Taiwanese Journal of Obstetrics & Gynecology 57 (2018) 885–889

Contents lists available at ScienceDirect

Taiwanese Journal of Obstetrics & Gynecology

journal homepage: www.tjog-online.com

Case Report

Prenatal diagnosis of coarctation of the aorta with ventricular septal defect: A case report

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ARTICLE INFO

Article history:
Accepted 5 October 2018

Keywords:
Aorta
Fetal
Prenatal
Ultrasound
VSD

ABSTRACT

Objective: To present an accurate prenatal diagnosis of coarctation of the aorta with ventricular septal defect and to illustrate how early diagnosis in prenatal period with proper referral and counseling can optimize management.

Case report: A case with coarctation of the aorta with ventricle septal defect was found to have an abnormal three vessel view at 12 weeks, and with close follow-ups, coarctation of the aorta with ventricle septal defect was diagnosed at 24 weeks. Following the support from a multidisciplinary team that provided counseling, diagnosis, and follow-ups, the pregnant woman decided to continue with the pregnancy and had a vaginal delivery at a medical center. The newborn made an uneventful recovery after undergoing cardiac surgery on day 9.

Conclusion: The case demonstrates the role a fetal medicine team plays in diagnosing, supporting, and seamlessly transferring the congenital heart disease case from the first line obstetrician to the cardiac surgeon. A multi-disciplinary team approach was able to lead to improved perinatal outcome of the congenital heart disease case.

Introduction

Coarctation of the aorta (CoA) is a common anomaly found in 5%–8% of newborns and infants with congenital heart diseases (CHD), and is commonly associated with ventricular septal defect (VSD) [1]. CoA involves a narrowing of the aortic arch, typically located at the isthmic region, between the left subclavian artery and the ductus arteriosus [2]. It is a life-threatening CHD that is commonly undiagnosed prenatally [1,3]. Infants with such a life-threatening heart defects may not initially have symptoms, or the clinical signs may be obscured upon routine physical examination, and serious conditions may not be recognized in the majority of cases [3]. In order to overcome this challenging disease, early diagnosis in the prenatal period, along with proper referral and counseling, can optimize its management. This case report presents an accurate prenatal diagnosis of CoA with VSD, followed by postnatal confirmation and surgical repairs.

Case presentation

Prenatal findings

A 31-year-old singleton pregnant woman, primigravida, attended a first trimester Down syndrome screening at 12 weeks and 5 days (12w5d). Her past history and pregnancy history were unremarkable. The first trimester Down syndrome risk based on: maternal age, nuchal translucency, fetal heart rate, beta-human chorionic gonadotropin (beta-hCG), pregnancy associated plasma protein-A (PAPP-A), nasal bone, and tricuspid regurgitation was 1 in 12,177.

It is an established screening protocol at our clinic to go beyond the required parameters for first trimester Down syndrome

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https://doi.org/10.1016/j.tjog.2018.10.020

screening and to check the four chamber view, three vessel view (3VV), and arch views of the fetal heart, when fetal position and time allow. The four chamber view appeared to be normal, but obtaining the Doppler signal of the aorta on the 3VV was difficult. The diameter of the aorta appeared to be slightly smaller than the pulmonary artery on the 3VV. The measurement of nuchal translucency was 2.0 mm, and tricuspid regurgitation was not present.

A follow-up scan was performed at 14 weeks and 5 days. The 3VV was still abnormal, with the aorta being smaller than the pulmonary artery, and the fetal anatomical screening was otherwise unremarkable. The case was managed as a high risk case and was assigned to our perinatal nurse for coordination of consultation to relevant specialties. Serial follow-ups were arranged at two to four week intervals (Fig. 1a–c).

At 23 weeks and 5 days, routine mid-pregnancy anatomical screening was normal, except that the diameter of the aorta (LVOT) was 2.9 mm (Z score: −2.94), and the pulmonary artery (RVOT) was 5.7 mm (Z score: 1.87). A tentative diagnosis of tubular hypoplasia of the aorta was made (Fig. 2a–d).

Consultation with a pediatric cardiologist was arranged at 24 weeks and 6 days. A repeat scan on the same day revealed an additional finding of a perimembranous VSD, 2.4 mm (Fig. 3a). The measurement of the isthmus was 1.5 mm (Z score: −3.7) on the sagittal section (Figs. 3b) and 1.5 mm (Z score: −4.3) on the 3VV. The aortic annulus was 2.6 mm (Z score: −3.0). A prenatal diagnosis of CoA with perimembranous VSD was made.

