Myocardial ischemia caused by an anomalous circumflex coronary artery

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The case of a 71-year-old male patient, with symptoms of dizzines and atypical chest pain and a positive isotopic exercise stress test, is reported. Coronary angiography demostrated an anomalous origin of the left circumflex coronary artery from right coronary ostium but no obstructive atherosclerotic coronary lesions. The posible relation between the congenital coronary anomaly and the clinical manifestations of the patient is discussed.

Key words: *Catheterization. Congenital heart defects. Ischemia. Scintigraphy.*

Isquemia miocárdica causada por una arteria circunfleja anómala

Presentamos el caso de un varón de 71 años con mareos y molestias torácicas atípicas, que presentó un test de esfuerzo con isótopos positivo. Se realizó una coronariografía que puso de manifiesto una arteria circunfleja que se originaba en el ostium coronario derecho y que no presentaba lesiones ateroscleróticas asociadas. Se discute la posible relación entre la clínica del paciente y la circunfleja anómala.

Palabras clave: *Cateterismo cardíaco. Cardiopatías congénitas. Isquemia. Gammagrafía.*

INTRODUCTION

The increasingly extended use of diagnostic coronariography is discovering numerous congenital anomalies of the coronary arteries. At first they were considered simple coronariographic findings and there was a tendency to characterize them as benign. However, this attitude was undermined by reports of cases of sudden death, AMI, angina, and syncope associated with their presence. We now think that a new attitude is needed and, although not all coronary anomalies should be considered malignant, we must begin to consider them «potentially malignant.»

CLINICAL CASE

The patient was a 71 year-old male watchmaker with no family history of ischemic heart disease or known coronary risk factors. He was undergoing rehabilitation for left cervicobrachyalgia due to C6-C7cervicoarthrosis. Occasionally and with no relation to effort, he referred dizziness without loss of consciousness and sweating followed by «chest discomfort.» Sublingual nitroglycerin resolved his dizziness and «discomfort.» He was admitted to our hospital with normal electrocardiograms and serial cardiac enzymes. Conventional stress testing following the Bruce protocol had to be interrupted for dizziness and emotional lability when 84% of theoretical maximum exercise heart rate (MEHR) was reached with no electrical changes being observed. A 24-h Holter study was normal. He was released with anxiolytic medication to complete studies on an outpatient basis.

In later follow-up visits he continued to have occasional dizziness and «chest discomfort» without losing consciousness «because he would lay down on the ground.» Several Holter studies were requested, but continued to be normal.

A tilt-test was negative. Given his reiterated symptoms a radionuclide myocardial perfusion test was made. After a conventional exercise stress test, according to the Bruce protocol, images at rest were obtained 3 h after intravenous administration of the radiodrug (Tc; 8 and 20-mCi doses). The conventional stress test was discontinued when dizziness appeared in 5 minutes, when the patient had reached 86% of

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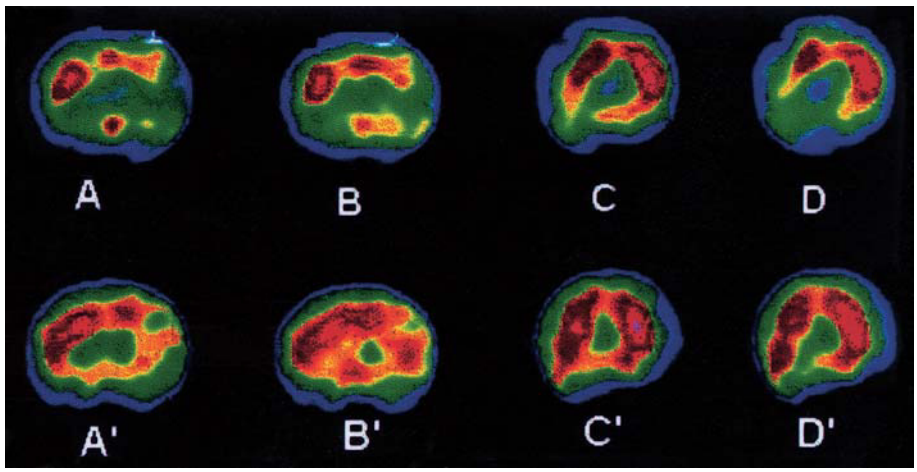

Fig. 1. Tomographies during effort (A, B, C, D) and rest (A', B', C', D') showing perfusion defects in the apical, inferior, and lower interventricular septal area.

MEHR without angor, electrical changes, and with a good pressor response. The study of myocardial perfusion after the ergometric test and at rest revealed small subsegmental perfusion defects in the apical, inferior, and lower interventricular septal area and positive redistribution to the lower septum and inferior face (Figure 1). These disturbances suggested ischemic changes induced by the test. In view of these results, cardiac catheterization was requested, which revealed right dominance and coronary arteries free of angiographic lesions. The circumflex coronary artery arose from the ostium of the right coronary and had a retroaortic path (Figures 2 and 3); LVF was normal. At follow-up 3 months later the patient referred occasional dizziness.

