Abstract

Mono-amniotic mono-chorionic pregnancies are uncommon among twin gestations. Further twin reversed arterial perfusion (TRAP) sequence is an extremely rare presentation. Acardiac acephalic fetus resulting as a result of TRAP sequence has an estimated incidence of 1 in 35,000 pregnancies. To the best of our knowledge, no such case has been reported till date which presented as obstructed labour. We are highlighting this case so as to alert the managing team of obstetricians regarding this rare cause of obstructed labour. Also to increase awareness regarding this presentation presenting in advanced second stage of labour with fetal malpresentation. Only diagnostic pointer in index case was malformed fetal toes visible at introitus of mother in advanced second stage of labour.

Introduction

Acardiac acephalic fetus is a very rare complication of monozygotic twin pregnancy. The estimated incidence of this complication is 1 in 35,000 pregnancies [1]. It results from abnormal placental vascular anastomosis [2]. Usually this condition is diagnosed on routine ultrasonographic examination. We report this rare condition in a tribal woman with no previous antenatal examination presenting as obstructed labour.

Case Report

A 24 year old woman gravida [2], para [1], at 37 completed week's gestation came to emergency room in labour. She was referred as a case of obstructed labour. She did not have any antenatal check-up or ultrasonic examination prior to this visit. Her previous delivery was an uncomplicated normal vaginal delivery. On examination she was dehydrated and exhausted. Her breath had acetone like smell. On abdominal examination uterus appeared to be over distended, tense and tender. Fetal heart could not be localized. Vulva was oedematous and swollen. Foot of the fetus was seen protruding out of the vulva (Figure 1).

However, this foot appeared deformed as only three toes were visible. On vaginal examination, cervix was fully dilated and effaced. An attempt was made to deliver the fetus vaginally but it leads to peeling of the fetal skin at the knees. With clinical diagnosis of obstructed labour an emergency sonogram was requested which revealed twin pregnancy with intra-uterine fetal death of both the twins. No inter twin membrane appeared to be over distended, tense and tender. Fetal heart could not be localized. Vulva was oedematous and swollen. Foot of the fetus was seen protruding out of the vulva (Figure 1).
had one toe whereas left foot had three digits. The lower spinal column and lower limb bones were apparently normal. The weight of the two foetuses was 2100 g (donor twin) and 4500 g (recipient twin) respectively; placenta was single and weighed 800 g. A diagnosis of monoamniotic monochorionic twin pregnancy with acardiac twin was made. Subsequently, both the fetuses and placenta were sent for autopsy, which confirmed the diagnosis of monoamniotic monochorionic twin pregnancy with acardiac accephalus fetus with twin reversed arterial perfusion (TRAP) sequence. Head, heart, lungs and upper limbs were absent in the acardiac twin. The soft globular mass (appearing as head) was actually edematous liver with multiple fluid filled cavities. Subsequently, mother had an uneventful post-operative period (Figure 2).

Figure 2: Acardiac acephalic twin along with donor twin seen (both still birth).

Discussion

Acardiac accephalus twin pregnancy is one of the rarest and unique complications of monozygotic twin pregnancy [1]. Four types of acardiac fetuses have been described in the literature [3].

- acardiac-acephalus (no cephalic development)
- acardiac-anceps (some cranial structures with or without neural tissue)
- acardiac-acormus (cephalic structures with limited or no truncal development)
- acardiac-amorphus (no cephalic or truncal differentiation)

The index case was acardiac accephalus. The TRAP sequence seen in the index case chiefly results from abnormal placental vascular anastomoses with consequent increase in arterial pressure in one twin leading to reversal of blood flow in the other twin. As a result, the “donor” twin pumps deoxygenated blood into the “recipient” (acardiac) twin [1]. A variety of anomalies can occur due to severe hypoxemia, which in the index case was acardiac accephalus twin.

Widespread use of sonography has resulted in early diagnosis of this condition even in first trimester. However, in the index case woman did not have any ante-natal ultrasonographic examination and presented in second stage as obstructed labour. Due to malformed externally visible foot (with only three toes; as shown in Figure 1), a high index of suspicion of some major malformation was suspected. Emergency ultrasound could detect only one fetal head, so a possibility of accephalic fetus was suspected. To the best of our knowledge, this is the first case report in literature where acardiac accephalus monochorionic pregnancy presented in second stage as obstructed labour.

This case is being presented to make the managing obstetricians aware regarding this extremely rare presentation presenting as obstructed labour. A high index of suspicion and timely intervention in terms of emergency caesarean section was detrimental in saving life of the mother.

Conflict of Interest

There is no conflict of interest with any individual or organization.

References