In order to plan for the transfer and to relieve parents' anxiety, consultations with the neonatologist and the pediatric cardiac surgeon were arranged by our perinatal nurse. Maternal care was managed by the case's local obstetrician, while fetal care was managed by our unit throughout the pregnancy.

A follow up scan at 30 weeks and 6 days at our unit revealed that the VSD measured 2.0 mm, the isthmus was 2.4 mm (Z score: −2.4) on the sagittal section (Fig. 4) and 1.8 mm (Z score: −4.8) on the 3VV, and the measurement of aortic annulus was 3.8 mm (Z score: −2.2).

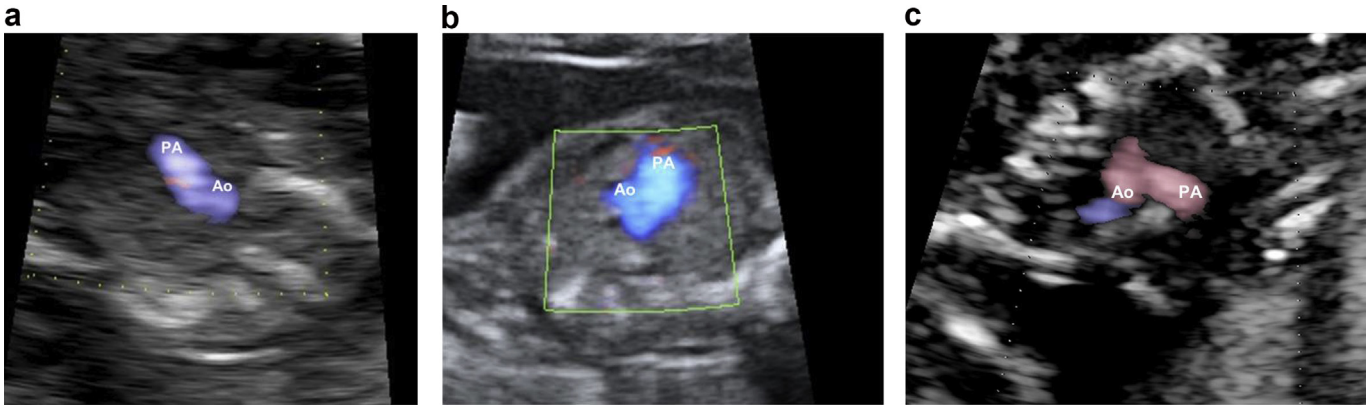

Fig. 1. a. Three-vessel-trachea view, 14 weeks and 5 days (14w5d). b. Three-vessel-trachea view, 16 weeks and 5 days. c. Three-vessel-trachea view, 18 weeks and 5 days.

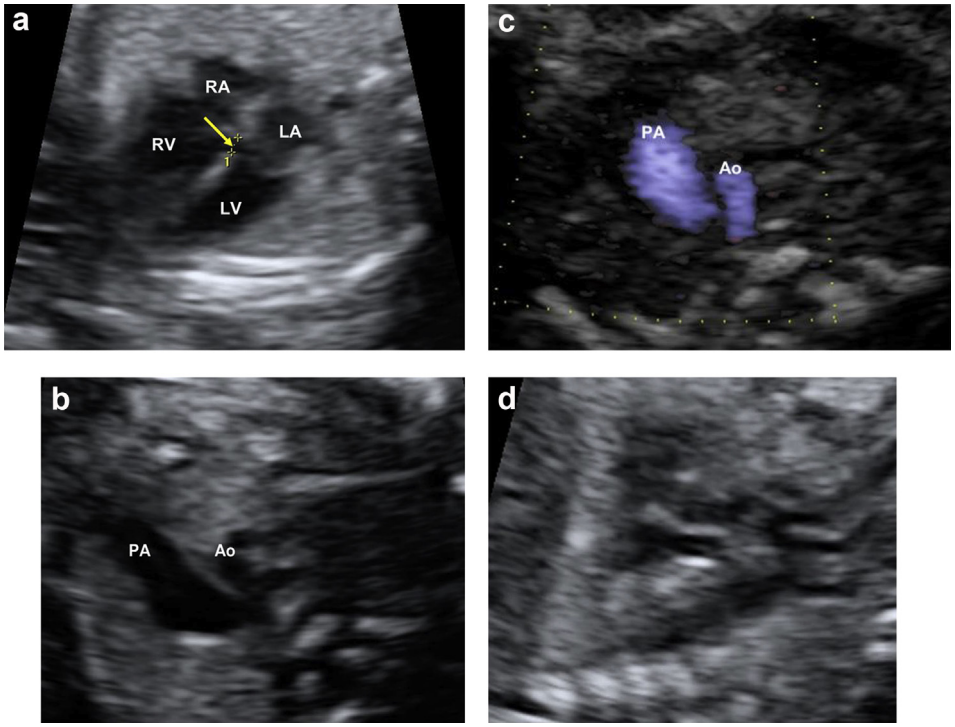

Fig. 2. a. Four-chamber view, ventricular septal defect with coarctation of the aorta, 23 weeks and 5 days. b. Three-vessel-trachea view, 23 weeks and 5 days. c. Three-vessel-trachea view, 23 weeks and 5 days. d. Longitudinal view of the aortic arch, 23 weeks and 5 days.

