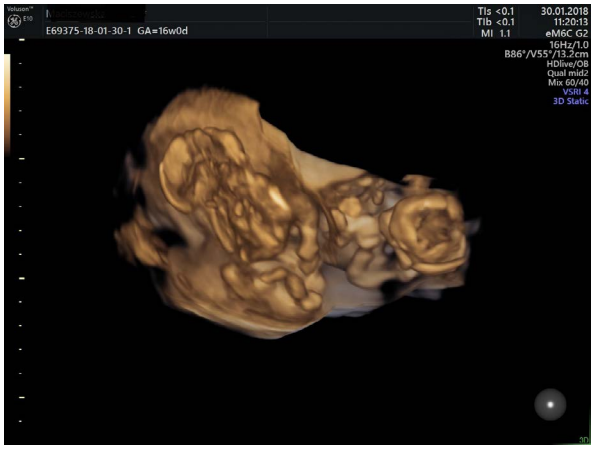

Figure 1. Ultrasonographic 3D reconstruction of conjoined twins and third fetus in 16th week of gestation

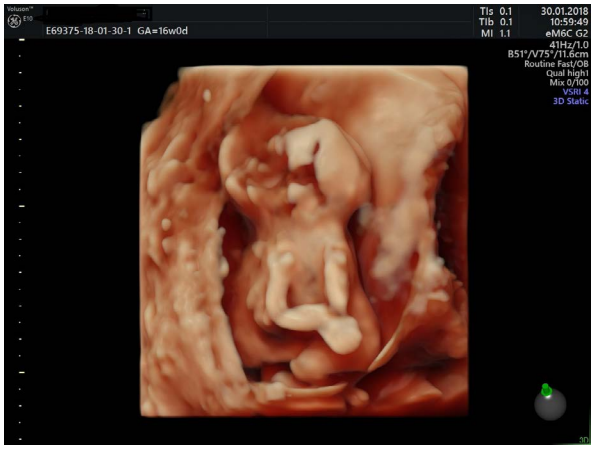

Figure 2. Ultrasonographic 3D reconstruction of conjoined twins

The live newborn was delivered in good condition (Apgar at 1 min 9 points, at 5 min 10 points, parameters of umbilical cord blood gasometry pH = 7.239, BE – 3.1). An umbilical cord blood culture was sampled for microbiological examination. An empirical antibiotic therapy was applied to the newborn in connection with the risk of intrauterine infection, which ended on the third day of the newborn's life in view of the correct result of microbiological examination and the exclusion of the infection features in the child. The neonatal period was complicated by jaundice of the conjunctiva and lacrimal sac and neonatal jaundice associated with premature delivery. On the second day of life, phototherapy of the newborn was introduced. The woman was discharged on the third day after the operation in good condition. The child was discharged on the 10th day of life in good condition with typical recommendations.

The stillborn CTs were ventrally conjoined, with the features of cephalo-thoraco-omphalopagus (from the height of the navel upwards; they had one face). A picture of the newborn was taken (Figures 3 A, B). The parents did not consent to an autopsy.

Discussion

Conjoined twins occur in about 1.5 per 100,000 births [2]. Considering the occurrence rate of triplet pregnancy, estimated at approx. 79 per 100,000 [3], the chance of encountering the case we are describing is extremely low. Therefore, there are no data in the literature suggesting possible models of management in the case of such a complication.

