Anomalous origin of the right coronary artery from the pulmonary artery combined with aortopulmonary window: a case report

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Introduction

Anomalous origin of the right coronary artery from the main pulmonary artery is a rare congenital cardiac malformation. We report a 13-day-old patient presenting with anomalous origin of the right coronary artery from the pulmonary artery with aortopulmonary window. We connected the aorta with the anomalous ostium, using closure of aortopulmonary window with the help of autologous pericardial patch.

Key words: Anomalous origin of the right coronary artery, aortopulmonary window, pericardial patch

Case Report

A 7-day-old male newborn was referred to the division of Pediatric Cardiology of the Baskent University Hospital for the assessment of a cardiac murmur. The infant was 3300 gr. The electrocardiogram showed sinus rhythm and right ventricular hypertrophy. Transthoracic echocardiography revealed good biventricular function, a 3.5-4 mm wide APW and anomalous origin of the right coronary artery from the pulmonary trunk. Cardiac catheterization was performed prior to surgical intervention and demonstrated mildly increased pulmonary pressures. Aortography revealed anomalous origin of the dominant right coronary artery from the pulmonary trunk and a large APW (Figure 1,2).

Surgical correction was performed with the cardiopulmonary bypass support, moderate hypothermia and cold antegrade crystalloid cardioplegic arrest. After longitudinal pulmonary arteriotomy, a 4-mm-wide APW and ARCAPA were identified. The anomalous
coronary ostium situated below the APW was derivated to the aorta through intrapulmonary baffle of autologous pericard. The pulmonary artery was closed primarily.

Total cardiopulmonary bypass time was 52 minutes. Weaning from extracorporeal circulation was unproblematic. The postoperative course was uneventful. The patient was discharged in an asymptomatic condition and control echocardiographies showed good right and left ventricular function, no residual shunt and no stenosis of the ascending aorta and pulmonary artery.

The right coronary artery blood flow was in normal range in the follow-up period of six months.

Discussion

APW with ARCAPA is a rare but important lesion (1-4). No genetic associations or environmental risk factors are known. The 2 competing embryologic theories are that APW is part of a spectrum of conotruncal abnormalities, which includes truncus arteriosus at one end of the spectrum, and that APW is unrelated to truncus arteriosus because the lesions associated with each defect are so dissimilar (5,6). Most patients remain asymptomatic because of the presence of well oxygenated blood in the pulmonary trunk owing to the left to right shunt through the APW (4,7). However, there are some cases presenting with angina, myocardial infarction, heart failure and sudden cardiac death (2,3,8). Our case was also asymptomatic and referred for assessment of a cardiac murmur. Because of the simultaneous filling of both great arteries, preoperative angiographic diagnosis could be difficult. Injection of the contrast into both the great arteries is the best way to diagnose APW and to determine the relationship between pulmonary and coronary arteries and the defect (4,8,9).

Echocardiography is often the only imaging obtained prior to the repair of many cardiac lesions. In our case initial diagnosis was also made by transthoracic echocardiography. This demonstrates that coronary artery evaluation should be performed at every new echocardiographic evaluation (7).

In general, a corrective operation is recommended in this malformation even for asymptomatic patients. To correct the aortopulmonary defect through a ligature in association with this coronary anomaly led to intraoperative problems that did not permit the use of this technique. Different techniques were employed for connection of the anomalous coronary ostium with the aorta. One of them is reimplantation of the right coronary artery from the pulmonary artery to the aorta (4).

In our case, we connected the aorta with the anomalous ostium, using APW with autologous pericardial patch as reported elsewhere (10,11).

Anomalous origin of the coronary arteries may be seen as isolated or associated with other cardiac defects, in particularity with conotruncal abnormalities. When a conotruncal abnormality is diagnosed, anomalous origin of the coronaries should always be considered. Exact preoperative diagnosis is very important for surgical correction, and it must be done early to prevent the development of irreversible pulmonary vascular disease.
References


