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Etiology and Management of Monoamniotic Twin Discordant for Anencephaly

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Abstract. A case of monoamniotic monochorionic (MAMC) female twins, with one twin being anencephalic, and the co-twin being normal, is reported. Twin to twin transfusion syndrome was also noted. The reasons for anencephaly in association with monozygotic (MZ) twinning are discussed. The management of such a case is presented.

Key words: Monoamniotic, Twin pregnancy, Discordance, Anencephaly

INTRODUCTION

MAMC twin pregnancy is a rare obstetric situation, with an approximate frequency of one in 30,000 births [18]. Management of such a pregnancy is a challenge for the obstetrician because of the very high incidence (60%) of perinatal mortality [23]. The reasons for increased perinatal mortality are cord accidents, higher rates of malformations, twin to twin transfusion and preterm deliveries. We report a rare case of MAMC pregnancy with one baby having anencephaly and other being normal. The phenomenon of discordance in malformations, i.e., one twin normal and the other having malformation, is also discussed. The dilemma faced in the management of such a case is presented.

CASE REPORT

A 31-year-old gravida 2 para 1 with previous one full term vaginal delivery ten years ago, attended antenatal clinic at 11 weeks of gestation. On pelvic examination, size of the uterus was found to be more than period of gestation. Ultrasonography revealed two fetuses in a single amniotic cavity without any intervening membrane.

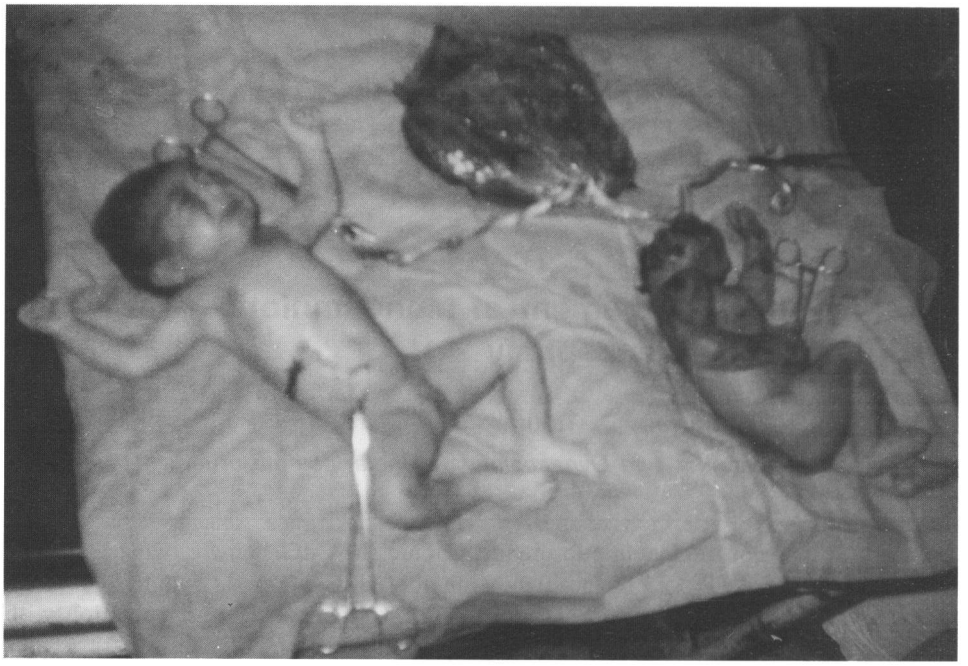

Fig. 1 - Monoamniotic monochorionic twins with one baby having anencephaly. The single placenta is showing insertion of two cords.

Repeated ultrasound done at 13 and 16 weeks of gestation showed two female fetuses lying in a single amniotic cavity. The presenting fetus was normal and the second twin had anencephaly. There were no other malformations noted. The single placenta was lying on the anterior wall of the uterus. She was admitted at 30 weeks of gestation for polyhydramnios. Ultrasonography revealed femur lengths of both fetuses corresponding with 30 weeks. Abdominal circumference (AC) of the normal fetus was 25 cms and 23 cms for anencephalic baby, corresponding to 31 and 28.5 weeks of gestation respectively. The findings were suggestive of twin to twin transfusion. She developed mild pregnancy induced hypertension at 34 weeks of gestation which was controlled on antihypertensive drugs. She was kept under close supervision till 38 weeks when she was induced with oxytocin. Continuous fetal monitoring was done throughout labour. She had spontaneous vaginal delivery after 12 hours of induction. Twin A was female, 2.54 kgs, with Apgar score of 9/10. Her hemoglobin was 17 gm% and hematocrit was 51%. Twin B was anencephalic female, 1.82 kgs, and died soon after birth (Figure 1). Her hemoglobin and hematocrit were 10 gms % and 30% respectively; thus confirming the diagnosis of twin to twin transfusion. Placenta was single without any intervening membrane and had insertion of two cords. It weighed 700 gms. The mother was discharged on second postoperative day with the live and healthy first-born twin.

