ESC European Society of Cardiology
European Heart Journal - Case Reports
doi:10.1093/ehjcr/ytab052

CASE REPORT
Congenital heart disease

Incidental finding and conservative management of left main coronary atresia in an adult patient: a case report

Giuseppe Talanas *, Giulia Corda, Guido Parodi , and Michele Portoghese

Department of Cardiology, Sassari University Hospital, Via Enrico De Nicola, 07100 Sassari, Italy

For the podcast associated with this article, please visit https://academic.oup.com/ehjcr/pages/podcast

Received 1 June 2020; first decision 23 June 2020; accepted 16 November 2020

Background Left main (LM) coronary atresia (LMCA) is a rare coronary anomaly where the LM is congenitally absent and a variable clinical spectrum can follow. The diagnosis of LMCA is generally made in youth because of the development of symptoms, but very rarely in adulthood. In symptomatic patients, surgical revascularization is recommended, whereas, in asymptomatic patients with LMCA and without inducible myocardial ischaemia, preventive surgical treatment is controversial.

Case summary A 58-year-old male patient with aortic ectasia detected during an echocardiogram performed to evaluate a hypertension-related preclinical cardiac damage and, due to this finding, an echocardiographic follow-up was suggested. Three years later, he was admitted to undergo coronary angiography (CA) after the computed tomography finding of a suspected occlusion of the LM with collateral circulation from right coronary artery (RCA) to left anterior descending and circumflex arteries. CA confirmed an LMCA and the RCA provided blood supply to the left coronary artery through collaterals whose calibre was similar to that of the target left-sided vessels. No obstructive coronary artery disease was detected. In order to detect potential myocardial ischaemia, a technetium-tetrofosmin cardiac single-photon emission computed tomography during maximal exercise-stress test was performed and it did not show a perfusion defect. Medical management with scheduled follow-up visits was deemed to be the best therapeutic option.

Discussion LMCA is a rare anomaly where LM is absent and the RCA provides collateral circulation for left coronary artery. In asymptomatic patients, preventive surgical treatment is controversial.

Keywords Left main coronary atresia • Computed tomography • Coronary angiography • Myocardial scintigraphy • Conservative management • Case report

Learning points

- Left main coronary atresia is an extremely rare coronary artery anomaly where the left main is absent and the left coronary artery is supplied through collaterals from the right coronary artery. The diagnosis is generally made in youth because of the presence of symptoms, although it is very rarely reported in adulthood.
- In symptomatic patients, coronary artery bypass surgery or surgical coronary repair are recommended. However, in adult patients without symptoms or evidence of inducible myocardial ischaemia, conservative management with optimal medical therapy is reasonable.

* Corresponding author. Tel: +39 079 2061521, Fax: +39 079 210512, Email: giuseppe.talanas@aousassari.it
Handling Editor: Monika Arzanauskaite
Peer-reviewers: Alberto Bouzas-Mosquera; Christoph Jensen and John Kanakakis
Compliance Editor: Daniel Tardo
Supplementary Material Editor: Vishal Shahil Mehta
© The Author(s) 2021. Published by Oxford University Press on behalf of the European Society of Cardiology.
This is an Open Access article distributed under the terms of the Creative Commons Attribution Non-Commercial License (http://creativecommons.org/licenses/by-nc/4.0/), which permits non-commercial re-use, distribution, and reproduction in any medium, provided the original work is properly cited. For commercial re-use, please contact journals.permissions@oup.com

Introduction

Left main (left main) coronary atresia (LMCA) is an extremely rare coronary artery anomaly where the LM is absent and left anterior descending (LAD) and circumflex (Cx) arteries, connected as usual, end blindly proximally and receive their blood supply from the right coronary artery (RCA) through collateral circulation.[1] The diagnosis of LMCA is generally made in childhood or adolescence because of the presence of symptoms, although it is very rarely reported in adulthood.[2] The clinical spectrum of the LMCA can range from asymptomatic forms to symptoms such as angina, syncope, or sudden cardiac death.[3] In symptomatic patients, coronary artery bypass graft (CABG) was the standard treatment of LMCA, but in more recent years, surgical anatomical coronary repair (i.e. osteoplasty) is gradually acquiring importance over CABG.[4] However, in asymptomatic patients with LMCA and without evidence of inducible myocardial ischaemia, preventive surgical treatment is controversial.

Timeline

3 years earlier	During a routine echocardiogram performed to evaluate a preclinical cardiac damage, dilation of the ascending aorta (45 mm) was found for the first time. Yearly echocardiographic follow-up was suggested.
2 weeks before hospital admission	Aortic computed tomography scan confirmed a stable dilatation of the ascending aorta and a suspected occlusion of the left main (LM). The patient was asymptomatic despite an active lifestyle.
Hospital admission	Coronary angiography showed an LM coronary atresia and the right coronary artery provided blood supply to the left coronary artery through collaterals whose calibre was similar to that of the target left-sided vessels.
3 days after hospital admission	Cardiac single-photon emission computed tomography during maximal exercise-stress test did not show a perfusion defect. Optimal medical therapy was recommended.
4 days after hospital admission	Hospital discharge with targeted medical therapy to prevent atherosclerosis of the single patent vessel.
9-months clinical follow-up	The patient is asymptomatic and in a stable clinical condition.

