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CLINICAL CASE

Cardiopulmonary exercise test findings in a patient with a coronary fistula

Hallazgos ergoespirométricos en un paciente con fístula coronaria

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Abstract

In medical practice, it is common to encounter challenges in the diagnosis of patients. The case report gives a detailed description of the clinical history of a young adult female patient who presented the cardinal warning signs of chest pain and longstanding dyspnea on exertion and was ultimately diagnosed with a rare presentation of a coronary fistula. The patient had previously undergone multiple cardiovascular tests described in the medical literature as a non-invasive means for identifying this disorder (conventional stress test, transthoracic echocardiogram, and myocardial perfusion), but with no definitive diagnosis. Finally, she underwent coronary arteriography, the gold standard procedure, due to her abnormal results on the cardiopulmonary exercise test, which was ordered due to dyspnea of unclear etiology.

Keywords: *Chest pain. Ergospirometry. Coronary fistula.*

Resumen

En la práctica médica es común encontrar retos en el diagnóstico de los pacientes. Este es un caso que realiza una descripción del cuadro clínico de una mujer, con síntomas de dolor torácico y disnea de esfuerzo con diagnóstico final de una rara presentación de fístula coronaria, a quien se le realizaron múltiples estudios cardiovasculares descritos, como métodos no invasivos para el hallazgo de dicha alteración, sin llegar a un diagnóstico, para finalmente, realizar el procedimiento estándar de oro como lo es la arteriografía coronaria, por los resultados anormales de la ergoespirometría indicada por disnea de etiología no clara.

Palabras clave: *Dolor Torácico. Ergoespirometría. Fistula coronaria.*

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Introduction

Clinical practice presents challenges in diagnosing patients. Chest pain is one of the most common symptoms seen in emergency rooms and is frequently associated with cardiovascular causes[1-3]. Some of the more infrequent diagnoses of chest pain include coronary fistulas, which are mostly congenital and explain the symptoms from a pathophysiological perspective in terms of coronary steal that may or may not be detected by means of myocardial stress tests or imaging. Of these tests, arteriography is considered the definitive diagnostic method.

Clinical case

A 49-year-old female patient presented with an approximately 18-month clinical history of episodes of moderately intense oppressive chest pain lasting up to 20 minutes at a time. The pain radiated to the neck and back and was associated with a feeling of shortness of breath unrelated to exercise, occasional diaphoresis, mucocutaneous pallor and palpitations. She had also experienced II/IV functional class reduction.

She had consulted on several occasions with the same symptoms, and therefore a coronary event was ruled out through biomarker (troponin) testing. In her family history she reported that her father had coronary disease, without specifying the age of onset or diagnosis, and her mother had an aortic aneurysm associated with arterial hypertension and type 2 diabetes mellitus. Her personal medical history included secondary anemia triggered by uterine fibroids requiring a hysterectomy two months prior. Her cardiovascular risk factors included a 20-year history of smoking one cigarette a day up to four months prior to the consult, overweight, and sedentarism.

Upon arrival at the emergency room, an electrocardiogram was performed which showed no signs of ischemia; troponin ruled out an acute coronary event. In light of the dyspnea and chest pain diagnoses under study, a D-dimer test was ordered, which was positive. This raised suspicion of a pulmonary embolism, which was ruled out by a thoracic angiotomography, with no evidence of other vascular abnormalities. The complementary transthoracic echocardiogram showed a 60% ejection fraction, normal biventricular function, mild mitral regurgitation, and trace tricuspid regurgitation. A submaximal stress test using the modified Bruce protocol showed a heart rate of 123 beats per minute (bpm) (71% of the HR_{max}), a low systolic peak pressure response, a normal double product of 21,608, 7 METs functional class (FC) IC; the test was suspended due to peripheral fatigue and was indeterminate for coronary insufficiency. Post-exercise myocardial perfusion was normal with preserved left ventricular systolic function with a 67% ejection fraction.

