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CASE REPORT

Cord entanglement in monochorionic monoamniotic twins

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Abstract: Monochorionic monoamniotic twin gestations have been associated with perinatal mortality rates as high as 28 % to 47 %. Umbilical cord entanglements and knots, twin-to-twin transfusion syndrome, congenital anomalies, prematurity and intertwin locking during labor are responsible for their high perinatal morbidity and mortality. We report here two cases of cord entanglements: One of them was associated with twin-to-twin transfusion syndrome with gross vascular anastomoses and a massive cord entanglement. The other one was associated with cesarean section due to dystocia of cord entanglement of the second fetus after vaginal birth of the first one. There is still no consensus in literature for the management and the mode of delivery of these rarely encountered cases *(Fig. 3, Ref. 13)*. Full Text in free PDF *www.bmj.sk.*
Key words: monochorionic monoamniotic twin gestation, umbilical cord entanglement.

Monoamniotic twins have a prevalence of less than 5 % of monozygotic twin gestations and 1 % of all twin pregnancies (1). They occur as a result of ovum division beyond eight days after fertilization and are characterized by a single amnion and single yolk sac. There may be two or one (conjoined twins) embryos present. They are at increased risk of preterm delivery and acute fetal death. Cord entanglement, malformations, twin-to-twin transfusion syndrome are responsible for their high perinatal morbidity and mortality (2). Literature presents perinatal mortality rates of 28 % and 47 % based mostly on case reports and small series.These high rates are suggested to be specifically due to cord accidents secondary to the entwining of the two umbilical cords and/or knotting and thus leading to occlusion (3). Earliness and reliability of diagnosis is essential if attempts to reduce the complication rates are to be made. Most typically, the diagnosis is made by ultrasound, with an inability to distinguish a dividing membrane between the fetuses. There are diagnostic pitfalls (oligohydramnios in one of the twins with closely adherent membrane) and the diagnostic accuracy is central to appropriate pregnancy management (4). Color flow Doppler is useful in the identification of umbilical cord entanglement in monoamniotic twin pregnancies and may provide a method of monitoring the fetuses for the evidence of cord compression (5).

In this study, we present two cases of monoamniotic twin gestations with umbilical cord entanglements.

Case 1

An 18-year-old primigravida was monitored as an outpatient up to 29 weeks when preterm labor occurred. Screening sonography revealed a monoamniotic twin pregnancy (without the division of amniotic membrane) and both fetuses were in cephalic presentation. The ultrasonographic assessment of the first fetus showed hydrops fetalis with massive ascites and fetal biometry was concordant with 29 – 30 weeks. The anatomic look of the second fetus, which developed in accordance with 26 weeks of gestation, was normal. Amniotic fluid was also normal and a single placenta was located on the posterior uterine wall. Twin-to-twin transfusion syndrome was observed and urgent delivery by cesarean section was performed due to the deterioration of heartbeats of the hydropic baby in external cardiotocogram (CTG).

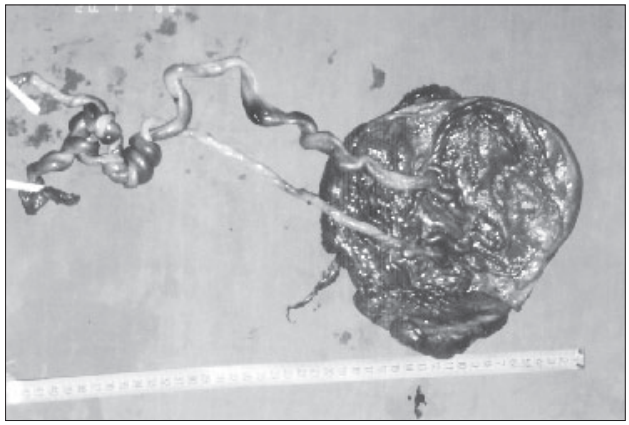

Fig. 1. Cord entanglement in case 1. Note the edema of the right cord belonging to the hydropic fetus.

A hydropic and asphyxiating female baby weighing 1650gr with Apgar scores of 3 and 5 and after that a female baby weighing 920 g with scores of 5 and 7 at one and five minutes, respectively were delivered. Ventilation was aided in both of the babies by a neonatologist. Umbilical cords of the twins had two true knots and 8 entanglements. Edema due to the cord of the

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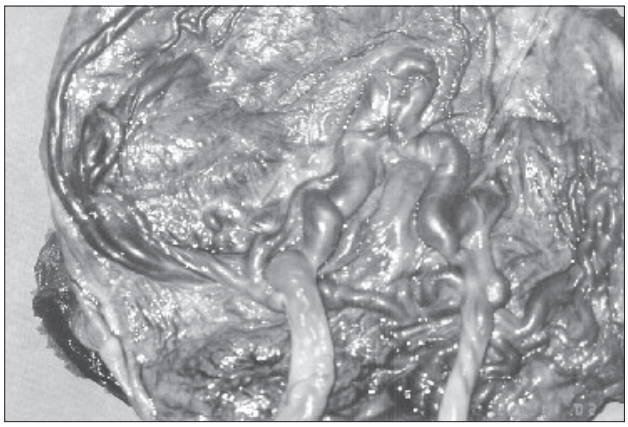

Fig. 2. Vascular anastomoses between the cord vessels.

