D-transposition of the great arteries with right-sided pulmonary hypoplasia

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ABSTRACT

Unilateral pulmonary agenesis or aplasia (UPA) in combination with congenital heart defects is rare and has not been reported in connection with transposition of the great arteries. This case demonstrated dextroposition of the fetal heart, and subsequent scans could not clearly visualize the right pulmonary artery. UPA should be considered in the workup and counseling for a family in the setting of fetal heart malposition, as there is a significant clinical impact.

Keywords: Cardiac malposition, fetal, pulmonary hypoplasia, transposition

INTRODUCTION

Unilateral pulmonary agenesis or aplasia in combination with congenital heart defects is rare and has not been reported in connection with transposition of the great arteries (TGA).

CASE REPORT

A 23-year-old woman (gravida 4, para 2) underwent a complete fetal ultrasound at 19-week gestation and subsequent fetal echocardiogram at 23-week gestation. The fetal cardiac diagnoses included dextroposition, left-sided superior vena cava (SVC), d-TGA, and ventricular septal defect (VSD) [Figure 1]. The right pulmonary veins and right pulmonary artery were not well visualized. At 38 2/7-week gestation, a follow-up ultrasound demonstrated premature atrial contractions, thickened myocardium, and decreased contractility. The woman underwent induction of labor later that day given the interval changes on the ultrasound. She delivered a female infant with a birth weight of 2510 g (<fifth percentile) and a length of 43 cm (<fifth percentile). The Apgar scores of eight at one minutes, and eight at five minutes

The postnatal echocardiogram confirmed the cardiac diagnoses of dextroposition, left-sided SVC, d-TGA, patent foramen ovale, large membranous VSD, large patent ductus arteriosus, and severely hypoplastic right pulmonary artery [Figure 2]. No right-sided pulmonary venous return was visualized. The initial postnatal chest X-ray demonstrated asymmetric hyperdensity of the right lung, hazy opacification of the right hemithorax, and rightward mediastinal shift [Figure 3]. The postnatal head ultrasound was notable for a small cystic structure in the caudothalamic groove. The renal ultrasound demonstrated mild right upper pole central calyceal dilation. Chromosomal microarray analysis confirmed a normal female karyotype.

Within the first 24 h after birth, the infant developed progressive hypoxia requiring intubation and escalation of ventilator support. The decision was made to

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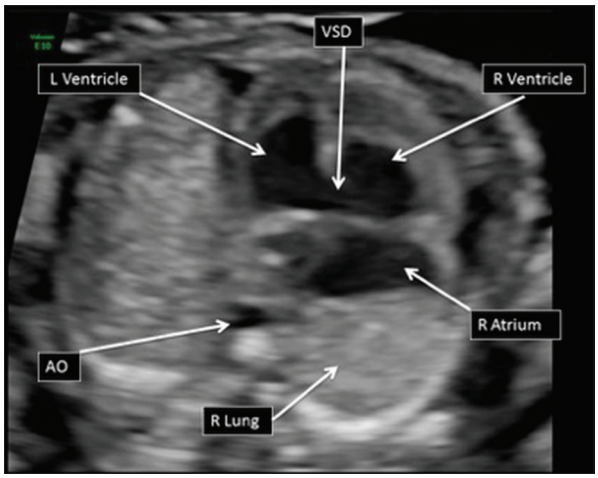

Figure 1: L Ventricle: Left ventricle, VSD: Ventricular septal defect, R Ventricle: Right ventricle, R Atrium: Right atrium, R Lung: Right lung, AO: Aorta

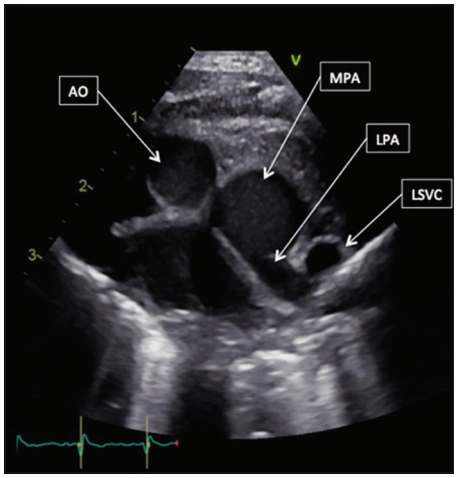

Figure 2: AO: Aorta, MPA: Main pulmonary artery, LPA: Left pulmonary artery, LSVC: Left superior vena cava

