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Percutaneous treatment of anomalous right coronary artery with malignant course. Report of three cases

Tratamento percutâneo de coronária direita anômala com trajeto maligno. Relato de três casos

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ABSTRACT – Coronary anomalies are rare congenital malformations that are associated with an increased risk of arrhythmias, ischemic events and sudden death. Many remain asymptomatic throughout the patient's life, and are diagnosed incidentally by imaging tests. The treatment is necessary when ischemia is confirmed or in the presence of symptoms, and surgical intervention is the method of choice. However, some studies allow percutaneous treatment to be used as an alternative, especially in anomalies of the right coronary artery. In view of this, the objective of this study was to report three cases of congenital malformation of the right coronary artery treated by percutaneous coronary intervention.

Keywords: Angioplasty; Hemodynamics; Coronary artery disease

RESUMO – Anomalias coronarianas são malformações congênicas raras, estando implicadas em risco aumentado de arritmias, eventos isquêmicos e morte súbita. Muitos permanecem assintomáticos durante toda a vida, sendo diagnosticados de forma incidental por exames de imagem. A abordagem se faz necessária quando a isquemia é comprovada ou na presença de sintomas, sendo a intervenção cirúrgica o método de escolha. Contudo, alguns trabalhos admitem o tratamento percutâneo como alternativa, especialmente em anomalias da artéria coronária direita. Diante disso, o objetivo deste estudo foi relatar três casos de malformação congênita da coronária direita abordadas por intervenção coronariana percutânea.

Descritores: Angioplastia; Hemodinâmica; Doença da artéria coronariana

INTRODUCTION

For proper cardiac performance, there must be an adequate myocardial blood supply, which is dependent on the right and left coronary arteries, which, in turn, normally have their origin in the right and left Valsalva sinuses, respectively, at the level of the sinotubular plane.¹

Coronary anomalies are congenital malformations considered rare. The anomalies are classified by their morphology into anomalies of origination and course, anomalies of coronary intrinsic anatomy and anomalies of coronary termination, and they may be benign or malignant.^{1,2} Depending on the study and the criteria used, their prevalence ranges from 0.3% to 5.6%. Most patients remain asymptomatic throughout their lives, often being diagnosed by tests conducted for other purposes, whereas others have catastrophic outcomes, such as myocardial infarction, cardiac arrhythmias and sudden death, especially in young athletes.³⁻⁵

After the anomaly is identified, it is prudent and recommended that a morphological study be performed by tomographic examination to establish, in a precise way, the location of the coronary ostium, the angulation, the course and the relationship of the anomalous coronary artery with adjacent structures that may be causing extrinsic compression and the associated atherosclerotic disease.⁶

The aim of this study was to report three cases of congenital malformation of the right coronary artery (RCA), with its ostium located on the contralateral side to

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the right Valsalva sinus, and its course between the main pulmonary artery and the aorta (interarterial) artery considered “malignant” due to its high risk of causing ischemia and sudden death. The Research Ethics Committee of the *Hospital de Urgências de Goiânia*, linked to the Plataforma Brasil, approved the present study (CAAE: 94882318.7.0000.0033).

REPORT 1

In February 2020, a 58-year-old sedentary woman, with arterial hypertension, diabetes mellitus, and dyslipidemia, on a regular drug regimen, without previous angina symptoms, underwent routine preoperative tests before a varicose veins surgery in the lower limbs. There were no abnormalities in the general and systemic physical examination. During the treadmill exercise test, a descending ST segment depression was found, measuring 1mm, in the recovery phase, in leads V5, V6, and CM5, and the test was considered positive for myocardial ischemia. The transthoracic echocardiogram was normal. Serum total cholesterol levels were 233mg/dL, triglycerides 108mg/dL, high density lipoprotein (HDL) 38mg/dL, low density lipoprotein (LDL) 173mg/dL, and fasting glucose 99mg/dL. She was referred for elective cardiac catheterization as part of the preoperative cardiovascular risk stratification.

She underwent a coronary angiography on March 19, 2020 via radial access, in which it was observed that the RCA ostium was located on the contralateral side to the right coronary sinus (Figure 1A). In addition, multiple severe

Figure 1. Coronary angiography images illustrating case 1. (A) Right coronary sinus with no right coronary artery ostium. (B) Severe lesions in the proximal and middle third of the right coronary artery. (C) Severe proximal lesion in the diagonal artery. (D) Severe lesion in the second branch of the left marginal artery.

coronary lesions were observed, two in the RCA (90% stenosis in the proximal third, and 95% stenosis in the middle third) (Figure 1B), and an 80% lesion in the proximal third of the first diagonal branch (Figure 1C), and a 90% lesion in the proximal third of the second left marginal branch (Figure 1D), both of great importance.

