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Case Report

About a case of a specific complication of monoamniotic twin pregnancy: umbilical cord entanglement

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ABSTRACT

Monoamniotic twin pregnancies are uncommon and often complicated by umbilical cord entanglement. It is important to investigate a umbilical cord entanglement during antenatal ultrasound examinations. The diagnosis is based on ultrasound with color and pulsed Doppler. Despite the high percentage of cord entanglement, the perinatal mortality rate remains very low. Rigorous ultrasound monitoring and recording of fetal heart rhythms can improve the prognosis. We report a case of umbilical cord entanglement in a monoamniotic twin pregnancy discovered during a caesarean section with a favorable outcome.

Keywords: Monoamniotic twin pregnancy, Cord entanglement, Perinatal complications

INTRODUCTION

Twin pregnancy is the simultaneous development of two embryos in the uterine cavity. A distinction is made between dizygotic twin pregnancies, which result from the fertilization of two separate oocytes, and monozygotic pregnancies, which result from the fertilization of a single oocyte to give an embryo that secondarily splits. This duplication can occur at different stages of embryogenesis. When it occurs between the 8th and 13th day after fertilization, a monoamniotic pregnancy is obtained. Monoamniotic pregnancies are uncommon and represent 1% of twin pregnancies.[1,2] These pregnancies have several risks: those related to monochorionicity and those specific to monoamnionicity such as conjoined twins and funicular accidents (cord entanglements and knots) which increase the risk of in utero fetal death[3-5] Even without growth restriction or twin-to-twin transfusion syndrome, monochorionic pregnancies have a higher rate of in utero fetal death than other twin pregnancies despite monitoring.[6] For this reason, it is recommended to interrupt this type of pregnancy before 37 weeks.[7] The delivery of monochorionic twins remains controversial. These pregnancies are reported to be a risk of acute intrapartum twin-to-twin transfusion, although evidence regarding its frequency is limited. As for all twin pregnancies, vaginal delivery should be allowed only if there are facilities for fetal monitoring during labor and the use of an emergency cesarean section without a delay. Nevertheless, the preferred mode of delivery for monoamniotic twin pregnancies is cesarean section. Even if the number of cases of cord entanglement is uncommon, this complication remains potentially fatal if the pregnancy exceeds 36 weeks or if vaginal delivery is performed.

We report a case of umbilical cord entanglement in a monoamniotic twin pregnancy discovered during a cesarean section with a favorable outcome.

CASE REPORT

This was a 28-year-old patient with a previous caesarean section two years ago, admitted for the management of twin pregnancy of 38 weeks of amenorrhea. The monoamniotic twin pregnancy was diagnosed at 20 weeks by ultrasound examinations and the fetal monitoring was done by a midwife with 3 antenatal visits. After her admission, a scheduled cesarean section was performed with a female twin A weighted 2465 grams had an Apgar score of 8 and 10. Then a twin B of the same gender, weighed 2950 grams with an Apgar score of 8 and 10. During the surgery, we had fortuitously discovered an entanglement of the umbilical cords (Figure 1A). Examination of the fetal annexes revealed a single placenta (Figure 1B) and a single amniotic sac with cord lengths of 52 and 53 cm respectively.

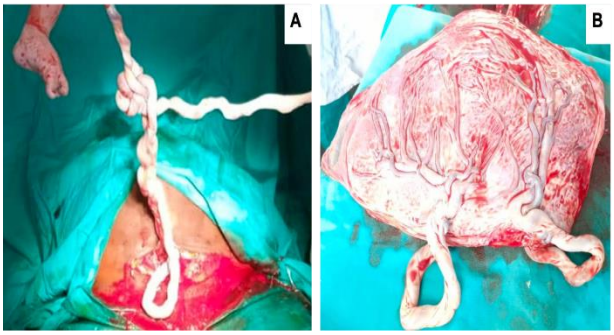

Figure 1: A) Cord entanglement discovered during caesarean section, B) fetal side of the placenta with peripheral vascular anastomoses between the 2 cords.

