Anomalous origin of the left circumflex artery from right sinus of Valsalva: a rare case but with great clinical relevance

Cristina Poleggi[a], Davide Restelli[a], Alfredo Luongo[a], Federica Cocuzza[a], Francesca Campanella[a], Mennili Sara[b], Alessio Currò[c], Antonino Recupero[a], Michele Scarano[b]

[a] *BIOMORF Department, Section of Cardiology, University of Messina, Messina, Italy*
[b] *Cardiology Unit of Madonna del Soccorso Hospital, San Benedetto del Tronto, Italy*
[c] *Cardiac Intensive Care, IRCCS-Piemonte Hospital, Messina, Italy*

ARTICLE INFO

Article history:
Submitted: 22. 5. 2022
Accepted: 29. 5. 2022
Available online: 20. 2. 2023

Klíčová slova:
Anomálie koronárních cév
CT angiografie
Ischemie myokardu

SOUHRN

Anomální odstup z a. circumflexa se vyskytuje vzácně a běžně není příčinou infarktu myokardu. Popisujeme případ 61letého muže, který se dostavil na vyšetření s paroxysmální supraventrikulární tachykardií a atypickou bolestí na hrudi. Po odeznění arytmie prokázal EKG záznam abnormality vlny T, přičemž hodnoty srdečních enzymů byly přechodně zvýšené. Koronarografie sice prokázala anomální odstup LCx z pravého Valsalvova sinu, avšak žádné obturující aterosklerotické změny. Kazuistika se zabývá možnou souvislostí mezi vrozenou koronární anomálií a klinickými projevy pacienta.

Keywords:
Computed tomography angiography
Coronary vessel anomalies
Myocardial ischemia

ABSTRACT

Anomalous origin of coronary artery (CA) is rare and does not generally lead to myocardial infarction. We report a case of a 61-year-old man presented with paroxysmal supraventricular tachycardia and atypical chest pain. After arrhythmia subsided, ECG showed T wave abnormalities and transient cardiac enzymes were found to be elevated. Coronary angiography demostrated an anomalous origin of the left circumflex artery (LCx) from the right sinus of Valsalva but no obstructive atherosclerotic coronary lesions. The possible relation between the congenital coronary anomaly and the clinical manifestations of the patient is discussed.

Introduction

Most CA anomalies are incidentally detected during coronary angiography. The anomalous connection of the LCx to the right coronary artery (RCA) or sinus is considered the most frequent CA anomaly with an angiographic incidence of up to 0.67%. Among them, only those with an interarterial course are regarded as hidden conditions at risk of myocardial ischemia and sudden cardiac death (SCD).[1–8]

We report an uncommon case of anomalous origin of LCx from the right sinus of Valsalva and a retroaortic path causing myocardial ischemia.

Clinical case

A 61-year-old Caucasian man presented to the emergency department complaining of palpitations and chest discomfort for an hour at rest. He had history of hypertension, diabetes and a single episode of atrial fibrillation. His medical treatment consisted of olmesartan 40 mg/die. On admission blood pressure was 140/70 mmHg, pulse rate 160 beats per minute (BPM), respiratory rate 20 per minute and body temperature 36 °C. The 12-lead ECG demonstrated atrial flutter with a 2:1 conduction ratio resulting in a ventricular rate of 157 BPM and ST segment depression in leads V_4–V_6 (Fig. 1). Transthoracic echocardiogra-

Address: Michele Scarano, MD, Cardiology Unit, Emergency Dept, Hospital "Madonna del Soccorso", Via Silvio Pellico n. 32, 63039 San Benedetto del Tronto, Ascoli Piceno, Italy, e-mail: m.braveheart1978@gmail.com
DOI: 10.33678/cor.2022.063

Please cite this article as: Poleggia C, Restellia D, Luongo A, et al. Anomalous origin of the left circumflex artery from right sinus of Valsalva: a rare case but with great clinical relevance. Cor Vasa 2023;65:133–136.

