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CASE REPORT

Anomalous origin of the right coronary artery from the pulmonary artery. Two case reports

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Abstract Anomalous origin of the right coronary artery originating from the pulmonary trunk (ARCAPA) is a rare but potentially fatal anomaly. We are presenting two cases of ARCAPA and reviewing the main previous published data on this lesion. The first patient presented at the age of 5 months with respiratory distress and severe chest infection. He was found to have heart murmur and cardiomegaly on chest X ray. Echocardiographic and angiographic data confirmed an ARCAPA associated to a large malalignment ventricular septal defect and distal pulmonary artery aneurysms. He underwent surgical closure of the ventricular septal defect and reimplantation of the ARCAPA on the aorta with good result.

The second case is an 11 year old male patient, complaining of dizziness and chest pain on exertion. Echocardiographic and angiographic data confirmed ARCAPA associated to a valvular pulmonary stenosis. He was operated on successfully. He got also direct reimplantation of the anomalous coronary artery on the aorta and a pulmonary valve commissurotomy.

An anomalous origin of the right coronary artery is a rare condition but may lead to myocardial ischemia and sudden death. Diagnosis is mainly made by echocardiography and confirmed by conventional coronary arteriography. Operative correction is the appropriate treatment for an anomalous coronary artery arising from the pulmonary trunk.

1. Introduction

Anomalous origin of the right coronary artery originating from the pulmonary trunk (ARCAPA) is a rare but potentially fatal anomaly. It can be isolated or associated to various other congenital heart defects. Different modalities are available to aid in establishing the diagnosis, including echocardiography, angiography, computed tomography coronary angiography and magnetic resonance angiography. We are presenting two cases of ARCAPA and reviewing the main previous published data on this lesion.

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1.1. Case n°1

A 5 month old boy was admitted to pediatric intensive care unit for severe respiratory tract infection with congestive failure leading to mechanical ventilation. The parents report history of failure to thrive since the end of the neonatal period. The body weight was 4.2 kg (−3 SD) and he had a systolic murmur along the left sternal border. The electrocardiogram was normal. Transthoracic echocardiogram revealed a large malalignment ventricular septal defect with a left to right shunt and mild sub pulmonary artery gradient, enlarged left cardiac chambers and good biventricular function. The coronary artery anatomy was not visualized on echocardiography. A Chest X ray showed cardiomegaly with cardio thoracic index of 0.6, congested lungs and multiple areas of atelectasis. A multidetector computed tomography of the chest was performed in order to study the pulmonary parenchyma and eliminate any respiratory tract compression. It revealed multiple small pulmonary artery aneurysms (Fig. 1). A cardiac catheterization was subsequently performed and confirmed the diagnosis of ventricular defect with increased pulmonary artery pressure and distal multiple pulmonary artery aneurysms. It also showed an anomalous right coronary artery arising from the pulmonary artery and filled via retrograde flow from the left coronary artery.

The infant was operated on after the infection had resolved. He underwent direct reimplantation of the anomalous right coronary artery into the aorta with closure of the large ventricular septal defect through a right atriotomy. The post operative period was uneventful. On the last routine follow up, 2 years after surgery, he was perfectly asymptomatic on acethyl salicylic acid therapy. He gained weight normally. The ECG was normal and the echographic study showed normal biventricular function with no residual shunt or residual pulmonary hypertension. The forward coronary flow from the aorta was well seen.

1.2. Case n°2

An 11-year-old boy presented to the pediatric cardiology clinic complaining of dizziness and non specific chest pain on exertion for several months. Physical examination revealed a 4/6 grade systolic murmur over the pulmonary area. Electrocardiogram was normal. An echocardiogram revealed normal segmental analysis, with normal biventricular contractility. A pulmonary stenosis with peak gradient of 58 mmHg was noticed. The left coronary artery was dilated. The origin of the right coronary artery could not be visualized on echocardiography. There was a diastolic flow into the main pulmonary artery, close to the pulmonary valve (Fig. 2). Angiographic images documented the right coronary artery arising from the pulmonary trunk, and selective left coronary arteriography showed retrograde filling of the coronary artery from collateral vessels (Fig. 3). Right ventricular angiography and pressure measurements confirmed pulmonary valvular stenosis with infundibular reaction. The patient underwent surgical reimplantation of the anomalous RCA into the aorta (Fig. 4) with pulmonary valve commissurotomy. The post operative course was smooth with no clinical or electrical sign of ischemia. The post operative echocardiographic study showed good biventricular size and function with good visualization of forward coronary flow from the aorta to both right and left coronary arteries. There was no residual pulmonary stenosis, and pulmonary regurgitation was mild. At 3-month follow-up, he is asymptomatic and has normal left ventricular function.

2. Discussion

Congenital coronary artery anomalies are rare in the general population with an incidence of only 0.3–0.9%[1] increasing up to 36% in patients with congenital heart disease.[2] Among these abnormalities, the anomalous coronary origin from the pulmonary artery is an extremely rare form. Four variations of this condition have been described: origin of the left coronary artery from the pulmonary artery (ALCAPA), origin of the right coronary artery from the pulmonary artery (ARCAPA), origin of an accessory coronary artery from the pulmonary artery, and origin of the entire coronary circulation from the pulmonary artery.[3] ARCAPA is extremely rare, with an estimated incidence in the general population of 0.002%.[3] Many patients are asymptomatic, and the true prevalence of ARCAPA is likely underestimated. In fact, Williams et al.[4] published in 1990 a review of 70 previously reported cases of ARCAPA. They noticed that this anomaly was diagnosed on the basis of a heart murmur, mostly continuous, in asymptomatic patients in 50% of cases. In the other patients, chest pain and congestive heart failure were the most frequent presentations, followed by palpitations, arrhythmias and myocardial infarction. Only 25–30% cases of ARCAPA are associated with structural heart defects[4] like

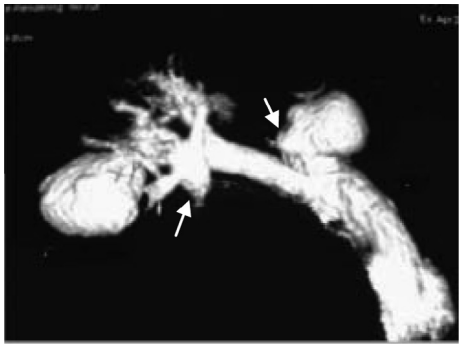

Figure 1 Small pulmonary artery aneurysms (arrows) on CT scan.

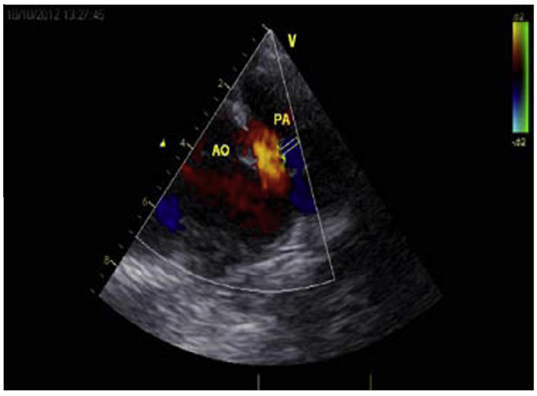

Figure 2 RCA from the pulmonary artery.