Case Report

Fetus Acardiac Amorphous Presenting as Placental Tumor: A Rare Case and Differentiating the Two

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Abstract

Fetus acardius is a rare manifestation of twin reversed arterial perfusion and is a parasite due to vascular circulation from donor twin and lacks any resemblance to human embryos. Antenatal diagnosis is challenging as there are no well-defined features. We report here a case which presented as placental mass, the diagnosis of which was evident after delivery. Antenatal diagnosis, review of the literature, and differential diagnosis from the placental mass are discussed. A primigravida in the late third trimester had ultrasonography (USG) showing a 7 cm × 5 cm mass adjacent to the placenta. She had no complaints and fetal biometry was normal. She delivered a healthy baby; placenta showed an attached mass without identifiable fetal body or limbs, and was covered with skin having scanty hairs, which was identified as an amorphous fetus. Early diagnosis is possible by early USG; an acardiac amorphous fetus may present as placental mass. Characteristic feature on USG supplemented by magnetic resonance imaging may help in reaching a correct diagnosis and optimal management.

Keywords: Acardiac twin, monochorionic gestation, monochorionic twin, parasite twin, placental tumor

Introduction

Acardiac twin is a rare complication of monozygotic twin pregnancies occurring in 1% of monochorionic twin gestation, with an incidence of 1:35,000 births.[1] The normal umbilical blood flow is reversed in acardiac twin and blood flows from the umbilical artery of the normal twin to the acardiac twin; this is known as twin reversal arterial perfusion (TRAP).[2,3] TRAP has mainly four phenotypes: acardius anceps, acardius acephalus, acardius acormus, and acardius amorphous.[3,4] The natural history of this condition diverges from *in utero* cardiac failure to fetal demise to normal-surviving twins.

Placental masses can be benign or malignant and have a varied presentation. If the patient presents late, diagnosis can be challenging. We report this case due to a challenging diagnosis and suggest some radiological features that may help in reaching the correct diagnosis to a keen clinician.

Case Report

A 25-year-old primigravida came for a routine antenatal checkup at 9 months of pregnancy. There was no pertinent past, medical, or family history. She did not have any ultrasonography (USG) in the first or second trimester. Third-trimester USG showed a single live intrauterine fetus of 35 + 2 weeks, with normal growth and adequate liquor amnii. A suspicious placental mass of 7.2 cm × 4.6 cm × 5.6 cm, well-defined with mildly echogenic border and central area of hyperechogenicity was seen adjacent to the placenta [Figure 1]. There were no color Doppler changes and no vascular connection was identified.

She delivered vaginally a healthy baby of weight 3010 g with a APGAR score of 9 and 10 at 1 and 5 min, respectively, and no anomalies. The placenta and membranes weighed 0.650 kg with three-vessel cord and a length of 51 cm. A mass of 8 cm × 6 cm × 4 cm covered with amnion was seen attached to the placenta [Figure 2]. The mass was soft in consistency, connected with the placenta with a thin cord. The mass was covered by skin with some hairs. It had few

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bud-like structures [Figure 2]. We could identify dorsal and ventral surfaces of the malformed and shapeless fetus, but not the cranial and caudal ends. Microscopy of the mass revealed normal-appearing skin with skin appendages, cartilage, endochondral ossification, and skeletal muscle on hematoxylin and eosin stain [Figures 3-5].

