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Case Report

Fetal Vascular Rings: Beyond The Anomalies of The Aortic Arch

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2. Key words

Ductus Arteriosus (DA), Right Ductus Arteriosus (RDA), Aberrant Right Subclavian Artery (ARSA), Left Aortic Arch (LAA), Sd.Di George.

1. Abstract

Different anomalies related to the inappropriate development of the ductus arteriosus or the aortic arch have been described, in some cases accompanied with chromosomal or morphological anomalies, and also being able to form a vascular ring that can compromise the postnatal life. For this reason, a complete fetal anatomy examination and cardiovascular study is needed to discard possible other malformations or ultrasound markers for fetal syndromes. A detailed prenatal diagnosis of the type of ductal arch anomaly and possible vascular ring can give us a postnatal prognosis and help pediatricians with the management of symptomatic neonates.

The abnormalities related to the position or branching of the ductal arch (ductus arteriosus) are very uncommon. We describe the first case reported of prenatal diagnosis of right ductus arteriosus, left aortic arch and aberrant right subclavian artery associated with 22q11 deletion.

3. Introduction

The fetalductus arteriosus (DA)is a vascular structure with functional importance in the fetal circulation. It connects the Main Pulmonary Artery (MPA) or its left branch to the aorta, diverting blood flow apart from the pulmonary vascular bed to the systemic circulation.

In the model of embryonic vascular development, six pairs of aortic arches communicate the ventral and dorsal aortas. The ductus arteriosus arisesfrom the sixth aortic arch. In a normally developed heart, the normal branching of the left aortic arch is, from right to left: brachiocephalic trunk (rising right common carotid and right subclavian), left common carotid and left subclavian artery [1]. In normal cardiac development, both right aortic and ductal arches regress at 4-7 gestational weeks [2] (Figure 1).

Different anomalies related to theinappropriate development of the aortic and ductal arches have been described, with a prevalence of 1-2% [3,4]. Arch anomalies refer to a variety of congenital abnormalities that are related to the position or branching of the aortic and/or ductal arches. Aortic arch anomalies are most common. However, other uncommon abnormalities of position/branching of the ductal arch may occur (right or double DA, unusual, tortuous or S-shaped ductus). Scarce data is published in the literature about right ductus arteriosus (RDA)[2-15]. Most of the papers describe RDA associated to Right Aortic Arch (RAA) or in the context of Congenital Heart Disease (CHD)[2]. The aim of this study is to report a new case of an uncommon type of RDA with Left Aortic Arch (LAA) without a major CHD and to review the literature.

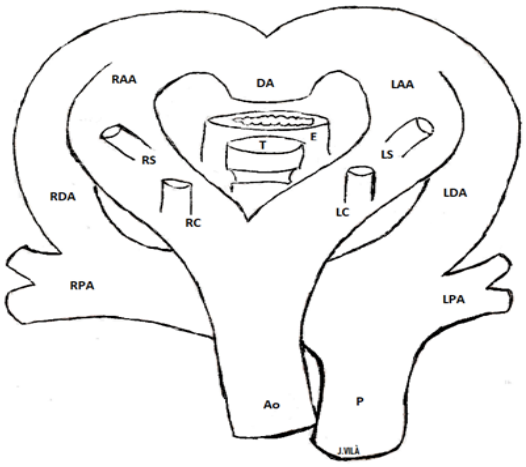

Figure 1: Hypothetical double aortic arch model of Jesse E.Edwards. Ao, ascending (ventral) aorta; DA, descending (dorsal)aorta; T, trachea; E, esophagus; LAA, left aortic arch; LC, left commoncarotid artery; LPA, left pulmonary artery; LS, left subclavianartery; P, main pulmonary artery; RAA, right aortic arch; RC, rightcommon carotid artery; RPA, right pulmonary artery; RS, rightsubclavian artery.