After analysis and joint discussion by the cath lab team, the attending physician and the patient, it was initially chosen to perform percutaneous treatment of the diagonal and marginal branches lesions, with implantation of everolimus-eluting stents with bioabsorbable polymer, which were successfully performed.

To trace and predict potential technical difficulties that could occur during the percutaneous coronary intervention (PCI) in the RCA, a morphological study was performed by computed tomography angiography of the coronary arteries, establishing, with accuracy, that its ostium was located on the contralateral side to the right Valsalva sinus, above the sinotubular junction (Figure 2A), with a compressed course between the main pulmonary artery and the aorta (interarterial) (Figure 2B).

RCA: right coronary artery; LMCA: left main coronary artery.
Figure 2. Coronary computed tomography angiography images with three-dimensional reconstruction. (A) Right coronary artery ostium on the contralateral side to the right Valsalva sinus, above the sinotubular junction. (B) Right coronary artery compressed course between the main pulmonary artery and the aorta (interarterial).

For treatment, the right femoral artery was punctured, for the passage of an XB guide catheter, since it was an anomalous coronary artery, and a 0.035"x180cm guidewire. Catheterization of the RCA ostium and transposition of the lesions were performed with a 0.014"x180cm guidewire. The lesion was pre-dilated with a 2.0x20mm balloon. The 3.0x32mm drug-eluting stent was implanted under 10 ATM final deployment pressure, in the middle third (Figure 3A), and the 3.0x20mm drug-eluting stent was implanted under 14 ATM final deployment pressure, at the origin and the proximal third (Figure 3B). The procedure was successfully performed, with no clinical or angiographic complications.

After 24 hours of observation on an intensive care bed, the patient was discharged for outpatient follow-up, asymptomatic.

RCA: right coronary artery.

Figure 3. Images of percutaneous coronary intervention in the right coronary artery illustrating case 1. (A) Drug-eluting stent implantation in the middle third. (B) Drug-eluting stent implantation in the proximal third. (C) Post-intervention angiographic result.

REPORT 2

A previously healthy 43-year-old male patient who had been practicing moderate-intensity physical exercises since young adulthood, with a negative family history of coronary artery disease (CAD). He was seen at the cardiology outpatient clinic with symptoms of chest pain during strenuous efforts, grade I by the Canadian Cardiovascular Society (CCS I). He underwent a treadmill exercise test with evidence of myocardial ischemia at 10 METs, and was referred to the cath lab for cardiac catheterization in February 2019. An RCA originating from the left coronary sinus (Figure 4A) with a separate ostium, different from the left main coronary artery (LMCA), a 20% lesion in the proximal portion of the left anterior descending artery (LAD), and absence of any other changes were observed (Figures 4A and 4B).

For a better anatomical evaluation of the RCA and its relation with adjacent structures, the patient underwent coronary computed tomography angiography (Figure 5), which confirmed the anomalous origin, slit-like opening, $<45^\circ$ angulation, and $>50\%$ proximal stenosis of the RCA, in addition to an anomalous interarterial course. No intervention

was performed at the time, due to the patient's non-consent. Then, clinical observation was proposed, and he was medicated with acetylsalicylic acid (ASA) and atenolol.

Nine months after diagnosis, the patient returned to the service, due to progression of symptoms to chest pain upon medium exertion. A myocardial perfusion scintigraphy (SPECT-MPI) with stress stage was performed using a treadmill exercise testing Ellestad protocol, which was interrupted by exhaustion, after reaching the maximum expected heart rate, indicating a regular physical capacity. During the examination, he presented a 3.5mm J-point depression, with slow rising ST segment and Y point at 2.5mm from the baseline in DIII, AVF, V4, V5, V6 and CM5, associated with frequent ventricular extrasystoles, which were considered positive for ischemia with a high-risk pattern. The scintigraphy result corresponded to mild transient hypoperfusion of minimal extent, involving the lower left ventricular myocardium (SSS=4, SRS=0 and SDS=4) with 3% of myocardium at-risk and a slightly reduced regional systolic thickening in the lower region during the stress stage (Figure 6A).

RCA: right coronary artery.

Figure 4. Coronary angiography images. (A) Dominant right coronary artery, originating from the left coronary sinus with a probable interarterial course. (B) Left anterior descending artery with a 20% proximal lesion and left circumflex artery without changes.

RCA: right coronary artery; LAD: left anterior descending artery; CX: circumflex artery.

Figure 5. Coronary computed tomography angiography, three-dimensional reconstruction. (A) Interarterial course of the right coronary artery. (B) Origin in the left coronary sinus with compressed proximal course.

Figure 6. Myocardial perfusion scintigraphy images. (A) Discrete transient hypoperfusion of minimal extent involving the lower left ventricular myocardial region. (B) Post-intervention image with normal result.