Case presentation

In a 58-year-old man with a history of arterial hypertension and mild hypercholesterolaemia, an aortic ectasia was detected during an echocardiogram performed to evaluate for hypertension-related preclinical cardiac damage and, due to this finding, echocardiographic follow-up was suggested. Three years later, the patient was admitted for coronary angiography (CA) after the computed tomography (CT) finding of a suspected occlusion of the LM with collateral circulation from RCA to LAD and Cx (*Figure 1A–D*). The indication for CT consisted of providing a more precise evaluation of the ascending aorta tracts which were not adequately explored by echocardiography. The patient was asymptomatic despite an active lifestyle and the physical examination was unremarkable. Cardiac auscultation revealed that cardiac sounds were rhythmic and no additional sounds (3rd or 4th sound) or bruits were audible; peripheral pulses were palpable, and no vascular bruits were heard as well as no clinical signs of heart failure were observed. Electrocardiogram was normal. Echocardiogram showed left ventricular concentric remodelling with normal systolic function (ejection fraction 65%), dilatation of the ascending aorta (45 mm) and a tricuspid aortic valve with mild aortic regurgitation. Routine laboratory tests were within normal limits.

CA showed LMCA as demonstrated by contrast injection into the left sinus of Valsalva (*Figure 1E*). The left coronary artery was retrogradely supplied from the RCA through Rentrop 3 grade coronary collaterals whose calibre was similar to that of the target left-sided vessels (*Figure 1F–G*, *Video 1*). No obstructive coronary artery disease (CAD) was detected. In order to exclude potential myocardial ischaemia, a technetium-tetrofosmin cardiac single-photon emission computed tomography (SPECT) was performed during maximal exercise-stress testing, which did not show a perfusion defect (*Figure 1H*). During weekly Heart Team meetings, preventative CABG was not deemed to be indicated since the patient was asymptomatic, without evidence of inducible ischaemia. The patient was discharged with targeted medical therapy to prevent atherosclerosis of this single coronary artery. Clinical follow-up with periodic transthoracic echocardiograms was planned; however, with the plan to follow-up earlier in the case of new symptom development. At 9-month follow-up, the patient was asymptomatic and in a stable clinical condition.

Discussion

LMCA is an extremely rare congenital disorder where LM is absent and the LAD and Cx are connected as usual but proximally end blindly. RCA provides collateral circulation for the left coronary artery.[1] This condition often coexists with supra- and subvalvular aortic stenosis or other congenital cardiac defects.[5,6] Usually patients develop symptoms during infancy, childhood, or adolescence, while, in adulthood, LMCA is very rarely reported and, when described, represents an incidental finding.[2,7] When symptoms are present, CABG was the historically recommended treatment, although, in more recent years, surgical coronary repair is increasingly being used.[3,4] However, when the patient is asymptomatic and inducible myocardial ischaemia has not been documented, the benefit/risk ratio of preventive cardiac surgery has not been demonstrated. Since our patient was asymptomatic, without CAD, the collateral circulation from RCA was well-developed and myocardial scintigraphy during maximal exercise-stress test did not show inducible myocardial ischaemia, medical management with targeted medical therapy including atorvastatin 80 mg to prevent atherosclerosis developing within this single coronary artery was deemed to be the best option. In a similar case, Rampin *et al.*[8] showed the absence of perfusion defects by cardiac

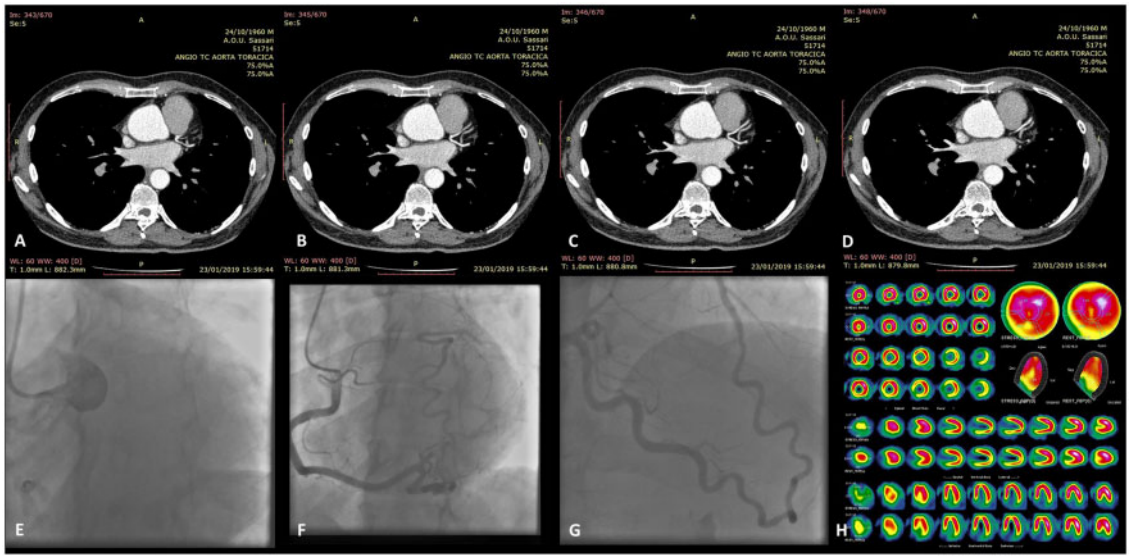
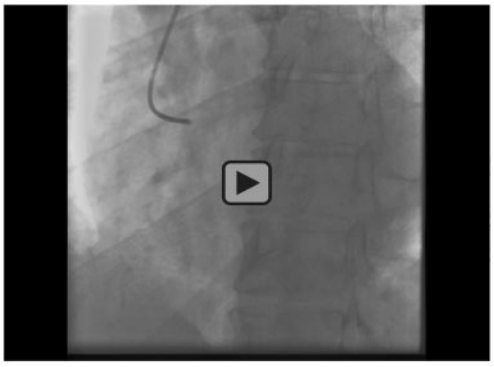


