Kardiologia Polska
2011; 69, 5: 505–506
ISSN 0022–9032

Left main coronary artery arising from right sinus of Valsalva: a rare congenital anomaly associated with distal vasospasm

Odejście pnia głównego lewej tętnicy wieńcowej z prawej zatoki wieńcowej — rzadka anomalia połączona z obwodowym kurczem tętnicy

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Abstract

A 50 year-old female patient was admitted to our outpatient clinic with a two year history of chest pain and dyspnoea on exertion. Echocardiography revealed apical hypokinesia with an ejection fraction of 50% on the left ventricle. Coronary angiography revealed that the left main coronary artery was arising from the right sinus of Valsalva and than coursing posterior to the aorta. There were significant stenoses at the proximal right coronary artery (RCA) and the proximal left anterior descending coronary artery (LAD). The RCA lesion disappeared after intracoronary nitroglycerine administration, and the LAD lesion disappeared the next day when the patient was due to undergo percutaneous intervention. Stress myocardial perfusion scintigraphy revealed anteroseptal ischaemia consistent with reversible ischaemia.

Key words: coronary anomaly, vasospasm

Kardiol Pol 2011; 69, 5: 505–506

INTRODUCTION

The left main coronary artery (LMCA) pursuing a retroaortic course has been considered a benign anomaly [1–3]. We report a case where a patient with this anomaly presented with evidence of myocardial ischaemia which was partly attributable to coronary artery vasospasm.

CASE REPORT

A 50 year-old female patient was admitted to our outpatient clinic with a two year history of chest pain and dyspnoea on exertion. She had no family history of ischaemic heart disease and no known coronary risk factors. Upon detailed examination, she was diagnosed as having chronic stable angina pectoris. Ventricular trigeminy was seen on ECG. On transthoracic echocardiography, apical hypokinesia was present, with a global ejection fraction of 50%. The size of the heart chambers were within normal limits. Biochemical parameters, including thyroid function tests, were also within normal ranges.

On coronary angiography, the LMCA was arising from the right sinus of Valsalva and than coursing posterior to the aorta (Fig. 1, Panel 1). There were significant stenoses at the proximal right coronary artery (RCA) and the proximal left anterior descending coronary artery (LAD) (Fig. 1, Panel 2 and 3). After intracoronary nitroglycerine administration, the severe lesion at the RCA disappeared (Fig. 1, Panel 4), but the LAD lesion was still present. Therefore, elective percutaneous coronary intervention (PCI) to the LAD was planned. The following day when the patient was taken into the cardiac catheterisation laboratory for PCI, the LAD lesion had also disappeared (Fig. 1, Panel 5). The site of the lesion was too far from the main stem to be a vasospasm due to catheterisation.

So the patient was considered to have intermittent vasospastic anginal attacks. A multislice computerised tomographic view of the patient was also obtained (Fig. 1, Panel 6). A single photon emission computed tomographic (SPECT) study was performed with 20 mCi of technetium-99m metho-

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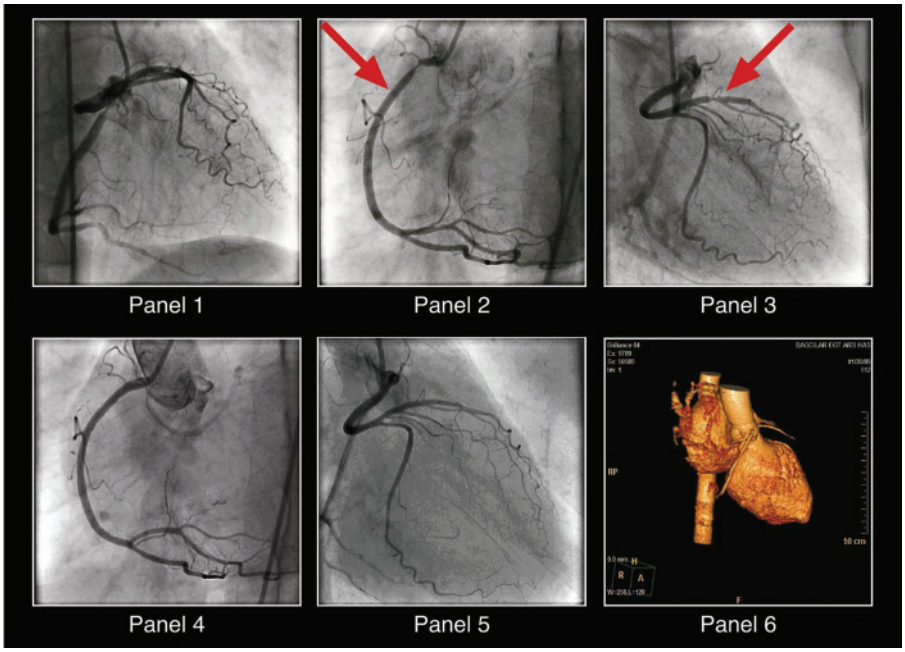

Figure 1. Panel 1. Proximal right coronary artery (RCA) and left anterior descending coronary artery (LAD) lesion due to vasospasm; **Panel 2.** RCA lesion; **Panel 3.** LAD lesion; **Panel 4.** Disappearance of RCA lesion after intracoronary nitroglycerine injection; **Panel 5.** Disappearance of LAD lesion; **Panel 6.** Retroaortic course of left main coronary artery on computed tomography angiogram

xyisobutylisonitrile after an exercise stress test, and again with the patient at rest. In the images taken after the stress test, hypoperfusion could be seen in the anteroseptal region, and perfusion of this segment returned to normal on the images taken with the patient at rest. We proscribed strenous physical activity for this patient and prescribed verapamil. She is currently free from anginal attacks.

DISCUSSION

The LMCA arising from the right sinus of Valsalva is an extremely rare congenital coronary anomaly [4, 5]. Origin of the LMCA in the right coronary sinus has been described at a frequency of 0.02% in autopsy series [1] and from 0.05 to 0.19% in angiographic series [2]. The LMCA either has a common ostium with the RCA, or arises independently to the ostium of RCA [1]. These cases are classified according to the course of LMCA into four categories [6]: (1) intertruncal or interatrial, between the aorta and pulmonary arteries; (2) anterior, in front of the right ventricular outflow; (3) posterior or retroaortic, behind the aorta; (4) intertruncal-septal or transseptal, through the supraventricular crest and interventricular septum. The retroaortic course of the LMCA is an uncommon entity.

This anomaly is serious and associated with sudden cardiac death and myocardial infarction, if anomalous LMCA passes between the aorta and the pulmonary artery [1, 5]. The anterior as well as the posterior course have been considered to be clinically insignificant [3, 5]. However, there have been isolated reports of ischaemia or sudden death associated with the retroaortic course of LMCA or one of its branches [7–9]. Although most patients with the anomalous LMCA arising from the right sinus of Valsalva are asymptomatic, the therapeutic approach must be individualised in each subject. In asymptomatic subjects, the age of the patient and the type of anomalous artery should be carefully evaluated. Surgical correction is generally indicated for patients who have symptoms or when these anomalies are identified in young people.

In our case, the retroaortic LMCA course and the patient's symptoms could be partly attributable to coronary vasospasm. Sudden cardiac death and ventricular fibrillation may occur even when LMCA courses retroaortically due to vasospasm.

To the best of our knowledge, this is the first case in literature reporting that the retroaortic course of anomalous LMCA is associated with documented ischaemia and vasospasm.

Conflict of interest: *none declared*

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