THE JOURNAL OF OBSTETRICS AND GYNECOLOGY OF INDIA

The Journal of Obstetrics and Gynecology of India (July–August 2013) 63(4):282–284
DOI 10.1007/s13224-012-0176-2

CASE REPORT

Diagnostic Dilemma in Twin-Reversed Arterial Perfusion Sequence

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Received: 15 March 2011 / Accepted: 12 October 2011 / Published online: 17 August 2012

Introduction

The twin-reversed arterial perfusion (TRAP) sequence is a rare complication of twin pregnancies. The anatomically normal twin is termed the pump twin; the recipient twin is malformed and is known as acardiac twin. It functions as a parasite to the pump twin and receives retrograde blood flow of poorly oxygenated blood from the pump twin via the abdominal aorta, resulting in a wide spectrum of malformations. The abnormal haemodynamics created in this situation can result in congestive heart failure, hydrops and consequent premature delivery of the pump twin such that the mortality rate may be as high as 50 %. We present a case of TRAP sequence in a twin pregnancy which was diagnosed at 35 weeks of gestation. It highlights the risk of monochorionicity-associated morbidity in multiple pregnancies.

Case Report

A 23-year-old primigravida with 35-week gestation was referred to our centre with extensive distension of abdomen. She had insignificant past and family history with no history of fever, drug intake, or X-ray exposure in the first trimester. Her earlier USG raised the possibility of a malformed foetus but did not give a confirmatory diagnosis. Ultrasound in the Radiology department showed twin pregnancy within the single sac with a live foetus and an acardiac foetus with large cystic spaces in the torso. It had only single identifiable limb attached to an amorphous soft tissue mass with no identifiable heart or kidneys. The foetus which showed normal activity had normal morphology and biophysical profile (Figs. 1, 2, 3).

The findings were confirmed post-delivery which was done transvaginally. The normal baby was delivered first, followed by acardiac twin which was delivered only after decompressing the large cystic spaces. The twin had no developed head, had round trunk like structure, and single limb. On autopsy, acardius acephalus with lymphangiomas was diagnosed.

Discussion

Acardia was first described in the sixteenth century. Early references refer to acardia as craniopagus parasiticus. It is now also called twin reversed arterial perfusion sequence, or TRAP sequence.

Acardiac twinning is a rare anomaly occurring in 1 in 35,000 pregnancies and up to 1 % of monozygotic twins. The risk is even greater in multiple pregnancies of a higher

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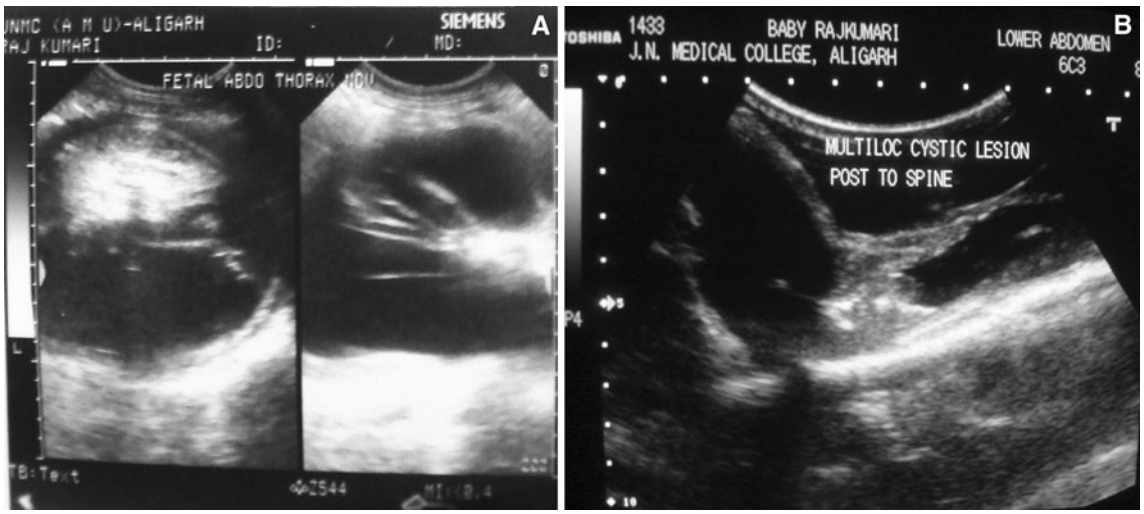