DISCUSSION

The incidence of congenital anomalies of the coronary arteries in different series ranges from 0.3% to 8.3%. The origin of the circumflex artery (Cx) in the right coronary sinus (from an ostium common to the right coronary or an independent ostium) or right coronary artery (as a proximal branch of this artery) is the most common anomaly of the origin of the coronary arteries. Thus, Effler in 1970 recommended calling it a «normal variant» rather than an anomaly.

The disposition of the Cx from its anomalous origin is always the same. From its origin the Cx goes backward and to the left, circling the aorta from behind, then passing between the posterior aortic wall and first

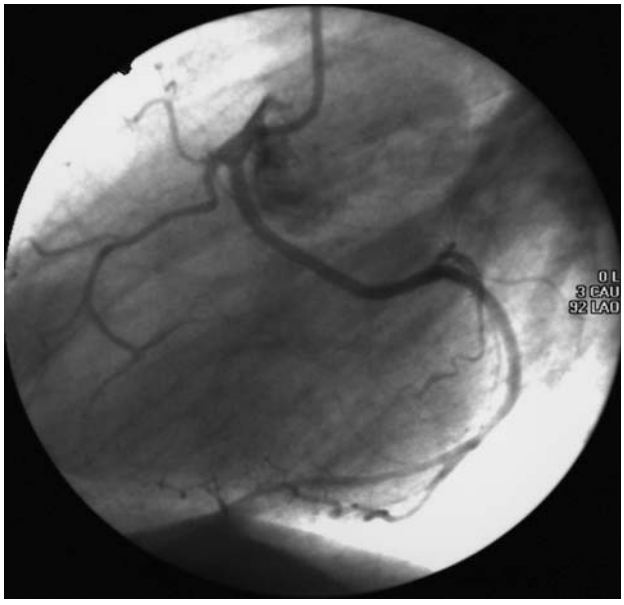

Fig. 2. Left anterior oblique coronariography view showing the circumflex artery originating in the right coronary ostium and following a retroaortic disposition to its normal distribution territory.

Fig. 3. Right anterior oblique coronariography view in which the origin of the circumflex artery in the coronary sinus is visible.

the anterior right atrial wall, then the left atrial wall, until it reaches its location in the left part of the atrioventricular sulcus, where it is covered by the left atrial appendage and has its usual disposition.

This anomaly has been and continues to be considered benign. Nevertheless, cases of association with sudden death, AMI, and angina pectoris in the absence of atherosclerotic lesions have been reported. The factor responsible for this pathogenicity could be repeated compression of this vessel by dilation of the aortic root or angling as a result of its retroaortic position, which would compress the coronary ostium into a «slit» that obstructs blood flow. It seems reasonable to carry out tests to detect possible myocardial ischemia before considering a coronary anomaly as benign. One of the tests most often used is the exercise stress test with thallium. However, for Piovessana, et al. and Molajo, et al. this test is not sensitive enough, as they have demonstrated in published reports of patients with Cx anomalies and positive conventional stress tests with negative thallium stress tests. This finding was attributed to a lack of sensitivity of this method in characterizing myocardial perfusion defects in these patients. In fact, Dunn, et al. questioned the sensitivity of the thallium exercise stress test in demonstrating ischemia in the territory irrigated by the Cx.

Our patient did not have clear clinical manifestations of angor. He presented dizziness and chest discomfort that remitted with occasional use of sublingual nitrates. His life was practically normal and in the last month he had only one episode of dizziness without loss of consciousness. We must also consider that his dizziness could be due to cervicoarthrosis, or that the «chest discomfort,» which could be caused by ischemia, could precipitate dizziness. We found no arrhythmic cause of dizziness in Holter studies and the tilt-test was negative. A finding suggesting a possible ischemic cause was the positivity of the radionuclide test, although it could be considered a false positive coinciding with the area of perfusion of an anomalous Cx.

In fact, an anomalous Cx artery, paradigm of the «benignity» of coronary anomalies, can sometimes be non-benign. Compression of the retroaortic segment of the Cx, or angling at its origin, could narrow the ostium to a slit and cause ischemia.

In this case we decide not to take an aggressive therapeutic approach for the moment. Taylor, et al has studied the main features of patients with coronary anomalies that cause sudden death. Age under 35 years and an interarterial path were the two factors most commonly related with this fatal outcome. The age of our patient and the fact that his clinical manifestations did not interfere with a normal life motivated us to use a conservative approach with anxiolytic drugs and sublingual nitrates.

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