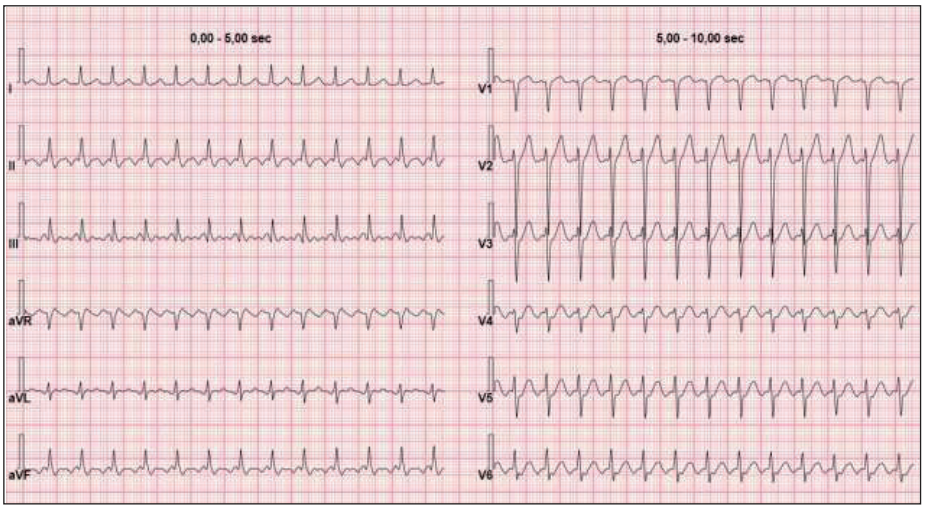

Fig. 1 – Initial 12-lead electrocardiogram (25 mm/s, 10.0 mm/mV).

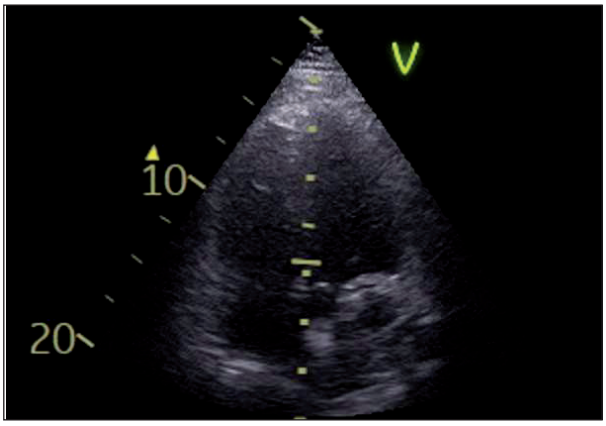

Fig. 2 – Transthoracic echocardiography in five-chamber apical view showing "RAC sign", a binary structure above the mitral valve plane into the atrioventricular groove overlapping the aortic root.

Fig. 4 – 3D volume rendered multi-detector CT image showing anomalous origin of LCx from right sinus of Valsalva with a retroaortic path.

phy revealed a preserved left ventricular ejection fraction and no segmental kinetic anomalies but a five-chamber apical view showed a "RAC sign", related typically to anomalous retroaortic course of the left coronary artery (Fig. 2).[9–28] Initial cardiac enzymes were in normal range. The patient was treated with intravenous infusion of amiodarone. He restored sinus rhythm in 2 hours and symptoms regressed completely, but the ECG taken after conversion