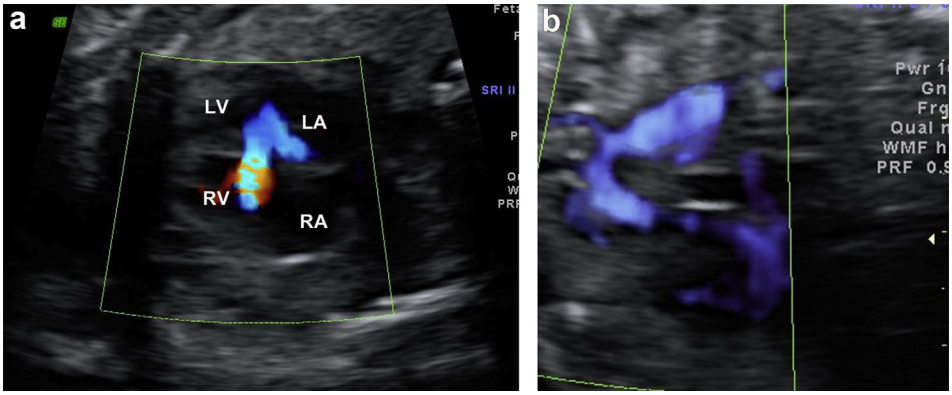

Fig. 3. a. Four-chamber view Ventricular septal defect, 24 weeks and 6 days. b. Longitudinal view of the aortic arch, 24 weeks and 6 days.

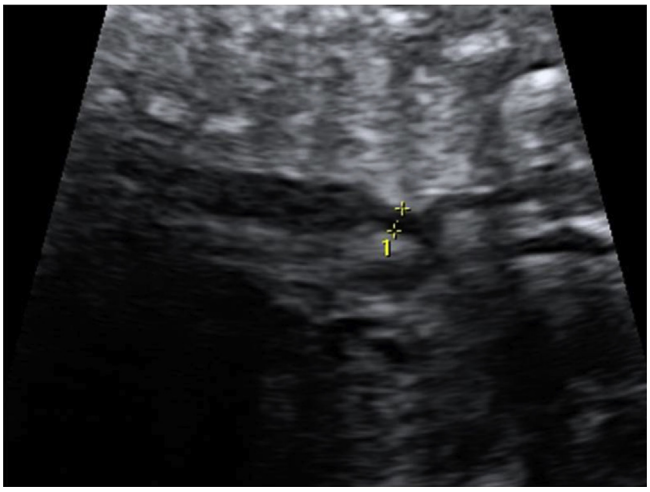

Fig. 4. Longitudinal view of the aortic arch, 30 weeks and 6 days.

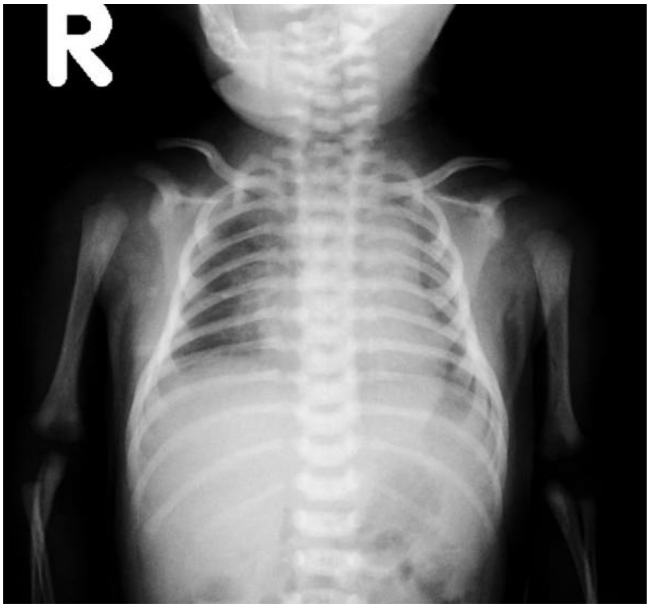

Fig. 5. First-day chest x-ray. Mild cardiomegaly.