In view of the clinical aspects and complementary exams, surgical treatment was proposed as the first option, in accordance with American and European guidelines, which was refused by the patient. As a therapeutic alternative, PCI was chosen after discussion by the Heart Team, based on some studies that allow for treatment with drug-eluting stents.⁷

Percutaneous coronary intervention was performed in January 2020 via the right radial artery, with a 6F XB guide catheter, and two 0.014" guidewires: an intra-arterial PT2 guidewire, going through the anomalous course up to its distal third, and a BMW guidewire, positioned freely in the aorta (floating wire technique) (Figure 7A) to prevent deep insertion of the guide catheter and provide the ideal positioning of the stent at the origin of the RCA. The everolimus-eluting drug-eluting stent was implanted with 4.0x20mm bioabsorbable polymer, inflated at 12 ATM without pre-dilation (Figure 7).

RCA: right coronary artery.

Figure 7. Percutaneous intervention in the right coronary artery. (A) Positioning of two guidewires (floating-wire technique). (B) Implantation of a drug-eluting stent at the origin and proximal third of the right coronary artery. (C) Post-intervention angiographic result.

There were no complications during and after the procedure. The patient was discharged for outpatient follow-up. After 3 months, the patient was asymptomatic, and a new SPECT-MPI was requested with a stress stage by treadmill exercise testing, to reassess the result of the intervention. The examination was performed using the ramp protocol and was interrupted by exhaustion at 7 minutes and 45 seconds (METs: 9.48), reaching 96% of maximum expected heart rate for age. There were no ST segment changes, arrhythmias or chest pain during the evaluation. After 99mTc-sestamibi infusion, the functional assessment of the left ventricle was considered normal, contrasting to the pre-intervention examination (Figure 6B).

REPORT 3

A 43-year-old woman was referred to the hospital for investigation of chest pain. According to her past medical history, she was sedentary, non-smoker and had no diagnosis of any diseases. Eight years before, she had burning chest pain radiated to the medial face of the left arm, triggered by strenuous efforts and of short duration, relieved by rest and with variable frequency. Episodes of syncope and an increase in the frequency of pain in the last 6 months, associated with dyspnea upon moderate exertion, prompted a more detailed investigation. The physical examination was normal. During the treadmill exercise testing, she had no symptoms, with no changes suggestive of ischemia on the electrocardiogram at rest and during exercise stress.

A coronary computed tomography angiography was requested, which identified anomalous origin of the RCA in the left coronary sinus with a “malignant” course and extrinsic compression (Figure 8). An anatomical study was indicated by coronary angiography. During the intervention, an RCA with anomalous origin in the left coronary sinus was observed, and by intravascular ultrasound imaging (IVUS), the presence of a slit-like origin was demonstrated,

with an exit angle of less than 45° and moderate dynamic compression during systole, with no atherosclerotic lesions (Figure 9).

For the assessment of non-atherosclerotic ischemia, a SPECT-MPI with pharmacological stress using dipyridamole was requested, and the patient had chest pain during the effort stage, which lasted for 50 minutes with spontaneous relief. Despite the symptoms, there were no changes in myocardial perfusion, which was considered negative for ischemia, with preserved ejection fraction in both stages.

Based on clinical findings and complementary exams, surgical treatment was proposed for decompression of the anomalous vessel course, with refusal by the patient. As an alternative, based on several cases reported in the literature and considering the experience of the service, PCI treatment was proposed.⁷ The treatment was performed with the implantation of a 3.5x16mm everolimus-eluting stent with bioabsorbable polymer (Figure 10).

The procedure was carried out without complications, with a favorable clinical course and complete remission of symptoms during follow-up. The patient continued on drug treatment regimen with ASA, clopidogrel, atorvastatin and metoprolol.

RCA: right coronary artery; LMCA: left main coronary artery; CX: circumflex artery; LAD: left anterior descending artery.

Figure 8. Coronary computed tomography angiography, three-dimensional reconstruction. (A and B) The origin of the right coronary artery is observed in the left coronary sinus, with an interarterial malignant course.

Figure 9. Coronary intravascular ultrasound images. (A) Right coronary artery distal to the extrinsic compression. (B) Slit-like origin of the right coronary artery. (C) Result after treatment with drug-eluting stenting.

Figure 10. Coronary angiography images illustrating case 3. (A) Right coronary artery originating from the left coronary sinus. (B) Drug-eluting stent implantation in the proximal portion of the right coronary artery. (C) Post-treatment angiographic result.

DISCUSSION

The reported cases illustrate the effective treatment via percutaneous access of RCA anomalies with an ostium located on the contralateral side to the right Valsalva sinus and a compressed course between the main pulmonary artery and the aorta.