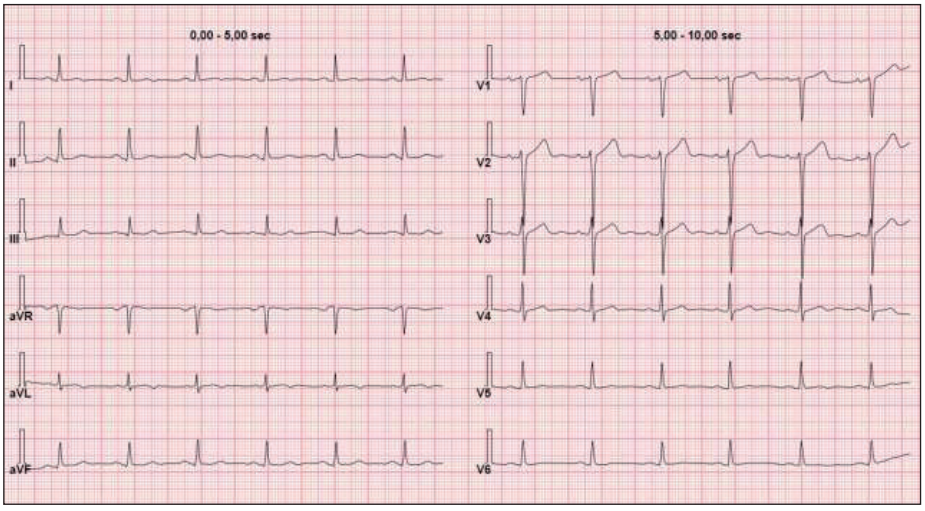

Fig. 3 – 12-lead electrocardiogram after sinus rhythm restoration showing negative T waves in I and aVL and flattened T waves in V_5 and V_6.

showed flattened T waves in leads V_5 and V_6, negative T waves in leads I and aVL (Fig. 3) and cardiac enzymes had transient increase: troponin T hs 95 pg/mL (n.v. <14), creatine kinase-MB 40 U/L (n.v. >20), myoglobin 96 ng/mL (n.v. 23–72). After the acute episode ended the patient underwent cardiac computed tomography angiography (CTA) with evidence of anomalous origin of LCx from the right sinus of Valsalva with a retroaortic course and a normal peripheral distribution (Fig. 4). Due to uncertain significance of ECG findings and cardiac enzyme increase, a coronary angiography was ordered and excluded obstructive atherosclerotic coronary lesions. Nuclear myocardial perfusion imaging revealed reversible small subsegmental perfusion defects in mid inferolateral wall and apical lateral wall. We established a medical treatment with beta-blocker and avoidance of intense physical activity. At follow-up 3 months later the patient did not refer any symptoms.

Discussion

Anomalous origin of a CA from the opposite sinus of Valsalva has been associated with myocardial ischemia and SCD. Our patient had an anomalous connection of the LCx branch to the right sinus of Valsalva with a retroaortic course which is considered the most frequent CA anomaly. Although this anomaly is usually considered benign, cases of association with SCD, myocardial infarction and angina pectoris in the absence of atherosclerotic lesions have been reported.[1,2] The factor responsible for this pathogenicity could be high orifice, ostial stenosis, slit-like/fish-mouth-shaped orifice, acute-angle take-off, intramural course and hypoplasia of the proximal coronary artery. Cardiac CTA did not reveal any of these characteristics in this patient. Thus, we hypothesized that the increased cardiac output and expansion of the great vessels during tachycardia or physical exertion could cause compression of the retroaortic segment or angling at its origin, narrowing the ostium to a slit and causing ischemia, a mechanism that has been reported in some studies.[29,30]

ECG abnormalities, especially ST segment depression, are well documented in literature during supraventricular tachycardia as a response to pacing-induced stress. These changes are usually diffused and disappear after conversion to sinus rhythm.[31] In this case, ST segment depression appeared in leads V_4–V_6 during tachycardia while flattened and negative T waves in leads V_5, V_6, I and aVL were seen hours later, accompanied by cardiac enzyme buildup. Due to these findings we decided to exclude any coronary stenosis with a coronary angiography. As the epicardial coronary arteries did not show any pathology, we suggest that the patient had transient ischemia due to LCx anomaly. Last step to confirm myocardial ischemia in case of CA anomalies is non-pharmacological functional imaging (we used nuclear study), as recommended by guidelines.[32]

As for the management of the anomalous aortic origin of a CA in adults, surgery is recommended as class IC in patients with typical angina symptoms who present with evidence of stress-induced myocardial ischemia in a matching territory or high-risk anatomy.[32,33] Our patient had evidence of stress-induced ischemia but he did not have clear manifestations of angina. He presented chest discomfort during supraventricular arrhythmia that remitted when sinus rhythm was restored. He did not refer any previous episode of angina. Among this type of CA anomalies it is known that interarterial path is most commonly related with fatal outcome while in retroaortic path SCD is rarely reported.[34] The age of our patient together with the retroaortic course of CA anomaly and the fact that his condition did not interfere with a normal life motivated us to use a conservative approach.

