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CASE REPORT

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PRENATAL DIAGNOSIS OF COARCTATION OF AORTA ASSOCIATED WITH DANDY WALKER ANOMALY- A CASE REPORT

Perriyanayaki @ Barani G and Adaikappan M

Department of Radiodiagnosis, Rajah Muthiah Medical College and Hospital, Annamalai University, Annamalai Nagar – 608 002

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A B S T R A C T

Coarctation of aorta is a constriction of aortic arch that ranges from a discrete shelf like lesion within the lumen to a segmental narrowing of a portion of the arch. The characteristic triad of Dandy-Walker malformation includes complete or partial agenesis of the vermis, cystic dilatation of the fourth ventricle and an enlarged posterior fossa with upward displacement of lateral sinuses, tentorium, and torcularherophili. Here we report a case of coarctation of aorta associated with Dandy walker anomaly diagnosed prenatally with antenatal ultrasound and fetal echocardiography.

INTRODUCTION

Coarctation of aorta is a constriction of aortic arch that ranges from a discrete shelf like lesion within the lumen to a segmental narrowing of a portion of the arch. This obstructive lesion may reduce the blood flow in the fetal aortic arch, leading to arch hypoplasia, although in some cases this may only be clinically evident after birth[1-3]. It can be classified as simple when it occurs without important intracardiac lesions and complex when it occurs in association with significant intracardiac defects.

The characteristic triad of Dandy-Walker malformation includes complete or partial agenesis of the vermis, cystic dilatation of the fourth ventricle and an enlarged posterior fossa with upward displacement of lateral sinuses, tentorium, and torcularherophili. This triad is typically found in association with supratentorial hydrocephalus, which should be considered a complication rather than part of the malformation complex.[4,5] Here we report a case of coarctation of aorta associated with Dandy walker anomaly diagnosed prenatally with antenatal ultrasound and fetal echocardiography.

Case Report

A 26 year old primigravida was referred at 22 weeks of gestational age to the Department of Radiodiagnosis with the diagnosis of posterior fossa malformation. The prenatal sonographic findings showed oligohydramnios with a large posterior fossa in communication with the fourth ventricle, upwardly rotated vermis and elevated tentorium, normal lateral ventricles measuring 8mm on right side and 7mm on left side at the level of atrium of lateral ventricle, normal third ventricle suggestive of Dandy walker anomaly. Corpus callosum appears normal. The fetus also showed left talipesequinovarus deformity and bilateral mild renal pelvis dilatation measuring 9 mm on both sides. Four chamber view of heart showed discrepancy in the size of ventricles with disproportionately small left ventricle. Hence detailed fetal echocardiography was done to rule out the presence of structural cardiac anomaly.

Fetal echocardiographic findings are as follows:

- Abdominal situs and cardiac situs were normal.
- The fetus had normal heart rate with regular rhythm. The cardiac size and axis were normal with concordant venoatrial, atrioventricular and ventriculoarterial connections.
- Four chamber view revealed a disproproportionately small and hypoplastic left ventricle with larger appearing right ventricle. Other cardiac chambers were normal. Outflow tract views shows smaller left ventricular outflow tract and proximal aorta. Three

Corresponding author:* **Perriyanayaki @ Barani G
Department of Radiodiagnosis, Rajah Muthiah Medical College and Hospital, Annamalai University, Annamalai Nagar – 608 002

vessel view shows narrowing of aorta when compared with the pulmonary artery. Persistent left sided superior vena cava is also noted draining into the coronary sinus.

- Colour Doppler also showed disproportionately small left ventricle with narrowing of aorta and persistent left sided SVC.

Parental counselling was given describing the condition of the fetus. The parents did not want to continue the pregnancy and opted for termination. Induction of labour done and pregnancy was terminated. The baby's parents refused for doing fetal autopsy and hence not done.

