Acardius Acephalus Monster - A Case Report.
Dhall U, Kayalvizhi I & Magu S*
Pt B.D.S. PGIMS, Rohtak, H.R.

Abstract: A case of an acardius acephalic monster is described. This case report is based on morphological and histological confirmation. Foetus acardius is a parasite for its vascular circulation from the donor twin. The twin reversed arterial perfusion syndrome is an extremely rare manifestation of fetofetal transfusion in twin pregnancy where the affected twin receives vascular supply retrogradely from the healthy twin. The affected twin, acardius acephalus represented with several malformations in association with absence of upper extremity. Placenta was monochorionic and diamniotic. This case was diagnosed prenatally by ultrasound examination.

Key Words: Acardius acephalus, Monozygotic twins, Human, Monster.

Introduction:
Acardia is a rare and severe congenital malformation usually found with monozygotic twin pregnancy, occurring approximately once in 35,000 births (James, 1977). It is characterized by the absence of functioning heart. Fetus acardius is a parasite as it receives its blood supply from the donor twin. But no two acardiac fetuses are alike due to the variability in the time of disruption of developmental process. This report adds to the literature for the typical example of an acardius acephalic monster.

Case Report:
A 24 years old gravida 4, delivered twins after 28 weeks of gestation. The first twin was fresh still born female, weighing 1100gm and had no obvious congenital malformation. The second twin was a well preserved monster weighing 1600 gm and had major congenital anomalies described below.

Anatomical Examination:
The fetus acardius was covered with thick, dark brown (darker than that of normal twin) skin that was soft and oedematous. Both the lower limbs were near normal except for equinovarus deformity of the foot. There was no head, the top of the monster coming to a blunted end. External genitalia were ambiguous (Fig.1). In the upper pole of monster there were subcutaneous cysts containing clear yellow fluid. Incision on the ventral surface of abdomen revealed parts of GI tract in the form of a tubular organ with a blind cranial end. At the cranial end of the abdominal cavity was a single mass of tissue in the midline identified histologically to be kidney (Fig.2). There was a cartilaginous bony skeleton and an incomplete spinal column. Heart, lungs, diaphragm, liver, spleen, stomach and pancreas could not be identified.

The organs identified with certainty after microscopical observation were, muscle, colon, kidney, urinary bladder (Fig. 3, 4 & 5) and ovary. Kidney showed...
four rows of glomeruli and collecting tubules. Skin from lower limb showed single layered epidermis, few rudimentary hair follicles, no sweat gland and numerous haemorrhagic areas.

Placenta:
Placenta was monochorionic and diamniotic. It weighed 440 gms. The normal umbilical cord was 25 cm long with a central attachment. The other umbilical cord was thin, atrophic, dark brown 18 cm long and attached on the margin of placenta. Each cord had one umbilical vein (UV) and two umbilical arteries (UA). One UV and one UA of normal fetus were anastomosing with similar vessels of abnormal fetus on the surface of placenta (Fig.3). Histology of placenta was quite normal.

Postmortem Radiological Study:
Radiological examination revealed a disproportionate mass of soft tissue and varying size of cavities present postero-superior to the rudimentary vertebral column. There was absence of ribs, skull and the upper limbs including the pectoral girdle (Fig.4). The pelvis and the lower limb bones were quite normal.

Discussion:
Acardia represents one of the most severe and rarest congenital anomalies. Among them the most common variety is the acardius accephalus in which the head is lacking and so are the upper extremities. The other types are acardius anceps, acardius acormus and acardius amorphous.

Acardius anceps is the most highly developed form possessing a partly developed head with remnants of cranial bones and brain tissue. The body and extremities are also developed. Acardius acormus is the rarest form of acardia. The monster is a head without a body. Placenta is attached to the head directly or via an umbilical cord ending in its cervical region. Acardius amorphous, the least developed monster not recognizable as a human form with minimally developed visceral organs. As there is no gross human form it is named acardius amorphous. (Napolitani and Schreiber, 1960).

Anatomical examination in present case showed lack of thoracic and superior abdominal organs and diaphragm. Subcutaneous necrosis with formation of fluid filled cysts present in the thickened edematous skin of the cranial region. Similar observation were also reported by Van Groeninghen et al (1985) and by James (1977). These authors explained that hypoxemia resulted in subcutaneous necrosis giving rise to pseudocysts filled with serous fluid in the thickened edematous skin.
Epidemiology:

Acardiac monsters have been known since 1533 when the condition was first described by Beneditti (Quoted by Napolitani and Schreiber, 1960). Acardiacs always occur in a multiple birth and two third are diamniotics (Spencer 2001). The occurrence rate is estimated to be less than one in 340 deliveries, 1% of monochorionic twin pregnancies and 1 in 30 monozygotic triplets (James 1977). Frutiger (1969) reported that acardiacs were predominantly female while Spencer (2001) found only 29% cases to be female. The present case in our study was also a female with monochorionic, diamniotic placenta. Two-third of acardiac fetuses are Acardiac acephalic, which is the most common variety among the various types of acardias mentioned in the literature, was also supported by Napolitani & Schreiber (1960).

Etiology:

The etiology of acardiac monster is still not known. Genetic defects have been reported to be the cause by some while others do not support this theory. Chromosomal abnormalities have been found in about half the cases by Van Allen et al. (1983). On the other hand Van Groeninghen et al. (1985) reported that most acardiacs showed normal karyotype.

Compressing of the cephalic pole of the embryo prohibiting curving and fusion of the primitive heart tube has been suggested to be the basic cause of this anomaly by Krause and Bejdl (1948). This results in nonformation of heart and heart dependent entodermal organs like thyroid, oesophagus, trachea, lung, liver, etc. as was also observed in the present specimen.

Most acardiacs are reported to have single umbilical artery indicating persistence of transitory single artery phase (which is normal up to Carnegie stage 12) (Monie, 1970). The present case, however, showed two umbilical arteries which also has been reported in literature suggesting pathology might have occurred late i.e. after Carnegie stage 12. Such acardiac fetuses are reported to be anceps variety which is the most developed type of acardiac fetus.

Presence of vascular anastomosis on the surface of placenta resulting in feto-fetal transfusion is a common feature of acardiac monsters. Whether this twin reversed arterial perfusion (TRAP) is the cause or the effect of acardia is still under controversy (Stephens, 1984; Seeds et al. 1987; Mohanty et al 2001).

Clinical Importance:

As the umbilical cord of acardiac twin is usually hypotrophic as in present case, it is likely to be torn during manipulation resulting in death of normal fetus due to blood loss through anastomosis. (Sanfilippo et al. 1979).

Hydramnios is a common feature in such pregnancy due to absent amniotic fluid swallowing. However, this was not the feature in present case.

Burden of providing circulation of two fetuses can result in cardiac hypertrophy, congestive failure (Simpson et al. 1983) hydrops foetalis or premature birth of normal fetus.

Improved imaging techniques like 2D ultrasonography, 3D ultrasonography and transvaginal Doppler ultrasonography have made the diagnosis of acardia possible even in the first trimester of pregnancy by detecting inversion of vascular flow in the recipient acardiac fetus (Bonilla Musoles et al. 2001). Early diagnosis may allow measures to be taken that may help to reduce the risk of such complications.

References: