CASE REPORT

Anomalous Origin of the Right Coronary Artery From the Pulmonary Artery and Mitral Regurgitation In a Newborn

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Anomalous origin of the right coronary artery (RCA) from the pulmonary artery (PA) is a rare congenital anomaly, and only 28 cases have been reported in the pediatric age group. We describe the case of an infant who had progressive mitral regurgitation and papillary muscle dysfunction in association with anomalous origin of the RCA from the PA. The diagnosis was made by color flow Doppler, confirmed by angiography, and the case was successfully corrected by reimplantation of the anomalous RCA to the aorta. This is only the second case of anomalous origin of the RCA from the PA diagnosed in infancy without an associated congenital anomaly of the heart and great vessels.

Key words: Anomalous right coronary artery, Neonatal mitral regurgitation, Papillary muscle dysfunction, Color Doppler diagnosis, Congenital heart disease

Anomalous origin of the RCA from the PA is a rare congenital anomaly. To our knowledge, only 61 cases of such anomalous coronary arterial origin have been reported in the medical literature and 28 of them are in the pediatric age group. This is the only reported case of an infant that presented with isolated mitral regurgitation and papillary muscle dysfunction in association with anomalous origin of the RCA from the PA.

Case History

A full term 2800g, 3day old Caucasian female infant was evaluated for a heart murmur. Prenatal and birth history were unremarkable and she was asymptomatic. The patient had a grade 2/6 soft systolic murmur, best heard at the cardiac apex and the rest of the cardiovascular examination was normal. The electrocardiogram and chest radiography were also normal. An echocardiogram showed mild mitral regurgitation with a left ventricular end diastolic diameter (LVEDD) of 1.7cm and a shortening fraction (SF) of 40%. Follow-up echocardiogram at the age of 6 months demonstrated moderate to severe mitral regurgitation, increased echogenicity of the papillary muscles, a LVEDD of 2.9 cm and a SF of 43%. Careful examination of the RCA origin was deceiving with 2-D imaging, yet with color flow Doppler the true origin of the RCA was determined (Figure 1). Up to this age, all follow up ECG's were normal and the girl remained clinically asymptomatic.

Cardiac catheterization at 8 months of age revealed a Qp:Qs of 1 the left ventricular end-diastolic pressure was 11mmHg and pulmonary artery pressures were normal. Angiography demonstrated a normal origin of the LCA, with retrograde filling through collaterals of a dilated RCA and connection of the RCA to the main pulmonary artery (Figure 2). The infant underwent successful transsection and reimplantation of the anomalous RCA to the ascending aorta one month later. After a post operative follow-up period of 6 months the infant was asymptomatic and with

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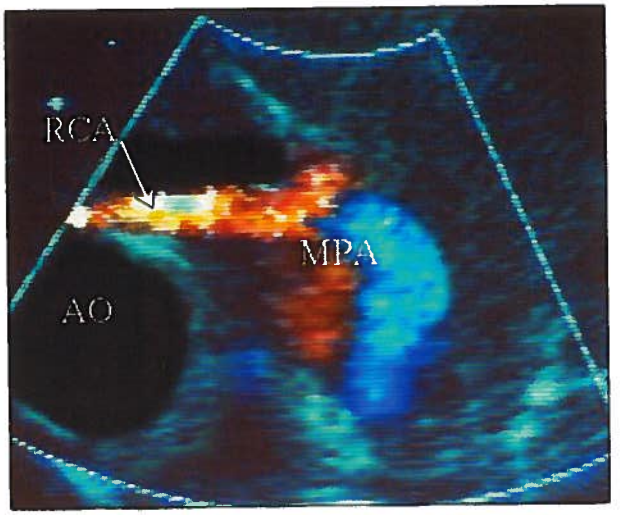

Figure 1. Parasternal short axis scan at the level of aortic and pulmonary valves. Color Doppler shows retrograde flow from the right coronary artery into the main pulmonary artery. AO= aorta; MPA= main pulmonary artery; RCA= right coronary artery.