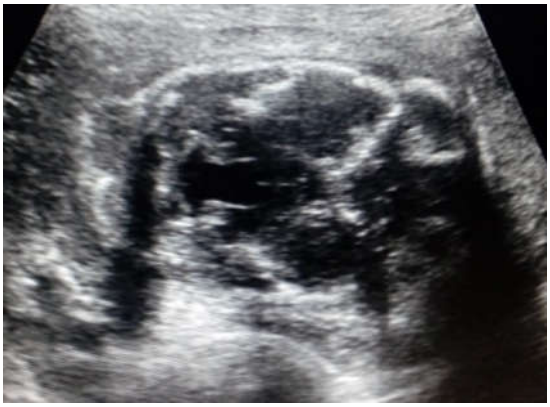

Figure 1. Shows posterior fossa communicating with the fourth ventricle

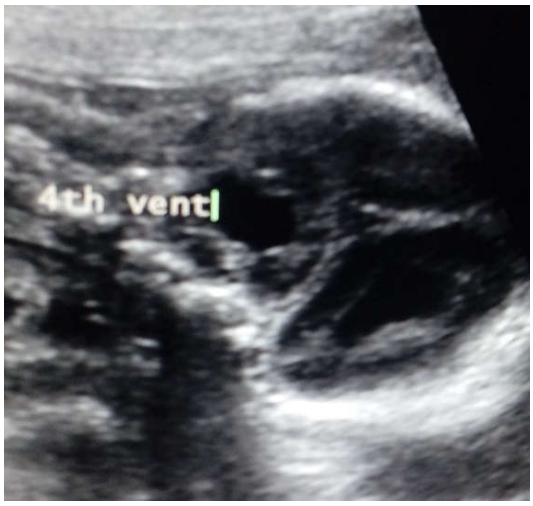

Figure 2. Sagittal image shows large posterior fossa with communication to the fourth ventricle.

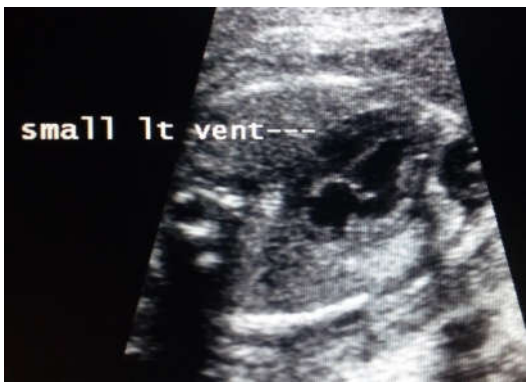

Figure 3. Four chamber view shows disproportionately small left ventricle when compared to right ventricle

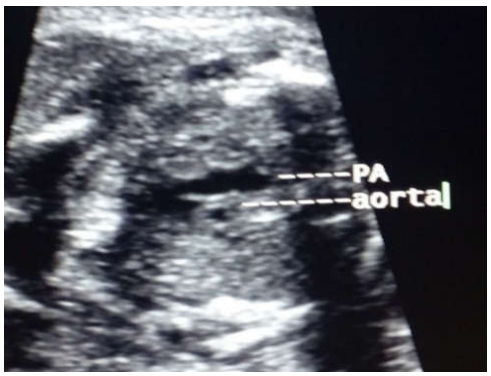

Figure 4 Three vessel view shows narrowing of aorta when compared to pulmonary artery and persistent left sided svc draining into coronary sinus.

DISCUSSION

Coarctation of the aorta is the fifth most common congenital heart disease, accounting for 6–8% of live births with congenital heart disease, with an estimated incidence of 1 in 2,500 births[6,7]. It affects male babies more than female[9].It is a narrowing of a segment of the aortic lumen along the aortic arch, which results in an obstruction to blood flow. Most commonly, this narrowing is located between the origin of the left subclavian artery and the ductus arteriosus. The severity ranges from slight narrowing of the distal end of the arch to severe hypoplasia of the entire arch.

Three types of coarctation are described depending on the location of the aortic narrowing in relation to the ductus arteriosus

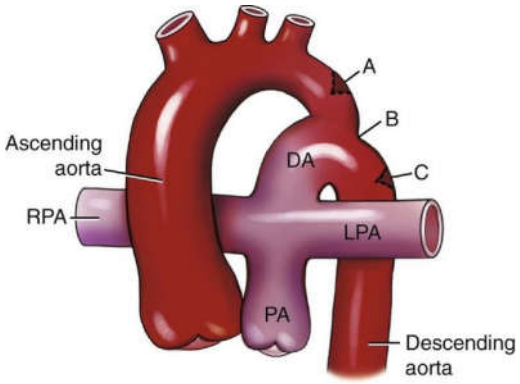

Figure 5. Diagram illustrating the different types of coarctation of the aorta. *A,* Preductal; *B,* ductal; *C,* postductal; *DA,* ductus arteriosus; *LPA,* left pulmonary artery; *PA,* main pulmonary artery; *RPA,* right pulmonary artery.

