Dextroposition of the fetal heart secondary to enlarged thymus. Coarctation of the aorta.

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The case shows mild disproportion between the sizes of the left and right heart chambers, with the left side slightly smaller, a perimembranous ventricular septal defect (VSD) and a hypoplastic transverse aortic arch in the three-vessels and trachea view, which led to the correct diagnosis of hypoplastic arch and coarctation of the aorta.

CASE PRESENTATION

The patient is a 31-year-old, G3P2 pregnant patient who was referred for fetal MRI at 32 weeks. She underwent a fetal echocardiogram at 31 weeks that showed: 1. an apparently bicuspid aortic valve with thickened leaflets, 2. smaller left heart chambers compared to the right, 3. a moderate size perimembranous VSD, and 4. dextroposition of the fetal heart thought to be cause by a mass in the left hemithorax. Fetal MRI was requested to: evaluate the left chest mass and the aorta.

INVESTIGATION

Fetal echocardiogram was performed at 31 weeks demonstrating mesocardia/dextroposition of the fetal heart with visceral and atrial situs solitus and normally related great vessels. Atrioventricular and ventriculoarterial connections were concordant. A mass with different echogenicity from lung tissue was seen adjacent to the fetal heart on the left side. A moderate size VSD was noted. The aortic valve appeared to be bicuspid, with thickened leaflets. The distal transverse arch and aortic isthmus appear to be hypoplastic but were poorly visualized. Fetal MRI showed that the mass seen by fetal echocardiography corresponded to an enlarged thymus *(Figure 1)*. Fetal cardiac MRI confirmed disproportion between the left and right sides of the heart, the moderate size VSD *(Figure 2)* and clearly showed a hypoplastic transverse arch of the aorta in the three-vessels and trachea view *(Figure 3)*. Sagittal views of the aortic arch could not be obtained.

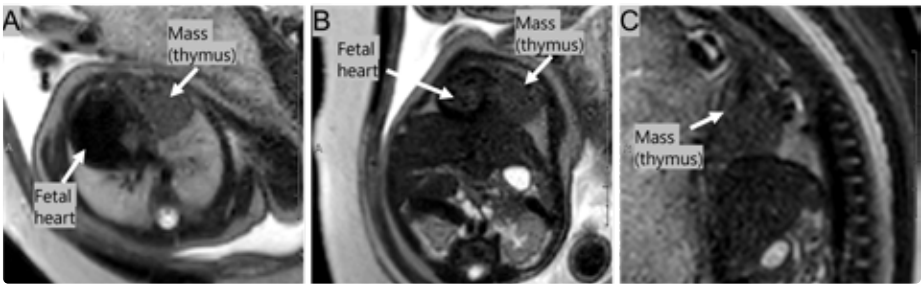

Figure 1: axial (A), coronal (B), saggital (C) T2-weighted images of the fetal chest on axial, coronal and sagittal planes show a hypointense T2 mass located in the left aspect of the anterior mediastinum, causing dextroposition of the fetal heart, with the black blood structure deviating to the right side of the chest.

DIFFERENTIAL DIAGNOSIS

Before the application of fetal MRI, the chest mass was thought to represent congenital pulmonary airway malformation (CPAM).

TREATMENT

The fetus was given prostaglandins immediately after birth to maintain patency of the ductus arteriosus and atrial septostomy to enlarge the foramen ovale. 3 weeks after birth an arterial switch procedure was performed.

OUTCOME AND FOLLOW-UP

A female baby was delivered full term via uncomplicated spontaneous vaginal delivery. Apgar scores were 8 and 9 and birth weight was 3170 grams. Moderate VSD, hypoplastic aortic arch and coarctation of the aorta were confirmed by echocardiogram, cardiac CT and surgery. The prenatal mass causing dextroposition of the fetal heart was confirmed to be the thymus by CT and surgery. Patient underwent successful surgical correction of the hypoplastic aortic arch/coarctation and underwent pulmonary artery (PA) banding. VSD being followed with planned removal of the PA band in the future and potential closure of the VSD if indicated at that time. The patient is 5 months old at the time of this report.

TAKE HOME MESSAGES

Fetal cardiac MRI was valuable in this case to better evaluate the aortic arch, which was difficult to visualize by fetal echocardiography. It correctly established the diagnosis of hypoplastic transverse arch/coarctation of the aorta which required postnatal surgical correction. A mass causing dextroposition of the fetal heart was also correctly diagnosed as an enlarged thymus.

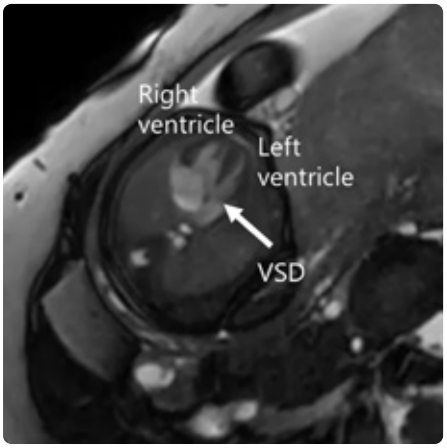

Figure 2: Axial views of the fetal heart show left < right disproportion of the cardiac chambers and a moderate perimembranous VSD

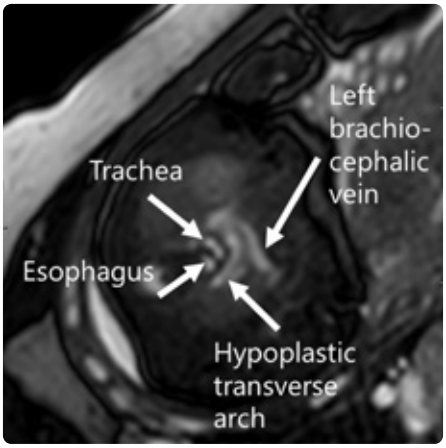

Figure 3: Axial view shows hypoplastic transverse arch, left brachiocephalic vein, TRACHEA and ESOPHAGUS