As symptoms persisted, and due to dyspnea of unknown origin, a treadmill CPET using the Naughton protocol was ordered, with results as shown in Table 1: HR_{max} 137 bpm (78.9% of the predicted value); initial blood pressure 120/70 mm Hg and peak exercise blood pressure 144/70 mm Hg, which decreased during exercise; peak VO_2 16.8 ml/kg^{-1}/min^{-1}, and threshold VO_2 of 7.3 ml/kg/min; FC 4.8 METs IIC; threshold ventilatory equivalent CO_2 44.1; ventilation – CO_2 slope (VE/VCO_2) 41 L/min; end-tidal CO_2 ($PETCO_2$) 23.7 mm Hg; end-tidal O_2 ($PETO_2$) 87.46 mm Hg; oxygen pulse 8.7 ml/beat; estimated indirect maximum cardiac output 7.4 L/min; stress electrocardiogram with no significant ST changes or arrhythmias; and, as symptoms, dyspnea and angina scored as 2/4 radiating to the back, which is relieved with rest. Based on these findings, the test was interpreted to be positive for coronary insufficiency and pulmonary vascular disorder, based on the following data: a respiratory quotient of 1.04 (submaximal test), a functional capacity with a peak VO_2 of 16.8 ml/kg/min, abnormal due to symptoms (chest pain), the behavior of the VO_2/WR slope (Fig. 1), the pressure response (falling systolic pressure despite increased load), a flat oxygen pulse (Fig. 2), an abnormal VE/VCO_2 slope, and low $PETCO_2$ and $PETO_2$.

As a result of this data, a coronary arteriography was ordered, which showed no obstructive lesions, but did show a posterior secondary vessel arising from the proximal OM1 constituting a high-flow AV fistula draining into the pulmonary artery (Fig. 3).

Discussion

This case report's significance and discussion consists of three points:

- Presentation and approach to a clinical history of long-standing chest pain and dyspnea in a young female patient with normal non-invasive cardiovascular testing.
- The role of an integrated cardiopulmonary stress test as a diagnostic tool in patients with chest pain and dyspnea of unknown origin.
- Detection of a rare coronary fistula.

Coronary fistulas, defined as the connection of one or more coronary arteries to other structures such as

Table 1. Cardiopulmonary exercise test results

Protocol																	
Naughton treadmill protocol. Two-minute warm-up at 1.3 km/h with variable increments in two-minute stages. Breath-by-breath measurement of exhaled gases, continuous 12-lead electrocardiographic monitoring, pulse oximetry, blood pressure measurement and subjective perception of physical effort recorded every two minutes.																	
Exercise												Recovery (min)					
Measurement	Rest	I	II	III	IV	V	VI	VII	VIII	IX	X	XI	0	1	3	5	8	
Km/h		1.6	3.2	3.2	3.2	3.2	3.2	3.2	4.8	4.8	4.8	4.8						
%		0	0	3.5	7	10.5	14	17.5	12.5	15	17.5	20						
HR	72	89	88	105	114	118	131	137	0	0			127	120	80	82	83	
systolic BP	120	136	144	148	144	140	140	144	0	0			130	128	110	110	108	
diastolic BP	70	70	70	70	70	70	70	70	0	0			70	70	70	70	70	
SO_2 %	94	95	95	95	95	96	95	95	0	0			94	96	97	96	94	
SPE		2	3	3	3	5	4	5	0	0								
Angina scale		0	0	1	2	2	2	2	0	0								

