Anomalous Origin of the Circumflex Coronary Artery in the Pulmonary Artery in Young Adult

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Introduction

The anomalous origin of coronary arteries is an accurately described clinical condition which comprises a set of clinical presentations. The sudden death is the most serious presentation, and is often associated with the origin of the left coronary artery in the pulmonary artery[1]. The circumflex artery originating in the pulmonary artery is an extremely rare anomaly, and is mostly found associated to coronary artery anomalies and/or congenital heart defects, without prior description of the presentation with sudden death[2,3]. We describe a young adult at the age of 24 with anomalous origin of the circumflex coronary artery in the pulmonary artery and the diagnosis made only after cardiac arrest.

Case Report

HCC, 24 years old, mechanic

After extenuating effort (a soccer match) while resting, he underwent sudden illness with subsequent cardiovascular circulatory collapse. He was attended by his colleagues, who found out it was a cardiac arrest and started administering resuscitation maneuvers which were continued by the team of paramedics which came to assist the patient; he needed around three defibrillation shocks until admitted to the hospital.

It was a patient which had a good functional capacity, used to play nearly 30 minutes of soccer thrice a week, period after which he was replaced due to fatigue. In the past disease history, neither the symptoms of angina, heart failure or arrhythmias, nor family history have been found.

The rest-electrocardiogram did not show significant abnormalities, as well as the blood count, hepatic and thyroid function tests. The chest radiograph revealed calcified perihilar lymph nodes. The heart magnetic resonance imaging revealed a mild increase in the volumes of left ventricle, a minimum pericardial and pleural effusion without functional abnormalities or late enhancement. He was discharged with mild motor and speech sequelae.

Six months after this, he was referred to our hospital with a cardioverter defibrillator implantation plan. During assessment, an echocardiography was conducted (GE Healthcare, Vivid 7 Dimension, USA), showing a slight increase in the left chamber volume indexes; the right coronary artery had an origin with normal topography and caliper, the proximal anterior descending coronary artery presented mild ectasia, of around 4.2 mm diameter (normal diameter for the patient's body surface between 2.11 and 3.84 mm)[4,5]. In left ventricle, a hypertrabeculation of the posterolateral apical walls with flow in the deep recesses. Diastolic laminar flows were also found outside these recesses, one of them being intramyocardial, in the interventricular septum directed at the posterior wall, and the epicardial flow in the posterior wall directed at the base of heart. Relying on these findings, the hypothesis of anomalous coronary artery was formulated, possibly the circumflex artery with retrograde flow and associated coronary fistulas.

The research was supplemented by a coronary artery angiotomography, which showed the anomalous origin of the circumflex artery from the right pulmonary artery, on its posteroinferior face. Its path was winding; after its origin, it descended until the right superior pulmonary vein, crossing the left atrium until it reached the interventricular posterior sulcus. The anterior descending sulcus presented mild ectasia. The angiography revealed a rich network of well-developed collateral branches of the right and anterior descending coronary arteries to the circumflex artery. A 24-hour ECG Holter was performed without abnormalities and an electrophysiological study have not induced any arrhythmia.

The patient was referred to heart surgery for correction. The right internal mammal artery was subject to inosculation anastomosis with the circumflex coronary arteries without complications. As a follow-up after one year of surgery, the patient preserves the normality of left ventricular function on pharmacologic stress echocardiography with dipyridamole, which was negative for ischemia.

Discussion

The normal coronary anatomy is characterized by two ostiums located centrally in the right and left sinus of Valsalva, being universally defined as follows: the coronary trunk originates in the left coronary sinus, typically below the sinotubular junction and usually dividing in anterior descending artery and circumflex artery. In 37% of individuals, the left coronary trunk presents a trifurcation in the Anterior Descending (AD) Artery, Circumflex Artery (Cx) and in a medial or intermediate branch.

The AD follows subsequently to the pulmonary trunk in the anterior interventricular sulcus and presents diagonal branches with path toward the anterolateral wall of the left ventricle.

Keywords

Anomalies of Coronary Vessels; Pulmonary Artery; Physical Effort; Cardiac Arrest; Young Adult.

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The circumflex artery follows in the posterior atrioventricular sulcus and changes in size and extension, depending on the degree of the coronary dominance. The Cx typically originates one to three obtuse marginal branches, which supply the free wall of the left ventricle. In only 1% of the population, the AD and Cx origins happen separately in the left coronary sinus[6,7].

Most of the anomalies of coronary arteries (81%) identified in angiographic series do not pose a major risk of impaired myocardial perfusion, thus being benign. Although not common, some anomalies are largely described as the cause of cardiac morbidity and mortability[8]. The origin of the left coronary artery in the pulmonary artery is the most dominant coronary anomaly associated to sudden death[1]. The occurrence of coronary anomalies ranges between 0.3% and 0.9% in patients without heart disease and up to 3% to 36% in those with structural heart diseases[9].