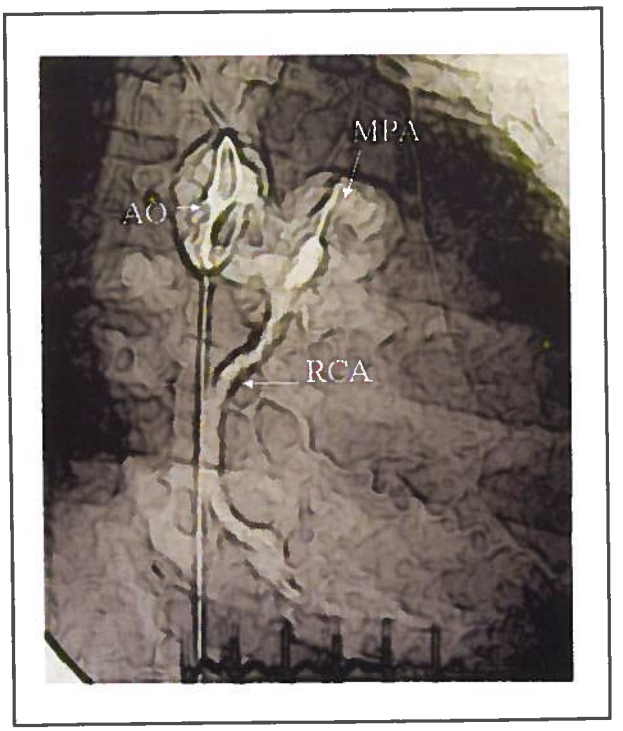

Figure 2. Retrograde aortic root angiogram. There is retrograde filling via collaterals of the dilated anomalous right coronary artery with eventual filling of the MPA. AO=aorta; MPA=main pulmonary artery; RCA= right coronary artery.

normal growth and development. Evaluation with color flow Doppler echocardiography showed only trivial mitral regurgitation.

Discussion

Congenital coronary artery anomalies account for approximately 1% of all congenital cardiac malformations. Anomalous origin of the RCA is extremely rare, as it accounts for only 5% of all coronary artery anomalies (1). Anomalous origin of the RCA from the PA was first reported by Brooks in 1885 as an incidental autopsy finding(2). Since then, there have been several case reports of patients with anomalous origin of the RCA from the pulmonary artery although mostly in the adolescent and adult population (3-5). To diagnose an isolated anomalous origin of the RCA from the pulmonary artery within the first months of life is very unusual. Vairo et al.(3) described a 2 months old infant who presented with early congestive heart failure due to anomalous origin of the RCA from the pulmonary artery not associated with a congenital anomaly of the heart or great vessels. Our patient is the second case of isolated anomalous origin of the RCA from the PA diagnosed so early in infancy.

Unlike anomalous origin of the left coronary artery from the pulmonary artery, most patients with anomalous origin of the RCA from the PA are asymptomatic. Nevertheless, there have been reports of patients with anomalous origin of the RCA from the pulmonary artery who had syncope, congestive heart failure, myocardial infarction, and even sudden death (4,5). The coronary steal phenomenon, where coronary blood flow is diverted from the myorcardium to the anomalous coronary artery, has been proposed as the possible mechanism for these symptoms (5,6). Our patient presented with mitral papillary muscle dysfunction at six months of age. We were able to document progressive mitral regurgitation, increasing echogenicity of the mitral papillary muscle and the development of intercoronary collaterals without any electrocardiographic sign of ischemia.

Associated congenital cardiac defects are described in up to 40% of patients with abnormal origin of the RCA (5). Aortopulmonary window is the most common malformation associated with abnormal origin of the RCA. Tetralogy of Fallot, double outlet right ventricle (Taussig-Bing type) and isolated valvular defects have also been associated with abnormal origin of RCA (5,7-9).

The diagnosis of anomalous origin of a coronary artery is usually made in symptomatic patients by either angiography or 2-D echocardiography with color flow Doppler. Our case was misleading, since with 2-D echocardiography alone the RCA appeared to connect to

either the aorta or pulmonary artery with minimal change in the angulation of the transducer. The application of color flow Doppler was essential in establishing the diagnosis, which was later confirmed by angiography.

As a general rule, surgical correction is recommended for this cardiac malformation, even when the patient is asymptomatic(1,5). The main reason to correct this malformation is to prevent the coronary steal phenomenon, which may cause ischemic myocardial damage(5,6). Although in some cases corrective surgery has failed to improve the clinical symptoms of the anomaly (5), our patient showed significant improvement of her mitral insufficiency six months after surgery.

Resumen

El origen anómalo de la arteria coronaria derecha de la arteria pulmonary es una cardiopatía congénita rara. Sólo se han reportado 28 casos en la edad pediátrica con este defecto. Se describe el caso de un infante con esta cardiopatía que se presentó con insuficiencia mitral progresiva desde el periodo neonatal y más tarde disfunción de los músculos papilares. El diagnóstico de origen anómalo de la arteria coronaria derecha se obtuvo mediante el uso de ecocardiografía con Doppler a color y luego fue confirmado por angiografía aórtica. Se implantó quirúrgicamente la arteria coronaria anómala a la aorta con resultados excelentes. Este es sólo el segundo caso informado en la literatura médica donde ésta anomalía coronaria sin otra cardiopatía asociada logra ser diagnosticada en la temprana infancia.

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