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4. Case Report

Patient of 35 years old, nulliparous, attended our hospital to pregnancy follow-up. First trimester scan was normal and combined test was at low risk of aneuploidy.

At the second trimester scanning (21+2w), an Aberrant Right Subclavian Artery (ARSA) was suspected. In the three-vessel and tracheaview (3VT) a complete vascular ring around the trachea and thymus hypoplasia were detected. Due to the suspicion of congenital heart disease, a detailed echocardiography was performed. The results were as follows: situs solitus, atrioventricular-ventriculoarterial connections and cardiac axis and size were normal. At 3VT view, right-sided ductus arteriosus was detected arising from the right pulmonary artery to the aortic isthmus of the ascending aorta located at the right side of the trachea, with a left-sided aortic arch and trachea in-between, having a U-shaped or ribbon-shaped confluence instead of V-shaped and the normal left aortic arch at the left of the trachea (Figures 2, Figures 3). The thymus was hypoplastic at 3VT view. ARSA was confirmed and RDA formed a complete vascular ring, described by some authors as the cross ribbon sign [12]. The rest of cardiac anatomy was normal.

Due to the diagnosis of ARSA, RDA and thymic hypoplasia, and invasive test was offered. An amniocentesis was performed at 27 weeks. Results revealed a normal karyotype with a 22q11 microdeletion at the microarray (Sd. DiGeorge). A 2940g fetus was delivered vaginally at term. The newborn presented an episode of neonatal hypocalcaemia and mild dysphagia that solved correctly. A postnatal echocardiography was performed confirming the LAA

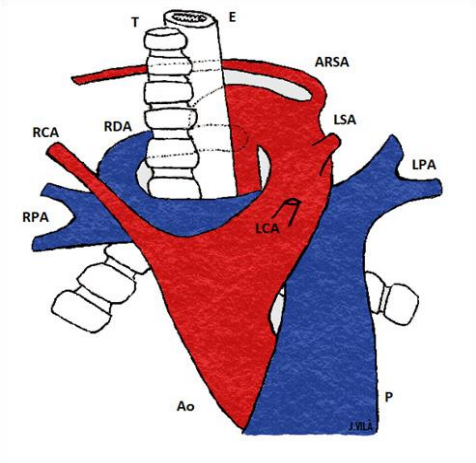

Figure 2: Developmental model of left aortic arch with right ductus arteriosus (RDA) and aberrant right subclavian artery (ARSA).Ao, ascending (ventral) aorta; DA, descending (dorsal)aorta; T, trachea; E, esophagus; LAA, left aortic arch; LC, left commoncarotid artery; LPA, left pulmonary artery; LS, left subclavianartery; P, main pulmonary artery; RAA, right aortic arch; RC, rightcommon carotid artery; RPA, right pulmonary artery; RS, right subclavian artery; ARSA, Aberrant right subclavian artery.

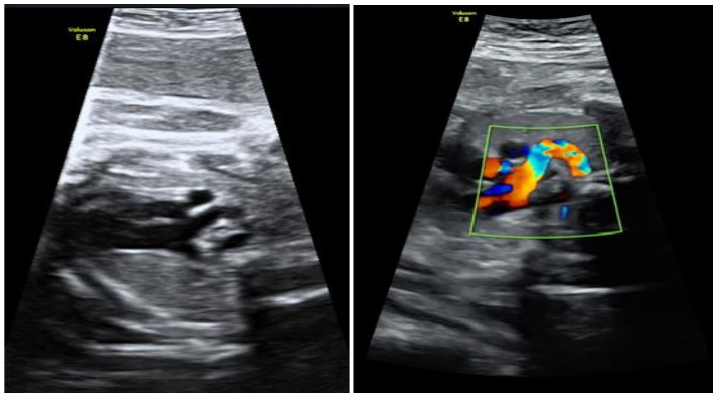

Figure 3: (A) Transversal thoracic 3VT view: Bifurcation of pulmonary artery with the origin of a complete vascular ring formed by the right DA. (B) Same view using colour doppler. R, right; l, left.

and a correct closure of the RDA. The 18-month old child is completely normal with no cardiovascular or respiratory symptoms.