The anomalous origin of Cx of the pulmonary artery is rare[10]. Its occurrence is usually associated with other congenital heart defects, such as Patent Ductus Arteriosus, aortic coarctation, subaortic stenosis and pulmonary valve stenosis[11], and isolated cases are infrequent[12,13]. The cases described include from newborns to adults, with diverse clinical presentations, reports of asymptomatic heart murmur[14], dyspnea[15] and angina[16,17]. The most severe forms found in the literature include myocardial ischemia, and cases of severe myocardial failure associated to this anomaly have few reports[18,19].

In older children and adults, relatively low pressures in the normal pulmonary artery create a gradient by which the blood flow is directed from the native coronary artery circulation, with the wide network of collaterals, to the anomalous artery and the pulmonary artery. This results in coronary-to-pulmonary artery fistula, entailing the phenomenon of coronary steal.

Symptoms and prognosis depend on the development of collateral vessels by the other two arteries. The presentation in adults may be as a new onset effort angina, dyspnea, ischemic abnormalities in the stress echocardiography or nuclear scintigraphy[20-22]. The sudden death is the most drastic presentation among the congenital coronary anomalies, and is strikingly prevalent in youngsters with sudden deaths connected to sports, with reports of 12% to 19% in this scenario connected to 1.2% in sudden death cases not connected to sports[2]. The area supplied by the pulmonary artery is profoundly ischemic[3], although the anomalous origin of the circumflex from pulmonary artery has not been associated with myocardial infarction, heart failure or cardiac arrest[18].

The treatment is surgical, either with the connection of Cx to its origin, connection with aortocoronary bypass or reimplantation of Cx in aorta[23]. It is recommended for children and adults to remove the phenomenon of coronary steal. Treatment failure of the anomaly may lead to congestive heart failure, angina, subacute bacterial endocarditis, myocardial

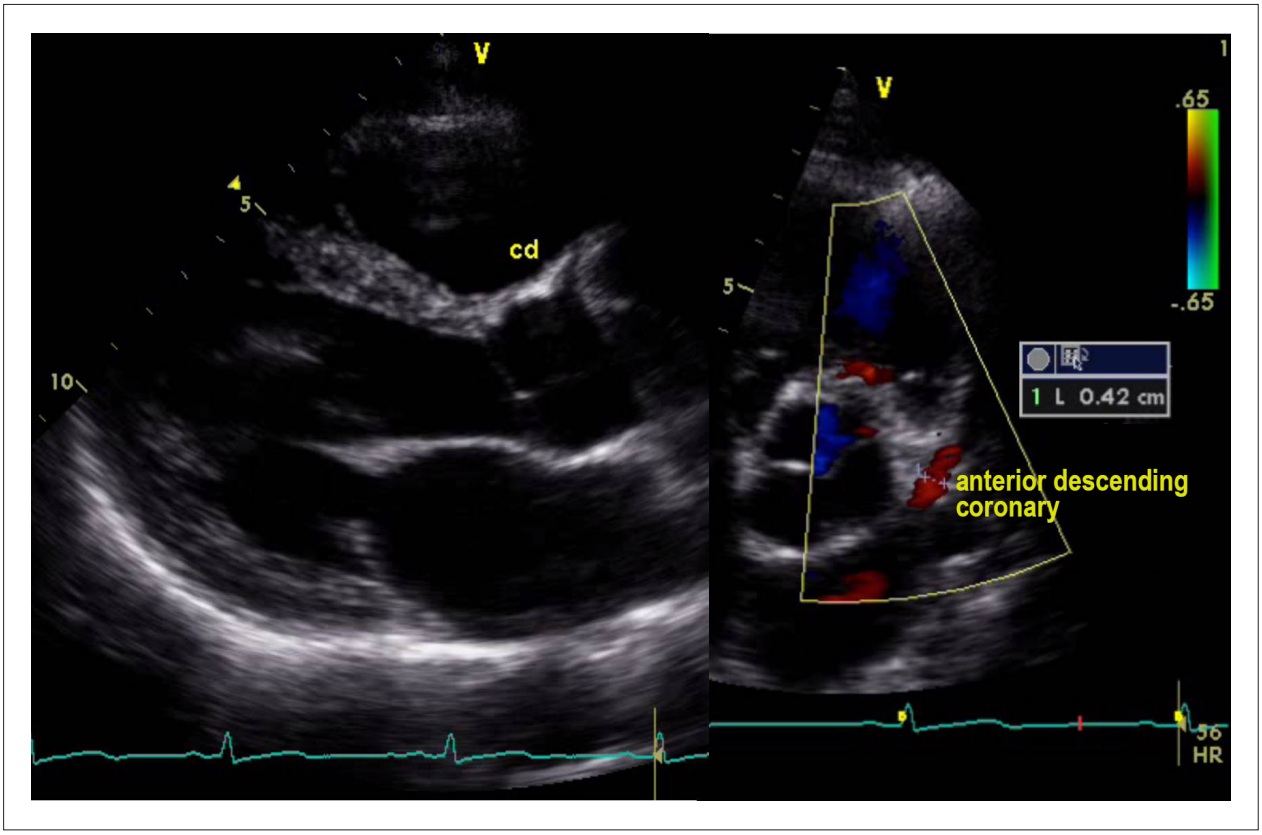

Figure 1 – *Right and anterior descending coronary artery with normal topography.*