	Rest	Predicted	Ventilatory threshold		Peak exercise			Rest	Predicted	Ventilatory threshold		Peak exercise	
				%		%					%		%
Heart rate (bpm)	72	174	109	63	137	78.9	Ventilation (lpm)	13	82	17	21	57	70
Oxygen pulse	3	9.7	4.2	50	8.7	90	Tidal volume (L)	0.751		0.611		1,377	
Cardiac output (L/min)	3.4		5.1		7.4		Respiratory rate	17	42	28	66	42	100
Ejection volume (ml)	37		46		54		VCO_2 (ml/min)	253		389		1,246	
Load (W)		108	48	45	136	127	Eq CO_2	43.1		35.6		42.3	
VO_2 (ml/min)	283	1615	521	32	1195	74	Eq O_2	38		26.6		44.1	
VO_2 (ml/kg/min)	4		7.3		16.8		Respiratory reserve B	86	86	82		38	
MET	1.1		2.1		4.8		Dyspnea index (VE/MVV)					60.4%	
RER	0.89		0.75		1.04		$PETCO_2$ mm Hg	23.36		28.5		23.7	
Peak exercise BP (mm Hg)					144/144		$PETO_2$ mm Hg	83.85		73.95		87.46	
Double product					19728		O_2 saturation (%)	94					
SPE (modified)					5/10		Minimum O_2 saturation (%)					93	
Chronotropic recovery index 1 min					17		O_2 desaturation during exercise (%)					1.1	

HR: heart rate; SPE: subjective perception of effort; % HR: % heart rate; BP: blood pressure; VO_2: oxygen consumption; $PETCO_2$-$PETO_2$: end-tidal partial pressure of CO_2 and O_2; BR: breathing reserve; RR: respiratory rate; VE/VCO_2 ($EqCO_2$) – VE/VO_2 (EqO_2): ventilatory equivalents for CO_2 and O_2; RER: respiratory exchange ratio; VE: ventilation; SO_2: oxygen saturation.

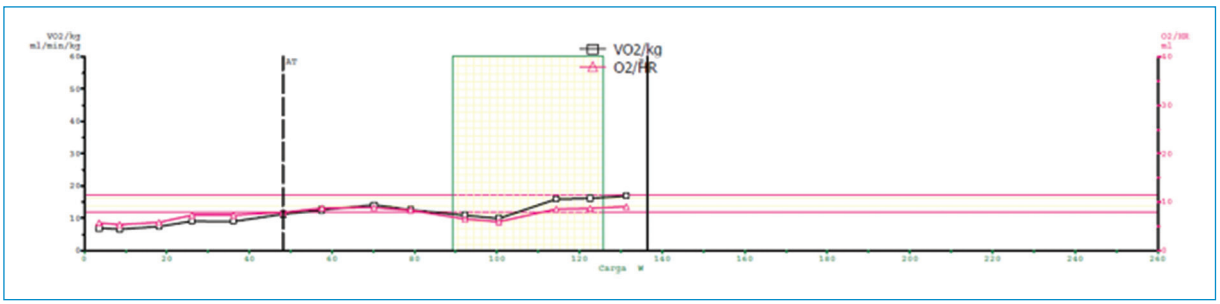

Figure 1. VO_2 reduction in relation to load in Watts.

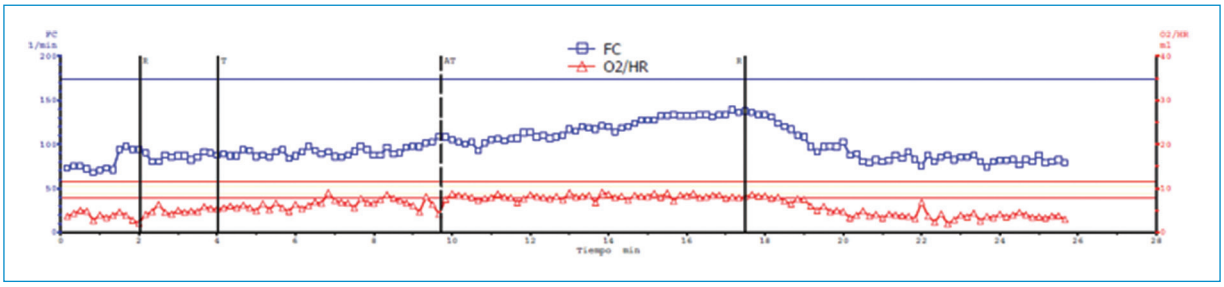

Figure 2. Flattened oxygen pulse (O_2/HR).

cardiac chambers or large vessels, are very rare anomalies with a 0.002%[2] prevalence in the general population and an incidence ranging from 0.08 to 1.2%, depending on the review[2,3]. They are classified based on a variety of characteristics including their morphology (origin, termination, number and size), etiology (congenital and acquired) or physiology (vascular short-circuits, ischemic disorders)[4].