DISCUSSION

MAMC twins are very rare. Only 1% of twins are MAMC. Once MAMC twin pregnancy is suspected, definite diagnosis of monozygosity needs to be made. Different authors have proposed various ultrasonographic criteria and invasive methods to diagnose monoamniocity. Rodis et al [17] gave the ultrasonographic criteria which had to be fullfilled on 3 different occasions by 3 different observers in order to diagnose MAMC twins i.e. (1) presence of single placenta, (2) absence of inter-ovular membrane, (3) twins of the same sex and (4) both fetuses moving freely within the amniotic cavity. The documentation of two umblical cords arising from two fetuses extending into a common tangled loops of cords also suggests monoamniocity [22]. Other invasive methods like (1) injection of dye into amniotic cavity followed by failure to obtain dye free amniotic fluid [19], (2) injection of air mixed with amniotic fluid to visualise the intervening membrane [20], (3) amniography with either plain X-ray [10] or computerized tomography have also been tried [7]. The management of MAMC twin pregnancy requires strict fetal surveillance throughout pregnancy and labour. The frequency of cord accidents i.e. cord knotting and entanglement leading to intra-uterine fetal demise is most common between 18-26 weeks of gestation [5, 21].The reason for this is the large space available for free fetal movements during this period. It is advisable to perform ultrasonography along with colour doppler at least once every two to three weeks to pick up cord tangling and knotting. On colour doppler, unusual flow velocity waveform pattern is seen in umblical artery in cases of cord entanglement. The systolic to diastolic (S/D) ratio is also very much high and diastolic notching is noted [10]. The suspicion of cord tangling on colour doppler alerts the obstetrician to supervise the pregnancy more closely and to be prepared for premature delivery.

Another dilemma in the management of MAMC pregnancy arises when one of the twins is found to have malformation, as happened in our case. Various hypothesis like ovopathy, aging of oocyte, in-vitro fertilisation, ovulation induction and anomalous X chromosome are linked with MZ twinning and increased rate of malformations. Discordance for anomalies has been reported in numerous pairs of MZ twins. Out of 87 pairs of MZ twins with malformations, discordance was found in 72% in a study by Loevy et al [12]. Discordance for central nervous system malformations was found in 3 of 4 pairs. Discordance of malformations in MZ twins raises provocative questions about the process of embryogenesis. The phenomenon of anomalous X chromosome inactivation has been invoked to explain the observed link between female zygote, MZ twinning and neural tube defect (NTD) [9]. This synthesis draws on two independent observations – one, that non-random X chromosome inactivation creates two separate clusters of cells which predisposes to twinning [4], and, second, that delayed X chromosome inactivation increases the risk of NTDs as the cells are aneuploid for a longer time [8].

Based on the above, we suggest a possible explanation for the observation of discordance of NTDs in MZ twins. It is conceivable that a cluster of cells in the zygote undergoes lyonisation later than others. This would create two separate clusters of cells and predispose to twinning and at the same time leave one of the cell clusters differentially vulnerable to develop NTDs as it would remain aneuploid for a longer time. It has also been suggested that the deleterious effect of anomalous X chromosome inactivation leading to discordance is mediated by environmental factors [15].

In our patient, the presenting twin was normal and the other was anencephalic. Though anencephaly is a lethal malformation, polyhydramnios is frequently encountered in such cases thereby increasing the risk of cord accidents and preterm labour. Such a situation has prompted attempts at selective termination of the affected fetus to prevent polyhydramnios and preterm labour and to save the parents from the psychological trauma of having a malformed child. There are reports in the literature of successful outcome after selective termination of a malformed fetus in diamniotic pregnancies, but it has a limited role in MAMC pregnancy as the injected toxin also harms the normal fetus because of the presence of a single placenta [6]. As the complications linked to the procedure are much more than the risk of carrying out pregnancy with a malformed fetus; the selective feticide is not routinely recommended in MAMC pregnancy.

Another potential source of complications in such pregnancies is twin to twin transfusion, because of the presence of intertwin vascular communication on the surface and within the parenchyma of the placenta. There are conflicting reports in the literature on the incidence of twin to twin transfusion in MAMC pregnancy. Few studies have reported it to be very rare [13, 24]. Others have reported the incidence to be high [5, 21]. However, the complication of twin to twin transfusion appears to be less frequent in MAMC as compared to diamniotic twins. The proposed hypothesis is the absence of deep arteriovenous anastomosis between the circulations of the fetuses of MAMC twins [16]. It has also been proposed that the presence of two sacs in a diamniotic pregnancy leads to a difference in intra-amniotic pressure that may influence feto-fetal transfusion differentially. This may not be the case in MAMC twins, as the single cavity may serve as a buffer for intertwin difference [2].

