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MONOCHORIONIC MONOAMNIOTIC TWIN PREGNANCY WITH EVANESCENCE OF A TWIN: A CASE REPORT AND LITERATURE REVIEW

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INTRODUCTION

A monochorionic monoamniotic twin pregnancy (MCMA) is the development of two fetuses within the same amniotic sac. This is the rarest type of twin pregnancy, representing about 1% of monozygotic twin pregnancies. This pregnancy results from a late division between the 8th and 13th day after fertilization. The frequency is rare as it is less than 1% of monozygotic pregnancies. This pregnancy is burdened with a very high morbidity either during pregnancy (death in utero, cord entanglement, prematurity) or at delivery. The morbidity of this pregnancy poses the problem of its ultrasound recognition given the rarity, the type of follow-up (frequency, ambulatory, hospitalization) and the optimal term of birth in order to avoid in utero deaths related to funicular causes and finally the mode of delivery.[1,4]

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We report in this work a case of monochorionic monoamniotic twin pregnancy with evanescence of a twin, through which and in the light of a review of the literature we insist on all the characteristics of this entity in particular the diagnosis, the means of monitoring and the modalities of therapeutic management.

Observation

Mrs. Z.R, 37 years old, G2P2: 1 delivery by cesarean section, 1 vaginal delivery, with no notable medical history, was referred to our hospital for the follow-up of a monochorionic monoamniotic pregnancy with evanescence of a twin in the first trimester with a live twin at 18 weeks of amenorrhea; her initial check-up was without particularities.

An ultrasound at 20 weeks of amenorrhea did not reveal any notable morphological abnormalities with a biometry corresponding to the gestational age and a cervical length of 35 mm, umbilical +cerebral Doppler and canal of Arantus without anomalies in the surviving fetus with degeneration of the second twin (Fig.1).

An Intensified follow-up of the surviving fetus by bi-monthly obstetrical ultrasound which showed good fetal growth kinetics, amniotic fluid quantity and fetal Doppler which remained normal throughout the pregnancy.

Biweekly recordings of the fetal heart rate from 30 days of age showed normal oscillating and reactive patterns without slowing down.

Maternal clinical and biological monitoring was without abnormalities.
Hospitalization was considered at 34 weeks of amenorrhea with administration of lung maturation.

A caesarean section was scheduled at 36 weeks of amenorrhea which allowed the extraction of a live tonic

reactive male newborn weighing 3050 g and an evanescent twin (Fig. 2-3).

The postoperative follow-up was simple.

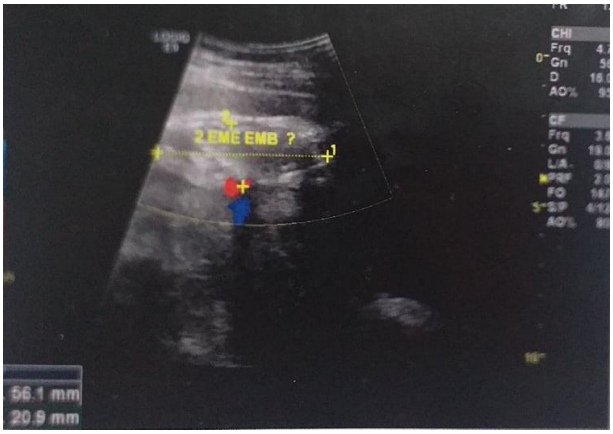

Fig. 1: Ultrasound image showing evanescence of a twin in the first trimester.

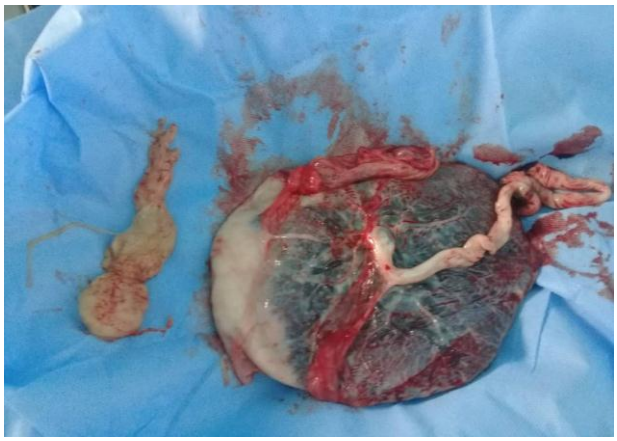

Fig. 2: Monochorionic monoamniotic placenta with evanescent twin.

