CASE REPORT

Acardius Amorphus: Magnetic Resonance Imaging (MRI) can be helpful in the Diagnosis when Ultrasound (US) is Inconclusive

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SUMMARY

An acardiac twin is rare and the diagnosis is commonly achieved from antenatal ultrasound (US) scans. However there have been cases where the appearances of the acardiac twin can be confusing and mimics a mass or tumour, for example, a teratoma. We experienced a case where the findings were unclear from the antenatal ultrasound scans and we had to resort to Magnetic Resonance Imaging (MRI), where we finally made the correct diagnosis based on the identification of two umbilical cords, supplying the normal fetus and the 'mass' (acardiac twin) respectively.

KEY WORDS:
Acardiac twin, ultrasound, magnetic resonance imaging

INTRODUCTION

An acardiac twin is a rare complication of multifetal pregnancy, with a reported incidence of 1% of monochorionic twin pregnancies, i.e. 1 of 35,000 pregnancies[1]. It occurs as a result of twin-reversed arterial perfusion (TRAP) sequence when both twins derive their blood supply from a single placenta. If one twin's cardiac function develops more slowly in early pregnancy, imbalance of blood pressure between the twins results in retrograde flow of poorly oxygenated blood to the heart of the abnormal twin. This interferes with the development of the heart so that it rarely progresses beyond a rudimentary stage of development. In most cases, the lower body and extremities develop to a certain degree but poor perfusion (and under oxygenation) to the upper body leads to underdevelopment of this area. The acardiac twin is a lethal condition while the normal twin is at risk of cardiac failure.

The pathologic appearance of acardiac twins varies considerably as described by Van Allen *et al*[2]. Four types are recognized: 1) Acardius anceps – head and face partially developed, 2) Acardius acephalus – no cephalic structures (most common), 3) Acardius acormus – head without body, 4) Acardius amorphous – formless blob, contains all tissue types but no recognizable organs. This differs from a teratoma only by its attachment to an umbilical cord.

Because the diagnosis is usually made from antenatal US scans by the obstetricians and MRI is rarely needed, radiologists may not be aware of the MRI appearances of acardiac twin. In fact as have been reported previously, the more severe forms of acardiac twin have been misdiagnosed antenatally as placental or uterine masses or teratoma by ultrasonography[3,4]. This is because a normal fetus is visualized and a mass (the acardiac twin) is seen intrauterine but separate from the normal fetus.

CASE REPORT

A 31-year-old woman, gravida 2 para 1, was referred to our hospital at 34 weeks gestation for further evaluation of the fetus with an intrauterine cystic mass. US examination revealed a single fetus, which was grossly normal. An intrauterine mass was seen, located anterior to the lower abdomen of the fetus. The entire mass measured 12 X 7 cm. It consisted of a homogenous soft tissue with what seemed to be a long bony structure within it (Figure 1a, arrowheads) and also multi-loculated cysts (Figure 1b, star). The mass was seen separate from the fetus but was not clear around the pelvic area. Colour Doppler US showed no increased vascularity. The placenta was large (hyperplacentosis). Liquor volume was normal. Since US could not clearly establish the origin of the mass, MRI was performed for further evaluation.

The MRI (Figure 2a-c) showed a fetus with cephalic presentation, which demonstrated normal appearance and internal structures consistent with gestational age. A well-defined oblong shaped mass clearly separate from the fetus and placenta was identified. As seen on US, this mass consists of a soft tissue with what seems like a bony structure in the center (Figure 2c, short double arrows). Multiloculated cystic component was observed on the other end of the mass (star). No recognized fetal parts are seen.

During the initial MRI assessment a diagnosis of a teratoma was considered. However on further scrutiny, two umbilical cords were identified emanating from the placenta; one to the fetus (Figure 2a, long arrow) and one towards the mass (Figure 2a, short arrow). The latter was not identified on US. These findings led us to believe that one of the umbilical cords was supplying a severely malformed fetus thus a diagnosis of acardiac twin (acardius amorphous) was made. The patient was appropriately counselled.

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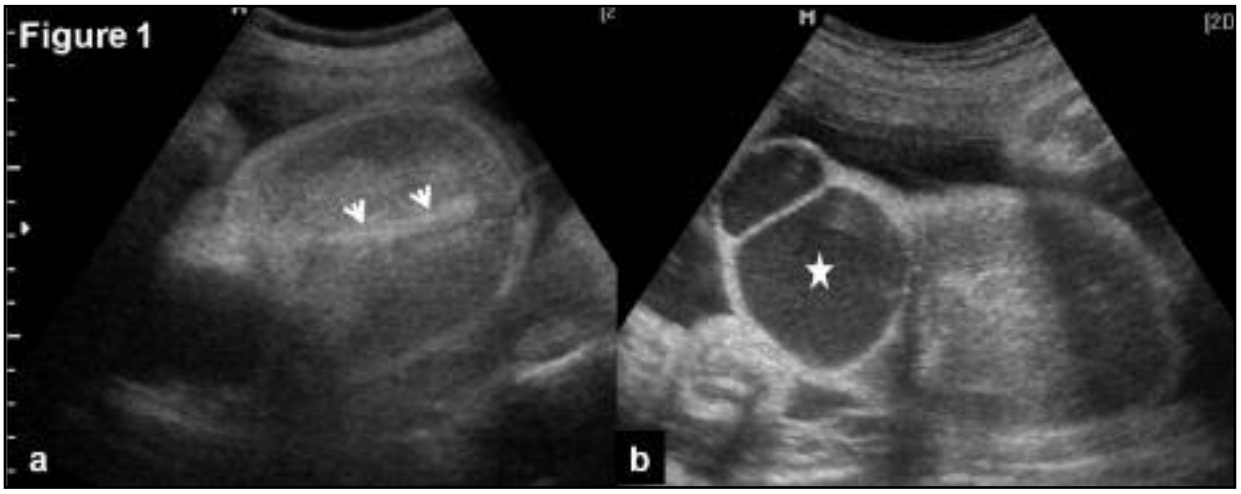

Fig. 1a-b: US scan showed an intrauterine mass which consist of a homogenous soft tissue with what seemed like a midline bony structure within it (arrowheads) and also multi-loculated cysts (star).