Fig. 1 **a**, **b** Antenatal ultrasound scan shows malformed twin showing parts of spine and large cystic spaces in the torso; these findings were confirmed on the post natal scan to be on the dorsal aspect of foetus and proved to be lymphangiomas on histopathology

Fig. 2 Shows characteristic dysmorphic appearance of the abnormal fetal mass in a TRAP sequence with only single formed limb and an amorphous soft tissue mass

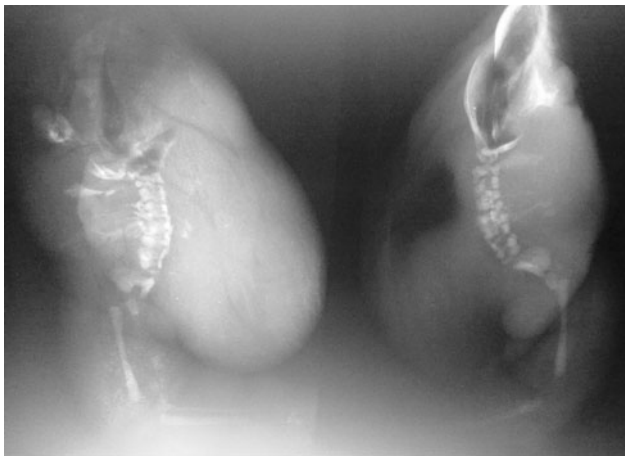

Fig. 3 Shows collapsed bones of the head post-craniotomy and malformed spine with few rudimentary ribs and bones of the single lower limb

order [1]. It is the extreme manifestation of the twin-twin transfusion syndrome (TTTS) spectrum. The acardiac twin (recipient) exists as a parasite, depending on the normal donor (pump) twin for its blood supply via transplacental anastomoses and retrograde perfusion of the acardiac umbilical cord. Perfusion of the malformed (acardiac) foetus occurs via artery-to-artery and vein-to-vein anastomoses between the foetuses. Umbilical arterial blood from the donor flows into the umbilical artery of the recipient, its direction reversed [2].

In an acardiac foetus, the malformation observed has characteristic features. Cardiac structures are absent or non-functioning, and the head, upper body and upper extremities are poorly developed. The lower body and lower extremities are, however, more or less normal [3]. Depending on the state of disruption, acardiac anomalies are divided into four categories: acardius anceps, acardius acephalus, acardius acormus and acardius amorphus.

Acardius acephalus is the most common type of acardiac twin. These twins do not develop a head, but may have an underdeveloped skull base. They have legs, but do not have arms. On autopsy, they are generally found to lack chest and upper abdominal organs. Acardius amorphus appears as a disorganized mass of tissues containing skin, bone, cartilage, muscle, fat and blood vessels. This type of acardiac twin is not recognizable as a human foetus and contains no recognizable human organs. Acardius anceps is the most developed form of acardiac twin. This form has

arms, legs and a partially developed head with brain tissues and facial structures. This type of acardiac twin is associated with a high risk for complications in the normal twin. Acardius acormus is the rarest type of acardiac twin. This type of acardiac twin presents as an isolated head with no body development [4].

For the pump twin, the prognosis is dismal, leading to a 50–70 % mortality rate due to congestive heart failure, polyhydramnios or preterm delivery. Doppler verification of reversed flow in the umbilical cord of the acardiac foetus confirms the diagnosis and antenatal ultrasonography is important in early diagnosis and optimal prenatal management. There are reports which describe acardiac twin who was missed on ultrasound and only detected at delivery. Hence, one should be cautious before diagnosing an unusual fetal malformation.

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