CASE REPORT

Unusual congenital coronary artery anomaly in a young adult presenting as sudden cardiac arrest

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SUMMARY

A previously asymptomatic young female with no previous medical or cardiac history collapsed during indoor exercise. A portable automatic external defibrillator showed a shockable rhythm. She received multiple electrical shocks with return to normal sinus rhythm without ischaemic ECG changes. Her troponin level was mildly elevated. A transthoracic echocardiogram revealed moderately reduced left ventricular ejection fraction with global hypokinesis. During emergent coronary angiography, the left main coronary artery could not be found. The right coronary artery was normal with robust collaterals to the entire left coronary circulation extending to the left main coronary artery, but did not fill the ostium. Coronary CT angiogram confirmed nearly complete absence of the left main coronary artery ostium. A diagnosis of left main coronary artery atresia was made. Patient underwent successful two vessel coronary artery bypass grafting. She continues to do well 1 year postoperatively.

BACKGROUND

Left main coronary artery atresia (LMCAA) is a very rare congenital anomaly.¹ Previously reported cases have shown a male predominance with a mean age of 54 years.² Given the low prevalence of LMCAA, with only 34 adult cases reported in literature and minimal understanding about this entity, it is important to report the demographics of new cases and describe their clinical presentation and management.^{1 2} In addition, there is limited data about post-treatment follow-up in patients with LMCAA.² We present a case of a young female with LMCAA manifested as sudden cardiac death (SCD) with 1-year follow-up and description of quality of life.

CASE PRESENTATION

A 28-year-old obese female with no previous medical or cardiac history was admitted after collapsing during indoor exercise. A portable automatic external defibrillator found a shockable rhythm. She received multiple electrical shocks and was successfully converted to normal sinus rhythm. She had no history of alcohol or drug abuse and had no family history of cardiac disease or sudden cardiac death.

INVESTIGATIONS

ECG on arrival showed only sinus tachycardia with a heart rate of 112 beats per minute. The

PR interval and QTc measured 118 ms and 436 ms, respectively. A transthoracic echocardiogram revealed moderately reduced left ventricular ejection fraction with global hypokinesis and no significant valve disease. During emergent cardiac catheterisation, the left main coronary artery could not be engaged and could not be identified despite proximal aortography (figure 1, video 1). However, the right coronary artery (RCA) was normal and revealed robust collaterals to the left circumflex and left anterior descending arteries extending to nearly the ostium of the left main artery (figure 2, video 2). No atherosclerotic coronary artery disease (CAD) was observed. A subsequent CT angiogram showed moderate distal RCA collaterals supplying left coronary artery (LCA) as well as the left main artery nearly absent, with only a thin sliver of contrast from the left coronary cusp, thus confirming the diagnosis of LMCAA (figures 3A, B and 4). Transoesophageal echocardiography was also performed and did not reveal any other structural heart abnormality or could the left main origin be identified with this imaging modality.

DIFFERENTIAL DIAGNOSIS

Given the patient's presentation and demographics, the initial differential diagnosis included congenital arrhythmias such as prolonged QT syndrome, hypertrophic cardiomyopathy or anomalous coronary artery. Premature CAD, acute myocarditis or idiopathic cardiomyopathy were also considered.

TREATMENT

The patient underwent a successful two vessel CABG with a left internal mammary artery to the left anterior descending artery and left radial artery graft to the obtuse marginal.

OUTCOME AND FOLLOW-UP

She recovered well and was sent home in stable condition. Four months postdischarge, the patient was evaluated in the hospital for chest pain and underwent exercise nuclear stress test which showed normal left ventricular ejection fraction of 67% and no evidence of infarct or ischaemia. One year post-CABG, she is asymptomatic and back to her normal activities of daily living including regular exercise.

DISCUSSION

LMCAA is an extraordinarily rare condition with only 34 adult cases reported in the literature.² Patients with this condition have absent left

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Figure 1 Aortogram showing the absence of ostial left main coronary artery (arrowhead) but clear the presence of ostial right coronary artery (arrow).

coronary ostium, or absent left main coronary artery trunk which on coronary CT can sometimes be seen as a string-like structure suggesting a rudimentary form of the left main coronary artery.⁵ The left coronary circulation is supplied by the RCA through collaterals. In paediatric patients, LMCAA usually coexists with other cardiac abnormalities such as valvular disease with the onset of symptoms early in life.⁴ In adults, LMCAA can be found in isolation and is usually asymptomatic early in adult

Video 1 Corresponding aortogram video of [figure 1](#) showing the absence of ostial left main coronary artery but clear presence of ostial right coronary artery.

Figure 2 Coronary angiogram in the left anterior oblique projection showing normal right coronary artery anatomy (arrow) with moderate right to left collaterals filling the left main coronary artery in retrograde fashion (arrowheads) primarily via the septal collateral system.

life.⁵ When symptomatic, the initial presentation is often sudden cardiac death. Because of its rarity, it is usually not part of the initial differential diagnosis for sudden cardiac death. However, when the condition is suspected, coronary angiography complemented with cardiac CT are diagnostic.^{3 6} The high risk of sudden cardiac death makes surgical correction with CABG as

Video 2 Corresponding left anterior oblique projection video of [figure 2](#) showing normal right coronary artery anatomy with moderate right to left collaterals filling the left main coronary artery in retrograde fashion primarily via the septal collateral system.