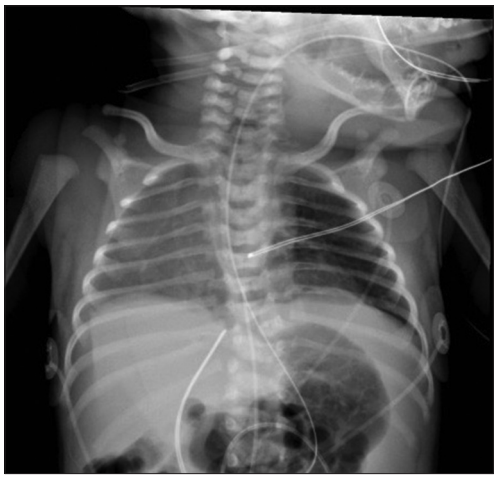

Figure 3: Anterior/posterior X-ray of a neonate. Asymmetric hyperdensity of the right lung, hazy opacification of the right hemithorax, and rightward mediastinal shift

perform a balloon atrial septostomy given the persistent hypoxia. The postseptostomy course was complicated by a right-sided pneumothorax necessitating chest tube placement, hypotension requiring inotropic support, persistent hypoxia, atrial tachycardia, and progressive lactic acidosis. Because of hemodynamic instability, the infant was unable to undergo a computed tomography angiogram for further evaluation of the cardiac and pulmonary anatomy. Given the infant's inoperable state and declining medical status, the family elected for comfort care including a terminal extubation at 10 days of age. The infant died shortly thereafter. The parents consented for an autopsy, and the findings were consistent with right-sided pulmonary hypoplasia. The right lung weighed 13.1 g and the left lung weighed 28.4 g. The right lung was markedly reduced in size and had a single lobe.

DISCUSSION

Pulmonary agenesis, where there is absence or severe hypoplasia of one or both lungs, complicates approximately 1 in 15,000 pregnancies.[1] When the hypoplasia or agenesis is unilateral and on the right, the prognosis appears to be worse, with perinatal and infant death occurring more frequently and at an earlier age.[1] Up to one-half of cases occur with associated anomalies, which can include genitourinary, cardiovascular, skeletal, or nonimmune hydrops.[2] Other anomalies or syndromes have been associated with cardiac dextroposition and right lung hypoplasia, such as diaphragmatic hernia or scimitar syndrome.[3]

While 1%–3% of d-TGA patients develop irreversible pulmonary vascular disease with subsequent pulmonary hypertension, there is no known association between d-TGA and pulmonary hypoplasia.[4] Wang *et al.* demonstrated that conotruncal defects with outflow tract obstructions had smaller main and branch pulmonary artery dimensions; however, these differences were not seen in those fetuses with d-TGA.[5] Ruchonnet-Metrailler *et al.* conducted a study on congenital heart defects and tested predictive variables such as lung weight, lung weight/body weight ratio, and pulmonary hypoplasia.[6] Lower lung weight/body weight ratios, pulmonary hypoplasia, and small pulmonary artery diameter correlated with right outflow obstructions but not d-TGA.[6]

Abnormalities of the cardiac axis and/or cardiac malposition may occur to the right (dextrocardia), midline (mesocardia), or extreme left and occur as a result of intrathoracic mass, pulmonary hypoplasia, or congenital heart disease.[7] Malposition of the heart, especially dextroposition in the absence of an intrathoracic mass, warrants further evaluation of the pulmonary anatomy and branch pulmonary arteries and veins.[3,8] Unilateral pulmonary artery atresia or

agenesis should be considered when assessing fetal cardiac deviation, especially if there are subjectively or objectively low lung volumes. Cardiac dextroposition is an indicator of pulmonary hypoplasia, and discrepancies in pulmonary artery sizes can be predictive of respiratory morbidity.[9] Unilateral lung hypoplasia can lead to pulmonary hypertension in the newborn, further complicating the postnatal clinical course.[3] Magnetic resonance imaging (MRI) has been utilized in cases of cardiac malposition to elucidate additional abnormalities after fetal echo.[10] One small study of 42 patients with an abnormal cardiac axis and/or malposition found that 23 of those cases (55%) were suspected of having lung or other extracardiac anomalies causing a shift in cardiac axis based on initial ultrasound. The addition of MRI identified 12 (29%) of them to have heterotaxy or an additional lung anomaly. Of the four patients suspected to have only an isolated cardiac anomaly on ultrasound, 3 (75%) were found to have both lung and cardiac anomalies after MRI.[7]

In a neonate who already faces cardiac anomalies requiring postnatal surgery, antenatal counseling would be impacted by discovery of a pulmonary abnormality that would further complicate the postnatal course and potentially affect surgical candidacy. A high suspicion for pulmonary hypoplasia and postnatal pulmonary hypertension should be maintained for the fetus found to have dextrorotation in the absence of an intrathoracic mass. There can be consideration for fetal MRI, especially in cases complicated by maternal obesity, oligohydramnios, or fetal lie not conducive to ideal scanning. These pregnancies should be delivered in a tertiary care center, with a multidisciplinary team caring for them including pediatric specialists for the neonate.

Declaration of patient consent

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

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Conflicts of interest

There are no conflicts of interest.

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