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DOI: 10.5603/FC.a2019.0083

Article type: Cardiology Investigation

Submitted: 2019-04-23

Accepted: 2019-05-01

Published online: 2019-07-02

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Reversible myocardial perfusion defect in a patient with anomalous origin of the left circumflex coronary artery from the right coronary sinus

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Abstract
This clinical note presents a 50-year-old woman with known anomalous origin of left circumflex coronary artery (LCx) from the right sinus of Valsalva, with retro-aortic course confirmed by coronary computed tomography angiography (CCTA). Due to progressive dyspnea and suspicion of ischemia the patient was referred to the nuclear medicine department for stress-rest myocardial perfusion scintigraphy. The imaging revealed inducible perfusion deficits mainly in lateral and infero-lateral wall involving 14% of the total left ventricular (LV) myocardium. Although, the detected coronary anomaly is considered nonmalignant, the extent of inducible ischemia exceeding 10% of the LV myocardium should be taken into consideration while managing the patient.

Key words: anomalous coronary artery; inducible ischemia

Introduction
Coronary artery anomalies (CAAs) are a group of variable congenital disorders with diverse pathophysiological mechanisms and clinical manifestations [1]. The incidence of CAAs is approximately 0.3–0.9% in patients without structural abnormalities of the heart and significantly more frequent (3–36%) in structural heart disease. Although in young athletes CAAs are the second most common cause of sudden cardiac death (SCD), in middle age or elderly individuals they are very rarely fatal. The type of coronary anomaly most commonly associated with SCD is the anomalous origin of a coronary artery (AOCA), especially the one with a course between the aorta and the pulmonary artery.

Clinical case
We present a case of 50-year-old female with no history of coronary artery disease (CAD), with known anomalous origin of left circumflex coronary artery (LCx) from the right sinus of Valsalva, with retro-aortic course confirmed by coronary computed tomography angiography (CCTA) (Fig. 1 and 2). The patient was referred to the nuclear medicine department for stress-rest myocardial perfusion scintigraphy to assess the degree of suspected ischemia due to progressive dyspnea. Treadmill stress test was not performed because of total hip replacement and post-surgery insufficiency. Single photon emission computed tomography (SPECT) was performed in 1-day protocol, with dipyridamole stress test (0.56 mg/kg of body weight) and under rest conditions, 1 hour after injection of 333 and 999 MBq of 99mTc-MIBI (methoxy-isobutyl-isonitrile), respectively. Imaging revealed inducible perfusion deficits in two regions: larger in lateral and infero-lateral wall, and smaller in anterior and antero-lateral wall, involving 14% of the total left ventricular (LV) myocardium (Fig. 3). The patient was submitted to medical treatment with valsartanum, amlodipine and bisoprololum.

Discussion
Reversible ischemia corresponding to limited coronary flow reserve (CFR) in lateral and infero-lateral wall of the LV — the region supplied by LCx, most probably resulted from the anomalous origin of this artery. Although, the detected coronary anomaly is considered nonmalignant and hemodynamically insignificant, the extent of inducible ischemia exceeding 10% of the LV myocardium is associated with high risk of cardiac death and myocardial infarct (annual mortality > 3%) [2], which should be taken into consideration while managing the patient. The smaller reversible perfusion defect in anterior (especially its apical segment) and antero-lateral wall was most probably induced by myocardial bridging of left anterior descending artery (LAD), also detected in CCTA.

Noteworthy, the patient was also diagnosed with mitochondrial myopathy with mutation of gene FBN1 (manifesting with muscle weakness), which could potentially influence the biokinetics of regional and/or global MIBI uptake in cardiomyocyte mitochondria [3–5]. MIBI is a cationic compound capable of passive diffusion into the cytoplasm and mitochondria and is trapped inside the mitochondria due to negative plasma and mitochondrial membrane potentials. We did not observe disturbances in regional myocardial tracer uptake other than described above. However, to quantitatively assess myocardial perfusion and estimate the absolute value of myocardial blood flow (MBF) in ml/min/g in all LV segments, one should perform positron emission tomography (PET) [6]. When considering the appropriate perfusion tracer for cardiac PET scan in this case, one
should avoid compounds which present with mitochondrial affinity, e.g. 18F-flurpiridaz (binds to mitochondrial complex 1 (MC-1)) and choose between ligands with non-mitochondrial mechanisms of uptake, e.g. 82Rb (potassium analogue), 13N ammonia (freely diffusing agent, which gets trapped inside the cell after conversion through glutamine synthase to 13N-glutamine) or 15O water (freely diffusing agent).

**Declarations of interest**
None.

**References**


**Figure 1.** Coronary computed tomography angiography (CCTA): cross-section (A) and 3-dimensional reconstruction (B) showing the anomalous origin of left circumflex coronary artery from the right sinus of Valsalva with retro-aortic course.

**Figure 2.** Coronary computed tomography angiography (CCTA): 3-dimensional color reconstruction showing precisely the anomalous origin of left circumflex coronary artery from the right sinus of Valsalva next to the normal origin of right coronary artery.

**Figure 3.** Myocardial perfusion scintigraphy — SPECT: (A) left ventricular (LV) slices, (B) bull’s eyes (polar maps). Inducible perfusion deficits in lateral and infero-lateral wall, and in anterior and antero-lateral wall, involving 14% of the total LV myocardium.