5. Discussion

Anomalies of the DA can be diagnosed prenatally (DA aneurism, absence DA), including abnormalities in DA branching [16,17].

RDAwith a LAAcould be suspected at the 3VT view, by a complete vascular ring described by some authors [12] as the cross ribbon sign. This vessel is a vascular structure that originates in the bifurcation of the MPA or in the origin of the right pulmonary artery. RDA crosses right to and behind the trachea and joins the descending left aorta. The persistence of RDA with LAA forming this vascular ring is very uncommon and rarely reported. Bronshtein et al. [2] reported a prevalence of 0.035%.The very low detection rate of this anomaly might be due to: (1) unawareness of the existence of this anomaly (2) difficulty of its diagnosis in the second trimester of pregnancy [2].

When a RDA is suspected, we should perform an echocardiography [13, 14] to exclude other congenital heart diseases and vascular rings. The examination must include the 5 transverse planes described by Yagelet al.[18]. Most recently a new sonographic view is suggested to the fetal heart exploration, particularly when a congenital heart disease or arches anomalies are suspected [4]: the subclavian artery view. In case of ductal/aortic arch anomalies, in the 3VT color image, Doppler demonstration of the usual blue or red 'V-sign' is replaced by a variety of patterns, depending on the etiology of the anomaly [19].

Differential diagnosis is important because arches anomalies can present similar patterns in the 3VT image. We should consider the relationship between the aortic and ductal arches with the trachea and the possibility of a vascular ring (complete or incomplete). Dif-

ferent combinations might occur: RAA with left ductus arteriosus (LDA), RAA with RDA, double aortic arch or LAA with RDA, as in our case. In the cases of RDA with LAA, 3VT view is completely abnormal. RDA with RAA, however, needs more attention and it could be easily overlooked: RAA with RDA form a V-shaped configuration like the normal fetal anatomy, but on the right side of the trachea [13].

3D/4D ultrasound can be very useful for a more precise diagnosis in case of vascular rings. In this case, 4D clarified the relationship between the vascular structures (Figures 4, Figures 5).

In addition, a complete fetal anatomy examination is needed to discard other malformations or ultrasound markers for fetal syndromes.

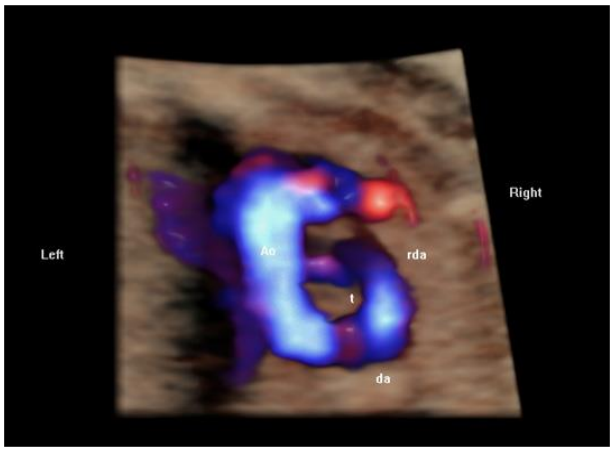

Figure 3: Transversal thoracic view, upper mediastinum using STIC technique: left aortic arch forming a complete vascular ring with the right ductal arch (cross ribbon sign). Ao, aorta; t, trachea; da, descending aorta; rda, right ductus arteriosus.

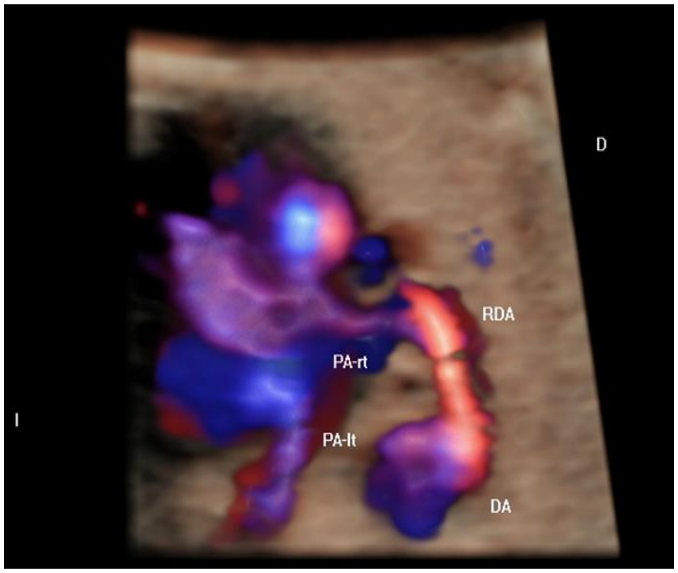

Figure 5: Transversal thoracic view, upper mediastinum: Bifurcation of the pulmonary artery, with the formation of the right DA originating in the right pulmonary artery and ending in the aortic isthmus. DA, descending aorta; RDA, right ductus arteriosus; PA-rt, right pulmonary artery; lt-PA, left pulmonary artery; r, right; l, left.

RDA with LAA is a very uncommon anomaly. We could find no reference to it in the main prenatal diagnosis textbooks and we found only 3 reports about RDA and LAA in the on line literature (Table 1). The lack of literature might be due to the rarity of this anomaly and also to the difficulty of its diagnose. A few more reports describe the association of RDA with RAA [2,13] diagnosed prenatally. All cases of RDA in pediatric literature are associated to other cardiac malformations [5, 8]. Despite of the limited literature, we would suspect that most cases of RDA are isolated, and remain undetected. We believe this because RDA is diagnosed after delivery only when it remains patent, and this occurs manly in association with CHD or other pathologic states.

To our knowledge this is the first case of prenatal diagnosis of RDA, LAA and ARSA associated with 22q11 deletion. DiGeorge Syndrome is associated with anomalies of the cardiac outflow tracts. In this case, DA was displaced to the right. Classically the anomalies of the aortic arch are associated with congenital heart defects, chromosomal anomalies, especially trisomy 21 or deletion 22q11 [20,21], but DA abnormalities are seldom mentioned.

In our case, the child had mild symptoms of compression after delivery but surgery was no needed. There isno consensus between pediatricians an obstetricianregardingesophagus or trachea compression symptoms. According to prenatal series most of the vascular rings are asymptomatic after delivery, with the exception of DAA and some cases of RAA with ARSA [22,23]. However, pediatricians report that between 30 and 60% of cases of anomalies in aortic arch and ARSA can cause obstructive symptoms due to an extrinsic compression of the esophagus and trachea [24,25]. It is described significant tracheal compression in asymptomatic infants with a RAA and left DA, considering an early airway investigation even in this type of patients[26]. Another sign recently described that should be taken into account is the high incidence of progressive stenos is in aberrant left sub clavian artery (ALSA) with RAA in asymptomatic newborns [27]. In our case, the RDA was associated to an ARSA that might worsen the prognosis.

6. Conclusion

In conclusion, ductal abnormalities are uncommon in prenatal life. Prenatal diagnosis of RDA is feasible and its diagnosis provides several benefits. Making a detailed prenatal diagnosis of the type of ductal arch anomaly and possible vascular ring can give us a postnatal prognosis and help pediatricians with the management of symptomatic neonates. Arch anomalies, aortic or ductal, can be either isolated or associated to other anomalies, commonly congenital heart diseases, and chromosomal abnormalities, including

22q11 deletion, as in our case. Based on this association invasive testing should be highly considered prenatally.

7. Aknowledgements

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