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Case Report

Vanishing acardiac twin with TRAP syndrome: A case report ☆

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ABSTRACT

Acardiac twin is a rare complication of monochorionic twin pregnancy. We present case of a 24 years-old primigravida with monochorionic pregnancy having an amorphous acardiac twin diagnosed during routine first trimester ultrasound scan. She was managed expectantly since there were no signs of hemodynamic compromise in the normal twin with close ultrasound fetal surveillance using gray scale and color Doppler ultrasound. Spontaneous regression of vascularity with reduced size of the acardiac twin was seen subsequently.

Introduction

Acardiac twin is a rare complication of monozygotic twin pregnancies occurring in 1% of monochorionic twin gestations with an incidence of 1 in 35,000 births [1]. Due to hemodynamic dependence of the acardiac twin on the pump twin, it is crucial to monitor the pump twin for signs of decompensation so that if indicated, timely intervention can be done to interrupt the vascular supply to the acardiac twin [2]. The goal of imaging is to maximize the chances of survival of the pump-twin. Serial ultrasound surveillance of the pump twin to detect any signs of cardiovascular impairment plays an imperative role in prognostication and in deciding appropriate management strategy for the pump twin.

Case presentation

A 24 year-old primigravida lady came to our department for her routine first trimester scan. This was her first pregnancy with spontaneous conception. She gave no prior history of any miscarriage. No prior antenatal ultrasound scans were done. On ultrasound, it was found that alongside a normal fetus corresponding to 13 weeks 3 days of gestation, an amorphous mass (2.4 × 2.1 cm) having umbilical vein, fundic bubble, liver and few echogenic bony rib and vertebrae like structures were noted; however, no detectable heart like structure or internal cardiac activity was evident on color Doppler. The amorphous mass was showing surface vascularity in the region of site of attachment of umbilical cord which on spectral Doppler

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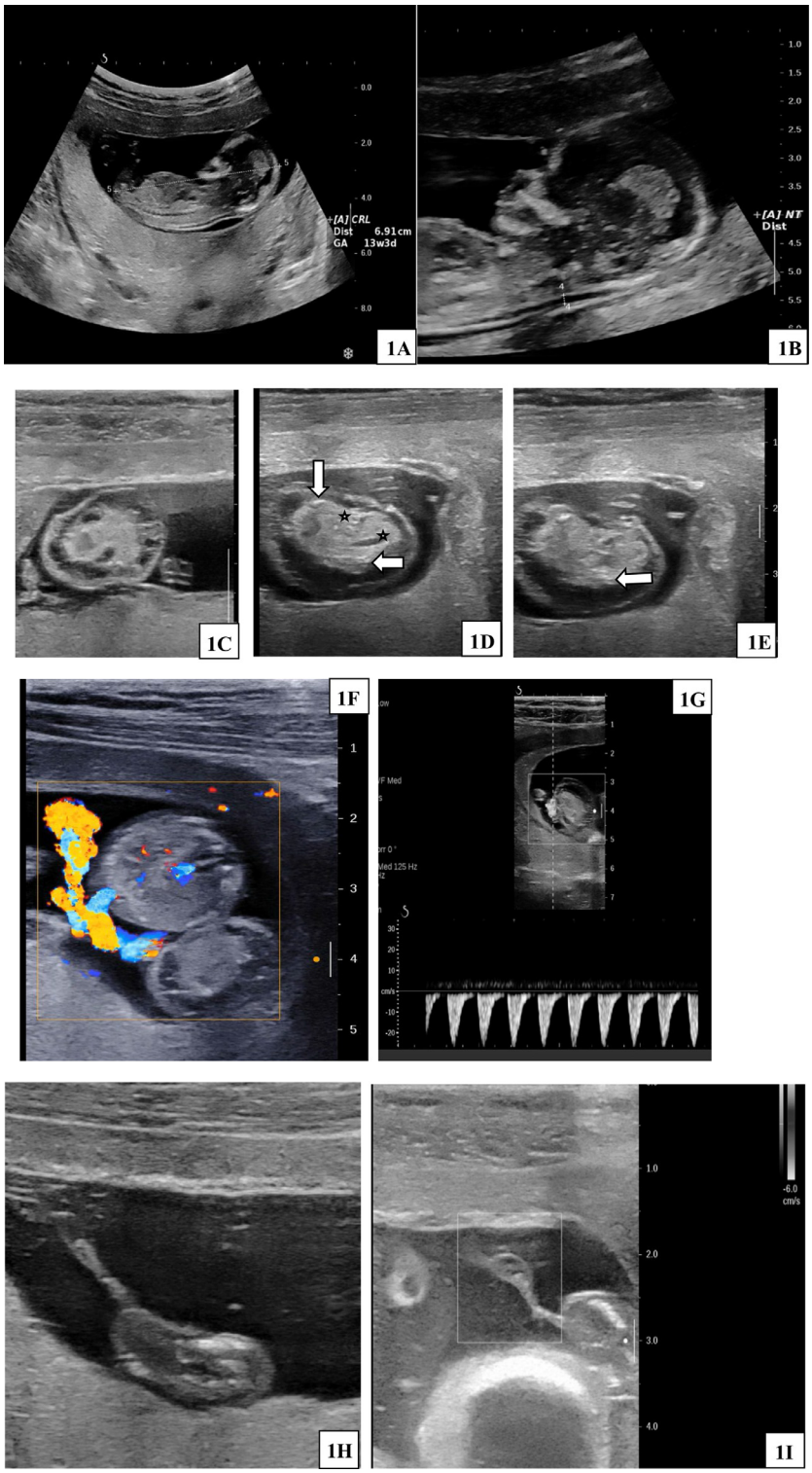

