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Case Report

Cardiocentro Ernesto Che Guevara Santa Clara Cuba

Coronary artery fistula to the right ventricle in a patient with systemic atherosclerosis

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Images from complementary tests are shown with patient's consent.

Abbreviations
ABI: ankle-brachial index

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ABSTRACT

Atherosclerosis is a systemic disease that affects a number of vascular beds. Clinical manifestations whether acute or chronic (acute myocardial infarction, stable angina, intermittent claudication, cerebrovascular disease, among others) start after long periods of progression; so it may present subclinically in patients with coronary artery disease. What is particularly interesting about this form of presentation is that within a series of cases with multivessel disease, associated with an ankle-brachial index (ABI)<0.9, after an acute coronary syndrome, we have identified, as an angiographic finding, the presence of a coronary artery fistula to the right ventricle in a patient with very low ABI and clinical intermittent claudication. This fistula led to the symptoms that hampered cardiovascular rehabilitation. It is an infrequent disease characterized by chest pain; with low reporting (0.3 to 0.8%), as an incidental finding in coronary angiographies.

Keywords: Coronary artery fistula, Coronary angiography, Peripheral artery disease, Critical limb ischemia, Ankle-brachial index, Rehabilitation

Fístula coronaria a ventrículo derecho en paciente con ateroesclerosis sistémica

RESUMEN

La ateroesclerosis es una enfermedad sistémica que afecta múltiples lechos vasculares. Después de períodos prolongados de progresión comienzan las manifestaciones clínicas, de forma aguda o crónica (infarto agudo de miocardio, angina estable, claudicación intermitente, enfermedad cerebrovascular, entre otras); por lo que puede cursar de manera subclínica en pacientes con enfermedad arterial coronaria. Lo interesante de esta forma de presentación es que dentro de una serie de casos con enfermedad multivaso, asociado a un índice tobillo-brazo (ITB) < 0,9, después de un síndrome coronario agudo, hemos encontrado, como hallazgo angiográfico, la presencia de una fístula coronaria a ventrículo derecho en un paciente con ITB muy bajo y clínica de claudicación intermitente. Esta fístula es la causa de los síntomas que interrumpieron la rehabilitación cardiovascular; es una enfermedad poco frecuente y causa de dolor torácico, que se informa solo de 0,3 a 0,8%, como hallazgo incidental en angiografías coronarias.

Palabras clave: *Fístula arterial coronaria, Angiografía coronaria, Enfermedad arterial periférica, Isquemia crítica de miembros, Índice tobillo-brazo, Rehabilitación*

INTRODUCTION

Atherosclerotic disease affects multiple vascular beds and does not occur in isolation, as several studies have demonstrated its systemic nature. The REACH Registry reports that lower-limb arterial disease, in patients with coronary artery disease, is associated with a worse prognosis[1,2]. Lower-limb peripheral artery disease is usually initially asymptomatic, so it requires a thorough search with far-reaching questioning, physical examination and ankle-brachial index (ABI) measurement to reach an accurate diagnosis[3].

The estimated prevalence in some ABI studies <0.90, is 25-40%, in patients hospitalized for coronary artery disease; while on clinical examination less than 10% would be diagnosed[4].

Several studies describe the presence of significant coronary vessel stenosis in patients with ABI <0.9. Apurva *et al*[5], reported a high prevalence of 3-vessel and multivessel disease in patients with peripheral artery disease and ischemic heart disease, associated with low ABI values[5-7]. An as yet unpublished series of cases studied at the Department of Cardiovascular Rehabilitation and Secondary Prevention of the *Hospital General Camilo* Cienfuegos in Sancti Spíritus, Cuba, assessed the degree of association between different variables and the presence or absence of multivessel disease. They found that, out of 503 patients previously diagnosed with acute coronary syndrome, 282 had multivessel disease. Some 260 (92.2%) had more than 50% stenosis in 1 or more major coronary vessels, and 22 (7.8%) had left main coronary artery disease. The ratio of multivessel disease to absence was found to be 3.06 (CI 2.0-4.67) times higher in patients with ABI≤0.9, which represented a statistically significant association.