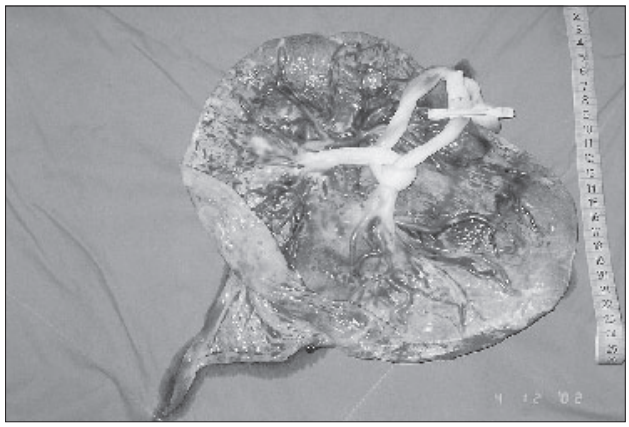

Fig. 3. Cord entanglement in case 2.

hydropic baby was notable (Fig. 1). On placental examination, two vascular gross anastomoses between the umbilical cord vessels were observed (Fig. 2). Pathology confirmed the diagnosis of monochorionic monoamniotic placenta with vascular anastomoses.

Case 2

A 28-year-old primigravida at 37 weeks of gestation by last menstrual period was admitted to our hospital clinic due to twin pregnancy and rupture of membranes. Sonographic evaluation revealed two male fetuses, both in cephalic presentation and well developed in accordance with their gestational age.Since her maternal pelvis was quite suitable it was decided to attempt vaginal delivery with closely monitored labor by means of external CTG.The first baby weighing 2,420 g with Apgar scores of 3 and 7 after one and five minutes, respectively was delivered by low forceps application for the indication of desceleration of heart rate (< 60/min).

However, the second fetus head was arrested at the inlet of pelvis and was not able to descend for about half an hour.Due to the desceleration of heartbeats, it was necessary to perform cesarean operation by way of which a baby weighing 2,650 g with Apgar scores of 3 and 4 after one and five minutes was delivered. Umbilical cords have been strangely entangled with each other (Fig. 3).

Pathologic examination of the placenta confirmed monochorionicity and monoamnionicity.

Discussion

Monochorionic monoamniotic twin gestations derive from a single blastocyst in which the zygotic division takes place for more than eight days after fertilization (6, 7). In these pregnancies, the fetuses are at high risk because of shared vascular areas (monochorionicity) as well as cord accidents and birth traumas due to monoamnionicity.Vascular anastomoses are rare in dichorionic placentas. They generally occur between fetuses and monochorionic placentas and can cause twin-to-twin transfusion, thus developing the risk of intrauterine fetal demise or prematurity as a cause of preterm delivery. Cord entanglements are the main risk of these pregnancies. They develop in very early gestational ages and become a crucial problem in later weeks (8).

In multiple gestations, widespread application of first trimester transvaginal sonography has enabled precise depiction of chorionicity and amnionicity. Sherer et al stated in a case report that the depiction of the branching sign of the umbilical artery by color Doppler imaging in the first trimester sonography was an indication of umbilical cord entanglement (2). On the other hand, Overton and his friends suggested that the branching sign was unlikely to be applicable during the first half of pregnancy when the cord vessels are too small for individual visualization (5). Initially, a prenatal sonographic diagnosis of entangled monoamniotic twins assessed in the second and third trimesters was reported by Belfort et al, who described an apparent branching of the umbilical artery in three such cases (9). Doppler flow velocimetry may reflect hemodynamic alterations in the fetoplacental circulation secondary to the narrowing of the umbilical vessels involved in cord entanglement, such as a ‘notch’ in the umbilical artery waveform (10).

Sherer and his friends presented a case in which prenatal sonography at 26 weeks of gestation depicted a monochorionic diamniotic twin gestation with concordant fetal growth and findings suggestive of a true knot of the umbilical cord (11). Cesarean delivery at 34 weeks of gestation revealed spontaneous antepartum septostomy with entanglement of two seperate umbilical cords. This case suggests that the differential diagnosis of findings considered consistent with a true knot of the umbilical cord in monochorionic diamniotic twin gestations, should include spontaneous antepartum septostomy and umbilical cord entanglement.

Twin-to-twin transfusion syndrome may also occur in these pregnancies, independent from amnionicity. The mechanism of this process remains to be clarified. Fetal survival rate is less than 10 % in case of conservative approach in this condition. In our first case, twin-to-twin transfusion syndrome with gross vascular anastomoses and a massive cord entanglement was seen and at 30 weeks of gestation, fetal distress indicated urgent cesarean delivery.

Clinical management of these pregnancies remains to be clarified. General thoughts have been focused upon the termination of pregnancy via cesarean section as early as at 32 weeks or after lung maturation verified by amniocentesis because of the risks of cord entanglement and birth trauma (12). In our second case, we had to apply cesarean section for the second baby due to its failure to descend after the vaginal birth of the first one. This shows dystocia originating from cord entanglement.

Carr et al described 24 cases of monoamniotic twins with no perinatal mortality after 30 weeks of gestation and suggested that prophylactic preterm delivery at 32 weeks of gestation could not be justified (8). Tessen and Zlatnik reported only one fetal death after 20 or more weeks of gestation in a retrospective controlled study of 21 monoamniotic pregnancies in which a double death occurred at 35 weeks of gestation (13).

In summary, monoamniotic monochorionic twin pregnancies have an increased risk of cord entanglement, twin-to-twin transfusion syndrome, prematurity and congenital malformations as seen in our case reports. In order to define the optimal route of delivery in these pregnancies, we need controlled and prospective studies. The research based on determinations of chorionicity and amnionicity via first trimester sonography would be helpful for planning fetal surveillance and neonatal follow-up.

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