The path of the fistula in this clinical case is unusual as it originates in the first obtuse marginal artery 1 (OM1), a branch of the circumflex artery, and drains into the trunk of the pulmonary artery. (Fig. 3)

Generally speaking, coronary fistulas are typically found on the right side of the heart[5]; studies show that 52-60% originate in the right coronary artery (RCA), 30% in the anterior descending artery (LAD) and 18% in the circumflex artery (Cx). Regardless of the point of origin, nearly 90% of fistulas drain into the right heart chambers: 40% into the right ventricle (RV) followed by the right atrium (RA), the coronary sinus (CS), and the trunk of the pulmonary artery (PA)[4,5].

Contrary to this, some case reports, such as the one presented by Canga[6] describing a sample of 49,567 arteriographies in patients over the age of 18, found that 0.1% had coronary fistulas. The most common originating artery was the LAD (50.8%) and the PA was the most common drainage point (53.7%). Altogether, 11.9% of fistulas originated in the circumflex artery.

Two case descriptions were found in Colombia, the first in relation to a fistula running from the right coronary to the pulmonary artery in a patient with cardiac syndrome X (angina and ST segment depression on a stress test, or exercise-induced ischemia with healthy coronary arteries)[7], and the second case involving a fistula from the LAD artery to the pulmonary artery, complicated by acute ST-elevation myocardial infarction[8].

The clinical presentation of the patient in this case, consisting of partially typical chest pain and reduced functional class, correlates with descriptions found in international literature. Most patients with fistulas are asymptomatic. Symptoms are present in 19-63% of cases, and most occur after the age of 18. The predominant physical finding is a continuous heart murmur, resulting from the systolic-diastolic flow in large fistulas. Congestive heart failure symptoms may develop when the fistula draining the right-sided system leads to pulmonary congestion and hypertension[2, 4-6, 8-11].

The challenge in this case was arriving at the final diagnosis of a coronary fistula. Clinical practice presents multiple daily challenges in patient diagnosis. Chest pain is one of the most frequently seen symptoms

