Anadolu Kardiyol Derg
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Thoraco-Omphalopagus conjoined

Torako-Omfalopaguslu yapışık ikizler

Introduction

Conjoined twins are rarely seen in medical practice, and the incidence is reported as 1 in 50.000 to 1 in 100.000 live births; 60% of these die in utero or are stillborn (1). The thoracopagus is the most common type, accounting for 40% of all conjoined twins cases (2). Thoracopagus conjoined twins have the highest incidence of cardiovascular abnormalities; they may have cardiac abnormalities in any type of fusion, and 90% of these twins share a common pericardial sac. The type of cardiac fusion is a critical factor for mortality (3).

Case Report

A 33-year-old mother presented a conjoined twin pregnancy at 35 weeks of gestation. Ultrasonographic examination had not been made until the week of the birth. Her pregnancy was spontaneous. There was no remarkable risk factor for congenital anomalies in her history. Female thoracoomphalopagus twins weighing 3000 g in total were delivered by cesarean section. The Apgar scores were 4 at 1 minute (min) and 6 at 5 min. The twins shared the thorax and abdomen (Fig. 1). Postnatal echocardiographic examination revealed that the babies shared a four-chamber heart with two ventricles and two atria (Fig. 2). The left ventricle (LV) connected to two aorta (Ao-1, Ao-2) and one pulmonary artery (PA-1) (Fig. 3, 4), while the right ventricle (RV) connected to only one pulmonary artery (PA-2) (Fig. 5). The pulmonary and systemic venous return could not be displayed. Multidetector computed tomography was planned to display the anatomical and vascular structure of the heart. However, the twins died shortly after birth due to cardiovascular collapse and progressive metabolic acidosis despite maximal inotropic and ventilatory support. The parents were informed that the infants had inseparable hearts and could not be saved. They refused the post-mortem examination.

Figure 1. Photo (A) and X-ray (B) images of the twins showing the shared thorax and abdomen

(Permissions to use the pictures of twins in publication are obtained from parents)

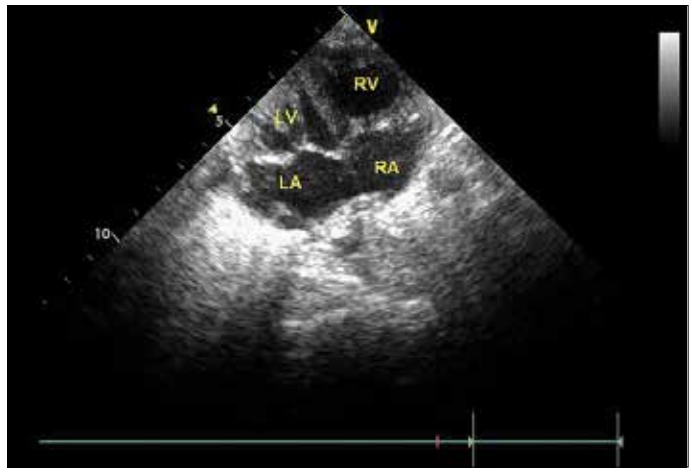

Figure 2. Echocardiogram showing a four-chambered heart with two ventricles and two atria

LA - left atrium, LV - left ventricle, RA - right atrium, RV - right ventricle

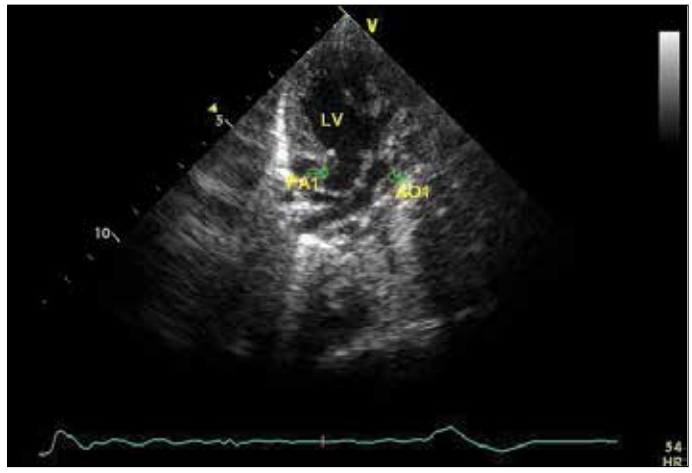

Figure 3. Echocardiogram showed aorta-1 and pulmonary artery-1 connecting the left ventricle

Ao - aorta-1, LV - left ventricle, PA - pulmonary artery-1

Discussion

Thoracopagus conjoined twins with a fused heart always have complex abnormal cardiac anatomy (4-7). The degree of cardiac fusion is most often the pivotal factor in the prognosis (6). Leachman et al. (4) classified twinning on the basis of cardiac fusion as follows: separate normal hearts (type A), fused atria with separate ventricles (type B), and fused atria and ventricles (type C). Andrews et al. (5) reported echocardiography results in the evaluation of 23 sets of conjoined twins with thoracic level fusion, and classified them into four groups as: group A: separate hearts, separate pericardium; group B: separate hearts, common pericardium; group C: fused atria, separate ventricles; and group D: atrial and ventricular fusion. Thoracopagus and thoracoomphalopagus twins have complex cardiac abnormal anatomy in both classifications. The present twins were thoraco-omphalopagus with a four-chambered heart. Three great arteries connected the left ventricle, while only one pulmonary artery connected the right ventricle. McMahon et al. (8) reported cardiac malformations in a review of 1262 cases of all types of conjoined twins. Of all the cases studied, 314 twins were thoracopagus and typically shared a single multi-chambered