DISCUSSION

Main finding

We report a case of umbilical cord entanglement in a monochorionic monoamniotic pregnancy with viable fetuses born by caesarean section.

Interpretation of findings

Umbilical cord entanglement is a specific complication of monoamniotic twin pregnancies. Its prevalence is estimated at 71-74%.[8-10] Some authors attest that cord entanglement almost always exists in monoamniotic pregnancies as reported by Dias with a 100% frequency of cord entanglement between 11 and 16 SA. Considering the rarity of monoamniotic pregnancies, cord entanglement remains an uncommon finding. The occurrence of these complex knots could be explained by the amniocity but above all by the movements of the foetus. There is two kinds of movements: passive and active. The first are communicated to the fetus by the influence of maternal movements, and the active movements which are specific of the fetus occur very early in the first months of foetal life.[11]

The prognosis of monochorionic monoamniotic pregnancies is poor with a high-rate perinatal complications.[12] These complications are linked to specific pathologies of monochorionicity such as twin-to-twin transfusion syndrome, malformations, spontaneous or intended prematurity and funicular incidents occurring before or at the time of delivery.[12] However, despite the high percentage of cord entanglement, the perinatal mortality rate remains very low. Indeed, a meta-analysis carried out in Italy with a cohort of 114 monoamniotic twin pregnancies found a mortality rate of 10%. Only two neonatal deaths were due to cord entanglement, the other cases were due to prematurity and major congenital malformations.[13] Moreover, in 90% of cases at least one twin survives and in 84% of cases both survive. Discordant birth weights between the two twins are not uncommon in monoamniotic twin pregnancies. Cord entanglement or vascular anastomoses may contribute to this weight difference. Roqué reports a weight difference in 70% of cases of cord entanglement. In our observation, the birth weight difference between the twins was 17%, in the normal range.

Impact of findings

Although the reported mortality is low, it is still unacceptable considering the possibilities offered by antenatal diagnosis.[13,14] All cases of monochorionic monoamniotic twin pregnancies should be assessed for entanglement. Diagnosis of cord entanglement is based on colour or 3D Doppler ultrasound with umbilical artery velocity. Its positive predictive value is 89%.[14,15] In the first trimester, the visualization of two different arterial flow patterns and different heart rates in the same pulse wave sampling gate is not only indicative of monoamnionicity but also of cord entanglement.[16] In the second and third trimesters, the diagnosis will be made by looking for apparent " ramification " of the umbilical artery on a colored Doppler. In addition, pulsed Doppler examination of the two " ramifications " allows the detection of different heart rates and resistance index.[17] Finally, the diagnosis can be fortuitous at birth, especially when the diagnosis of chorionicity has not been made as was the case in our observation. In conclusion, it is important that the diagnosis of monoamnionicity is made earlier in the pregnancy because the prognosis and appropriate management depend on it.

Once the diagnosis of cord entanglement has been made, the discussion turns to the timing and mode of delivery. Several authors recommend delivery between the 32nd and 34th week of amenorrhoea.[12-18] Although several teams recommend caesarean section as the mode of delivery, cases of vaginal delivery have been reported with a rate of complications and perinatal mortality similar to other types of twin pregnancy.[19,20] These discussions open up avenues of research to determine whether scheduling delivery at a predefined time or choosing a delivery route influences the neonatal prognosis of monoamniotic twin pregnancies with

umbilical cord entanglement. It should be noted, however, that given the rarity of monoamniotic pregnancies, studies with larger sample sizes will remain difficult to implement. In the meantime, the clinical judgement of the practitioner and caution should prevail.

CONCLUSION

Monoamniotic twin pregnancies are uncommon. They are associated with several specific complications such as cord entanglement. This one should be detected during the antenatal period and ideally in the first trimester of pregnancy. It is useful to remind the importance of ultrasound in the first trimester of pregnancy during which the diagnosis of chorionicity and amnionicity is established. Such information is crucial in the monitoring and prognosis of any twin pregnancy.

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