Discussion

TRAP is unique to monochorionic gestations in which there is a normal pump twin and a recipient twin which is acardiac. TRAP is more common in triplet pregnancies, though clinically more frequently seen in twins due to a higher incidence of twinning. TRAP results in early tissue hypoxia, leading to secondary atrophy of heart and other major organs.[4,5] Others explain it by severe genetic or major cardiac embryogenesis defect, leading to failure of the heart development in one twin which later forms vascular connections.[3,6] Associations of TRAP with other anomalies do point to the possibility of defects in early embryogenesis. The acardiac twins are dependent on the pump twin for its perfusion, and the pump twin suffers from high-output failure making both of them at-risk of mortality. The upper half of the body –head, cervical spine, and upper limbs – is very poorly developed or not developed at all in acardiac twins which is invariably lethal. There is a marked difference in the size of the umbilical cord and inconsistent membrane development.[5]

Acardiac twins might be acephalus (absent head), anceps (poorly formed head), acormus (presence of head only), and amorphous (amorphous unrecognizable mass).[6]

USG and Doppler are helpful in the diagnosis of TRAP; discordance in growth and malformations with the retrograde flow in acardiac twin can be seen. Anencephaly, conjoint twins, single fetal demise, and placental tumors are often a differential diagnosis. Our case presented with amorphous mass, without identifiable vascular flow. If there are no fetus-like structures, it is difficult to diagnose this condition in the absence of previous USG. However, a central hyperechoic area with echoes similar to bone and well-defined outer hyperechoic area and Doppler flow changes in the cord should raise suspicion for this condition.

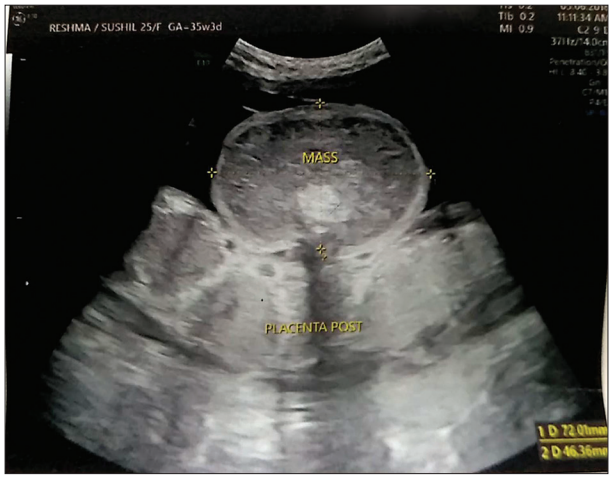

Figure 1: Ultrasonography showing mass adjacent to the placenta

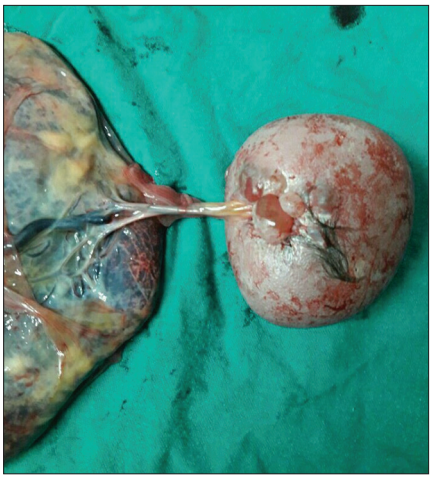

Figure 2: The placenta with cord and mass connected to it; bud-like structures and hairs can be seen

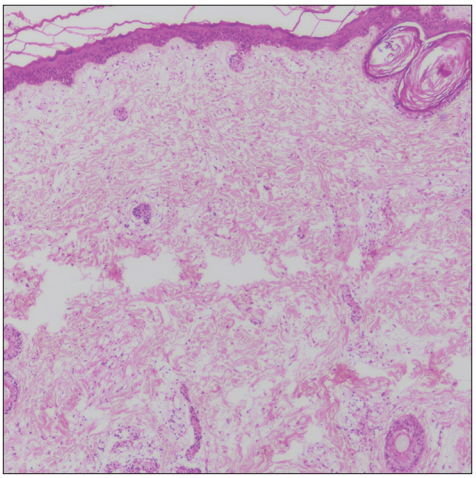

Figure 3: The skin of parasitic twin with skin appendages (H and E stain, ×4)