Mode of delivery in these cases is also a controversial issue. Cesarean section is preferred by many authors [14, 17]. However, where the presenting fetus is normal and the aftercoming twin has the lethal malformation (as happened in our case), vaginal delivery can be safely given. This is because the risk to the aftercoming twin due to cord prolapse is not relevant in such a situation. Other selected cases where vaginal delivery can be given are those where both the fetuses have normal growth, and umbilical doppler does not show any signs of cord compression, and both the fetuses are in cephalic presentation. It is prudent to avoid early amniotomy to prevent cord accidents, the major cause of mortality in such pregnancies. It is also very important to identify the exact origin of the cord before clamping and cutting, especially when nuchal cord is present. Accidental cutting of the cord of a second twin after the delivery of the first twin has been reported in the literature [14].

In conclusion, owing to the rarity of MAMC twin pregnancy, more studies are required to definitively outline the management of MAMC twins.

REFERENCES

1. Annan B, Hutson RC (1990): Double survival despite cord entwinement in monoamniotic twins. Case report. Br J Obstet Gynecol 97: 950-951.
2. Baldwin VJ (1994): The pathology of monochorionic monozygosity. In Baldwin VJ (eds): "Pathology of multiple pregnancy". Springer- Verlag New York 199-213.
3. Belfort MA, Moise KJ, Kirshon B (1993): The use of color flow doppler ultrasonography to diagnose umbilical cord entwinement in monoamniotic twin gestation. Am J Obstet Gynecol 168: 601-604.

4. Burn J, Povey S, Boyd Y, Munro EA, West L, Happer K, Thomas D (1986): Duchenne muscular dystrophy in one of monozygotic twin girls. J Med Genet 23: 494-500.
5. Carr SR, Aronson MP, Coustan DR (1990): Survival rates of monoamniotic twins do not decrease after 30 weeks gestation. Am J Obstet Gynecol 163: 719-722.
6. Evans MI, Goldberg JD, Dommergues M, Wapher RJ, Lynch L, Dock BS, Horenstein J, Golbus MS, Rodeck CH, Dumez Y, Holzgreve W, Timor-Tritsch H, Johnson MP, Isada NB, Monteagudo A, Berkowitz RL (1994): Efficacy of second trimester selective termination for fetal abnormalities: International collaborative experience among the world's largest centers. Am J Obstet Gynecol 171: 90-94.
7. Finberg HJ, Clewell WH (1991): Definitive prenatal diagnosis of monoamniotic twins: swallowed amniotic contrast agent detected in both twins on sonographically selected CT images. J Ultrasound Med 10: 513-516.
8. Hall JG (1986): Neural tube defects, sex ratios, and X inactivation. Lancet ii: 1334-1335.
9. James WH (1988):Anomalous X chromosome inactivation: the link between female zygote, monozygotic twinning, and neural tube defects? J Med Genet 25: 213-216.
10. Kofinas AD, Penry M, Hatjis CG (1991): Umblical vessel flow velocity waveforms in cord entanglement in a monoamniotic multiple gestation: a case report. J Reprod Med 36: 314.
11. Lavery JP, Gadwood KA (1990): Amniography for confirming the diagnosis of monoamniotic twinning: a case report. J Reprod Med 35: 911.
12. Loevy HT, Miller M, Rosenthal IM (1985): Discordant monozygotic twins with trisomy 13. Acta Genet Med Gemellol 34: 185-188.
13. Lumme RH, Saarikoski SV (1986): Monoamniotic twin pregnancy. Acta Genet Med Gemellol 35: 99-105.
14. Mac Leod FN, Mac Coy DR (1981): Monoamniotic twins with an unusual cord complication. Case report. Br J Obstet Gynecol 88: 774-775.
15. Osathanondh R, Driscoll SG, Naftolin F (1974): Discordant severe cranial defects in monozygous twins. Am J Obstet Gynecol 122: 301-305.
16. Pessonier A, Ko-Kivok-Yun P, Fournie A (1994): Grossesses gémellaires monochoriales monoamniotiques. Problèmes diagnostiques et risques évolutifs. J Gynecol Obstet Biol Reprod 23: 299-302.
17. Rodis JF, Vintzileos AM, Campbell WA (1987): Antenatal diagnosis and management of monoamniotic twins. Am J Obstet Gynecol 157: 1255-1257.
18. Salerno LJ (1959): Monoamniotic twinning: a survey of the American literature since 1935 with a report of four new cases. Obstet Gynecol 14: 205-213.
19. Sutter J, Arab H, Manning FA (1986): Monoamniotic twins: antenatal diagnosis and management. Am J Obstet Gynecol 155: 836-837.
20. Tabsh K (1990): Genetic amniocentesis in multiple gestation: a new technique to diagnose monoamniotic twins. Obstet Gynecol 75: 296-298.
21. Tessen JA, Zlatnik FJ (1991): Monoamniotic twins: a retrospective controlled study. Obstet Gynecol 77: 832-834.
22. Townsend RR, Filly RA (1988): Sonography of nonconjoined monoamniotic twin pregnancies 7: 655.
23. Wensinger JA, Daly RF (1962): Monoamniotic twins. Am J Obstet Gynecol 83: 1254-1256.
24. Wharton B, Edwards JH, Cameron AH (1968): Monoamniotic twins. J Obstet Gynecol Br Cwlth 75: 158-163.

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