Figure 3 Cardiac CT angiography showing no ostial left main coronary artery (arrowheads in A and B) but the presence of ostial right coronal artery (arrows in A and B).

an adequate treatment strategy.^{1 2 4 7} A reasonable therapeutic alternative is ostioplasty with a patch in young patients with close proximity of the left coronary to the aorta, as in this case.^{8 9}

This case presented two major challenges. One was to determine the underlying cause of the patient's sudden cardiac arrest. The fact that the QTc interval was normal and no further arrhythmias postcardiac arrest were detected suggested that a primary arrhythmogenic arrest was less likely. Both transthoracic and transoesophageal echocardiograms ruled out other major structural heart issues; however, it was notable for moderate left ventricular dysfunction. Possible explanations for this last finding were acute myocardial stunning postcardiac arrest, acute myocarditis or idiopathic cardiomyopathy. Her repeat echocardiogram showed normalisation of left ventricular function with normal wall motion 1 week after admission and before CABG surgery, supporting postcardiac arrest stunned myocardium. The second challenge arose after coronary angiogram, which did not identify the ostial left main coronary artery. The question was then to determine if the condition was congenital versus acquired due to single vessel obstructive CAD. Given her lack of atherosclerotic risk factors and that coronary angiogram failed to find any other atherosclerotic disease, we favoured the diagnosis of LMCAA. Coronary CT angiography confirmed this diagnosis.

Figure 4 Coronal view of the CT angiography showing the absence of the left main coronary artery ostia and normal course of the left anterior descending artery.

Little is known about the embryology of this condition and the development of coronary collaterals. The presentation of LMCAA in neonates suggests that at least basic collaterals are present already at birth,⁷ but the effectiveness of the collaterals may be the key to survival into adulthood. In our patient, collateral circulation to the LCA was provided by distal branches of the RCA and primarily via the intramyocardial septal perforating arteries. As in two prior reported cases in literature, this pattern is frequently seen in young patients, but unlike both of those reported cases, the symptom at onset in our case was SCD and not angina. This feature makes our case the first of its kind.^{2 10 11} The biological pathway of collateral development would certainly be of interest not only in this rare condition but also in more common scenarios of acquired CAD. At this point, there is limited understanding about which patients will present with symptomatic ischaemia versus sudden cardiac death and the importance of collateral patterns. Some patients may be asymptomatic throughout their lives.

Learning points

- ▶ Left main coronary artery atresia (LMCAA) is a rare congenital coronary anomaly that can manifest as sudden cardiac death in a young female.
- ▶ Exercise-induced ischaemia may be the most common pathogenesis of sudden cardiac death in patients with LMCAA.
- ▶ Emergent coronary angiography complemented by CT angiography are diagnostic.
- ▶ Coronary artery bypass grafting is an adequate treatment strategy for LMCAA.
- ▶ Ostioplasty with a patch is a reasonable alternative.

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REFERENCES

- 1 Musiani A, Cernigliaro C, Sansa M, *et al*. Left main coronary artery atresia: literature review and therapeutical considerations. *Eur J Cardiothorac Surg* 1997;11:505–14.
- 2 Unzué L, García E, Parra FJ, *et al*. Congenital atresia of the left main coronary artery in an adult: a rare anomaly with an unfavorable prognosis. Review of the literature. *Cardiovasc Revasc Med* 2015;16:498–502.
- 3 Saito T, Motohashi M, Matsushima S, *et al*. Left main coronary artery atresia diagnosed by multidetector computed tomography. *Int J Cardiol* 2009;135:e27–e29.
- 4 Tanawuttiwat T, O'Neill BP, Schob AH, *et al*. Left main coronary atresia. *J Card Surg* 2013;28:37–46.
- 5 Rubio Alonso B, Jurado Román A, Alonso Charterina S. Left main coronary artery atresia in an asymptomatic elderly adult. *Rev Esp Cardiol* 2015;68:436.
- 6 Levisman J, Budoff M, Karlsberg R. Congenital atresia of the left main coronary artery: cardiac CT. *Catheter Cardiovasc Interv* 2009;74:465–7.
- 7 D'Souza TF, Samuel BP, Vettukattil JJ, *et al*. Surgical treatment of neonate with congenital left main coronary artery atresia. *Ann Thorac Surg* 2016;101:352–5.

- 8 Kaczorowski DJ, Sathanandam S, Ravishankar C, *et al*. Coronary ostioplasty for congenital atresia of the left main coronary artery ostium. *Ann Thorac Surg* 2012;94:1307–10.
- 9 Ohba M, Matsuo K, Kabasawa M, *et al*. Surgical angioplasty for ostial atresia of left main coronary artery in child. *Gen Thorac Cardiovasc Surg* 2015;63:239–41.
- 10 Nishida N, Chiba T, Ohtani M, *et al*. Two adult cases of congenital atresia of the left coronary ostium-comparison of a sudden death case with a long-term survival case. *Virchows Arch* 2005;447:742–6.
- 11 Fortuin NJ, Roberts WC. Congenital atresia of the left main coronary artery. *Am J Med* 1971;50:385–9.

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