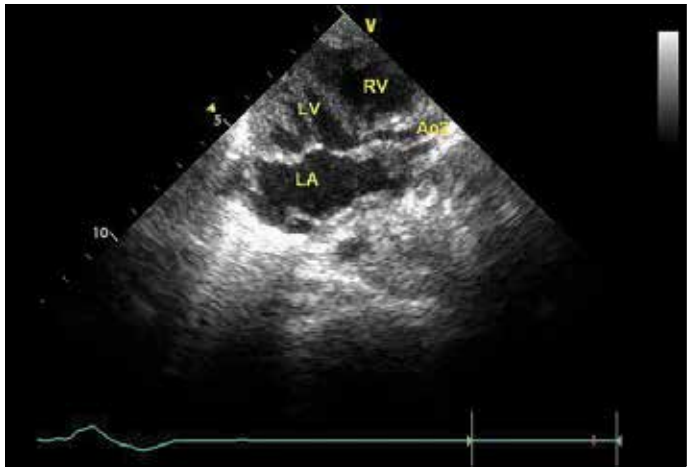

Figure 4. Echocardiogram showing aorta-2 connecting the left ventricle
Ao2 - aorta-2, LA - left atrium, LV - left ventricle, RV - right ventricle

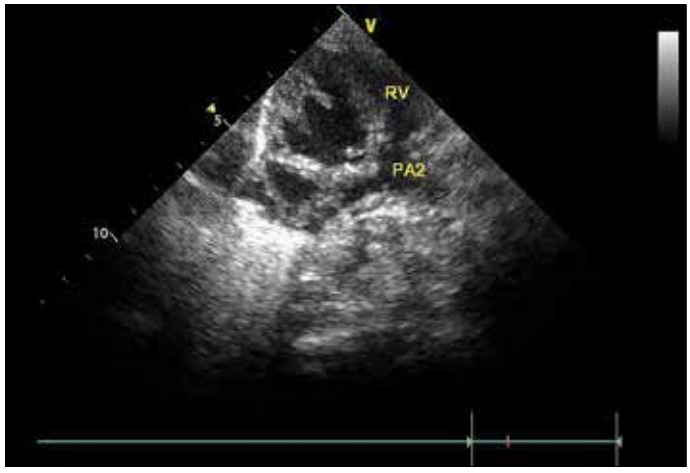

Figure 5. Echocardiogram showing pulmonary artery-2 connecting the right ventricle
PA2 - pulmonary artery-2, RV - right ventricle

heart; there were from one to four atria and from one to four ventricles, with several variations of the great vessels, and triple-outlet connection was observed in only three cases of all 1262 twins. The present case had a triple-outlet connection from the left ventricle; two aorta and one pulmonary artery were visualized as connecting the left ventricle. Spitz et al. (2) classified management of 17 conjoined twins in three distinct categories. The nonoperative management group includes twins with inseparable hearts as with our case. The present twins also had only a single four-chambered heart and triple-outlet left ventricle. This also distinguishes our case from the other cases of cardiac fusion in conjoined twins reported in the literature; however, the prognosis was not different (1-8). Our twins died on the first day due to heart failure and progressive metabolic acidosis. For inseparable twins, early diagnosis may be offered to the parents if they wish to consider pregnancy termination (9).

Conclusion

Although conjoined twins, especially thoracopagus type, are known to present several cardiac anomalies, a shared single four-chambered heart with triple-outlet left ventricle is a rare condition. The abnormalities can be determined in the fetal period with regular monitoring of the pregnancy and fetal echocardiographic examination.

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References

1. Spitz L, Kiely EM. Conjoined twins. JAMA 2003; 289: 1307-10. [CrossRef]
2. Spitz L, Kiely EM. Experience in the management of conjoined twins. Br J Surg 2002; 89: 1188-92. [CrossRef]
3. Gilbert-Barness E, Debich-Spicer D, Opitz JM. Conjoined twins: morphogenesis of the heart and a review. Am J Med Genet 2003; 120A: 568-82. [CrossRef]
4. Leachman RD, Latson JR, Kohler CM, McNamara DG. Cardiovascular evaluation of conjoined twins. Birth Defects 1967; 3: 52-65.
5. Andrews RE, McMahon CJ, Yates RW, Cullen S, de Leval MR, Kiely EM, et al. Echocardiographic assessment of conjoined twins. Heart 2006; 92: 382-7. [CrossRef]
6. McMahon CJ, Mullins CE, Vick GW 3rd, Caville CC, Bezold LI, Nihill MR. Cardiac catheterization in diagnosis and management of congenital heart disease in thoracopagus conjoined twins. Catheter Cardiovasc Interv 2000; 51: 159-67. [CrossRef]
7. Liu HC, Lo CW, Weng ZC, Hwang B, Lee PC. Various modalities for evaluation of a fused heart in conjoined twins. Pediatr Cardiol 2012; 33: 192-200. [CrossRef]
8. McMahon CJ, Spencer R. Congenital heart defects in conjoined twins: outcome after surgical separation of thoracopagus. Pediatr Cardiol 2006; 27: 1-12. [CrossRef]
9. Baldwin VJ. Pathology of multiple pregnancy. New York: Springer-Verlag; 1994. [CrossRef]

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