Kinking of the Aorta in the Third-trimester Ultrasound Scan

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Section 2 – Answer

Case

A 40-year-old healthy woman, gravida 2, para 1 (with a vaginal delivery 7 years ago), was referred to our Obstetrics Department for ultrasound surveillance due to advanced maternal age. The first pregnancy was uneventful. Family history was unremarkable.

The first ultrasound scan was normal, but since it was performed after the ideal timing (at 16 weeks), aneuploidy screening was carried out with cell-free fetal DNA scanning, which presented low risk.

The second-trimester ultrasound scan at 20 weeks and 1 day of gestation showed a live fetus with no apparent fetal abnormalities; nevertheless, because cardiac examination was difficult, fetal echocardiography was performed at 21 weeks, which showed to be normal.

The patient was diagnosed with gestational diabetes at 26 weeks, and glycemic control was achieved with diet.

At 28 weeks, a third-trimester ultrasound scan was carried out, showing a live fetus in a cephalic presentation with an estimated fetal weight of 1392 g (90th percentile) and normal anatomy for the gestational age.

The third-trimester ultrasound scan was repeated at 34 weeks and 4 days of gestation, showing a live fetus in a cephalic presentation with an estimated fetal weight of 2480 g (50th percentile). Throughout the examination, an irregularity of the cardiac rhythm (suggestive of cardiac extrasystoles) was detected, and upon a cardiac examination, an abnormal course of the aorta was identified: a prominent kinking without frank aneurysm formation in the proximal descending aorta with no significant stenosis [Figure 1].

The fetal echocardiography at 35 weeks showed situs solitus and levocardia with normal cardiac axis and normal atrioventricular concordance. The four-chamber view revealed a left ventricle/right ventricle proportion of 1.46 [Figure 2a].

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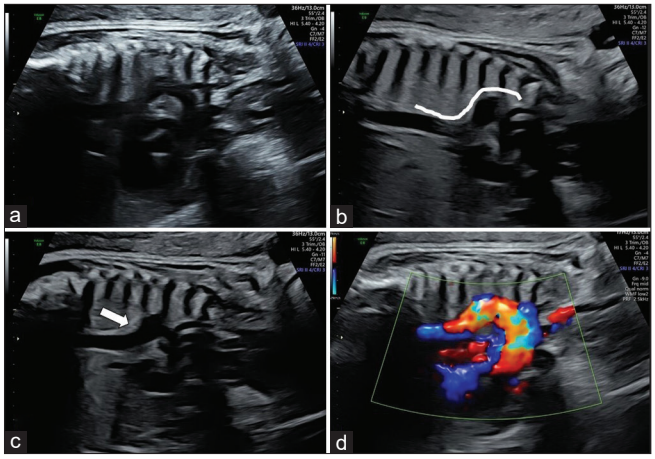

Figure 1: (a-c) A two-dimensional echocardiography sagittal view of the aortic arch at 34 weeks of gestation, showing an abnormal course of the aorta (white line) with a prominent and focal kinking (arrow) without frank aneurysm formation in the proximal descending aorta; (d) A sagittal view of the aortic arch with color Doppler revealing a slight aliasing effect in the kinking/bulking area

The ventricular outflow tract views, three-vessel view, and three-vessel and trachea views were normal. The sagittal arch view revealed an elongation of the distal aortic arch, a focal kinking at the proximal descending aorta, and an absence of significant stenosis or enlarged collateral arteries [Figure 2b]. In addition, frequent supraventricular extrasystoles were visualized. Maternal assessment was unremarkable (no cardiac or medication history and normal laboratory tests, including normal thyroid-stimulating hormone).

A healthy female newborn was delivered at 40 weeks. At 1 day of life, a transthoracic echocardiogram was performed, confirming the focal kinking described with no stenosis or collateral arteries [Figure 3].

