¹ Hospital ENCORE, Aparecida de Goiânia, GO, Brazil.

Incidental finding of an ARCAPA during angiography

Achado incidental de ARCAPA durante angiografia coronária

Adriano Gonçalves de Araújo¹^{ID}, Victor Eduardo de Almeida e França¹, Pedro Arthur Ferreira Borges¹, Álvaro de Moraes Júnior¹, Maurício Lopes Prudente¹, Giuliano Gardenghi¹

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ABSTRACT – Anomalous coronary arteries are rare conditions, especially when the origin of the right coronary artery comes from the trunk of the pulmonary artery. The manifestations of the disease can occur at an early stage, leading to sudden death, myocardial infarction or ischemic cardiomyopathy. Many reported cases have been asymptomatic for long periods. Surgery is always indicated as seen in most of the literature. We describe an angiographic case of an oligosymptomatic 76-year old patient referred for elective coronary angiography for preoperative evaluation of prostate surgery. Although surgical treatment is the first choice, especially in early diagnosis, this patient was referred for a conservative treatment.

Keywords: Congenital abnormalities; Coronary vessels; Coronary angiography

RESUMO – Artérias coronárias anômalas são patologias raras, especialmente quando a origem da artéria coronária direita se dá no tronco da artéria pulmonar. As manifestações da doença podem ocorrer precocemente, levando a morte súbita, infarto agudo do miocárdio ou cardiomiopatia isquêmica. Muitos casos descritos permanecem assintomáticos por longos períodos. A cirurgia é sempre indicada, como visto na maior parte da literatura. Descrevemos um caso angiográfico de um paciente de 76 anos, oligossintomático, encaminhado para cinecoronariografia eletiva para avaliação pré-operatória de cirurgia de próstata. Embora o tratamento cirúrgico seja a primeira escolha, especialmente no diagnóstico precoce, esse paciente foi referido para seguimento em tratamento conservador.

Descritores: Anormalidades congênicas; Vasos coronários; Angiografia coronária

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Corresponding author:

Giuliano Gardenghi
ENCORE Hospital
Rua Gurupi, Quadra 25, Lote 6 a 8
Vila Brasília
Zip code: 74905-350 – Aparecida de Goiânia, GO, Brazil
E-mail: ggardenghi@encore.com.br

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INTRODUCTION

Congenital coronary artery malformations are rare and are usually diagnosed incidentally by coronary angiography or acute cardiovascular events, such as sudden death and myocardial infarction. Among the congenital heart diseases, the anomalous coronary artery origin is even more uncommon, and one of the most severe conditions, ranging from 0.2 to 1.5% in the general population,¹⁻³ while the anomalous origin of the right coronary artery from the pulmonary artery (ARCAPA) is even rarer, with a prevalence of 0.002%.^{1,4-9}

ARCAPA was first described in 1885 by St. John Brooks, in two cases of autopsy.⁵⁻⁸ The natural course may be insidious if there is collateral coronary circulation, which, due to the low pulmonary resistance, allows the flow from the left to the right.² In general, the main symptoms of the anomalous origin of the coronary arteries are angina, heart failure, dyspnea, myocardial infarction and sudden death.^{1,5-8} This anomaly can be present in association with other structural and congenital heart diseases, such as tetralogy of Fallot or aorto-pulmonary window in 22% of cases.⁹

This case report describes an incidental finding of an ARCAPA during angiography in an oligosymptomatic 76-years-old male patient. This case report was evaluated by the Research Ethics Committee of the *Hospital de Urgências de Goiânia*, linked to the Brazil Platform, and was approved under number CAAE: 85497418.2.0000.0033.

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CASE REPORT

A 76-year-old male, coming from the rural area of the Midwestern region in Brazil, was referred to the Interventional Cardiology Department for risk evaluation of a prostate surgery. From the cardiovascular point of view, he had only dyspnea upon moderate exertion, and he also suffered from advanced Alzheimer's disease. The presence of dyspnea upon moderate exertion was initially reported by the patient's son, starting in the last 12 months before the angiographic evaluation, according to information from the clinical cardiologist. Electrocardiogram showed sinus rhythm and unspecified alterations of ventricular repolarization. No echocardiographic evaluation was performed before the angiographic procedure. Although there have been attempts to perform treadmill stress tests to assess the presence of myocardial ischemia, they were not successful due to the lack of collaboration of the patient associated to Alzheimer's disease. Thus, the attending clinical cardiologist chose to perform invasive angiographic investigation, under sedation. The patient was not able to perform any other stress test due to his cognitive deficit, although he did not present motor restriction for it.