Our article presents the case of a woman who was undergoing scheduled cardiovascular rehabilitation, with a diagnosis of previous acute coronary syndrome, without multivessel disease, and intermittent lower limb claudication due to proven peripheral artery disease; whose outcome was sadly, unsatisfactory, due to a relatively rare coronary angiographic finding: a right ventricular coronary fistula.

CASE REPORT

We hereby report the case of a 55-year-old woman with a history of smoking, controlled hypertension and sedentary life, who was admitted to the Department of Cardiology at *Hospital General Camilo Cienfuegos* in Sancti Spíritus, Cuba, diagnosed with non ST-segment elevation acute coronary syndrome. The electrocardiogram, performed while in pain, showed: sinus rhythm of 84 bpm, QRS axis-11 degrees and horizontal ST-segment depression<2 mm of V_3-V_6.

The patient had a good outcome, clinically and hemodynamically stable. A subsequent echocardiogram showed no regional motility disorders and the presence of preserved global systolic function (left ventricular ejection fraction [LVEF] of 56.7%).

Aiming to accomplish an adequate risk stratification and before including her in the cardiovascular rehabilitation program, she was decided to undergo a treadmill test for assessment; yielding unconclusive results (the programmed submaximal heart rate for age, 4:34 minutes workout, speed of 2.7 km/h and slope of 10% was not achieved) since she stopped due to pain in the gluteal region and lower left limb, at the level of the gastrocnemius muscle (intermittent claudication); symptoms she did not refer to previously.

Left lower limb pulses were confirmed to be markedly reduced: at the femoral level and, to a lesser extent, at the pedal and posterior tibial pulses, prior to hemodynamic studies. Ankle-brachial index results were low in the compromised limb (pedal

Table. Ankle-brachial index measurement results.

Location	BP (mmHg)	ABI
Left lower limb		
Pedal pulse	40	0,33
Posterior tibial pulse	50	0,41
Right lower limb		
Pedal pulse	98	0,81
Posterior tibial pulse	90	0,75

ABI, ankle-brachial index; BP, blood pressure.

pulse 0.33 and posterior tibial pulse 0.41), although they were also pathological (ABI<0.9) in the right lower limb (**Table**). Blood pressure in the right arm was 120/80 mmHg.

A conventional lower-limb angiography was subsequently performed, showing severe obstructive lesion at the proximal left iliac artery, and a milder one −less than 50%− in the left femoral artery (**Figure**). Peripheral artery disease rehabilitation program was initiated following consultation with the Department of Angiology. But during low intensity exercises, the patient presented dyspnea events accompanied by sweating, reason why rehabilitation was suspended.

In view of the severity of her atherosclerotic disease and high probability of significant coronary artery disease, a coronary angiography was performed at the *Cardiocentro Ernesto Che Guevara* in Villa Clara, with the following results: short left main coronary artery, left anterior descending (LAD) artery and dominant circumflex artery, with no significant angiographic lesions; and poorly developed right coronary artery, without lesions. Besides, a fistula from circumflex artery to a low pressure chamber (right ventricle) was found.

COMMENTS

Ankle-brachial index's predictive value for determining the severity of coronary artery disease has been confirmed in several studies, hence making it a useful, simple, affordable and inexpensive method for predicting multivessel disease[8,9]. However, in this case there is no correlation between low ABI and severity of coronary artery stenosis; instead, a coronary fistula was found, which is a rare anomaly, with an estimated incidence in the general population of 0.002%[10].

