AN ACARDIUS AMORPHUS IN A TWIN PREGNANCY

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SYNOPSIS

A case of acardius amorphus occurring in a monozygotic twin is described. The most important features are the absence of a heart and its existance as a complete parasites on the normal twin. The aetiology, incidence and associated clinical problem are discussed.

INTRODUCTION

An acardius amorphus was first described by Benedictus in 1539. It consisted of a mass of connective tissue covered with skin, containing rudiments of the skeleton and sometimes of the viscera. It was the least developed form of a fetus acardius of which four types had been recognized. We described below the first such case seen at the University Hospital, Kuala Lumpur.

CASE REPORT

An Indian primigravida, aged 28 years, was booked at 29 weeks gestation. Her blood pressure was 160/100 mmHg and she had mild ankle oedema; there was no proteinuria. The uterine size corresponded to the period of amenorrhoea. The foetal presentation was breech and there was no clinical evidence of hydramnios. Three days after admission, her blood pressure settled to 130/80 mmHg.

At 36 weeks gestation, she went into labour after spontaneous rupture of membranes. An assisted breech delivery was carried out and a live female infant weighing 2110 gms was delivered. Subsequent examination of the uterus showed it to be unusually large suggesting the presence of an undiagnosed twin. Exploration of the uterus revealed an amorphus mass; this was pushed down into the pelvis with the aid of the abdominal hand, and subsequently delivered with application of forceps; its short cord was torn at delivery. The placenta and membranes were delivered intact. The pre-term infant was normal and discharged two weeks later.

GROSS APPEARANCE

The ovoid acardius amorphus mass, measured 15 × 8.5 × 5 cm and weighed 420 gms. The cranial end was identified by a few tuffs of hair and two cystic swellings measuring 3.5 × 2.5 cm in diameter. The caudal end was identified by the presence of a rudimentary lower limb which measured 3 × 1.5 cm (Figure 1).

X-ray features (Figure 2) revealed a soft tissue mass, rudimentary skeletal frame of vertebrae and a femur.

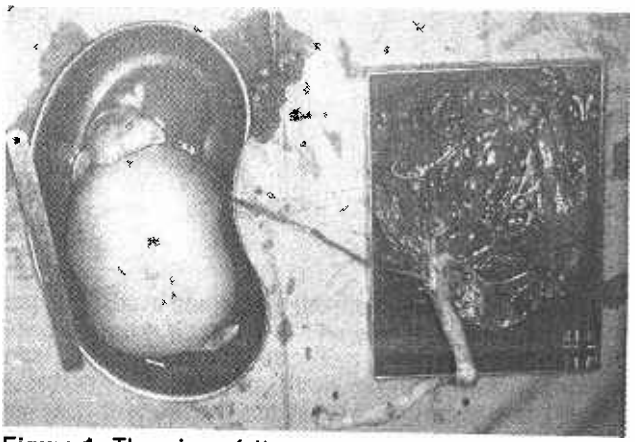

Figure 1. The size of the acardius amorphus is comparable to that of the placenta. Its cord was attached to the normal umbilical cord.

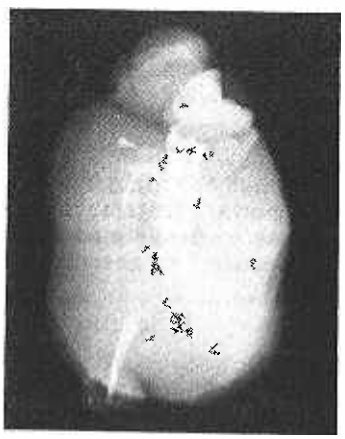

Figure 2. A rudimentary frame of vertebrae and a femur measuring 3 × 0.5 cm in the acardius amorphus.

Figure 3. The structures are adipose, fibrous and muscle tisue covered by skin.

On sectioning (Figure 3) the mass is composed of essentially adipose, fibrous and muscle tissues surrounding an ossens bone and covered externally by skin. The vessels from the umbilical cord are seen to ramify within the mass in a haphazard manner. There is no fetal heart or pleural structures identified. A well defined femur is identified and measures 3 × 0.5 cm.

Microscopic examintion confirmed the presence of remnants of the following:-

(a) adrenal glands with distinct cortex and medulla

(b) large intestine with mucinous lining epithelium and smooth muscle wall

(c) embryonal lymphoid and liver tissue

(d) mature adipose tissues, cartilage, bone with marrow and skin with underlying dermal appendages. There were no neural elements or cardiac muscles seen.

PLACENTA AND CORD (FIGURE 1)

The placenta weighed 406 gm and it was monochrionic and diamnionic. The normal umbilical cord of the first twin had a velamentous insertions. The slender cord of the amorphus mass measured 12 cm length and was attached to the normal umbilical cord about 3 cm from the site of its insertion.

The normal cord contained two arteries and one vein. The thin cord of the monster contained one vein and only one artery. This single artery and vein was shown to be continuous with the umbilical artery and vein of the normal foetus.

DISCUSSION

Based on the variable stages of development, four types of acardiac monsters have been described (1,2).

(a) Acardius anceps

The head is rudimentary but the trunk and limbs develop normally.

(b) Acardius acephalus

The head, upper limbs and upper part of the trunk are absent, but the lower limbs and pelvis develop normally.

(c) Acardius acormus

The monster is only a partially developed head without a body.

(d) Acardius amorphus

The monster consists of a mass of connective tissue covered with skin, containing rudiments of the skeleton and sometimes of the viscera.

The frequency of occurrence is estimated to be 60-75% in acardius acephalus, 20% in acardius amorphus and 10% in acardius anceps (3).

In a review of the world literature of acardius monsters (4), the incidence of its occurrence was calculated as one in every 34,600 deliveries. Until 1983, 149 cases had been reported in the world literature, seven of which occurred in triplets deliveries. Recently, Tauchi (5) reported 88 cases in Japan with three occurring in triplets.

An acardius amorphus is a complete parasite obtaining its blood supply from the normal fetus through the umbilical artery which is poor in oxygen and nutritions; further, the blood flow is in a reverse direction. The existenance of a direct arterial communications between the twin remains speculative.

Genetic factors have not been established as an underlying aetiology. Ross (6) had refuted such a possibility as an acardius amorphus was noted to occur in triplets where two normal infants developed from the same ovum.

Epidermiologically, the occurrence of acardiac monsters is unrelated to maternal age or parity. The incidence of congenital anomalies in such a twin is the same as that in monozygotic twinning 8.3% (7).

Clinically, polyhydramnios is a common occurrence though not demonstrated in our patient. Premature delivery is also frequent. As in our case the presence of an acardius amorphus may remain unsuspected antenatally in the absence of polyhydramnios. Ultrasound appearance of a gradually growing mass without any heart beat gives a clue to the diagnosis of acardiac monster (5). Labour is often uncomplicated as most often the normal twin is delivered first followed by the acardius twin.

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