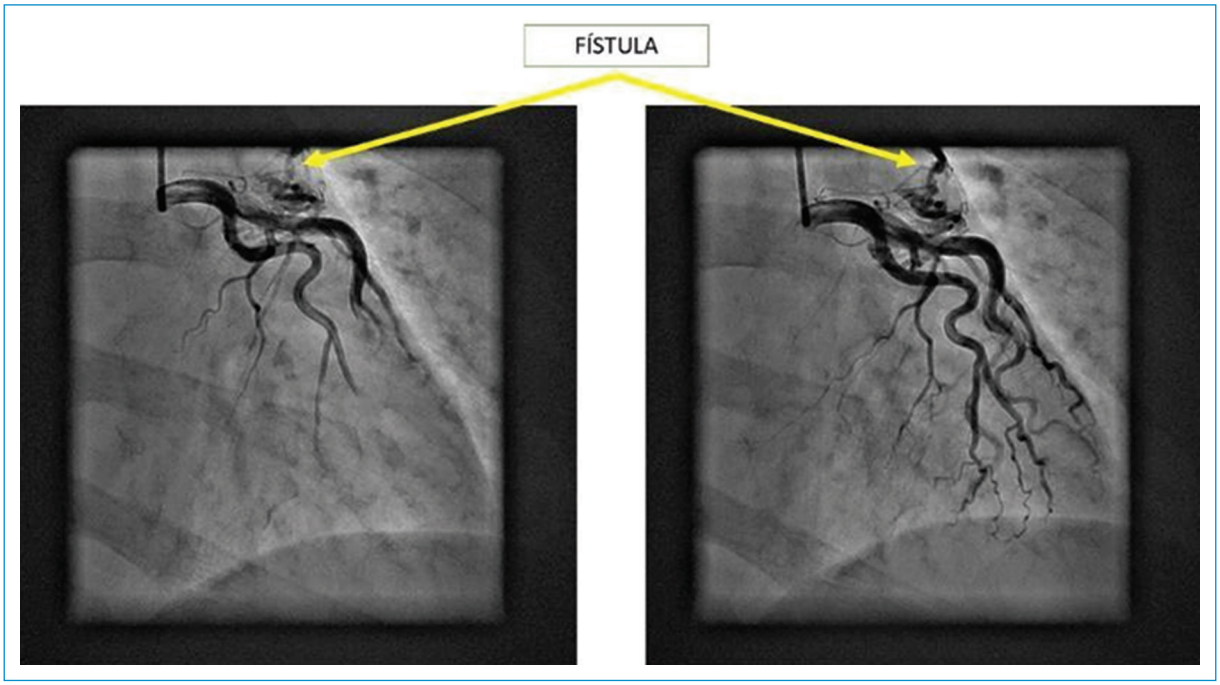

Figure 3. Evidence of a posterior secondary vessel arising proximally from MO1 and constituting a high-flow AV fistula draining into the pulmonary artery, according to the arteriography report.

Table 2. Suggested protocol for patients at risk for coronary anomalies

Suggestive symptoms (detailed clinical history)				
Mild				**Severe**
Non-athlete		**Athlete**		
Clinical follow-up (EKG and CXR)		EKG, CXR, TTE, TEE, stress test		EKG, CXR TEE, exercise test
Positive	**Negative**	**Negative**	**Positive**	
TTE		Clinical follow up	Cardiac catheterization/IVUS	Cardiac catheterization/IVUS
Stress test		Intramural		
Cardiac catheterization/IVUS		Yes	No	
		If severe narrowing consider intervention	Medical treatment and follow-up	

EKG: electrocardiogram, CXR: chest x-ray; TTE: transthoracic echocardiogram; TEE: transesophageal echocardiogram; IVUS: intravascular ultrasound.
Modified version of Challoumas's proposal as adapted from Anguelini[11].

in emergency rooms and is often associated with cardiovascular causes[1].

As previously stated, uncommon chest pain diagnoses include coronary fistulas, which may or may not be detected through imaging studies with or without myocardial stress, with arteriography being the definitive diagnostic method[2-4,5,10]. Table 2 presents the framework proposed by Challoumas, adapted from Anguelini[11], in which the diagnostic algorithm is arranged according to the severity of symptoms and even depending on whether or not the patient is an athlete.

None of the articles reviewed reported the use of ergospirometry, or cardiopulmonary exercise testing, as a diagnostic tool in patients with fistulas. However, one

Table 3. Summary of paraclinical tests and possible findings in the literature, correlated with clinical case findings[2,11]

Diagnostic tool	Usefulness or findings described in the literature	Clinical case findings
EKG	Signs of volume overload and some ischemic changes. 50% may be normal.	Sinus rhythm; no sign of ischemia
Chest x-ray	Generally normal, cardiomegaly.	Normal
Transthoracic echocardiogram	Enlarged cavities or regional or global dysfunction.	Preserved biventricular function LVEF: 60%. Slight mitral regurgitation with traces of tricuspid regurgitation
Transesophageal echocardiogram	Able to identify fistulas and their anatomical and functional characteristics.	
Multidetector computed tomography	Anatomical detail and the presence or absence of complications such as obstructions or coronary steal.	Negative CT angiography.
Cardiac nuclear magnetic resonance	Anatomical and functional detail.	-
Myocardial perfusion	Presence or absence of ischemia.	Normal. Preserved ventricular function.
Stress test	Determine functional capacity and signs of coronary insufficiency as well as arrhythmias.	Clinically and electrically indeterminate for coronary insufficiency. Submaximal 71% of FCmax. Halted due to peripheral fatigue and dyspnea.
Intracoronary Doppler echo	Determine function	-
Coronary arteriography	Definitive diagnostic method. Evaluates and establishes anatomical and functional details as well as complications; indicates intervention.	Normal ventriculogram 75% EF, RC dominant. LC: AD normal. Cx normal. Evidence of high flow A-V fistula originating in MO1 and draining into the pulmonary artery
Cardiopulmonary exercise test	-	Submaximal test RER 1.04, VO_2 16.8 ml/kg/min, abnormal due to symptoms, abnormal blood pressure response, flattened oxygen pulse. Positive for coronary insufficiency and pulmonary vascular abnormalities.

