Right coronary anomaly in a patient with myocarditis and cardiac arrest: a case report

Lisa S. Oberli¹, Laurent M. Haegeli¹,², and Bettina Heidecker¹*

¹University Hospital Zurich, Rämistrasse 100, 8091 Zurich, Switzerland; and ²Division of Cardiology, Kantonsspital Aarau, Tellstrasse 25, 5001 Aarau, Switzerland

Introduction

Management of coronary anomalies continues to be a controversial topic in medicine, for which only in specific clinical scenarios recommendations for management are clearly defined. We are presenting a previously healthy 18-year-old patient who survived sudden cardiac death (SCD). Multiple potential aetiologies were evaluated, including malignant coronary anomaly, acute myocarditis, potential Brugada type 3 electrocardiographic pattern, and urine drug screening positive for lysergic acid diethylamide (LSD).

Case presentation

Malignant right coronary anomaly with interarterial course and acute angle takeoff was diagnosed with coronary computed tomography angiography. Signs of acute myocarditis were detected in cardiac magnetic resonance imaging and endomyocardial biopsy. Due to potential Brugada type 3 electrocardiographic pattern flecainide provocation testing was performed to rule out Brugada Syndrome. Confirmatory chromatography revealed that prior LSD drug screening was false positive. Ultimately, the patient underwent cardiothoracic surgery with unroofing of the right coronary artery. Subsequent clinical course was favourable.

Discussion

Right coronary artery anomalies are more prevalent than left coronary artery anomalies but less often associated with SCD. Interarterial course and acute angle takeoff are risk factors for unfavourable outcomes. Myocarditis is a potential trigger of arrhythmias and SCD. In patients with Brugada type 2 and 3 electrocardiographic pattern (saddleback ST-segment elevation), provocation testing with flecainide, ajmalin, or procainamide can be used to unmask Brugada type 1 electrocardiographic pattern. Due to the proarrrhythmic potential of many recreational drugs, screening for these substances can be useful in young adults presenting after cardiac arrest; cross-reaction of substances as in our patient have to be considered.

Keywords

Case report • Coronary anomaly • Myocarditis • Brugada ECG
**Timeline**

| Day | Timeline diagnostic workup and therapy |
|-----|----------------------------------------|
| Day 1 | Out-of-hospital resuscitation. Presentation of a vasopressor-dependent, intubated patient. Electrocardiogram: right bundle branch block morphology. T wave inversions and ST-segment elevation in V3 and V4. Cranial, cervical, and thoracic computed tomography: no signs of intracranial bleeding, cervical spine injury or pulmonary embolism. Transthoracic echocardiography: diffuse hypokinesia, severely reduced left ventricular ejection fraction (LVEF). Mechanical ventilation and antibiotic therapy (due to aspiration and acute respiratory distress syndrome) |
| Day 2 | Coronary angiography and endomyocardial biopsy: negative for coronary artery disease in the left coronary system, unable to engage the right coronary system. Transthoracic echocardiography: Diffuse hypokinesia, LVEF 29% |
| Day 3 | Coronary computed tomography: anomalous right coronary artery arising from the left sinus of Valsalva with acute angle takeoff and interarterial course between aorta and pulmonary artery |
| Day 5 | Successful weaning and extubation |
| Day 10 | Cardiac magnetic resonance imaging: Borderline myocarditis, anterolateral hypokinesia with circular pericardial effusion, normalized LVEF |
| Day 16 | Negative Brugada provocation testing |
| Day 20 | Successful cardiothoracic surgery |
| Day 25 | Start of cardiac rehabilitation with a wearable cardioverter-defibrillator |
| Day 157 | Implantation of subcutaneous implantable cardioverter-defibrillator |

**Case presentation**

A healthy 18-year-old woman lost consciousness while trying to catch the bus on her birthday. She was found with pulseless ventricular fibrillation (VF) upon arrival of emergency services. After four defibrillations with 200 Joules, adrenaline 3 mg, and amiodarone 300 mg intravenously (IV), the patient developed return of spontaneous circulation 15 min later. She was intubated and brought to the emergency department.

Cardiovascular exam findings revealed a patient with cool, mottled skin, euvolemic with normal heart sounds, and no murmurs on auscultation. Lungs were fully expanding and clear to auscultation. The patient was vasopressor-dependent and intubated on assist-control ventilation. Physical cooling procedures were started immediately.

A 12-lead electrocardiogram (ECG) was obtained (Figure 1) and demonstrated sinus rhythm at approximately 110 b.p.m. with right bundle branch block morphology, negative T-waves, and ST-segment elevation in V3 and V4. Laboratory tests revealed leukocytosis (17 G/L, normal 3.0–9.6 G/L), elevated CRP (37 mg/L, normal <5 mg/L), and peak high sensitivity troponin T of 274 ng/L (normal <14 ng/L). Urine drug screening was positive for lysergic acid diethylamide (LSD). Confirmatory chromatography revealed that this was a false positive result due to cross-reaction with fentanyl, which had been administered several hours prior upon arrival to the emergency department. Initial transthoracic echocardiography was consistent with diffuse hypokinesia and reduced left ventricular ejection fraction (LVEF) of 29%. The patient underwent computed tomography to exclude intracranial bleeding, cervical spine injury and pulmonary embolism. Coronary angiography was negative for obstructive coronary artery disease in the left coronary system. Since the right coronary system could not be engaged via catheter, a coronary computed tomography was ordered to evaluate for coronary anomalies. Indeed, an anomalous right coronary artery (RCA) was identified arising from the left sinus of Valsalva with acute angle takeoff and interarterial course between aorta and pulmonary artery (Figure 2).

