CASE REPORT

Coronary spasm as the cause of myocardial ischaemia in a patient with anomalous origin of the left anterior descending artery from the proximal right coronary artery

Jun Nakazato, Kazuhito Hirata, Minoru Wake

Department of Cardiology,
Okinawa Chubu Hospital,
Uruma, Japan

Correspondence to

Dr Jun Nakazato,
jun-na@rk9.so-net.ne.jp

Accepted 25 May 2014

SUMMARY

A 49-year-old woman developed angina at rest. A CT of the coronary artery revealed that the left anterior descending artery arose from the right coronary artery, and traversed between the aorta and pulmonary trunk. An exercise stress myocardial scintigraphy did not reproduce myocardial ischaemia or anginal symptoms. A coronary angiography did not show any atherosclerotic changes. Finally, an ergotamine provocation test for vasospasm revealed diffuse severe spasm in the right coronary artery and the left anterior descending artery. Surgical correction of the anomaly was deferred and the patient was managed with medications to control spasm with good clinical outcome.

BACKGROUND

Anomalous origin of a coronary artery is a well-established cause of sudden death. The mechanism of myocardial ischaemia or sudden death has been attributed to a slit-like coronary orifice, and systolic squeezing or compression of the anomalous coronary artery, which courses between the aorta and pulmonary trunk. In the present case, myocardial ischaemia was not reproduced with an exercise stress scintigraphy but a coronary vasospasm appeared to be an important cause of her resting angina. We consider that the coronary vasospasm is one of the important causes of myocardial ischaemia and sudden death in patients with anomalous origin of a coronary artery.

CASE PRESENTATION

A 49-year-old woman was taken to the emergency room (ER) reporting of acute onset of dyspnoea, chest discomfort and cold sweat at rest. This was the first episode of chest pain for her. The patient had no significant risk factors for coronary artery disease except for positive family history (her father died of acute myocardial infarction at the age of 70).

Initial vital signs showed a blood pressure of 80/- mm Hg and a heart rate of 108 bpm. An auscultation of the heart and lungs were normal. There was neither jugular venous distention nor peripheral enema.

A 12-lead ECG on admission showed normal sinus rhythm with a ST segment depression in II, III, aVF and V4–6 (figure 1). A chest X-ray showed neither cardiomegaly nor enlargement of the

mediastinum. A bedside transthoracic echocardiography in the ER showed no wall motion abnormalities in the left ventricle, and valvular structures and function were normal. Thirty minutes later, ST segment returned to baseline and her symptom was relieved spontaneously. Creatine kinase was 68 IU/L on admission and was 48 IU/L 7 h later. Troponin T was not elevated either.

After admission, a CT of the heart was performed, and revealed that the left anterior descending (LAD) coronary artery arose from the proximal portion of the right coronary artery (RCA) and it traversed between the aorta and the pulmonary artery (figure 2). There was no significant atherosclerotic narrowing in major epicardial coronary arteries. An exercise stress myocardial scintigraphy to identify possible compression between the aorta and the pulmonary trunk did not show any evidence of exercise-induced myocardial ischaemia

Figure 1 The initial ECG showing ST depression in II, III, aVF leads in the emergency room.

CrossMark

To cite: Nakazato J, Hirata K, Wake M. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2014-204408

Figure 2 A CT angiography of the heart showing the left anterior descending (LAD) artery arising from right coronary artery. Arrows indicate that LAD courses between the ascending aorta and the pulmonary trunk. Abbreviations: Left circumflex artery (LCX), left anterior descending (LAD), aorta (AO), pulmonary artery (PA), right coronary artery (RCA).

(figure 3). Because her symptom occurred at rest, the possibility of vasospastic angina was suggested. At the time of coronary angiography, an ergotamine provocation test was performed after confirming that there was no significant atherosclerotic narrowing. A diffuse severe spasm was induced in the RCA and the LAD (figure 4), and typical chest pain with ST depression on 12-lead ECG were reproduced (figure 5).

The patient was diagnosed as having coronary vasospastic angina superimposed on anomalous origin of the left anterior descending artery.

TREATMENT

Surgical management was deferred and the patient was followed up with medications (isosorbide, nicorandil and benidipine) to control vasospastic angina.

OUTCOME AND FOLLOW-UP

Since the initiation of medications to control vasospasm, the patient has been stable and asymptomatic for 18 months.

DISCUSSION

The most common type of anomalous aortic origin is the left circumflex coronary artery arising from the right sinus of Valsalva or the RCA (0.37%).¹ The LAD originated from the RCA in 4 of 16 573 patients (0.02%) undergoing cardiac catheterisation.² Anomalous aortic origin of a coronary artery is established as a cause of sudden death. Frequently quoted autopsy reports have suggested 57% mortality for an anomalous left coronary artery (LCA) originating from the right coronary sinus, and 25% mortality rate for RCA originating from the left coronary sinus.³ Several potential mechanisms have been proposed to explain myocardial ischaemia and sudden death in patients with anomalous aortic origin of a coronary artery. The oblique take-off of the anomalous artery produces a slit-like coronary orifice.^{3–5} Furthermore, the course of anomalous coronary artery between the aorta and pulmonary trunk has been proposed to lead to compression by these two great vessels.^{3–6}

However, these hypotheses explain only exercise related ischaemic events. Our patient experienced her symptoms of cardiac ischaemia at rest and not during exercise. The cause of angina at rest proved to be due to coronary vasospasm as suggested by strongly positive provocation test. There has been several case reports in which coronary vasospasm was documented in patients with anomalous aortic origin of the coronary artery.^{7–12} These results, as well as our observation in the present case, suggest that the coronary vasospasm is also one of the important causes of myocardial ischaemia and sudden death in patients with anomalous origin of a coronary artery. In

Figure 3 A stress myocardial scintigraphy did not show any evidence of myocardial ischaemia.

