CASE REPORT

Acardius Acephalus: “Headless–Heartless Nine-Month Life”

Ray Sabyasachi · Basak Suvadip · Ghosh Prakash Ch. ·
Pal Mallika · Laga Madhusudan · Pradhan Mamata

Received: 19 September 2009 / Accepted: 5 June 2012 / Published online: 22 February 2013
© Federation of Obstetric & Gynecological Societies of India 2013

Introduction

Acardiac twin is an extremely rare and bizarre complication of monochorionic twin pregnancy occurring at an incidence of 1 in 34,600 deliveries or 1 % monozygotic twins [1]. It is also referred to as TRAP Sequence (Twin Reversed Arterial Perfusion) where as a result of arterial–arterial and venous–venous anastomoses without any arterial–venous connection, the acardiac (since it has no heart of its own) thrives as a parasite by getting blood supply from the donor hydropic twin [2]. Acardius is commonly associated with acephaly and failure of development of most of the thoracic organs and upper limbs.

We present a case report of Acardius acephalic twin which was retrospectively diagnosed following twin delivery of an unbooked pregnant woman admitted to our hospital.

Ray S. (✉), Assistant Professor · Basak S., Clinical Tutor ·
Laga M., Medical officer · Pradhan M.,
Professor and Head of the Department
Department of Obstetrics and gynaecology, Midnapore Medical
College, 25/H/1 Upendra Chandra Banerjee Road, Kolkata,
West Bengal 54, India
e-mail: subhadipmullick@hotmail.com

Ghosh P. Ch., Clinical Tutor
Department of Paediatrics, Midnapore Medical College, Kolkata,
West Bengal, India

Pal M., Assistant Professor
Department of Pathology, Midnapore Medical College, Kolkata,
West Bengal, India

Case Report

A 22-year-old unbooked Hindu female, gravida 2 para1, with a previous normal delivery, was admitted to our hospital at 37 weeks of gestation with labor pains. Cesarean section was decided for this patient due to non-progress of labor. She delivered a live healthy male child having apparently no congenital anomaly, with a birth weight of 2.5 kg. The other twin was a fleshy mass without any head-like structure weighing 1.7 kg. The liquor that drained during delivery was adequate for both the twins.

Anatomic Examination

The fetus acardius weighing about 1.7 kg had rudimentary upper limbs with identifiable fingers, 4 in each limb. The lower limbs were also short with an equinovarus type of deformity. It had no head, the top of the monster coming to a blunt head with a tuft of hair at the center of the upper end. The external genitalia were male with maldescended testis (Figs. 1, 2, 3).

Autopsy Report

A pathological autopsy revealed no heart, lung, liver, spleen, and kidney. The mediastinal cavity was present containing yellow colored fluid. Loops of intestine were also found to present in the mass.

Fig. 1 Picture of acardia acephalus

Postmortem Radiologic Report

Radiologic examination revealed normal pelvic and lower limb bones with complete spine and ribs. The upper limb bones were rudimentary. There was an absence of the skull as well as the pectoral girdle.

Placenta

It was monochorionic and diamniotic, weighing 600 g. The umbilical cord of the normal twin was 20 cm with central attachment, while the other umbilical cord was 12 cm long and atrophic.

Fig. 2 Autopsy on the acephalic mass

Fig. 3 Radiologic picture of acardiac acephalus

The imaging investigations and autopsy report were consistent with our diagnoses of acardiac twin.

Discussion

TRAP sequence arises from extensive artery to artery and vein to vein anastomoses in the twin placenta, leading to preferential perfusion of the parasitic twin's lower body with low pressure hypoxemic blood coming from the normal donor twin. This results in a “living” thing which does not have a heart, together with varying degrees of upper body regression in the recipient twin. The burden of providing circulation for two fetuses can result in cardiac hypertrophy, congestive failure, hydrops fetalis, or premature birth of the normal fetus. This was not the case in our patient, who delivered a full-term healthy child.

Acardiac monsters have been known since 1533, when the condition was first described by Beneditti [3]. Two-thirds of acardiac fetuses are acardiac acephalic, which is the most common variety, and they are predominantly female [3].

Based on the degree of cephalic and truncal maldevelopment, four types are recognized:

1. Acardius anceps—head and face partially developed.
2. Acardius acephalus—no cephalic structures.
3. Acardius acornus—there is a head, but no body; rarest type.
4. Acardius amorphous—formless blob containing all types of tissue, but no recognizable organs.

Our case was Acardius acephalus, having two arteries and two veins. In over 50 % of cases, the umbilical cord of the acardiac twin contains only a single umbilical artery.

Diagnosis was not made during the antenatal period as this mother had never asked for a USG scan. USG with Doppler has made the diagnoses of acardia possible even in the first trimester of pregnancy by detection of inversion of vascular flow in the recipient acardiac fetus. So, as soon as the USG detects a twin pregnancy, chorionicity should be determined and if monochorionic, Doppler should be carried out to detect such a TRAP sequence. Early diagnosis may allow measures to be taken to reduce risk of complication, as the mortality rate is reported to be between 50 and 70 % for the pump twin [2].

Fetal echo is also essential to evaluate signs of heart failure for the pump twin.

Differential diagnoses are mainly with the amniotic band syndrome.

Management aims at interrupting the blood flow from the pump twin to the acardiac twin without harming the normal twin. The in utero treatment modalities that have been attempted include medical therapies, selective delivery of the acardiac twin in the 2nd trimester followed by prolongation of pregnancy for the normal twin, and umbilical cord blockage [3]. The most promising one

seems to be endoscopic ligation of the acardiac twins' umbilical cord with a success rate of 70–80 % [4]. USG-guided thrombosis of the umbilical artery using absolute alcohol/thrombogenic coils/fibrin has been successfully attempted by Grab. In other cases, a high energy device is used to destroy the blood vessels and surrounding tissue at the site where they enter the acardiac twin, a procedure termed radio frequency ablation.

References

1. Kim Tae Jin, Kim Chong Jai, Park Sung Hye, et al. Holoacardius Hemisomus Acephalus: A Case Report. *Kor J Path.* 1989;23: 487–9.
2. Nik Lah NAZ, Che Yaakob CA, Othman MS, et al. Twin Reverse arterial perfusion sequence. *Singap Med J Case Report.* 2007;48: e335–7.
3. Dhall U, Kayalvizhi I, Magu S. Acardius Acephalus. Monster—A Case Report. *J Anat Soc India.* 2005;54:26–8.
4. Krishna LG, Uppoor R, Shankar JS. Amorphous acardiac parabi-otic twin. *Indian J Radiol Imaging.* 2005;15:267–70.