A frequency of 0.3-0.8% has been reported as an incidental finding in coronary angiographies performed for any indication, with a 1.9:110.11[10,11] male/female ratio. In general, they are small in size and lack clinical significance, although they may cause arteriovenous short circuits with further hemodynamic implications. Coronary artery fistula has been defined as a direct communication between one or more coronary arteries with another major vessel or with a heart chamber (vena cava, right or left ventricle, pulmonary veins or artery). It may arise from any major epicardial vessel: right coronary artery (33-55%), left anterior descending artery (35-49%) and circumflex artery (17-18%). Furthermore, they can also drain into low pressure structures such as the right ventricle (40%), right atrium (26%), pulmonary artery (17%), superior vena cava (1%) and coronary sinus (7%)[10]. Drainage to other heart chambers (atrium [5%] and left ventricle [3%]) is less frequent. Fistulas may be congenital (most) or ac-

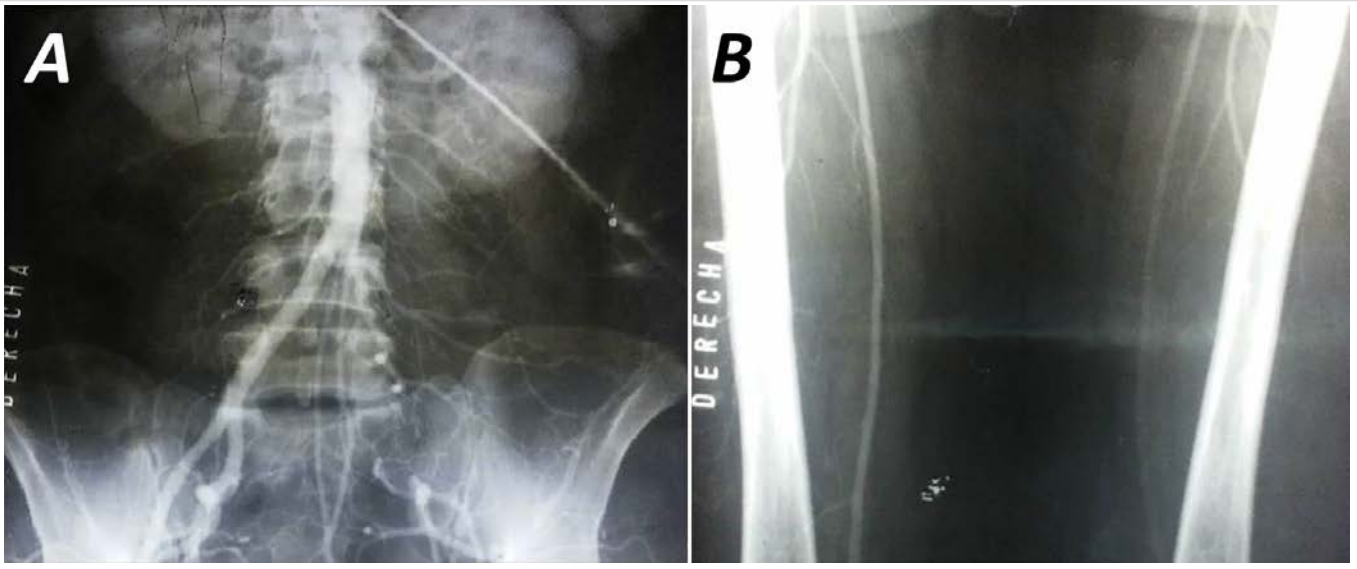

Figure. A. Aorto-iliac angiography showing extensive occlusion in the proximal segment of the left common iliac artery. The distal collector is filled by collateral blood flow. **B.** Distal blood flow at the level of femoral arteries, with decreased left contrast intensity.

quired, as a result of invasive procedures, septal myomectomy, chest trauma, catheter guidance during balloon angioplasty, among others[10-12].

Coronary fistulas are divided into five types, according to the chamber or vessel they drain; I) right atrium, II) right ventricle, III) pulmonary artery, IV) left atrium and V) left ventricle[11]. Physical examination shows no findings that are typical of this anomaly. A diastolic, systolic or continuous murmur, with or without pectoral fremitus, can be heard only in patients with high flow fistulas[12].