Due to recent rhinovirus infection and a history suggestive for myocarditis, an endomyocardial biopsy was obtained on day 2 and cardiac magnetic resonance imaging (CMR) on day 10, revealing borderline myocarditis, anterolateral hypokinesia with circular pericardial effusion and normalized LVEF. Only mild signs of inflammation without recurrence of arrhythmias during telemetry monitoring suggested that myocarditis was less likely the sole aetiology of the patient’s cardiac arrest. However, the inflammation could have lowered the threshold for arrhythmias and may have been the trigger for cardiac arrest on the grounds of an already predisposing condition—coronary anomaly.

Given suspicion for potential Brugada type 3 ECG pattern (<2 mm saddleback ST-segment elevation), the patient underwent Brugada Syndrome provocation testing with flecainide (2 mg/kg continuous IV infusion over 10 minutes, Figure 3). No obvious signs for Brugada type 1 ECG pattern were revealed with this test.

Acute respiratory distress syndrome after aspiration was treated in the intensive care unit. Ultimately, after all reversible causes for the patient’s cardiac arrest had been ruled out, the patient underwent cardiothoracic surgery 20 days post admission with unroofing of the RCA and a wearable cardioverter-defibrillator (WCD) was installed to bridge until implantation of a subcutaneous implantable cardioverter-defibrillator (S-ICD) for secondary prevention.

Neurological course was favourable with no remaining neurological deficit and cardiothoracic surgery was successful. An S-ICD was implanted 5 months after the cardiac arrest.

Due to persistent, haemodynamically not relevant pericardial effusion, oral therapy with non-steroidal anti-inflammatory drug (naproxen 500 mg twice a day), and colchicine (0.5 mg once a day) was initiated.

**Discussion**

Anomalous origin of a coronary artery from the opposite sinus of Valsalva has been associated with SCD in young adults. The prevalence of RCA arising from the left sinus of Valsalva is higher than the left coronary artery (LCA) branching from the right sinus of Valsalva, but less frequently associated with SCD. It was
hypothesized that SCD may occur due to sudden occlusion of the affected coronary artery because of its unusual course with consecutive myocardial ischemia, ventricular tachycardia or fibrillation. Young adults <30 years old are at highest risk and given a possible genetic element for coronary anomalies first degree relative screening should be considered.

An interarterial course of an anomalous coronary artery between aorta and pulmonary artery is a class IB indication for surgical repair in documented LCA anomaly, whereas in documented RCA anomaly it is with evidence of ischaemia. Therapy in asymptomatic young patients with RCA arising from the left sinus of Valsalva with acute angle takeoff and interarterial course and without evidence for stress induced ischaemia is controversial. While some experts recommend medical therapy (beta and calcium antagonists, nitrates, antiarrhythmic drugs) in older patients with or without restriction from vigorous exercise, others generally recommend surgery in particular in teenagers with a coronary anomaly. The risk of SCD has to be balanced against perioperative risks. Since there are no clear guidelines for these clinical circumstances, the decision about further management continues to be on an individual basis. Preventive ICD-implantation has the limitation of not protecting against ischaemia.

In our patient, given the presence of a right dominant coronary system, interarterial course and status post cardiac arrest we decided for surgical correction. There are no guidelines for ICD implantation after surgical correction in patients with preserved LVEF. While anomaly of the coronary artery may be the reversible cause for SCD, it could be possible that it is only a bystander and part of a syndrome that predisposes to arrhythmias, for example a syndrome entailing a channelopathy. Subcutaneous implantable cardioverter-defibrillator has been proven effective in primary and secondary prevention with no risk of lead-related long-term complications making it particularly valuable in young age. While awaiting an ICD implantation or if the risk of SCD is transient in well-selected patient a WCD is a non-invasive bridging modality.

This case report presents a rare case with a broad spectrum of differential diagnoses. A limitation of this case is that we could not definitively exclude another non-reversible cause of SCD despite extended diagnostic workup.

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

**Figure 2** Coronary computed tomography: right coronary artery (arrow) arising from the left sinus of Valsalva with acute angle takeoff and interarterial course between aorta and pulmonary artery.
Other investigated differential diagnoses were Brugada syndrome, myocarditis, and LSD-intoxication. Clinical manifestations of Brugada syndrome are VF, SCD, syncope, and palpitations. Brugada syndrome is diagnosed in the presence of type 1 ST-segment elevation (coved ST-segment elevation >2 mm in >1 of lead V1–V3 followed by a negative T-wave) spontaneously or after application of a sodium channel blocker. In patients with Brugada type 2 and 3 ECG pattern (saddleback ST-segment elevation), provocation testing with flecainide, ajmalin, or procainamide can be used to unmask Brugada type 1 ECG pattern. Due to its proarrhythmic potential, testing requires a monitored setting.12

Also in myocarditis rhythm monitoring is recommended as clinical course may be complicated by tachyarrhythmia and SCD. There are infectious and non-infectious triggers of myocarditis. Due to inflammation and resultant cell break down of cardiomyocytes, troponins, and inflammatory markers are often elevated. Evaluation for myocarditis includes endomyocardial biopsy or CMR.13

Furthermore, cardiovascular complications have been reported in the context of LSD.14,15

**Conclusion**

Coronary anomalies have been associated with SCD in young adults. Right coronary artery anomalies are more prevalent than LCA anomalies but less often associated with SCD. Moreover, interarterial course and acute angle takeoff are risk factors of unfavourable outcomes. Acute myocarditis is associated with increased risk of arrhythmias and SCD.

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**Consent:** The author(s) confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

**Conflict of interest:** none declared.

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