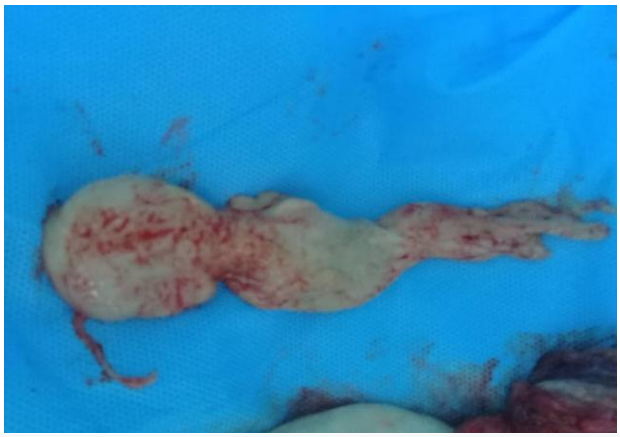

Fig. 3: Picture showing the evanescent twin.

DISCUSSION

The diagnosis of monochorionic monoamniotic twin pregnancy (MCMA) must be made in the first trimester of pregnancy in order to define the management to be adopted according to the type of twin pregnancy.

The optimal time for this diagnosis is between 11 and 14 weeks' gestation.[5]

The diagnosis of monoamniotic pregnancy is based on specific criteria:

- The presence of a single placenta,
- The absence of an inter amniotic membrane and therefore of the lambda sign,
- A close insertion of the two umbilical cords with sometimes visualization of an entanglement.[6,7]

Diagnosis by the number of vitelline vesicles has been questioned since the discovery of postnatal confirmed MCMA twin pregnancy with visualization of two yolk bladders on first trimester ultrasound.[8,9]

The surveillance of MCMA twin pregnancies is based on

- The study of External fetal heart monitoring

The gold standard of intensified surveillance of MCMA GGs is the performance of FHR recordings. The purpose of the FHR study is to detect fetal distress and thus prevent IUGR.

- Antepartum ultrasound

Ultrasound monitoring includes:

- The search for signs of anastomotic syndromes,
- Assessment of fetal growth, amniotic fluid quantity, placental status, cord appearance, fetal well-being (Manning score)
- The study of umbilical arterial Dopplers as well as the velocity of the middle cerebral artery.

Cord entanglement can be visualized by 2D ultrasound with color Doppler or 3D ultrasound, which allows better visualization in space.

This type of twin pregnancy (MCMA) exposes to maternal-fetal complications

Indeed, maternal-fetal morbidity can be divided into three groups according to the complications. We distinguish complications related to:

- Multiple pregnancy such as preeclampsia, threat of premature delivery, cholestasis, thrombosis, delivery hemorrhage for the maternal side and intrauterine growth retardation, weight discrepancy between twins, malformations, prematurity for the fetal side. A weight discrepancy greater than or equal to 20% increases early neonatal morbidity in newborns.
- Monochorionicity, namely anastomotic syndromes (TOPS and TAPS), malformations and congenital anomalies.
- Monoamnionicity, represented by malformations and IUFD due to cord anomalies, entanglements or knots.[10,11]