- Preductalcoarctation: narrowing seen proximal to the ductus arteriosus
- Ductal coarctation: narrowing seen at the level of the ductus arteriosus
- Postductalcoarctation: narrowing seen distal to the ductus arteriosus.

Preductal coarctation accounts for approximately 2% of all coarctations and are seen most commonly in infants and are commonly associated with other intracardiac abnormalities. Preductal coarctations occur early in embryological development and are thought to result from

decreased blood flow through the left side of the fetal heart.

Ductal and postductal coarctations account for the remaining 98% of coarctations. They are usually an isolated finding, but may be associated with aortic valve abnormalities. Ductal and postductal coarctations occur as a result of the presence of abnormal muscular-ductal tissue. This type of coarctation occurs after birth when the ductus arteriosus closes.

Sonographic findings in fetus with COA

Ventricular disproportion- Suspicion for coarctation of the aorta usually arises when there is ventricular disproportion in fetus with a smaller left ventricle than the right ventricle. It is important to compare the z-scores to ensure that the right ventricle is of normal size, and the left ventricle is smaller[10]. The left ventricle mid-cavitary dimension during systole to right mid-cavitary dimension remains an important clue. LVmc/RVmc< 0.6 has shown a sensitivity of 70%, a specificity of 67% and a positive predictive value of 73% for neonatal intervention for critical coarctation of aorta.

Discrepancy of the great vessels- Aortic coarctation is almost always associated with a discrepancy of the great vessels where the diameter of the pulmonary artery is bigger than the diameter of the aorta. This is probably related to blood flow redistribution because of the increased resistance of the left ventricular outflow tract. Slodki et al. (2009)[11] investigated the utility of analyzing prenatal mediastinal measures of the great arteries in the third trimester by measuring the diameters in the three vessel and trachea view. They found that the main PA: Ao diameter ratio can be a helpful tool for distinguishing true from false Coarctation of Aorta. Rizzo et al. (2010)[12] proposed to measure the sizes of both pulmonary artery and aorta in the three vessel view by using four dimensional sonography with spatiotemporal image correlation (STIC). They also found that PA: Ao ratio was significantly higher in fetuses with CoA compared to those with a normal heart. The three vessel and trachea view allows comparison of the aortic arch and the ductal arch and assessment of the fetal isthmus. Isthmus/ductal diameter, isthmus/ductal angle and z-scores are important measurements to be made when coarctation is suspected.

Aortic arch - Hypoplasia of the aortic arch affects the proximal arch, most commonly between the left common carotid artery and the left subclavian artery or the isthmus, and may extend into the brachiocephalic vessels. Sonographically a small aorta at the level of the valve is present in most fetuses due to hypoplasia of the isthmus and transverse arch. Sagittal view of the arch may be normal. Hypoplasia of the transverse arch and isthmus may be detected in 80-100% of cases when adequate images of the distal arch can be obtained[13].

Color Doppler may demonstrate normal flow in the aorta with normal velocities, increased or decreased velocities distal to the coarctation[14], retrograde flow proximal to the coarctation[15],high velocity jet that is present within the narrowed segment or just proximal to it and turbulent flow.

Doppler flow has been shown to be a useful tool in assessing retrograde blood flow through the foramen ovale and the isthmus.

When there is a persistent left superior vena cava, it is important to keep in mind the association with aortic coarctation[15].

Associated anomalies*:* In 35%–45% of cases, Coarctation of Aorta is an isolated heart defect, but in the remaining 55%–65% of cases, other cardiac anomalies (Ventricular Septal Defect, aortic and mitral valve anomalies, bicuspid aortic valve, transposition of the great arteries, and Double Outlet Right Ventricle) are associated. The presence of multiple left-sided obstructive cardiac lesions associated with coarctation of the aorta has been referred to as the Shone syndrome. The presence of a persistent left sided superior vena cava has also been reported in association with coarctation of the aorta in a certain number of cases. Extra cardiac malformations are frequently associated with Coarctation of Aorta in 25%–35% of cases. These include urinary, gastrointestinal, central nervous system, and facial anomalies. Risk of chromosomal anomalies is high, reaching 30%. They are mainly monosomy X, but also trisomies 21 and 18 and 22q11 microdeletion can occur.

