Case Report

Acardiac Anceps Twin: A Case Report

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ABSTRACT

Acardiac twin is an anomaly, which rarely occurs in monochorionic twin pregnancies. The normal twin is also called as pump twin as it pumps blood to the acardiac twin. Acardiac twin live like a parasite with normal twin. In acardiac twin, heart does not develop properly or presents with rudimentary heart, due to which other upper structures of the body do not develop. This often results from placental vascular anastomoses. As heart does not develop, survival of acardiac twin is impossible. A 26-year-old woman referred as a case of uterine inversion after delivery of stillborn female fetus at home and placenta at some local hospital. The mass coming out of vagina turned out to be a acardiac twin with poorly developed head with few hairs, unformed trunk, poorly developed both lower limbs and centrally attached umbilical cord. As woman was unbooked and uninvestigated, the diagnosis of acardiac twin was made after delivery of the fetus.

Keywords: Monochorionic twin, Acardiac twin, Pump twin.

INTRODUCTION

In recent study, it was estimated that in monochorionic twin pregnancies incidence of acardiac twin is 2.6%. [1]Twin reversed arterial perfusion (TRAP) is the most severe form of twin-to-twin transfusion syndrome. The normal twin is also called as pump twin because it pumps blood to the acardiac twin. Acardiac twin live like a parasite with normal twin, in which heart does not develop properly or presents with rudimentary heart and due to which other upper structures of the body also not develop. This often results from placental vascular anastomoses. As heart is not developed, survival of acardiac twin is impossible. Blood perfused to the acardiac twin from the pump twin is apparently deoxygenated. Acardiac twin and normal twin grow together, but due to inadequate supply of oxygen to acardiac twin, necessary structures or organ of life does not develop and acardiac twin therefore presents with multiple structural and developmental deformities. In acardiac twin, if heart does not develop at all, it is called as holoacardia, and if some rudimentary cardiac tissue develops then called as pseudocardia.

Acardiac twins can be classified on the basis of degree of malformation:[2,3]

- Acardiac acephalus: This is the most common variety, in which head and upper extremities are not well developed.
- Acardiac anceps: This is the most developed variety. In this type some cranial structures and neural tissues are present. Trunk with extremities are also developed.
- Acardia acormus: It is the rarest form in which cephalic structures present but truncal structures are not developed and umbilical cord is attached to the head.
- Acardiac amorphous: It is the least developed form in which cephalic or truncal structure are not distinguishable.

CASE REPORT

A 26-year-old female P4L3 was referred for suspected uterine inversion in view of some irregular fleshy mass came out of vagina after the delivery of stillborn female baby at home and placental delivery at nearby hospital. At the time of admission to labor room, she was conscious, oriented and severely anemic. On abdominal examination, uterus size was 22–24 weeks. On local examination of external genitalia, irregular mass of approximately 10 cm × 12 cm was seen coming out of vagina. On pulling the mass downwards, cord is seen attached to central part of the mass. Presence of cord attachment confirmed that mass was malformed fetus. The fetus was delivered by downward and backward traction. The fetus was diagnosed as acardiac twin

with poorly formed head with few hairs, undeveloped trunk, poorly formed both lower limbs (Figure 1), and centrally attached cord. The weight of acardiac twin was 1200 grams. Further evaluation of fetus was done by X-ray which revealed poorly developed cranial bones, ribs, spine, pubic bone, ilium , right and left femur with right incomplete tibia. On autopsy, heart was absent and other thoracic and abdominal organs were also absent. Some neural tissue was present at cephalic pole and vascular tissue was also present in body. All these findings further confirm it as a case of acardiac anceps twin.

DISCUSSION

Acardiac anceps is the most developed variety among all forms of acardiac twins. The blood system of acardiac twin is connected with normal twin. Normal twin drives blood through both fetuses because of which in acardiac twin blood flows in a reverse direction therefore it is called as reversed arterial perfusion. In this case, woman had spontaneous conception and diagnosis was made after delivery of both fetus as she was unbooked and uninvestigated. Such cases should be diagnosed as early as possible to prevent further complications of pump twin.

Pathogenesis can be explained by two theories:[2,4] According to first theory malformation occurs in acardiac twin due to deep placental anastomoses which occurs during early embryogenesis.

The other hypothetical theory explains that primary defect occurs during embryogenesis which causes cardiac development failure in one twin as a result of artery-artery anastomoses through which normal twin perfuses the acardiac twin.

Acardiac twin can be diagnosed by combined pulsed and color Doppler ultrasound studies. These studies documents perfusion of arterial blood flow in a retrograde manner.

The management options available to improve the outcome of normal twin are:

1. **Umbilical cord ligation:** This technique is carried out by ultrasound or by ultrasound endoscopy.
2. **Laser therapy of the placental vessels** is used to treat twin-twin transfusion syndrome.
3. **Laser umbilical cord occlusion:** In this endoscopic laser occlusion of umblical artery and vein is done.
4. **Radiofrequency ablation:** In this technique vessels are occluded by use of high frequency alternating current.
5. **Transection of the umbilical cord of the acardiac twin:** This is used in monochorionic twins for prevention of pump twin demise.

In a procedure like umbilical cord occlusion chance of normal twin survival will be 85–90% with high risk of neurological injury (5%).[5]

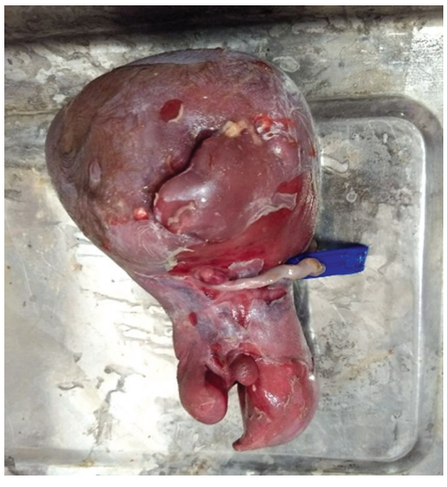

Figure 1 Acardiac twin having few hairs, undeveloped trunk and poorly developed lower exrtremities. Size: 10 cm × 12 cm

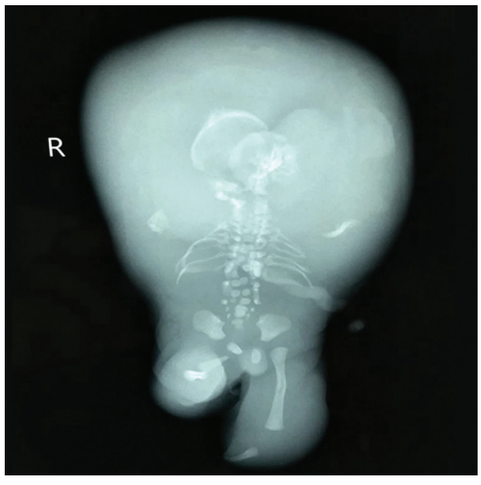

Figure 2 X-ray of acardiac twin showing different bones-cranial bones, ribs, spine, pubic bone, ilium, right and left femur with right incomplete tibia

CONCLUSION

Early diagnosis of this TRAP sequence helps to delivery of pump twin in time before development of severe complications. Timely intervention in this case could have saved the life of female fetus.

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