Coronary Artery Disease Associated with Coronary Anomaly and Situs Inversus Totalis in Man Submitted to Angioplasty

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Introduction

Dextrocardia associated with situs inversus totalis is a rare condition with an incidence of 1:10,000 in the general population.1,2 It is characterized by a complete left-to-right reversal of the abdominal organs, including the abnormal position of the heart, resulting from changes in embryological development during the gastrulation stage (third week), characterized by the establishment of the cranio-caudal, dorso-ventral and right-left axes in the embryo.

This condition is transmitted by autosomal recessive genes, located in the long arm of chromosome 14, which affects the genetic cascade responsible for left-right differentiation. It should be noted that most affected individuals can have normal lives.3

The association of situs inversus totalis with coronary atherosclerotic diseases have similar incidence to the general population. However, the surgical approach in an event of acute infarction has significant statistical difference, since in patients with the anatomical condition mentioned before, there are more reports in the literature of myocardial revascularization surgery than percutaneous coronary intervention.4 The exceptional nature of the case was more evident after the coronary angiography showed right coronary artery originating from the left coronary sinus and anomalous pathway, which is a rare congenital coronary anomaly. Coronary artery anomalies can be found in 0.3% to 5.6% of the population.3 Congenital changes are associated with early morbidity and mortality in young adults, and there are reports of sudden deaths either during or after extenuating physical activity.5 It is estimated that this is the second more frequent cause of sudden death of cardiovascular origin among athletes, occurring between 12.2% to 17.2% in Europe and in the United States and dividing expert opinion, especially in regards with the therapeutic approach.7

Case report

A 46 year-old white male patient, natural from Rio de Janeiro, previously healthy, with a Framingham score of 11.7% (dyslipidemia, previous history of CAD) and dextrocardia associated with situs inversus totalis, reported that in September 2017 he started with dyspnea to medium efforts, which subsequently evolved to small efforts. On October 14, 2017, when he was making physical effort while repairing his house, he started with oppressive chest pain radiating to interscapular region associated with dyspnea and palpitation. Thus, he went to the emergency care unit, where he had a syncopal episode. Laboratorial tests were performed and the results showed positive myocardial necrosis markers, with a significant curve and a peak CK-MB 17.4 ng/mL and a troponin of 0.6 ng/mL. He was diagnosed with acute ST-segment elevation myocardial infarction (STEMI), presenting reperfusion criteria, and treated with thrombolytic therapy.

He was transferred to the State Institute of Cardiology Aloysio de Castro (IECAC), on October 16, 2017, where he underwent coronary angiography, which showed a severe 90% (ninety percent) lesion in the proximal...
segment of the right coronary artery and also revealed the anomalous origin of the right coronary artery from the left sinus. On the following day, the patient underwent successful angioplasty with stent implantation. He evolved with clinical improvement and was discharged with a prescription for the following medications: ASA, atenolol, enalapril and simvastatin. Two weeks after this procedure, the patient was in good general condition and asymptomatic. To the physical examination, he presented with a heart rate of 53 bpm, a respiratory rate of 18 breaths/min and blood pressure of 135/90 mmHg. The heartbeat was regular with two clicks and normal sound without murmurs. The conventional electrocardiogram (ECG) performed after the angioplasty showed typical dextrocardia pattern and electrocardiographic changes characteristic of inferior wall infarction. A nuclear magnetic resonance (NMR) imaging of the abdomen confirmed situs inversus totalis, with the liver on the left side and the spleen on the right side.

An important aspect to point out is the patient’s lack of risk factors for coronary events.

Discussion

The importance of the electrocardiographic diagnosis of dextrocardia is essential in the acute phase of coronary heart disease, especially in cases of clinical emergency, in which there is a need for immediate treatment with direct prognostic implications. Other aspects of importance are clinical reasoning and semiologic diagnosis, essentially by physical examination, in pathologies that involve dextrocardia and/or situs inversus totalis.

In these specific cases, we highlight the relevance of the epidemiology, clinical features, hemodynamics and anatomic abnormalities, through which we verify: frequency and disease-association in the population in general; the presence of coronary artery disease; acute coronary events and primary percutaneous intervention and, finally, right coronary artery originating from left coronary sinus and anomalous pathway. In other words, the association of benign and malignant anomalies in the same patient. Hence the need for early identification and the importance of clinical reasoning.

Coronary angiography and percutaneous coronary intervention (PCI) in these patients are technically difficult and require certain modifications, such as mirror image angiographic angulation, proper catheter selection and catheter manipulation for selective cannulation of coronary arteries. Regardless of the unusual anatomy, percutaneous coronary intervention in patients with coronary artery disease and dextrocardia is normally successful.

PCI can be safely performed using femoral or radial approaches, although as shown by previous case reports of PCI for AMI in patients with the mirror-image dextrocardia, it is conventionally performed with femoral arterial access.

In this case, the coronary angiography was performed via the right radial artery. Contrast injection into the aorta revealed dextrocardia and anomalous origin of right coronary artery from left coronary sinus. Catheterization of left coronary artery was performed using a JL4 catheter and showed absence of severe obstructive atherosclerotic lesions. Due to the anomalous origin of the right coronary artery, an AL2 catheter was used for characterization, which showed a 90% lesion in the proximal segment of the right coronary artery. We decided to perform a coronary angioplasty and to implant a conventional stent, 3.5/14 mm with a pressure of 14 atm, followed by dilating force at the target lesion with a 4.0/10 mm balloon, inflated to a pressure of 12 atm. Control angiography shows a good final angiographic result.

Dextrocardia accompanied by atherosclerotic coronary disease has been poorly described in the literature, but it can be treated with percutaneous coronary intervention. The procedure was performed with movements contrary to the usual, showing that coronary angioplasty can be used to treat these patients.

Author contributions

Conception and design of the research: Fuchs A. Acquisition of data: Cunha IAT, Mourão JRC, Lora LB, Duarte FCC. Analysis and interpretation of the data: Cunha IAT, Mourão JRC, Lora LB, Duarte FCC. Statistical analysis: Catanheda CRO. Writing of the manuscript: Cunha IAT, Mourão JRC, Lora LB, Duarte FCC. Critical revision of the manuscript for intellectual content: Fuchs A, Catanheda CRO. Supervision / as the major investigator: Catanheda CRO.

Potential Conflict of Interest

No potential conflict of interest relevant to this article was reported.
Figure 1 - PA Chest x-ray – Dextrocardia.

Figure 2 - A: anomalous origin of the right coronary artery from the left sinus; B: pre-angioplasty; C: positioning of stent; D: post-angioplasty.
Figure 3 - A: ECG leads and electrodes in standard position; B: ECG leads and electrodes in inverted position.

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Ethics approval and consent to participate
This article does not contain any studies with human participants or animals performed by any of the authors.
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