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ACARDIAC HEADLESS TWIN: CASE REPORT AND LITERATURE REVIEW

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ABSTRACT: The fetus acardiac headless is a rare anomaly characteristic of twin monochromic pregnancies. Its frequency is 1/35000 births. Its evolution is constantly lethal for the affected fetus. The healthy fetus is exposed to a high risk of death in utero or complications. Among the different pathophysiological theories proposed, only the vascular theory is retained. Arterio - arterial and venous anastomoses are the source of vascularization against the current TRAP syndrome (Twin Reversed Arterial Perfusion disorder) of the abnormal fetus which receives the desaturated blood of oxygen and low in nutrients from healthy fetus.

KEYWORDS: acardiac headless, twin, fetus.

1 INTRODUCTION

The monochorionic twin pregnancies are characterized by the presence of vascular connections between the twins. These can be at the origin of pathologies as "Twin Reversed Arterial Perfusion Syndrome" which is defined as the combination of an acardiac, headless and a healthy twin. [1] This syndrome is very rare and is associated with high mortality of healthy twin anemia and heart failure. Antenatal diagnosis is possible for the establishment of appropriate monitoring and therapeutic means to interrupt the vascular anastomoses between the twins in order to achieve a selective feticide. Obstetric management varies between abstentionism and interventionist attitude that depends on the healthy twin prognosis dominated by prematurity and heart failure. We report a case of twin acardiac headless diagnosed during cesarean section which was done to the patient who was poorly followed during her pregnancy.

2 CASE PRESENTATION

Here we present a 25 years old female patient, gravida 1, Para 1, without medical or surgical history admitted in our department in labor. The patient was not followed during her pregnancy. She was transferred from a health center for better management of an obstructed labor.

On admission, Blood pressure was 120/70 mm Hg, pulse = 84 bpm, Tº = 37.3^{o}C, weight = 69kg, height = 162 cm.

On obstetrical examination fundal height was = 32cm, Uterine Contractions present, fetal heart beat present with severe bradycardia for the first twin, the other twin there was no cardiac beat. On vaginal examination, the patient was at full dilation, but the presentation was not engaged.

The ultrasound was not done because there was a technical problem.

The cesarean section was indicated in emergency and the patient was quickly sent in operating room.

The cesarean section was performed without complications, and the first born was extracted by cephalic extraction, and was in posterior variety, with a tight circular cord APGAR 8-10-10 birth weight 3360 grams. The second one, was a abnormal

female fetus, without the head, 2 small legs (Fig1, Fig2), a small chest, two drafts of the upper limbs, a right side blank and an umbilical cord unrelated to the placenta in contrast to that of the healthy twin that fits the placenta .We note the presence of anastomoses with the vessels of malformed twin.

We note the presence of anastomoses with the vessels of malformed twin. Microscopic examination of the membranes of the cord and the placenta reveals no particular anomaly. Pathological examination of the fetus malformed confirmed the diagnosis of twin acardiac and headless weighing 830 grams measuring 16cm. The two legs each have a foot of 5cm.

Fig. 1. Acardiac and headless twin (anterior view)

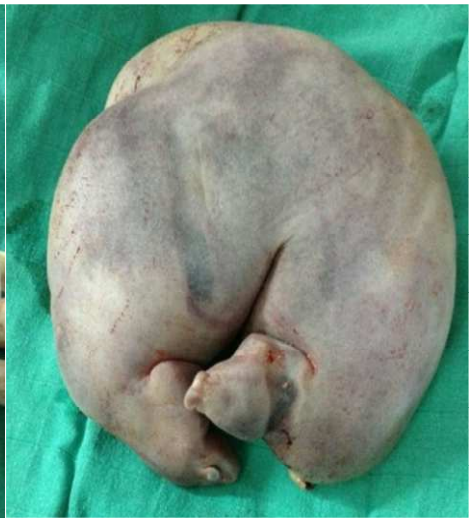

Fig 2: Posterior view

3 DISCUSSION

The acardiac headless twin is a very rare malformation which is found in monozygotic mono-amniotic pregnancies rarely bi-amniotics where acardiac twin is not vascularized by the placenta but by his twin through an umbilical artery against the current [2]. This phenomenon is called TRAP syndrome (twin reverse arterial perfusion) which is a special form of TTS syndrome where the normal fetus as donors giving blood for himself and for the recipient who is acardiac twin. Venous return is via the umbilical vein of normal twin [1-2]. Deoxygenated blood from the donor pass directly with low pressure into the twin acardiac instead of returning to the placenta [2] .This is the cause of the interruption of organogenesis, which generating abnormal cephalic pole members and abdominal organs. The incomplete morphogenesis including the fetal circulatory system would be explained by the reversal of blood flow [3] .The infusion of acardiac fetus is against the current by desaturated blood oxygen at low pressure from an umbilical artery more often single (50%). The incidence of this syndrome is estimated at 1/35000 births and affects approximately 1% of monozygotic twin pregnancies. [4] An abnormal karyotype was found in 50% of cases that could not be achieved in our observation.

The diagnosis is most often posed by prenatal ultrasound found that a twin pregnancy with severe malformations of the twins .The Doppler can show the level of the umbilical artery of twin acardiac a stream with an abnormally high resistance . In contrast, pregnancy outcome seems to be better correlated with the difference in resistance indices between the two twins.

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The diagnosis should be retained in certain abnormalities but not specific: anencephaly, the absence of facial bones, holoprosencephaly, anophthalmia, the diaphragm atresia, the absence of limbs or rudimentary limbs, the absence of chest and acardiac the esophageal atresia, absence of liver and gallbladder, single umbilical artery and skin edema .While in the twin pump can be signs of heart failure in type of ascites, pleural effusion, skin edema or hydramnios [1-8].

The differential diagnosis is fetal death in-utero of a normal twin, anencephalic or associated with a large cystic hygroma, a placental tumor or intra-amniotic and a bi-amniotic pregnancy by treating the skin of an acardiac as in hydrops inter-amniotic membrane. The definitive diagnosis must therefore appeal to the color and pulsed Doppler showing retrograde perfusion of acardiac twin.

To ensure the infusion of acardiac fetus, its healthy co-twin provides significant cardiac stress with increased risk of premature delivery, intrauterine growth restriction, cardiomegaly, heart failure or even hydrops fetalis. The mortality rate the healthy twin varies between 50% and 75% explained by the significant effort required to myocardial perfusion two fetuses [5]. The prognosis of healthy twin depends on the pulsatility index in umbilical artery infusing the fetus acardiac and size. If acardiac twin weighs less than 25 % of the weight of the donor, evolution is often favorable as is the case of our publication.

The treatments which is proposed is to interrupt the vascular communications between the two twins, selective feticide, endoscopic laser coagulation ultrasound guided , selective reduction of the twin acardiac radiofrequency at the umbilical cord or coagulation at the clamp the umbilical cord acardiac fetus [6-7] .

If acardiac twin is small, an ultrasound monitoring is necessary in search of complications in healthy twin .Otherwise, if it is big or it quickly grows, invasive treatment is required by coagulation or by interruption radiofrequency the abdominal aorta or its acardiac pelvic vessels.

4 CONCLUSION

The twin acardiac headless is an extremely rare and special shape of the to-twin transfusion syndrome complicating monochorionic twin pregnancy. Prenatal diagnosis is based on ultrasound coupled with color and pulsed Doppler. The mortality rate of the healthy twin by heart failure remains high and hence the interest of a close monitolling. The treatments offered so far are intended to interrupt the vascular communications between the two co-twins to achieve selective feticide.

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