The next repeat follow-up scan at 34 weeks and 6 days revealed that the VSD measured 3.1 mm, the isthmus was 2.7 mm (Z score: −2.5) on the sagittal section and 2.6 mm (Z score: −3.5) on the 3VV, and the measurement of aortic annulus was 4.0 mm (Z score: −3).

The case was referred to a medical center for delivery at over 37 weeks. The mother had a spontaneous vaginal delivery of a female baby at 38 weeks and 2 days, weighing 2863 g, and had Apgar scores of 8 and 10 at one and 5 min, respectively.

Postnatal findings

First-day chest x-ray showed pulmonary congestion and mild cardiomegaly (Fig. 5). Echocardiography showed CoA, a posteriorly malaligned perimembranous VSD (2.22–3.44 mm) with bidirectional shunt, small patent ductus arteriosus (PDA) (1.61 mm) with left-to-right shunt. The diameter of the ascending aorta was 4.4 mm, the diameter after the first branch was 3.7 mm, and after the second branch it was 2.89 mm. The isthmus was 1.64–2.21 mm. The pressure gradient across the isthmus was 3.97 mmHg. Prostaglandin E1 (PGE1) infusion was started soon after the neonate was born, with a dosage of 0.01 mcg/kg/min to keep the ductus arteriosus open. Follow-up echocardiography on the third day showed a PDA of 5.47 mm with bidirectional shunt under continuous PGE1 infusion, a secundum type atrial septal defect (ASD) (3.88 mm) with left-to-right shunt, dilated main pulmonary artery, and a normal left ventricle ejection fraction (79.6%) (Fig. 6a, b, 6c). Cardiac computed tomography scan showed aortic arch hypoplasia, and an enlarged PDA (6.03 mm). The diameter of the aortic isthmus was 1.2 mm, while the ascending aorta and the distal aorta measured 5.2 mm and 3.1 mm, respectively (Fig. 7a and b). The size of VSD was 3.9 mm. The postnatal diagnosis was consistent with the prenatal impressions.

Cardiac surgery

Surgery was performed on the ninth day. A single-stage operation for total correction of CoA and division of PDA and repair of VSD and ASD was performed. The operative findings further confirmed the prenatal and postnatal diagnoses.

Discussion

CoA, frequently associated with VSD, is still a major congenital heart disease threat. Prenatal diagnosis remains challenging and is affected by high false-positive diagnosis rates

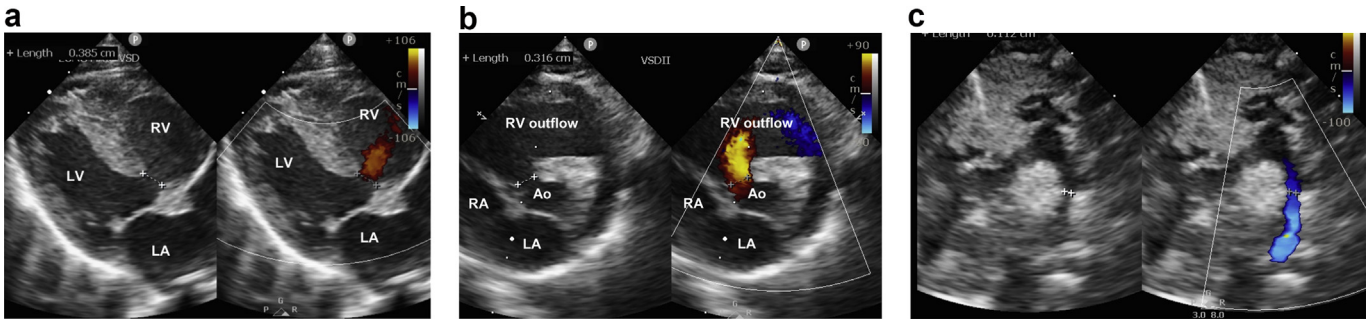

Fig. 6. a. Echocardiogram at first day. Parasternal short axis view, Perimembranous type ventricular septal defect. b. Echocardiogram at first day. Parasternal short axis view, Perimembranous type ventricular septal defect. c. Echocardiogram at first day. Suprasternal notch view.