Conclusion

The fact that CA anomalies include many different entities and that any group has collected a large enough series to clarify the natural prognosis of each entity may contribute to our difficulty in the clinical identification and management of these conditions. We report a case of anomalous origin of LCx from right sinus of Valsalva causing transient myocardial ischemia in a patient that has always been asymptomatic. This anomaly has been and continues to be considered benign, nevertheless we suggest to judge the clinical significance of this kind of CA anomaly on a case-by-case integrated approach after exclusion of all other possible causes of signs or symptoms.

References

1. Corrado D, Penelli T, Piovesana P et al. Anomalous origin of the left circumflex coronary artery from the right aortic sinus of Valsalva and sudden death. Cardiovasc Pathol 1994;3:269–271.
2. Piovesana P, Corrado D, Verlato R, et al. Morbidity associated with anomalous origin of the left circumflex coronary artery from the right aortic sinus. Am J Cardiol 1989;63:762–763.
3. Sovová M, Sovová E, Sluka M, et al. Complicated course of anomalous origin of right coronary artery in the left sinus of Valsava in a professional ice hockey player. A case report. Cor Vasa 2020;62:431–434.
4. Fendrychová V, Ondrášek J, Zatočil T, et al. Anomalous left coronary artery from the pulmonary artery: A rare case diagnosed in an adult. Cor Vasa 2018;60:e412–e417.
5. Dattilo G, Lamari A, Messina F, et al. The chance finding at multislice computed tomography coronary angiography of an ectopic origin of the left circumflex coronary artery from the right sinus of Valsalva. Int J Cardiol 2011;149:e43–e46.
6. Dattilo G, Carerj S, Lamari A, et al. The chance finding at multislice computed tomography coronary angiography of myocardial bridging. Int J Cardiol 2012;154:e21–e23.
7. Gallo M, Rizzati F, Padalino M, et al. Anomalous origin of right coronary artery from pulmonary artery with aneurysmal coronary arteries. Cor Vasa 2016;58:e515–e517.
8. Zahradníková S, Buděšínský T, Knot J. An anomalous origin and course of left anterior descending artery. Cor Vasa 2020;62:452–456.
9. Vatrano M, Orlando L, Cassadonte F, et al. Triple oral combination therapy in patients with idiopathic pulmonary arterial hypertension and recurrent vessel dissection of inoperable pulmonary artery aneurysm. Cor Vasa 2021;63:726–731.
10. Widimský J. The role of arterial hypertension in the primary prevention of stroke. Cor Vasa 2016;58:e279–e286.
11. Imbalzano E, Scarpelli M, Mandraffino G, et al. Combination therapy with aliskiren versus ramipril or losartan added to