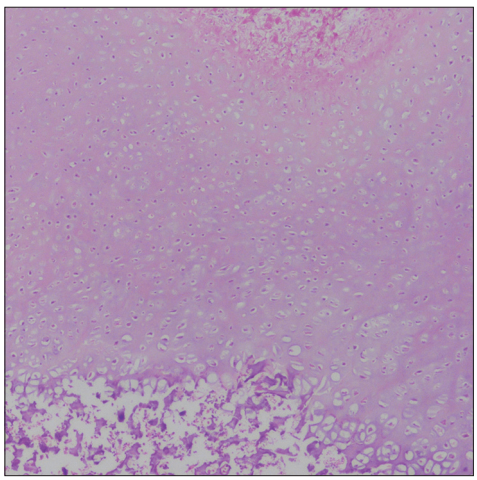

Figure 4: Cartilage and endochondral ossification in the mass (H and E stain, ×4)

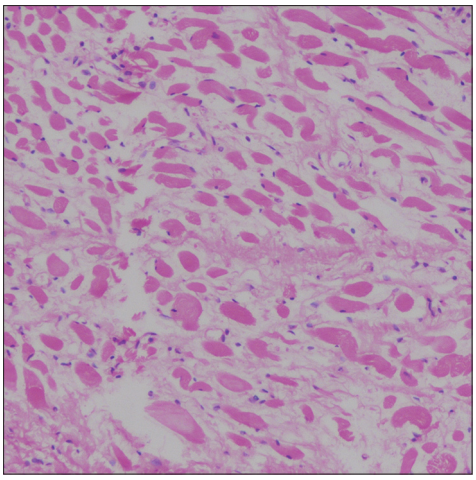

Figure 5: Fascicles of fetal skeletal muscle (H and E stain, ×10)

Placental tumors are rare, the most common being chorioangioma of the placenta.[7] This tumor is clinically silent but can become symptomatic if >5 cm in size. Abnormal vascular connections with fetal vessels may lead to fetal anemia, cardiomegaly, and congestive heart failure. Being highly vascular, it can be easily detected by Doppler USG. Teratomas of the placenta are benign tumors and lie between amnion and chorion. They carry an excellent prognosis. Tissues of variable echogenicity demonstrating fat, calcifications, and fluid can be seen in radiologic investigations. It can be confused with fetus acardiacus. In the absence of fetus-like structures, it becomes difficult to identify at times. Placental mesenchymal dysplasia is a rare vascular anomaly and can be diagnosed by multicystic mass and villous hyperplasia; associated fetal growth restriction, preterm delivery, and fetal anomaly have been reported.[8] Malignant melanomas of the mother can metastasize to the placenta, as also metastasis from hematological malignancy, breast, and lung. Fetal neuroblastoma can also have placental metastasis. Both maternal and fetal metastasis are obvious by their clinical features.[7]

In our case, as no fetal structure was identified, and no vascular abnormality was seen, placental teratoma was suspected based on the USG picture. However, central bone-like echogenicity with echoic capsule raised suspicion. Magnetic resonance imaging may show mixed high-, intermediate-, and low-intensity signals, ribs, and other bony structures and vascular supply can be identified and has minimal image degradation with movement.

Spencer's criteria[8] for the diagnosis of acardiac twins are: (1) enclosed by the amnionic sac, (2) covered by normal skin. (3) anatomic parts recognizable grossly, and (4) attachment to the placenta by the umbilical cord and the presence of vascular anastomosis. Our case fulfilled the criteria of diagnosis as it was covered by amniotic sac, covered by normal skin, attachment to the placenta by the thin cord; amorphous fetus does not has a recognizable anatomic part.

Treatment for this condition if diagnosed early includes ligation or obstruction of the blood flow to the acardiac twin for a better prognosis for the pump twin. Laser coagulation gives good results if detected and managed in time.

Obstetricians and radiologists should be aware of this entity for timely diagnosis and management, as the chances for the survival of pump twin are dependent on early management. The importance of early USG and anomaly scan is again stressed upon, more so in developing countries.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal the identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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