The occurrence of in utero fetal death, the most serious complication as in our case, in one of the twins of a monochorionic pregnancy is associated with a significant

increase in morbidity and mortality of the surviving twin. Older series have reported a risk of co-twin mortality of 10% to 26% and risks of neurological complications by brain damage of 26% to 51%.[12,13] A recent meta-analysis of 19 studies and 904 twin pregnancies estimated the risk of fetal death of the surviving twin at 12%. When comparison was possible in these studies, it was estimated that the risk of in utero fetal death in MC pregnancies after 20 SA was 6 times greater than in BC pregnancies.[14] It would also seem that the gestational age of onset of Intrauterine fetal death (IUFD) and the time between the IUFD and the birth of the surviving twin are the elements that have an impact on the risk of cerebral sequelae of the co-twin. For Nicolini, when death occurs in the 1st trimester, 92% of survivors are lesion-free. In contrast, when death occurs in the 2nd and 3rd trimesters, only 60% of surviving fetuses are unharmed. Nicolini also showed that in surviving fetuses without sequelae, the time interval between IUFD and birth was longer.[15] It is the anastomoses present on the chorionic plate of MC pregnancies that are responsible for the mortality and morbidity of the surviving twin. These anastomoses are characteristic of MC pregnancies and are almost always present on pathological examination.[16] It is the interdependence between the 2 circulations of the twins, via the anastomoses, that is at the origin of the disruptive vascular lesions observed.

Two theories to explain these lesions on the surviving twin have been proposed. The first is the theory related to an embolic phenomenon, because the first lesions described were multifocal, affecting the brain with multicystic leukomalacia, porencephaly, hydranencephaly, but also the digestive system with intestinal atresia, the kidneys with diffuse cortical necrosis and the skin. This theory is based on the hypothesis of the release of tissue thromboplastin from the dead twin to the surviving twin via the placental anastomoses leading to vascular occlusions and DIC.[17-15] This theory could never be proven by fetal/neonatal or pathological investigations and relies entirely on the finding of necrosis and infarction in the fetal brain and kidneys postmortem. In addition, the coagulation profile of surviving fetuses after fetal cojumatic death has been found to be normal on fetal blood draws.[19-21] The other theory is the hemodynamic theory which emphasizes the sudden hemodynamic variations after the IUFD of a MC twin. Indeed, several studies highlight that the morbidity of the surviving twin could be due to an acute and brutal hemodynamic decompensation just before or at the time of the IUFD of the co-twin when its arterial pressure drops, leading to an exsanguination of the living fetus towards the dead fetus through the placental anastomoses. This results in a profound hemodynamic change with hypovolemia and anemia in the survivor that may lead to death or ischemic injury. This abrupt phenomenon is probably more pronounced in MC pregnancies complicated by transfusion-transfusion syndrome and depends on the number and size of anastomoses present. Several authors have demonstrated the validity of this concept by reporting deep anemia of the surviving twin by performing fetal blood puncture within 24 hours of the co-twin's death,[19,20,22-25] Moreover, this exsanguination has been documented twice in vivo using color and pulsed Doppler.[26,27]

The management of an IUFD in a monochorionic pregnancy varies with the assumed length of the interval between death and its ascertainment. If anemia is suspected by measurement of peak systolic velocity in the middle cerebral artery within 24 hours[28] after the IUFD of a MC twin pregnancy, some teams propose to perform an in utero blood transfusion in order to avoid profound hypovolemia and thus to treat the fetal anemia causing morbimortality.[22,24, 25] This attitude could decrease the mortality of the cochlea but has not shown any benefit on the risk of developing brain damage in the survivors.[25] If the diagnosis is made more than 24 hours after the accident, it is accepted that the survivor should not be extracted because the lesions classically appear within three weeks to one month after the accident occurred. The fine and unusual semiology of these lesions requires ultrasound monitoring by abdominal and endovaginal means by experienced teams (germinolysis cysts, polymicrogyria, intracranial hemorrhage, delayed gyration and periventricular leukomalacia). This follow-up should be completed by a brain MRI ideally performed around 32 weeks after the twin's IUFD but the date will depend on the term of death.

CONCLUSION

Monochorionic monoamniotic twin pregnancy remain a rare entity, however, they are high-risk pregnancies requiring close monitoring; ultrasound and External fetal heart monitoring; in order to prevent and detect complications early, especially the death of a twin which is the most feared complication and the date of its occurrence must be specified in order to prevent the prognosis of the second twin.

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