Ultrasonography is the primary diagnostic tool. In the first trimester, in addition to the features characteristic of monochorionic pregnancy (single yolk sac, no separating amniotic membrane, and single placenta) [4], the ultrasound features that should arouse suspicion are as follows: more than 3 vessels in the umbilical cord, hyper-reflexion of the spine, and no relative change in the position of the twins towards each other, especially during consecutive ultrasound assessments [4]. These features refer to a case in which only conjoined foetuses are present in the uterine cavity. In the case described by us, the diagnosis was more difficult due to the presence of an additional foetus in a separate amniotic sac. The second circumstance hindering the diagnosis was its commencement in the second trimester of pregnancy. In such cases, ultrasound examination using three-dimensional (3D) imaging may be particularly useful for diagnosis. Beginning the diagnostics by obtaining a 3D volume from the surface rendering of fused foetuses allows for obtaining initial orientation as to their position towards each other. In the presence of an additional foetus, there is a considerable risk of occurrence of artefacts which can be removed with a virtual scalpel software. Once these have been removed, an additional advantage is the possibility to rotate the figure in any direction and to perform a multiplanar evaluation with the assessment of foetal anatomy at each level of the obtained figure [5]. Such diagnostics can be performed as early as in the first trimester of pregnancy but may be subject to a false-positive error before the amniotic sac expands [6]. Early diagnosis is important due to the possibility of making a decision on the termination of pregnancy. On the other hand, it also furnishes more time for consulting a potential foetal separation surgery and for choosing an appropriate centre in which the patient will give birth. Additional diagnostic indications can be provided by 3D physical models (3DPM) [7] using a three-dimensional printer. Producing such a model may serve as a dummy model during prenatal consultation with parents. It may also be helpful for the neonatal team in planning assisted ventilation.

There are also reports of 3EPM being used prior to separation surgeries. Such models enable visualisation of the spatial and actual correlation between foetal organs and vessels during the qualification and planning of separation surgery [8].

In our case, considering that the third foetus had a separate amniotic sac and placenta, it seems natural to inform the patient about the possibility of selective foeto-reduction due to the CTs' inability to survive. Such a procedure may also prevent complications in later pregnancy affecting the healthy foetus, such as preterm birth resulting from uterine overdistension.

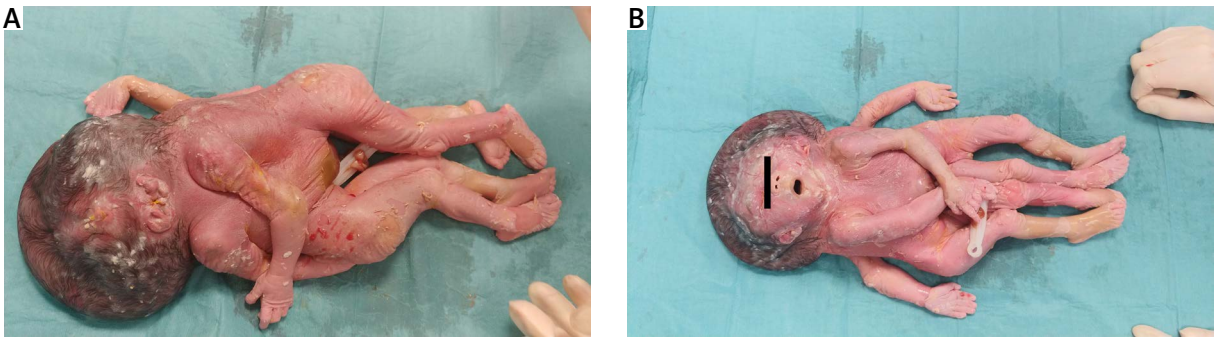

Figure 3. Post mortem photo of conjoined twins

Cases of such a procedure in similar situations have been described in the literature. In a series of cases from 2021, three pregnancies were [9] described in which such a procedure was performed between the 15th and the 17th week of pregnancy. In all cases, pregnancies resulted in the birth of healthy newborns between the 34th and the 37th week of pregnancy. Selective terminations were performed by injecting potassium chloride (KCl) to the CTs in 1 case and by conducting ultrasound-guided intrathoracic injection of KCl in 2 cases [9]. The literature also presents cases with a less favourable outcome, such as premature birth at 26 weeks of gestation after administering an intrathoracic injection of KCl in the first trimester of pregnancy [10]. We also found 3 cases of deaths after selective foeticide in advanced pregnancy. Two cases occurred after microwave ablation of the umbilical cord [11, 12] and one after endoscopic laser occlusion [13]. In our case, the parents decided to continue the pregnancy, which resulted in the spontaneous death of CTs at 30 weeks of pregnancy and favourable outcome with the birth of the late premature baby at week 36 of gestation. The disadvantage of our description is the lack of a pathomorphological report due to the lack of parental consent to perform an autopsy. The newborn was delivered in good health is developing normally.

Acknowledgments

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Conflict of interest

The authors declare no conflict of interest.

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