Their prevalence varies among published works, and this is attributed to the different criteria used to define and classify a coronary anomaly.^{3,4} The most used criteria are those used by Angelini et al., who define as normal any morphology found in >1% of general population; as a variant of normality when uncommon, but found in >1%, and as abnormal when found in <1% of the population. They were classified into anomalies of origination and course, anomalies of coronary intrinsic anatomy and anomalies of coronary termination.⁸

As for the potential risk involved in causing hemodynamic changes and symptoms, in a study by Yamanaka and Hobbs, 81% of cases were classified as benign, whereas 19% had malignant anomalies, namely ectopic coronary origin of the pulmonary artery; origin in the opposite aortic sinus; single coronary artery, and large coronary fistula.^{1,2}

Most patients with coronary anomalies do not show signs, symptoms or complications, and the diagnosis is made incidentally in catheterizations.² When present, they may have catastrophic results. The main complications are chest pain, cardiomyopathy, syncope, dyspnea, ventricular fibrillation, myocardial infarction and sudden death, especially in young athlete patients.^{4,8}

Viewed from the frontal plane, the RCA usually appears at 90° in relation to the aortic sinus, whereas the LMCA appears at an angle of around 45°, and can vary from 25° to 90°. In patients with anomalous origin of the coronary arteries, those with an interarterial course have a higher risk of myocardial ischemia and sudden death.¹ When performing the necropsy of these patients, in the majority of cases an intracoronary thrombus was not evidenced; however

they had histological findings of an ischemic event, suggesting that the adrenergic stimulus of intense exercise could be associated with spasm of the anomalous artery.⁹

Several hypotheses regarding the pathophysiological mechanism involved in the increased risk have been postulated: the increase in cardiac output during physical exercise and the consequent expansion of the vascular lumen of the main pulmonary artery and the aorta causing extrinsic compression of the proximal portion of the coronary artery; the reduction of the angle formed between the coronary artery and the aorta during physical activity, generated in cases of anomalies with origin above the sinotubular plane; spasm or torsion; and proximal portion with intramural aortic course.¹

For the treatment of coronary anomalies, conservative (beta-blockers and avoidance of competitive exercises), percutaneous and surgical approaches can be considered, depending on clinical aspects and complementary exams.¹⁰ A guideline published in 2008 by the American College of Cardiology (ACC) and the American Heart Association (AHA) recommended revascularization surgery in patients who have been diagnosed with anomalous right coronary artery coursing between large arteries with documented ischemia.¹¹ However, with the advent of new techniques and drug-eluting stents in recent years, percutaneous treatment has been performed with satisfactory results and low mortality in late follow-up.⁷

Percutaneous coronary intervention can be considered, especially in RCA anomalies, when there are symptoms and/or high risk of sudden death; >50% stenosis with the vessel on a distal position to the normal lesion assessed by IVUS; >1/3 of the myocardium irrigated by the affected vessel, and documented reversible ischemia. Surgery remains the main indication in patients with stenoses that affect large areas, especially in LMCA anomalies.¹²

During the procedure, technical difficulties are often observed in catheterization and in the passage of the guide

due to anatomical differences, generating an increase in the procedure time with prolonged exposure to radiation and a greater amount of contrast used, a fact that can be minimized by performing an anatomical study by coronary computed tomography angiography (or magnetic resonance imaging) - which is highly recommended.¹⁰

Using the Scheme classification, based on the location of the coronary ostium, Sarkar et al. assessed the importance of choosing the guide catheter for the efficacy of the PCI. In addition to the anatomical aspect, other factors can also influence the selection of the catheter, such as operator experience and preference.¹³ In the first case, an XB guide catheter was used, and in the second and third cases we used a JR guide catheter, which provided the necessary coaxial alignment and adequate support for the intervention.

Despite the limitations of the case series, recent publications have shown that PCI is a safe option for selected patients.¹² We report three rare cases of RCA anomalies, with compressed interarterial course, treated with PCI and drug-eluting stenting. The success of the procedure was objectively evidenced by regression of the ischemic changes seen in scintigraphy and treadmill exercise tests, in addition to the improvement of symptoms. Overall, PCI was safe and could be considered as a treatment in specific situations.

A limitation of this study was the absence of long-term follow-up of the patients, which would be important to reveal potential late complications.

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None.

DECLARATION OF CONFLICTS OF INTEREST

The authors declare there are no conflicts of interest.

CONTRIBUTION OF AUTHORS

Conception and design of the study: Prudente ML; Guimarães HL; Gardenghi G; data collection: Prudente ML; Guimarães HL; data interpretation: Prudente ML; Rocha DF; Barbosa FP; Nacrueth FC; text writing: Prudente ML; Guimarães HL; Gardenghi G; approval of the final version to be published: Prudente ML; Guimarães HL; Rocha DF; Barbosa FP; Nacrueth FC; Gardenghi G.

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