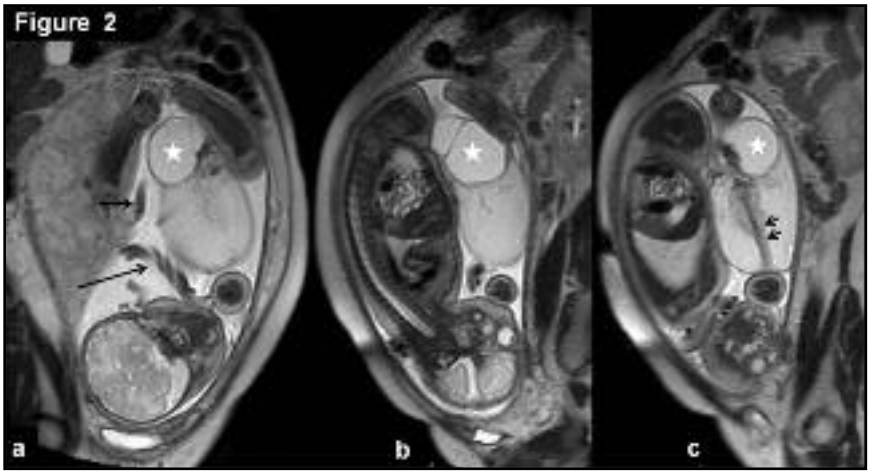

Fig. 2a-c: Fetal MRI showed the normal fetus with cephalic presentation and well-defined mass clearly separate from the fetus and placenta. The bony-like structure within the mass (short double arrows) and multiloculated cystic component was observed (star). Two umbilical cords seen emanating from the placenta; to the fetus (long arrow) and another towards the mass (short arrow).

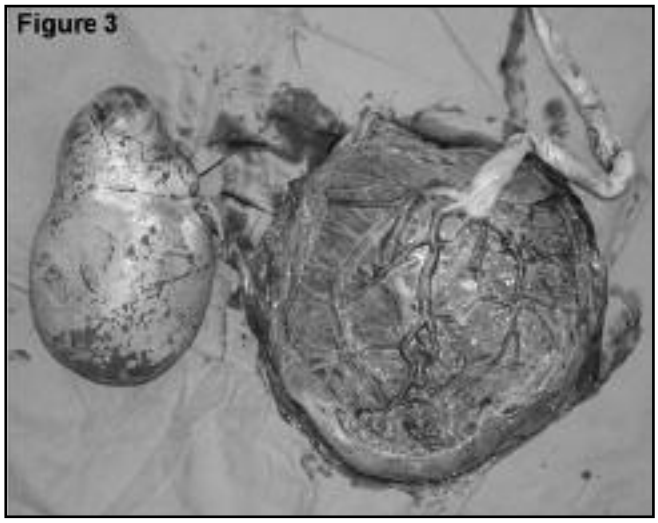

Fig. 3: Gross appearance of the acardius amorphous attached to the placenta via its umbilical cord.

The patient was seen two weeks later and the normal fetus showed no signs of cardiac failure (a known complication from twin-reversed arterial perfusion) while the acardiac twin had not increased in size. Despite explaining to the patient and her husband that normal vaginal delivery is possible, they requested for a Caesarean section, which was performed uneventfully at 37 weeks gestation. A healthy 2.7 kg baby boy was delivered. There was only a single placenta and single amniotic membrane (monochorionic monoamniotic twins). The acardiac twin (Figure 3) was delivered still attached to the placenta via its umbilical cord. The umbilical cord contained only 2 vessels; a small artery and a large vein. The patient and her newborn son were discharged well on the third postoperative day.

DISCUSSION

Prenatal diagnosis of acardiac twin is usually made with a high degree of sensitivity using US scan. However like in our case, when the patient was referred and presented to us for

the first time in the 3rd trimester, US diagnosis becomes technically more difficult due to the fetus being less mobile from limited space. MRI maybe useful as an adjunct in already identified cases to assess the presence of retrograde blood flow in the acardiac fetus and umbilical artery[5]. In our case, we had to resort to fetal MRI to help with the diagnosis as it was unclear and inconclusive from US. As MRI is not the standard method used for the diagnosis, case reports on MRI appearance of acardiac twin is scarce from our review of the literature; other than a technical report of a non-gated fetal MRI of umbilical blood flow in an acardiac twin[5].

The clue to the correct diagnosis in our case was the identification of separate umbilical cords supplying the normal fetus and the 'mass' (acardiac twin) respectively. We hope the illustration of the MRI appearance of the acardius amorphous in our case will be useful as a reference to radiologists and obstetricians and also to other health professionals generally as reports on MRI of this condition is scarce.

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