Symptoms are related to either the extent of the short circuit or the time of progression of the malformation. Most patients remain asymptomatic, but if the short circuit is significant, then it could lead to asthenia and heart failure over the years. Other clinical manifestations may be precordialgia (7%), dyspnea (40%) or, at worst, myocardial infarction (3%)[13].

There is consensus that it should only be treated in patients who are symptomatic or at risk of complications, such as in cases of coronary steal, aneurysm or major arteriovenous short circuit; which may cause myocardial ischemia[14]. Coronary fistulas can be diagnosed by angiography and may sometimes be detected by Doppler ultrasound. Medical facilities with sufficient technology prefer non-invasive imaging studies for diagnosis (multislice computed tomography and magnetic resonance imaging)[15]. A stress test or myocardial scan could be rather useful in patients with typical symptoms of coronary insufficiency. However, in most cases these tests prove negative. Electrocardiograms during pain events show non-specific data without describing electrocardiographic characteristics typical of this disorder.

Percutaneous intervention is the method of choice, less invasive and with shorter hospital stays. Therefore, surgery is reserved for cases with: 1) multiple fistulas, 2) major branches involvement as a complication during coils embolization (endovascular devices for fistula or other vascular closure) or 3) when the fistulous pathway is narrow, restrictive and drains into a heart chamber[16-18]. According to Díaz de la Llera *et al*[18], the percutaneous coil occlusion has been performed safely and effectively for more than two decades reaching the point of becoming the first therapeutic option. These same authors[18] found, in a total of 3075 coronary angiographies, 4 adults (0.13%) with coronary fistulas draining into vessels of the pulmonary region. Percutaneous closure by means of coils release was successfully carried out in all of them.

In the case reported, no therapeutic procedure was performed as the patient was reluctant to undergo interventional or surgical treatment for her peripheral artery disease. She thus continues her medical treatment since the coronary fistula, in this case, has no indication for closure.

REFERENCES

1. Mukherjee D, Eagle K. The importance of early diagnosis and treatment in peripheral arterial disease: insights from the PARTNERS and REACH registries. Curr Vasc Pharmacol. 2010;8(3):293-300.
2. Agnelli G, Cimminiello C, Meneghetti G, Urbinati S; Polyvascular Atherothrombosis Observational Survey (PATHOS) Investigators. Low ankle-brachial index predicts an adverse 1-year outcome after acute coronary and cerebrovascular events. J Thromb Haemost. 2006;4(12):2599-606.
3. Aboyans V, Ricco JB, Bartelink ME, Bjorck M, Brodmann M, Cohnert T, *et al*. Guía ESC 2017 sobre el diagnóstico y tratamiento de la enfermedad arterial periférica, desarrollada en colaboración con la European Society for Vascular Surgery (ESVS). Rev Esp Cardiol [Internet]. 2018 [cited 22 Ago 2019];71(2):111.e1-e69. Available at: https://doi.org/10.1016/j.recesp.2017.12.015
4. Alves-Cabratosa L, Garcia-Gil M, Comas-Cufí M, Blanch J, Ponjoan A, Martí-Lluch R, *et al*. Role of low ankle-brachial index in cardiovascular and mortality risk compared with major risk conditions. J Clin Med [Internet]. 2019 [cited 22 Ago 2019];8(6):870. Available at: https://doi.org/10.3390/jcm8060870
5. Badheka AO, Rathod AD, Bharadwaj AS, Bhat S, Kizilbash MA, Veeranna V, *et al*. Outcomes and risk prediction model for peripheral arterial disease in patients with stable coronary artery disease. Angiology. 2011;62(6):473-9.
6. Arroyo-Rodríguez C, Brito-Zurita OR, Sandoval-Navarrete S, Solis-Vásquez R, Ornelas-Aguirre JM, Olea-Hernández C, *et al*. Risk factors for three-vessel coronary artery disease in patients of Northwest México. Arch Cardiol Mex. 2018;88(5): 423-31.
7. Saleh A, Makhamreh H, Qoussoos T, Alawwa I, Alsmady M, Salah ZA, *et al*. Prevalence of previously unrecognized peripheral arterial disease in patients undergoing coronary angiography. Medicine (Baltimore) [Internet]. 2018 [citado [cited 29 Ago 2019];97(29):e11519. Available at:

http://dx.doi.org/10.1097/MD.0000000000011519
8. Basyouni MW, Shabana AM, El Kilani WM. Prevalence of lower extremities peripheral arterial disease among Egyptian ischemic patients attending cardiac rehabilitation unit. Egypt Heart J. 2018; 70(4):295-299.
9. Cordero A, Morillas P, Bertomeu-González V, Quiles J, Soria F, Guindo J, *et al*. Pathological ankle-brachial index is equivalent of advanced age in acute coronary syndromes. Eur J Clin Invest. 2011;41(12):1268-74.
10. Davidson CJ, Bonow RO. Cateterismo cardiaco. En: Mann DL, Zipes DP, Libby P, Bonow RO, eds. Braunwald Tratado de Cardiología: Texto de Medicina Cardiovascular. 10ma ed. Filadelfia: Elsevier Saunders; 2015. p. 442-72.
11. Abdelmoneim SS, Mookadam F, Moustafa SE, Holmes DR. Coronary artery fistula with anomalous coronary artery origin: a case report. J Am Soc Echocardiogr [Internet]. 2007 [cited 31 Ago 2019];20(3):333.e1-4. Available at: http://dx.doi.org/10.1016/j.echo.2006.09.012
12. Branco VGC, Ramos LM, Coelho LCA, Amorim RO, Borges RM. Semiologia do aparelho cardiovascular. Anatomia e fisiologia. Rev Cad Med [Internet]. 2018 [cited 1 Sep 2019];1(1):55-65. Available at: http://www.revista.unifeso.edu.br/index.php/cadernosdemedicinaunifeso/article/view/753/438
13. Gascueña Rubia R, Hernández Hernández F, Tascón Pérez JC, Albarrán González-Trevilla A, Lázaro Salvador M, Hernández Simón P. Isquemia miocárdica demostrada secundaria a fístulas coronarias múltiples con drenaje en el ventrículo izquierdo. Rev Esp Cardiol. 2000;53(5):748-51.
14. Vicario J, Licheri A, Gerardo L, Lofeudo C. Sinusoides y fístulas coronarias a cavidad ventricular izquierda. Presentación de un caso clínico. Rev Fed Arg Cardiol. 2004;33(2):236-9.
15. Duran A, Michelis V, Díaz P, Lujambio M, Kuster F, Lluberas R, *et al*. Evaluación de pacientes portadores de fístulas coronario-ventriculares múltiples. Rev Méd Urug. 2003;19(3):237-41.
16. Urmeneta Ulloa J, Molina Borao I, Ochoa Rea ME, Sánchez-Insa E. Embolización percutánea en fístula de arteria coronaria previo al trasplante pulmonar. Arch Bronconeumol. 2017;53(9):520-1.
17. Jama A, Barsoum M, Bjarnason H, Holmes DR, Rihal CS. Percutaneous closure of congenital coronary artery fistulae: Results and angiographic follow-up. JACC Cardiovasc Interv. 2011;4(7):814-21.
18. Díaz de la Llera LS, Fournier Andray JA, Gómez Moreno S, Mayol Deya A, González García A, Pérez Fernández-Cortacero JA. Fístulas de arterias coronarias en adultos. Oclusión percutánea mediante *coils*. Rev Esp Cardiol. 2005;58(1):93-6.