Fig. 1 – (A and B) showing normal twin with a crown rump length corresponding to 13 weeks 3 days normal nuchal translucency for gestation age. Figure (C, D, & E) showing an amorphous solid cystic mass (white arrow in figure C & D) separate from the normal twin measuring approximately 2.4 x 2.1 cm in size. Umbilical vein, liver (star in Fig. D) along with few echogenic bony structures were noted within the amorphous mass (white arrows in Fig D & E) suggesting amorphous acardiac twin. Figure (F and G) showing color and spectral Doppler images of the amorphous mass with surface vascularity near the attachment site of umbilical cord which on spectral (Fig G) Doppler showed persistent reversed flow in the umbilical artery. Figure (H and I) Subsequent USG done after 1 month showed reduction in size of the acardiac twin (1.8 x 1 cm) with absence of flow within umbilical cord on color Doppler (Fig I).

tracing showed reversal of flow in the umbilical artery. There was a single placenta with normal attachments of both the umbilical cords. The other fetus had normal nuchal translucency of 1.6 mm, a normal nasal bone length for gestation, 3 vessel umbilical cord and normal Ductus venosus Doppler parameters with no signs of hemodynamic decompensation.

A provisional diagnosis of monochorionic pregnancy with acardiac twin with twin reversed arterial perfusion (TRAP) was given and the patient was referred to dedicated fetal medicine unit for further management. The patient was managed expectantly since no ominous signs of heart failure or growth restriction were noted in the normal fetus. The patient was kept under weekly sonographic surveillance.

On subsequent ultrasound done 1 month later, no structural anomaly was noted in the normal fetus, however there was significant reduction in size of acardiac twin (1.8 × 1 cm) with no detectable internal or surface vascularity implying spontaneous demise of the acardiac twin without any intervention.

Discussion

Acardiac twin is a rare complication of monozygotic twin pregnancies occurring in 1% of monochorionic twin gestations with an incidence of 1 in 35,000 births [1]. It can be considered as the most severe form of twin-twin transfusion syndrome where abnormal vascular connections from the normal pump twin feed the amorphous acardiac twin. The normal pump twin carries a high risk of perinatal mortality due to risk of cardiovascular decompensation. There are various schools of thought for optimal management of acardiac twin pregnancies. Management options include elective termination, observation with close antepartum surveillance (serial cardiotocography, ultrasonography and echocardiography) and surgical interventions [2,3].

Antenatal diagnosis of this condition hinges on the gray scale and color Doppler findings in a monochorionic pregnancy where one of the fetuses is dysmorphic, has no cardiac activity and is being parasitically supplied by its vascular connections with the normal twin. Doppler study is essential for early diagnosis of acardiac twin so as to plan a timely and effective intervention to salvage most of the pump twins. Spontaneous cessation of blood flow in the acardiac twin is seen in 60% of cases, also seen in our case. In about 50% of these there is demise of the pump twin [4,5]. Expectant management with serial ultrasound surveillance, including fetal echocardiography, is a reasonable approach in management of these cases if there are no poor prognostic features. Delivery is indicated if signs of cardiac decompensation are noted at a viable gestational age.

The other entity in the differential diagnosis of this condition is an intrauterine fetal demise of 1 twin. Continued growth of the abnormal “presumed dead-twin” and presence of blood flow in the presumed dead twin by color Doppler favors the diagnosis of an acardiac twin. A retrograde pattern of fetal perfusion can be elicited on Doppler of the umbilical artery of the acardiac twin [6].

Presence of an acardiac twin is not always intended to risk the normal pump twin. The pump twin is, however at an increased risk for in-utero cardiac failure-necessitating close fetal surveillance for signs of decompensation. Unfavorable prognostic factors for the surviving twin include a relatively higher weight of the acardiac twin (particularly when more than 70% of the donor twin). A high resistance index in the umbilical artery of the acardiac twin is considered favorable for the pump twin [6].

In our case during the first trimester scan, surface vascularity was present near the cord attachment site of the amorphous acardiac twin with no demonstrable heart-like structure or internal cardiac activity. The umbilical artery in acardiac twin showed reversal of flow on color and spectral Doppler. On subsequent ultrasound which was done later after 1 month, the normal fetus showed no structural anomaly or any signs of hemodynamic decompensation, however the size and surface vascularity of the acardiac twin got significantly reduced- the vanishing acardiac twin. The patient was kept under weekly ultrasound surveillance. The size of the acardiac twin got significantly reduced in the subsequent scans and it became papyraceous. A healthy 2400 gm child was delivered via cesarean section at 35 weeks period of gestation.

Conclusion

Monochorionic pregnancies with acardiac twin having TRAP syndrome need close sonographic fetal surveillance. If no signs of hemodynamic compromise are seen in the normal twin during surveillance, the patients can be managed expectantly by keeping a close watch on the size and vascularity of the acardiac twin utilizing gray scale and color Doppler ultrasound.

Patient consent

Written detailed informed consent was obtained by the author from the patient included in the case report.

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