conventional therapy in patients with type 2 diabetes mellitus, uncontrolled hypertension and microalbuminuria. J Renin Angiotensin Aldosterone Syst 2015;16:956–964.
12. Imbalzano E, Vatrano M, Mandraffino G, et al. Arterial stiffness as a predictor of recovery of left ventricular systolic function after acute myocardial infarction treated with primary percutaneous coronary intervention. Int J Cardiovasc Imaging 2015;31:1545–1551.
13. Imbalzano E, Saitta A, Lamari A, et al. Echo-Doppler evaluation of recent onset chronic venous insufficiency in elderly patients: does the heart have a role? Recenti Prog Med 2013;104:569–573.
14. Vícha M, Skála T, Benešová K, et al. Review of non-insulin antidiabetic pharmacotherapy in patients with heart failure diabetes mellitus in the Czech Republic in 2018. Cor Vasa 2022;64:20–24.
15. Kubíčková M, Bis J. Type 2 diabetes and heart failure – how to optimize cooperation of cardiologist and diabetologist. Cor Vasa 2021;63:373–377.
16. Vadivelu R, Vijayvergiya R. Panvascular risk factor – Diabetes. Cor Vasa 2018;60:e18–e29.
17. Dattilo G, Lamari A, Crosca S, et al. Correlation between insulin resistance and endothelial dysfunction assessed by flow-mediated dilation. Recenti Prog Med 2012;103:328–332.
18. Apsite K, Lurina B, Tupahins A, et al. Atrial fibrillation, oral anticoagulants and health related quality of life. Cor Vasa 2018;60:e597–e602.
19. Patanè S, Marte F, Dattilo G. Intermittent changing axis deviation during acute myocarditis. Int J Cardiol 2010;145:e13–e16.
20. Casale M, Mezzetti M, Gigliotti De Fazio M, et al. Usefulness of Sacubitril/Valsartan in reduction of atrial fibrillation burden in a patient with ICD delivering inappropriate therapies. A new possibility? Cor Vasa 2020;62:336–339.
21. Casale M, Correale M, Laterra G, et al. Effects of Sacubitril/Valsartan in Patients with High Arrhythmic Risk and an ICD: A Longitudinal Study. Clin Drug Investig 2021;41:169–176.
22. Casale M, Mezzetti M, Gigliotti De Fazio M, et al. Novel active fixation lead guided by electrical delay can improve response to cardiac resynchronization therapy in heart failure. ESC Heart Fail 2022;9:146–154.
23. Dattilo G, Falanga G, Casale M, et al. Oral Anticoagulants: Old and New Therapy. In: Berhardt LV (Ed.). Advances in Medicine and Biology. Volume 83. New York: Nova Science Publishers, Inc., 2015:13–70.
24. Patanè S, Marte F, Sturiale M, et al. Atrial flutter, ventricular tachycardia and changing axis deviation associated with scleroderma. Int J Cardiol 2011;153:e25–e28.
25. Patanè S, Marte F, Dattilo G, et al. Acute myocardial infarction and left bundle branch block with changing axis deviation. Int J Cardiol 2012;154:e47–e49.
26. Arseniou AA, Nikas DN, Stamatis KV, et al. A groove crossing the aortic root. Hellenic J Cardiol 2022;64:101–103.
27. Lo Gullo A, Rodríguez-Carrio J, Aragona CO, et al. Subclinical impairment of myocardial and endothelial functionality in very early psoriatic and rheumatoid arthritis patients: Association with vitamin D and inflammation. Atherosclerosis 2018;271:214–222.
28. López VC, Blanco P. Retroaortic Anomalous Coronary Artery Visualization on Transthoracic Echocardiogram. J Cardiovasc Echogr 2021;31:179–180.
29. Barriales Villa R, Díaz Molina B, Arias Castaño JC, et al. [Myocardial ischemia caused by an anomalous circumflex coronary artery.] Rev Esp Cardiol 2002;55:200–202. [Article in Spanish]
30. Aydin M, Ozeren A, Peksoy I, et al. Myocardial ischemia caused by a coronary anomaly: left circumflex coronary artery arising from right sinus of Valsalva. Tex Heart Inst J 2004;31:273–275.
31. Bukkapatnam RN, Robinson M, Turnipseed S, et al. Relationship of myocardial ischemia and injury to coronary artery disease in patients with supraventricular tachycardia. Am J Cardiol 2010;106:374–377.
32. Baumgartner H, De Backer J, Babu-Narayan SV, et al. 2020 ESC Guidelines for the management of adult congenital heart disease. The Task Force for the management of adult congenital heart disease of the European Society of Cardiology (ESC). Eur Heart J 2021;42:563–645.
33. Warnes CA, Williams RG, Bashore TM, et al. ACC/AHA 2008 Guidelines for the Management of Adults With Congenital Heart Disease. Circulation 2008;118:e714–e833.
34. Pérez-Pomares JM, de la Pompa JL, Franco D, et al. Congenital coronary artery anomalies: a bridge from embryology to anatomy and pathophysiology – a position statement of the development, anatomy, and pathology ESC Working Group. Cardiovasc Res 2016;109:204–216.