Differential Diagnosis - The marked prevalence of the right heart sections over the left on the four-chamber view, present in the critical form of coarctation, should be differentiated from hypoplastic left heart syndrome (HLHS) and from total anomalous pulmonary venous connection (TAPVC). However, the former is characterized by a sealed or atretic mitral valve, which is mainly normal in coarctation. In addition, color Doppler confirms normal left ventricular filling in diastole and thus differentiates aortic coarctation from HLHS. As far as TAPVC is concerned, the recognition of a single pulmonary vein entering the left atrium on color Doppler rules out this diagnosis. On the aortic arch views (transverse and longitudinal), severe coarctation should be differentiated from interrupted aortic arch (IAA). In the latter case, the aortic arch cannot be demonstrated and a malalignment VSD is almost always associated.

The Dandy-Walker malformation is a term which represents not just a single entity, but several abnormalities of brain development which coexist. This disorder is a congenital brain malformation typically involving the fourth ventricle and the cerebellum which was first described in 1914 by W Dandy and K Black fan and was designated as Dandy-Walker syndrome in 1954 by C Benda, who also reported familial occurrence[16]. The Dandy-Walker malformation has an estimated prevalence of about 1:30,000 live births with a slight female preponderance and is responsible for 4-12% of infantile hydrocephalus[17].

Dandy-Walker malformation is frequently associated with other intracranial anomalies such as corpus callosal agenesis, holoprosencephaly, occipital encephaloceles and ocular abnormalities. Extra-cranial anomalies include polycystic kidneys, cardiovascular defects, polydactyly and cleft palate. Postnatal studies indicate that the incidence of associated malformations ranges between 50 and 70%.

Dandy-Walker malformation is recognized sonographically by an enlarged posterior fossa, cystic posterior fossa mass communicating with the fourth ventricle and varying degrees of vermian hypoplasia/agenesis. In this condition, borderline to overt ventriculomegaly and other neural/extra neural defects are generally present. Distinction between Dandy-Walker variant and megacisterna magna in the fetus is difficult as definitive criteria have not been firmly established. The former condition should be suspected when a thin communication is found between the fourth ventricle and the cisterna magna, the latter when the cisterna magna has a depth greater than 10 mm[18]. In the midtrimester, a cisterna magna measuring more than 10 mm with an 'open' fourth ventricle should alert to the possibility of Dandy-Walker complex. The transcerebellar view, that is recommended as a part of the standard sonographic examination of the fetal brainwill not detect all cases of the Dandy-Walker complex. The available experience suggest that a standard exam may not reveal even severe forms of the Dandy Walker complex.

CONCLUSION

Even with good visualization of the fetal aortic arch, prenatal diagnosis of isolated coarctation is very difficult. However, when associated with tubular hypoplasia of the entire arch, the ultrasound diagnosis is less difficult and can be reasonably made during the prenatal period. Sonographic findings possibly suggesting the presence of coarctation include direct and indirect signs. Indirect signs are seen on the four-chamber view, the out flow views and on the three-vessel view. The right ventricular-to-left ventricular width ratio has been reported as 1.69 in fetuses with Coarctation of Aorta and 1.19 in normal fetuses. Direct signs of CoA, when possible, can be found by assessing the three-vessel and trachea view. On this plane, the ductal arch appears significantly larger than the aortic arch. The transverse part of the aortic arch is severely reduced in size, especially if tubular hypoplasia of the whole arch is present. On the longitudinal view of the aortic arch, the narrowing is usually located at the isthmus; if tubular hypoplasia of the whole arch is present, the entire transverse arch is elongated and narrow, and the left subclavian artery appears to arise at the junction of the ductus arteriosus with the descending aorta. Z-scores for the measurement of the size of the aortic isthmus, the transverse arch and the angle between the isthmus and the ductus arteriosus were proposed to improve the accurate description of this anomaly. On color Doppler, a complete or partial inversion of the flow in the distal part of the arch and/or in the ascending aorta and a left-to-right interatrial shunt can be seen. However, it should be recalled here that prenatal diagnosis of Coarctation of Aorta remains challenging, with a high rate of false positive and false negative diagnosis. Four-dimensional echocardiography may be used to assess the discrepancy in the size of the aortic and ductal arches.

Antenatal sonography allows a definitive diagnosis of only the severe anatomic varieties of the Dandy-Walker complex, those characterized by both a large posterior fossa cystic mass and a wide defect in the cerebellar vermis, referred to as classic Dandy-Walker malformation. It is difficult to solve prenatally the doubt of either a mega-cisterna magna or a small inferior defect of the vermis as in Dandy-Walker variant and this can only be resolved by post natal imaging studies.

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