Figure 1 (*A–D*) Computed tomography images with progressive filling of contrast are shown; in the left main coronary atresia, the left anterior descending artery, and circumflex artery are connected as usual but proximally end blindly. (*E*) Injection of contrast in the left sinus of Valsalva confirming left main coronary atresia. (*F*) The right coronary artery provides a collateral circulation for the left coronary artery. (*G*) The calibre of coronary collaterals was similar to that of the target left-sided vessels. (*H*) Myocardial single-photon emission computed tomography during maximal exercise-stress test did not show perfusion defects as compared to rest.



Video 1 Angiography of the right coronary artery allowed to visualize the left coronary artery through a developed collateral circulation.

SPECT in a patient with LMCA; however, in that case, since the patient was symptomatic for exertional dyspnoea, CABG was performed. Moreover, a conservative strategy with optimal medical therapy in a further patient with LMCA and the absence of substantial perfusion defect at cardiac SPECT has been described with a favourable mid-term follow-up.[9]

Conclusion

LMCA is an extremely rare coronary artery anomaly and the diagnosis is hardly reported in adult patients. Since our patient was asymptomatic, without CAD and inducible myocardial ischaemia, CABG was deemed to not be indicated by the Heart Team. Targeted medical therapy to prevent atherosclerosis of this single coronary artery with yearly follow-up visits including echocardiogram was deemed to be the best therapeutic option.

Lead author biography

Giuseppe Talanas earned his medical degree at the University of Sassari, and Specialization in Cardiology at the University of Parma. From 2009, he is working as an interventional cardiologist in Sassari (Italy). His principal field of interest is percutaneous coronary intervention, intracorononary imaging, and CTO-PCI. Among structural heart interventions the field of interest is TAVI and left atrial appendage occlusion.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

Acknowledgements

We are indebted with Anna Piga for CT images.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: none declared.

Funding: none declared.

References

1. Fortuin NJ, Roberts WC. Congenital atresia of the left coronary artery. *Am J Med* 1971;**50**:385–389.
2. Koh E, Nakagawa M, Hamaoka K, Sawada T, Oga K. Congenital atresia of the left coronary ostium: diagnosis and surgical treatment. *Pediatr Cardiol* 1989;**10**:159–162.
3. Musiani A, Cernigliaro C, Sansa M, Maselli D, De Gasperis C. Left main coronary atresia: literature review and therapeutical considerations. *Eur J Cardiothorac Surg* 1997;**11**:505–514.
4. Albadi W, Martin A, Kreitmann B, Roubertie F. Anatomic repair of a left coronary artery main stem atresia. *Can J Cardiol* 2019;**35**:1419.e5–1419.e7.
5. Sabzi F, Heidari A, Faraji R. Congenital combined atresia of the left main coronary with supravalvar aortic stenosis. *Ann Card Anaesth* 2018;**21**:423–426.
6. Vidne BA, Nili M, Aygen M, Levy MG. Congenital atresia of the left main coronary ostium. *Scand J Thorac Cardiovasc Surg* 1979;**13**:37–40.
7. Rubio Alonso B, Jurado Roman A, Alonso Charterina S. Left main coronary atresia in an asymptomatic elderly adult. *Rev Esp Cardiol* 2015;**68**:436.
8. Rampin L, Rinuncini M, Zuin M, Rigatelli G, Roncon L, Colletti PM et al. Left main coronary artery atresia in young man examined with 99mTc-Tetrofosmin SPECT: a rare and challenging anomaly. *Clin Nucl Med* 2016;**41**: e452–e453.
9. Graidis C, Dimitriadis D, Psifos V. Congenital atresia of the left main coronary artery in an adult: an extremely rare anomaly. *J Invasive Cardiol* 2012;**24**: E325–E327.