

Acardius Acephalic Monster

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ABSTRACT

Dermoid cyst in postmenopausal women is a rare entity. It is seen most commonly in young reproductive age group. It constitutes about 10 to 20% of all ovarian tumors in pregnancy. Chance of malignancy is about 1-2%, torsion is common.

Here, we have presented an unusual case of dermoid cyst in a postmenopausal woman who presented with complaints which were not directly related to the cyst. A 16-week mass per abdomen was not bothering the patient at all, rather she presented in the OPD with a mass coming out per vaginum, which was a third degree uterocervical descent. Patient was posted for vaginal hysterectomy and the cyst was removed perabdominally.

Keywords: Acardius acephalus, Monozygotic twins, Human monster.

INTRODUCTION

Acardiac monsters have been known since 1533 when the condition was first described by Beneditti.¹ Acardiacs always occur in a multiple birth and two-third are diamniotics. The occurrence rate is estimated to be less than one in 340 deliveries, 1% of monozygotic twin pregnancies and one in 30 monozygotic triplets.²

A case of an acardius acephalic monster is described. This case report is based on morphological and sonological confirmation. Fetus 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 retrograde 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.

CASE REPORT

A 24-year-old gravida 2, delivered twins after 28 weeks of gestation. The first twin was a female, weighing 1000 gm and had no obvious congenital malformation with severe respiratory distress and was admitted in NICU for low birth weight but inspite of resuscitative measures could not survive. The second twin was a well-preserved monster, weighing 700 gm and had major congenital anomalies described below.

The fetus acardius was covered with thick, dark brown (darker than that of normal twin) skin that was soft and edematous. 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). 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 to be kidney. 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 and ovary.

Placenta: Placenta was monochorionic and diamniotic. It weighed 440 gm. 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. Histology of placenta was normal.



Fig. 1: Acardius acephalic monster

DISCUSSION

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.³ On the other hand, some have reported that most acardiacs showed normal karyotype. Compression 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. This results in nonformation of heart and heart dependent endodermal organs, like thyroid, esophagus, 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. 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 fetofetal 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.⁴

CLINICAL IMPORTANCE

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.⁵ Early diagnosis may allow measures to be taken that may help to reduce the risk of such complications.

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