Figure 4 (A) Left anterior oblique view of the left anterior descending artery from right coronary artery. (B) An ergotamine provocation test for vasospasm revealed diffuse severe spasm in the right coronary artery and left anterior descending artery. (C) Coronary spasm was relieved with the administration of intracoronary nitroglycerin. Abbreviation: Left anterior descending (LAD)

addition, Nathan *et al*⁹ reported a case in which, after successful anatomical correction of the anomalous coronary artery, postoperative course was seriously complicated due to diffuse coronary vasospasm resulting in haemodynamic instability necessitating mechanical support in the early postoperative period. In view of this case report, surgical therapy and medical therapy to control vasospasm appeared to be very important. One possible mechanism of vasospasm is an accelerated

Figure 5 The 12-lead ECG during ergotamine testing showing ST depression in II, III, aVF V5 and V6 leads.

atherosclerosis at the site of mechanical stresses induced by compression between the aorta and pulmonary trunk, which may lead to a tendency to develop vasospasm.¹⁰

In conclusion, in addition to anatomical causes, a coronary vasospasm is an important cause of myocardial infarction and sudden death in patients with anomalous origin of the coronary artery. Therefore, provocation test for vasospasm should be considered in evaluation of those patients and medical management should include medications to control vasospasm.

Learning points

Anomalous aortic origin of coronary artery is established as a cause of sudden death. In addition to anatomical causes, vasospasm of the coronary artery is also an important cause of myocardial ischaemia and sudden death. We recommend performing a provocation test and using medications for vasospasm while evaluating and treating patients with anomalous origin of the coronary artery.

Contributors JN identified and managed the case. KH and MW advised the management of this case.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Yamanaka O, Hobbs RE. Coronary artery anomalies in 126,595 patients undergoing coronary arteriography. *Cathet Cardiovasc Diagn* 1990;21:28–40.
- 2 Yuksel S, Meric M, Soyulu K, *et al*. The primary anomalies of coronary artery origin and course: a coronary angiographic analysis of 16573 patients. *Exp Clin Cardiol* 2013;18:121–3.
- 3 Taylor AJ, Rongan KM, Virmani R. Sudden cardiac death associated with isolated congenital coronary artery anomalies. *J Am Coll Cardiol* 1992;20:640.
- 4 Cheitlin MD, DeCastro CM, McAllister HA. Sudden death as a complication of anomalous left coronary origin from the anterior sinus of Valsalva, a not-so-minor congenital anomaly. *Circulation* 1974;50:780–7.
- 5 Roberts WC, Siegel RJ, Zipes DP. Origin of the right coronary artery from the left sinus of Valsalva and its functional consequences: analysis of 10 necropsy patients. *Am J Cardiol* 1982;49:863–8.
- 6 Barth CW, Roberts WC. Left main coronary artery originating from the right sinus of Valsalva and coursing between the aorta and pulmonary trunk. *J Am Coll Cardiol* 1986;7:366–73.
- 7 Kubota Y, Monji T, Nakagawa H, *et al*. Anomalous origin of the left main coronary artery from the right aortic sinus of Valsalva with vasospastic angina. *Chest* 1991;100:1167–8.
- 8 Kaku B, Kanaya H, Ikeda M, *et al*. Acute inferior myocardial infarction and coronary spasm in a patient with an anomalous origin of the right coronary artery from the left sinus of Valsalva. *Jpn Circ J* 2000;64:641–3.
- 9 Nathan M, Singh M, Lakdawala N, *et al*. Anomalous left coronary artery from aortic with Prinzmetal's angina: a postoperative dilemma successfully managed. *Pediatr Cardiol* 2007;28:400–2.
- 10 Mercado JR, Coble WL Jr, Thompson PD. Premature atherosclerosis: a possible mechanism for symptoms in anomalous coronary arteries-report of case. *Conn Med* 2010;74:91–2.
- 11 Maddoux GL, Goss JE, Ramo BW, *et al*. Angina and vasospasm at rest in a patient with an anomalous left coronary system. *Cathet Cardiovasc Diagn* 1989;16:95–8.
- 12 Rossi PA, Rapeport K, Fisher EA, *et al*. Coronary artery spasm as a cause of chest pain in a patient with anomalous origin of the right coronary artery from the left sinus of Valsalva. *Tex Heart Inst J* 1988;15:124–7.

Copyright 2014 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow