

Prenatal Diagnosis of Isolated Right Aortic Arch: A Case Report

Emre Ekmekci*, Emine Demirel and Sefa Kelekci

Department of Obstetrics and Gynaecology, Faculty of Medicine, Izmir Katip Celebi University, Turkey

Abstract

Detection of aortic arch anomalies prenatally is rare. Right aortic arch is a subgroup of aortic arch anomalies. In this paper, we report an isolated right aortic arch case that is detected in the 22nd week of pregnancy. Prenatal diagnosis is crucial for prenatal genetic counselling and approach in neonatal life.

Keywords: Aortic arch; Right sub-clavian artery; Left sub-clavian artery; Ultrasonography

Introduction

Aortic arch anomalies are various and most common are; right aortic arch (RAA); double aortic arch (DAA); circumflex retroesophageal aortic arch or left aortic arch with an aberrant right subclavian artery (ARSA) [1]. Two types of right aortic arch anomalies are defined: mirror-image branching type and aberrant left Subclavian Artery (LSCA) type[2]. Incidence of right sided aortic arch is reported as 1/1000 in a low risk pregnancy population [3]. The diagnosis of RAA is primarily based on imaging of three vessels-trachea (3VT) plane at fetal cardiac examination on ultrasonography. In normal foetal cardiac and aortic anatomy, the aorta arises from the left ventricle upward to the right, forms aortic arch, and runs backward to the left passes by the trachea through the diaphragm. The first branch of it is innominate artery, which divides into the right common carotid and right subclavian arteries, whereas the second branch is left carotid artery; and the third is LSCA. In normal, at 3VT plane, the superior vena cava, ascending aorta, and main pulmonary artery line up from right to the left. The trachea is located right and posterior to the aorta [4]. Diagnosis of aortic arch anomalies are based on the abnormalities at the arrangement of the great vessels on 3VT plane.

Case Report

A 31-year-old, gravida 3, parity 1, abortion 1 pregnant woman underwent prenatal routine second mid-trimester ultrasonography in the 22nd week of her pregnancy for fetal anatomic scan. Her pregnancy has continued uncomplicated until now and there was no prominent feature in her history. First trimester screening was normal and nuchal translucency was in normal limits. At sonographic fetal examination fetal anatomy is evaluated normal and fetal biometrics was compatible with gestational age. Four chamber views is defined normal, left ventricular and right ventricular output were defined normal at fetal cardiac examination. At 3VT plane aorta was located at the right side of trachea. Arrangement of the vessels was pulmonary artery, trachea, aorta, superior vena cava from left to the right (Figure 1). In color Doppler, U shaped vascular ring is visualized around the trachea (Figure 2). No other abnormality is detected on cardiac examination beside right sided aortic arch. Patient is informed about the risks about fetal karyotype abnormality and fetal Di George Syndrome. So cordocentesis is recommended to the patient for prenatal invasive testing. Cordocentesis is performed and karyotype was normal. 22q11.2 deletion is not detected. Patient is informed about probable problems in neonatal life. Pregnancy continued until term without any complication and at the 39th week of gestation a 3650 gr, 52 cm, healthy, male infant baby is delivered by cesarean section due to previous cesarean section history. Baby did not have any respiratory problem, stridor or dysphagia problem in neonatal life. Baby is being followed by pediatrics now.

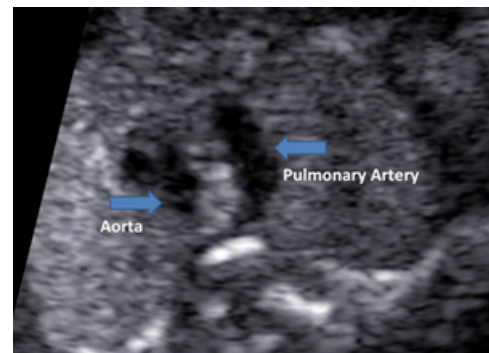


Figure 1: Arrangement of great vessels and trachea on gray scale sonography. Trachea is located right to the pulmonary artery and it's between aorta and pulmonary artery (abnormal arrangement).

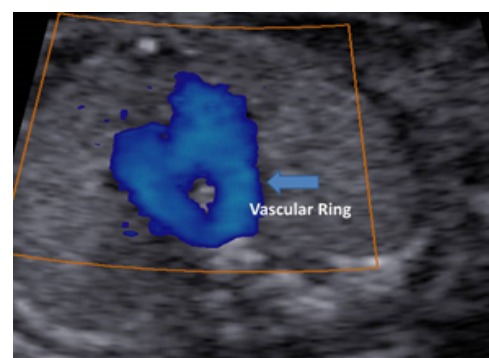


Figure 2: Vascular ring and compressed trachea on Doppler sonography. Colour Doppler shows the compressed trachea between left ductus arteriosus and aorta.

Discussion

The abnormalities of aortic arch can be proved better with the basic embryological development aortic arch, which is defined by Edwards. In this theory, ascendant aorta divides into two aortic arcus and joins

***Corresponding author:** Emre Ekmekci, Department of Obstetrics and Gynaecology, Faculty of Medicine, Izmir Katip Celebi University, Turkey, Tel: +90 232 329 3535; E-mail: dr.ekmekci@hotmail.com

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again at the descending aort level in the embryonic period. Right and left common carotid arteries and subclavian arteries come from each right and left aortic arches. Also right and left ductus arteriosus arise from each pulmonary arteries and join to each arches afterwards each subclavian arteries. In normal development, right aortic arch regresses at the distal level of right subclavian artery. And normal left sided aortic arch develops. Abnormalities in this regression results with aortic arch abnormalities [5,6]. If left aortic arch regresses at the distal level of left subclavian artery, right ductus arteriosus persists and left one regresses. Left subclavian and carotid arteries arise as the first branches of aort and goes to left. This arrangement results mirror-image branching of right aortic arch and this is often associated with other cardiac abnormalities (90%). This association is only 10% in aberrant left subclavian artery [7]. At aberrant left subclavian artery with right aortic arch, left aortic arch regresses at the level the between left subclavian artery and left carotid artery right ductus arteriosus regresses and left one persists. This arrangement results with right aortic arch, left persisted ductus arteriosus and a U shaped vascular ring around the trachea. Rarely aberrant left subclavian artery arises from directly descending aorta by a branch called Kommerell's diverticulum [8].

We detected right sided aortic arch on grey scan sonography at 3VT plane. Then we have detected U shaped vascular ring around the trachea on colour Doppler sonography. Ductus arteriosus was surrounding trachea from the left side and the right aortic arch from the right side. So our patient was aberrant left subclavian artery type of RAA. Even right aortic is an isolated finding on sonography fetal invasive karyotyping is necessary for excluding the karyotyping abnormalities like trisomy 21 and microdeletion at 22q11.2. Associated cardiac anomalies are more common for right aortic arch at mirror-image branching than left aberrant subclavian artery (U shaped vascular ring) [9]. Associated cardiac anomalies with right aortic arch are Tetralogy of Fallot, common arterial truncus, absent pulmonary valve syndrome, tricuspid atresia and double outlet right ventricle. The risk for microdeletion 22q11 is higher if right aortic arch is associated with a conotruncal cardiac anomaly. Rarely it can be associated with extracardiac anomalies [10,11]. Our our patient was aberrant left subclavian artery type and we did not detect any associated cardiac anomaly. Berg et al detected 2 associated cardiac anomalies of 22 patients with aberrant left subclavian artery type of RAA in their series. However, they detected associated cardiac anomalies at all of 23 patients with mirror-image branching type. Conotruncal malformations have a clear association with 22q11 deletion syndromes (CATCH-22). Rauch et al. showed that in patients with conotruncal malformations, the anomalies of the subclavian arteries are the most important anatomical marker for the presence of monosomy 22q11, independent of the laterality of the aortic arch. We performed cordocentesis for karyotyping and 22q11.2 microdeletion. Karyotype was normal microdeletion is not detected. Aortic arch anomalies may cause clinical problems due to tracheobronchial compression, esophageal compression. Compression

to these organs usually is associated with ring shaped vascular anomalies. Most associated vascular ring forms are double aortic arch, right aortic arch with U shaped ring and Kommerell's diverticulum [12]. In summary, aortic arch anomalies can appear in various types in prenatal sonography. Although they don't have a significant morbidity if isolated, they can be associated with abnormal karyotype and microdeletion at 22q11.2. So obstetricians should be rigorous for them at 3VT plane at fetal ecocardiography.

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