Coronary angiography was performed and showed dominant right coronary artery (RCA) with anomalous origin from the trunk of the pulmonary artery, with no obstructive lesions, which irrigates the inferior and posterior wall of the left ventricle, with multiple opacifications due to collateral circulation from the left main coronary artery (LMCA) (Figures 1 and 2). Left main coronary artery, left anterior

Figure 1. Coronary angiography showing dominant right coronary artery with anomalous origin from the trunk of the pulmonary artery. PA: pulmonary artery; RCA: right coronary artery; CLT: collaterals.

Figure 2. Coronary angiography evidentiating a dominant right coronary artery with anomalous origin from the trunk of the pulmonary artery. LMCA: left main coronary artery; LCx: left circumflex; LDA: left anterior descending; RCA: right coronary artery; CLT: collaterals.

descending (LAD) and left circumflex (LCx) arteries with usual and appropriately origin and course, ectasias, did not show obstructive lesions. Intense collateralization between left coronary and RCA was observed. From the manometric point of view, left ventricular, aorta and pulmonary circuit normotension were observed. The patient was discharged after the procedure.

DISCUSSION

We reported the case of a 76-year-old male with Alzheimer's disease in the last 10 years. During a complementary coronary angiography, a diagnosis of ARCAPA was made, as well as diffuse coronary ectasias, and left to right coronary fistulas. The symptoms reported by the patient characterize him in Functional Class II by the New York Heart Association, an unusual fact compared to the described findings.

In the literature, four different types of anomalous origin of coronary artery from pulmonary artery are described, including ARCAPA, the anomalous left coronary originating from the pulmonary artery (ALCAPA), the origin of the circumflex artery from the pulmonary artery and the complete origin of both coronary circulation systems from the pulmonary artery. Among the malformations, ALCAPA is the most common and fatal in childhood if not treated.⁵ Symptoms are usually related to the direction of coronary flow (usually from the coronary to the pulmonary artery, left to right, due to pressure difference) and to collateral circulation.^{5,7}

The main symptoms described in the literature are sudden death, ischemic cardiomyopathy and acute myocardial infarction, especially in younger population (also referred to as child). The onset of most symptoms is in the first decade of life, or there are no symptoms throughout the whole life, characterizing, in some cases, the malformation as a casual finding.²⁻⁹ In the aforementioned case, the patient is oligosymptomatic from the cardiovascular point of view except for family complaint due to the advanced dementia. It is worth mentioning that he remained completely asymptomatic until the sixth decade of life, an uncommon fact to individuals with such findings.

The diagnosis of such malformation should be investigated by means of other methods, such as echocardiography and electrocardiography, under observation of signs of ischemia. However, the final diagnosis is made primarily by coronary angiography or computed tomography angiography.³ Definitive treatment is indicated for all individuals, and can be performed by ligation of the right coronary and bypass with a saphenous vein graft originating from the aorta, or sectioning the right coronary artery and reinserting it into the aorta.^{5,7} Conservative treatment was chosen by the assistant physician taking into account the clinical characteristics of this patient, being corroborated by the literature as exception cases.^{2,5,7}

This case in question was not accompanied by in-hospital staff due to the outpatient status of the examination, and follow-up by an attending physician with no professional link with the service. After contact made by the service with the patient's family, we were informed about the proposal of conservative treatment by the attending physician, and no scheduling of the proposed surgery. Although rare to see asymptomatic elderly patients in these ARCAPA condition, this 76-year-old patient showed it is possible, mainly concerning flow direction and collateral circulation from normal coronaries, which resulted in a good or acceptable myocardial perfusion. Such combinations of anatomic coronary circulation make it possible to follow up ARCAPA patients in a conservative way.

SOURCE OF FINANCING

None.

CONFLICTS OF INTEREST

The authors declare there are no conflicts of interest.

CONTRIBUTION OF AUTHORS

Conception and design of the study: AGA; data collection: AGA, AMJ and MLP; data interpretation: AMJ and MLP; writing of the text: AGA, VEAF, PAFB e GG; approval of the final version to be published: AGA, VEAF, PAFB, AMJ, MLP and GG.

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