Anomalous origin and extrinsic stenosis of left coronary principal artery. Beware of sudden death

Anomalia odejścia i zewnętrzne zwężenie lewej tętnicy wieńcowej. Ostrzeżenie przed nagłym zgonem

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A 47-year-old man with a history of hypercholesterolaemia, a former smoker, reported atypical chest pain. The electrocardiograms, blood analysis, biomarker seriation, chest radiography, and echocardiography showed no alteration. We performed a stress test that was inconclusive because it did not reach the programmed heart rate, showing no evidence of ischaemia. We performed a coronary computed tomography (CT), which revealed a congenital anomaly at the output of the left coronary artery (LCA). This artery originated from the Valsalva right sinus (Figs. 1, 2) with an initial path between the root of the aorta and pulmonary trunk, and with evidence of compression of the LCA between the two structures (Figs. 3, 4). We indicate performing coronarography, which revealed extrinsic compression of the LCA in its proximal portion, without other coronary lesions, and consequently the patient was referred to cardiac surgery. Anomalous origin of the LCA in the right Valsalva sinus is rare, with an estimated prevalence of 0.3–1.3% and an elevated incidence of death before the second decade of life. The course between the aorta and the pulmonary artery can cause coronary artery compression and can even lead to sudden death, especially if the subject exercises intensely, due to reduced coronary flow. The diagnosis can be complicated because the initial exams and ischaemia detection tests are frequently normal. The preferred diagnostic test to examine this anomaly seems to be CT coronary and magnetic resonance angiography. The CT provides higher spatial resolution to visualise the entire path of the coronary arteries and its route. Surgical correction is the elective treatment for patients with this anomaly, due to high risk of sudden cardiac death. Reimplantation of the LCA into the left coronary sinus, coronary artery bypass grafting, or unroofing of the intramural component are the surgery options.

Figure 1. Cardiac computed tomography reconstruction, three dimensional, with suppressed image of pulmonary artery. The origin of the right and left principal coronary arteries is the Valsalva right sinus. The left anterior descending artery (LAD) and left circumflex coronary artery (CX) are less developed compared with right coronary artery (RCA); LCA — left coronary artery; AR — aortic root

Figure 2. Cardiac computed tomography reconstruction, three dimensional, with suppressed image of pulmonary artery. Axial image of the birth of the left and right principal arteries. Notable stenosis of left coronary artery (LCA); RCA — right coronary artery

Figure 3. Cardiac computed tomography. The left coronary artery (LCA) is compressed by the pulmonary artery and aorta root (AR); PAT — pulmonary artery trunk; RCA — right coronary artery

Figure 4. Cardiac computed tomography. Three veils of aortic valve are observed, with both Valsalva sinuses. The two principal coronary arteries arise from the right coronary sinus, left principal coronary artery compression by the pulmonary artery trunk (PAT) and aortic root (AR) can also be seen; LCA and RCA — left and right coronary artery; LSV and RSV — left and right sinus of Valsalva

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