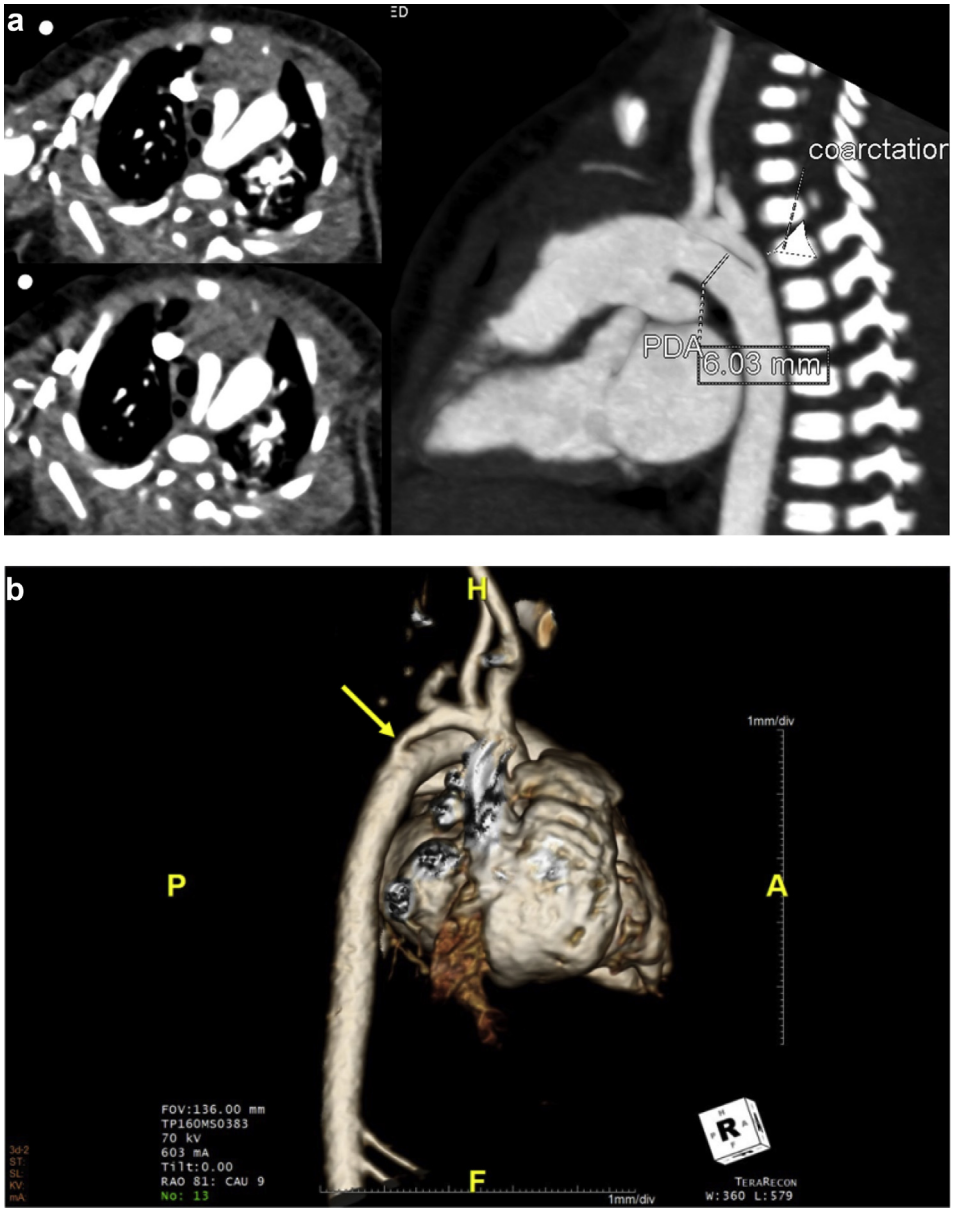

Fig. 7. a. Cardiac CT scan. Aortic arch hypoplasia and patent ductus arteriosus 6.03 mm. b. Cardiac CT scan, 3D image. Aortic arch hypoplasia.

[4]. Screening of CHD in the first trimester may be feasible but difficult. Fetal heart biometry and nuchal translucency measurements provides a chance to identify patients with high risk for CHD [5,6], who require a further detailed scan or referral. In cases prenatally diagnosed with CoA, serial follow-up ultrasound is recommended to check whether there is progression to

hypoplastic left heart syndrome [7]. Essentially, patient counseling with sufficient information and multi-disciplinary medical expert support can improve perinatal outcomes. Our case demonstrated the feasibility of first trimester screening of congenital heart disease at the time when nuchal translucency screening is performed. After confirming the diagnosis in mid-pregnancy, vertical integration was coordinated by a perinatal nurse, and collaboration among local obstetricians, fetal medicine specialists, neonatologists, pediatric cardiologists, and pediatric cardiac surgeons was achieved seamlessly. The multi-disciplinary medical team secured maternal care during pregnancy and fetal care with timely treatment immediately after birth.

Conflict of interest

The authors state that they do not have any conflict of interest.

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