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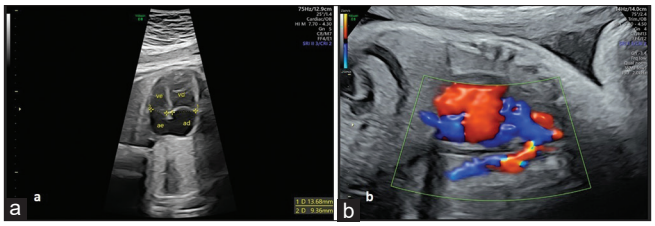

Figure 2: (a) A two dimensional echocardiography four chamber view at 35 weeks, showing a left ventricle/right ventricle proportion of 1.46 (13.68/9.36) – normal value <1.5[1]; (b) A sagittal arch view with color Doppler, revealing a focal kinking without frank aneurysm formation in the proximal descending aorta with no significant stenosis or enlarged collateral arteries and a slight aliasing effect across the lesion, and with no major hemodynamic abnormalities

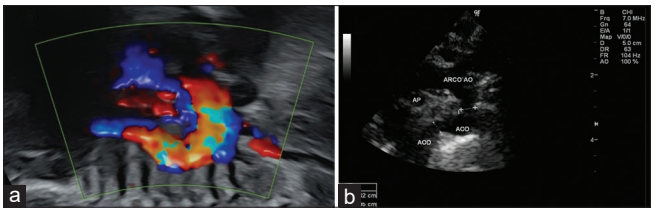

Figure 3: Comparison between prenatal and postnatal findings: A two-dimensional echocardiography sagittal arch view at 35 weeks of gestation (a) and at day 1 of life (b) showing the abnormal course of the aorta with a prominent and focal kinking in the proximal descending aorta, without aortic stenosis. ARCO AO: Aortic arch, AOD: Descending aorta, AP: Pulmonary artery

Interpretation

The findings from the fetal echocardiography were consistent with apparent aortic pseudocoarctation, which was confirmed at day 1 of life by the transthoracic echocardiogram. The follow-up after 1 and 2 months was unremarkable.

Discussion

Aortic pseudocoarctation is a very rare congenital anomaly of kinking, or buckling, of the aorta without a pressure gradient across the lesion, hence there is an absence of hemodynamic abnormalities.[2]

Its exact etiology is not well known. One proposed embryological cause is the failure of compression of the third through the seventh segments of the dorsal aortic roots and the fourth arch segment.[3]

Aortic pseudocoarctation can coexist with other congenital heart diseases, such as bicuspid aortic valve.[4]

Misdiagnosis is common, especially because it mimics true coarctation; nevertheless, it can be differentiated from it by recognizing a high, elongated arch with a kinking which lacks luminal narrowing, and unlike coarctation of the aorta, there is no obstruction of blood flow nor measurable pressure gradient across the area of redundancy/narrowing. Coarctation of the aorta is usually associated with collateral flow; however, this is absent with pseudocoarctation.[5-7]

The diagnosis of this condition usually occurs during childhood or in adults who are resistant to or have difficulty in treating hypertension, or as an incidental finding in imaging modalities.[4] To our knowledge, there are no reported cases where this condition has been diagnosed or suspected prenatally.

Echocardiography is a useful initial diagnostic tool to assess associated congenital defects. Computed tomography angiograms and/or magnetic resonance imagings of the chest are also important imaging modalities to assess the narrowed segment of the aorta and to rule out associated aortic aneurysm or dissection. Novel modalities, such as flow-sensitive cine imaging and three-dimensional imaging, are valuable and highly accurate in assessing the gradient across the constricted area.[4] Cardiac catheterization and angiography provide a decisive diagnosis for this condition.[8]

There is no published data regarding the diagnosis or management of this condition in fetal patients, and as such, from the moment of suspicion, a fetal echocardiography is recommended and a multidisciplinary discussion with pediatric cardiology and neonatology is essential to define if the delivery should be in a tertiary hospital and how the postnatal surveillance should be performed.

Pseudocoarctation has been shown to be complicated due to the aneurysm formation of the thoracic descending aorta, leading to sudden aortic rupture or dissection.[9] Therefore, close follow-up is important for asymptomatic patients without associated anomalies, while surgical treatment should be recommended for all symptomatic patients.[2]

In conclusion, aortic pseudocoarctation is a rare anomaly mimicking true coarctation, which can be suspected during routine pregnancy ultrasound scans. Hence, awareness of this condition can modify the follow-up and improve future surveillance.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the forms, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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