Case Report

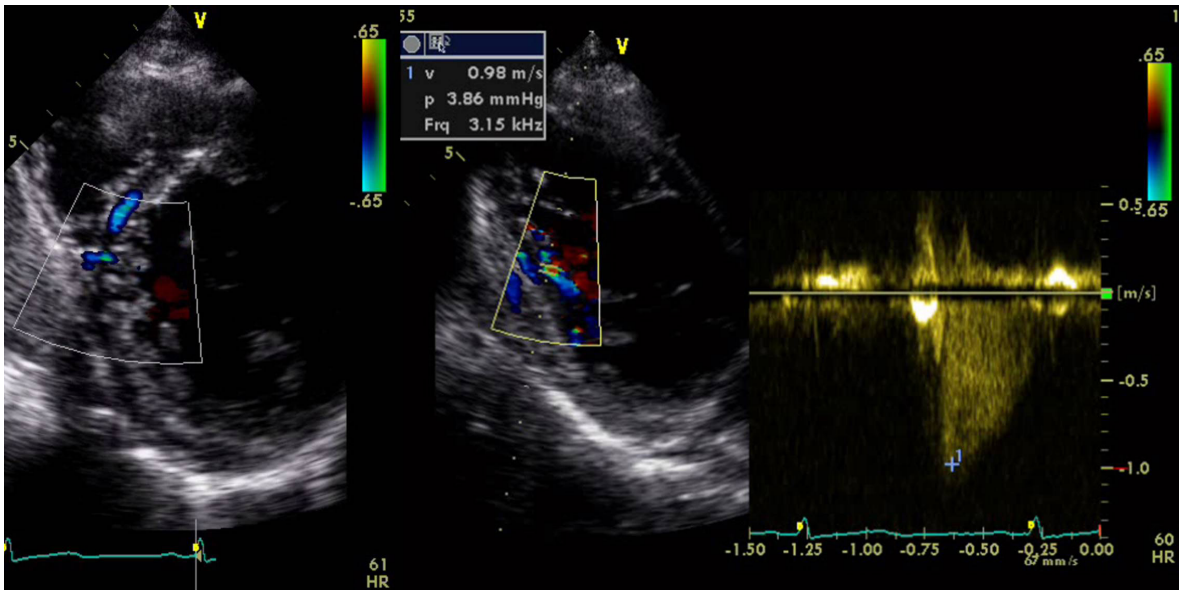

Figure 2 – *Flow in the interventricular septum directed at the posterior wall and the base of heart.*

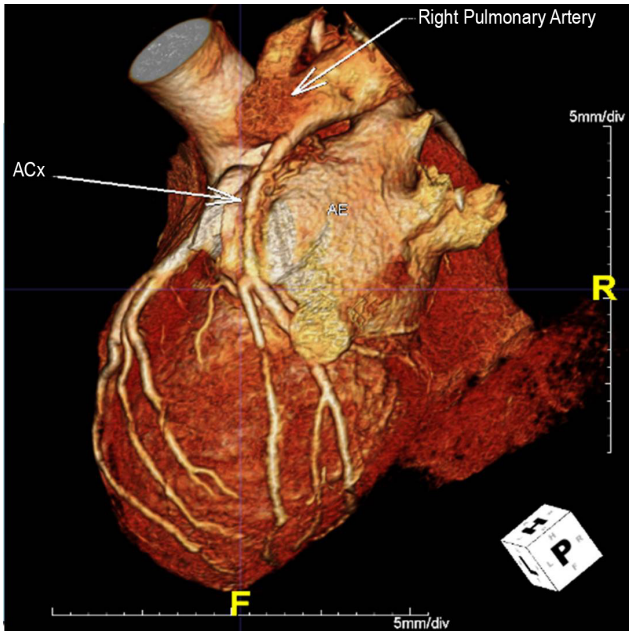

Figure 3 – *Tomography showing the anomalous origin of the circumflex artery and its path.*

infarction and coronary artery aneurysms with embolism and subsequent rupture[19]. The surgical correction helps restructure the myocardial perfusion and improves the left ventricle function.

Clinical findings of the retrograde flow in pulmonary artery of a single coronary fistula may lead to a wrong diagnosis of Patent Ductus Arteriosus[24].

The clinical recognition of this condition is masked by a good collateral circulation and a relatively small area supplied by this vessel. However, potentially severe complications detected with anomalous origin in other coronaries of the pulmonary artery may happen, and finding and correcting this defect is necessary[2].

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