case report and subsequent topical and concept review by Reddy et al. mentions that stable, asymptomatic fistulas should be monitored on a regular basis and a detailed clinical history kept. Should hidden symptoms be suspected, a stress test or cardiopulmonary exercise test would be indicated. New symptoms, or paraclinical findings, would mandate an arteriography[4]. Table 3 summarizes the primary diagnostic tools for coronary fistulas[2,5,10,11], correlating them to our patient´s findings.

From a pathophysiological point of view, there are four factors involved in the effect and impact on the patient:

1. The presence or absence of overload at the fistula´s termination site.
2. Coronary involvement.
3. Valvular and endocardial complications.
4. Extracardiac complications[10].

Where there is no obstruction, coronary steal is the main cause of ischemia[2,9].

Ergospirometry is a widely-used procedure in clinical practice, and is considered the standard test for determining functional capacity. It uses a stress test together with the analysis of hemodynamic, ventilatory, gas exchange, and electrocardiographic variables, as well as symptoms, to extrapolate the systemic events that lead to the progressive increase in oxygen consumption due to the increased metabolic demand produced by exercise[12-15].

In cases where the oxygen uptake process is abnormal, analysis of these variables makes it possible to establish which system may be affected[13].

Variables related to coronary insufficiency have been found[15].

In 2012, the European Society of Cardiology and the American Heart Association published clinical recommendations for the cardiopulmonary exercise test in specific populations. Within the medical conditions described, they specify the ergospirometry variables for suspecting

myocardial ischemia, including oxygen pulse and VO_2/WR (the oxygen uptake to work rate relationship) that, under normal conditions, increase progressively during maximal exercise. Left ventricular dysfunction induced by myocardial ischemia results in prematurely flattened or decreasing trajectories. A stratification of patients with a possible diagnosis of myocardial ischemia was proposed, including these variables along with symptoms, electrocardiographic changes, and blood pressure behavior[14], which was reaffirmed in the 2016 update.

The specific finding in this clinical case was abnormal oxygen consumption associated with and related to variables that indicate coronary insufficiency. This was indicated by symptoms, a flattened oxygen pulse curve and an abnormal VO_2/WR curve starting out with a normal progressive increase that, however, falls after a certain work rate, relating in turn to an anomalous blood pressure response in which systolic blood pressure diminishes in spite of an increased workload. (Figs. 1 and 2).

This is related to Forman's[13] report, who described that an initial pattern of increased VO_2/WR during exercise followed by an abrupt flattening may reflect the onset of left ventricular dysfunction induced by ischemia in a patient with coronary insufficiency.

Conclusion

The interesting thing in this case is that, as part of complementary testing, ergospirometry was ordered for a young adult female patient with symptoms of angina and long-standing functional class reduction, but without cardiovascular risk factors, and that the values obtained helped orient to a specific method for a definitive diagnosis of the issue at hand.

The case helps consider the potential usefulness of, or ability to identify, the behavior of ergospirometric values as a diagnostic tool in patients with coronary fistulas, and stimulates interest in expanding inquiry into this field of knowledge, potentially becoming a starting point for future related research.

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Conflicts of interest

The authors declare no conflicts of interest.

Ethical disclosures

Protection of human and animal subjects. The authors declare that no experiments were performed on humans or animals for this study.

Confidentiality of data. The authors declare that they have followed the protocols of their work center on the publication of patient data.

Right to privacy and informed consent. The authors have obtained approval from the Ethics Committee for analysis and publication of routinely acquired clinical data and informed consent was not required for this retrospective observational study.

Use of artificial intelligence for generating text. The authors declare that they have not used any type of generative artificial intelligence for the writing of this manuscript, nor for the creation of images, graphics, tables, or their corresponding captions.

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