Heliyon 10 (2024) e23766

Contents lists available at ScienceDirect

Heliyon

journal homepage: www.cell.com/heliyon

Recurrent cardiac arrest and complete atrioventricular block due to idiopathic coronary vasospasm: A case report

Wei Gao [a,b,c,d,*], Yingwu Liu [a,b,c,d], Tong Li [a,b,c,d,**]

[a] Department of Heart Center, The Third Central Hospital of Tianjin, 83 Jintang Road, Hedong District, Tianjin, 300170, China
[b] Tianjin Key Laboratory of Extracorporeal Life Support for Critical Diseases, China
[c] Artificial Cell Engineering Technology Research Center, Tianjin, China
[d] Tianjin Institute of Hepatobiliary Disease, Tianjin, China

ARTICLE INFO

Keywords:
Coronary spasm
Complete atrioventricular block
Implantable cardioverter defibrillator
Cardiopulmonary resuscitation
Case report

ABSTRACT

Recurrent complete atrioventricular block induced by coronary spasm is rare. We present a case of a 47-year-old woman who suffered from two episodes of out-of-hospital cardiac arrest within one year due to complete atrioventricular block caused by coronary vasospasm. No implantable cardioverter defibrillator was implanted after her first episode. As for the second episode, permanent brain injury was left behind despite successful cardiopulmonary resuscitation. She underwent a challenging rehabilitation process and an implantable cardioverter defibrillator was implanted before discharge. We captured the dynamic changes of the electrocardiogram during the episode with high temporal resolution. This case illustrates the importance of recognizing coronary spasm as a potential cause of complete atrioventricular block and highlights the need for implantable cardioverter defibrillator in such patients to improve survival and quality of life.

1. Introduction

Complete atrioventricular block (CAVB) is a life-threatening condition that requires prompt diagnosis and treatment. The most common causes of CAVB are degenerative changes of the conduction system, myocardial infarction, cardiac surgery, and drugs [1]. Coronary spasm is a rare cause of CAVB that can be triggered by various factors such as emotional stress, hyperventilation, cold exposure, smoking, and cocaine use. Coronary spasm can cause transient ischemia of the sinoatrial node, atrioventricular node, or His-Purkinje system, resulting in various types of bradyarrhythmias(2). However, recurrent CAVB induced by coronary spasm is extremely uncommon and only a few cases have been reported in the literature [2,3].

Here, we present a rare case of a 47-year-old woman in good condition who suffered from two episodes of out-of-hospital cardiac arrest within one year due to CAVB caused by coronary vasospasm.

* Corresponding author. Department of Heart Center, The Third Central Hospital of Tianjin, 83 Jintang Road, Hedong District, Tianjin, 300170, China.
** Corresponding author. Department of Heart Center, The Third Central Hospital of Tianjin, 83 Jintang Road, Hedong District, Tianjin, 300170, China.
E-mail addresses: 2466727266@qq.com (W. Gao), litong3zx@sina.com (T. Li).

https://doi.org/10.1016/j.heliyon.2023.e23766
Received 1 June 2023; Received in revised form 11 December 2023; Accepted 13 December 2023
Available online 16 December 2023

2. Case presentation

A 47-year-old woman with controllable hypertension presented with an episode of neck and back pain, loss of consciousness, and CAVB. She had her first similar episode one year before this episode and was associated with ST-segment elevation in the inferior leads and CAVB on prehospital ECG (Fig. 1A). The patient had her first episode at home and lost consciousness. The prehospital emergency medical service (EMS) personnel arrived and performed the first ECG (CAVB, heart rate 45 bpm). The EMS personnel did not perform chest compressions (considering that the above ECG finding was not lethal), but administered 1 ampoule of adrenaline intravenously. Then, the patient was rapidly transported to our emergency room by ambulance (about 18 minutes en route). In her first episode, she regained sinus rhythm after successful cardiopulmonary resuscitation with intact central nervous system function in our emergency room (Fig. 1B). There, she was given close ECG monitoring, peripheral blood collection for routine laboratory tests, intravenous infusions of polarizing solution and vasopressors, to maintain circulation stability. Then, she was soon transferred again by ambulance to another hospital based on the wishes of her family, where coronary angiography (Fig. 2) and Holter monitoring showed no significant abnormalities. Acetylcholine provocation test was not performed, and she was diagnosed with coronary spasm and discharged without an implantable cardioverter defibrillator there. She took calcium channel blocker (CCB) for about one month after the first episode.

The second episode was witnessed by her family who did not perform chest compressions but called the paramedics. The initial ECG at the scene showed junctional escape rhythm with a heart rate of 32 bpm (Fig. 1C) and she received cardiopulmonary resuscitation. The repeat ECG in the emergency department showed junctional rhythm with a heart rate of 50 bpm and ST-segment elevation in the inferior leads (Fig. 1D). She restored sinus rhythm with a heart rate of 139 bpm after 8 minutes of advanced cardiopulmonary resuscitation (Fig. 1E). She was admitted to the coronary care unit for further management.

On admission, she was unconscious with a body temperature of 37.5 °C, heart rate of 68 bpm, and blood pressure of 145/50 mmHg. Her pupils were equal at 3.5 mm with sluggish light reflex and absent corneal reflex. Her lung auscultation was normal. Her head and chest CT scans were unremarkable. Her left ventricular ejection fraction was 59 %. Her laboratory tests showed normal hematology, mild hyperglycemia (11.92 mmol/L), and elevated transaminases (alanine aminotransferase 146 U/L, aspartate aminotransferase 89 U/L). Her arterial blood gas analysis showed metabolic acidosis with pH 7.213, PCO_2 35.8 mmHg, PO_2 396.1 mmHg, HCO_3^- 14.1 mmol/L, oxygen saturation 99.7 %, and lactate 11.53 mmol/L. Her troponin I, D-dimer, and B-type natriuretic peptide levels were normal. She was diagnosed with coronary spasm with CAVB, aborted sudden cardiac death, hypoxic-ischemic encephalopathy, liver injury, stress hyperglycemia, and gastrointestinal ulcer. She received invasive mechanical ventilation, therapeutic hypothermia, continuous renal replacement therapy, optimal medical therapy, and parenteral nutrition support.

Her blood tests on day two showed elevated troponin I (9.34 ng/mL), BNP (545 pg/mL), D-dimer (1800 ng/mL), and transaminases (alanine aminotransferase 367 U/L, aspartate aminotransferase 252 U/L). Her left ventricular ejection fraction was 58 % with segmental wall motion abnormalities. She developed limb rigidity and clonus, and repeat head CT scan showed severe cerebral edema. She was treated with oral sodium valproate, topiramate, levetiracetam, and intermittent intravenous mannitol and albumin, and her

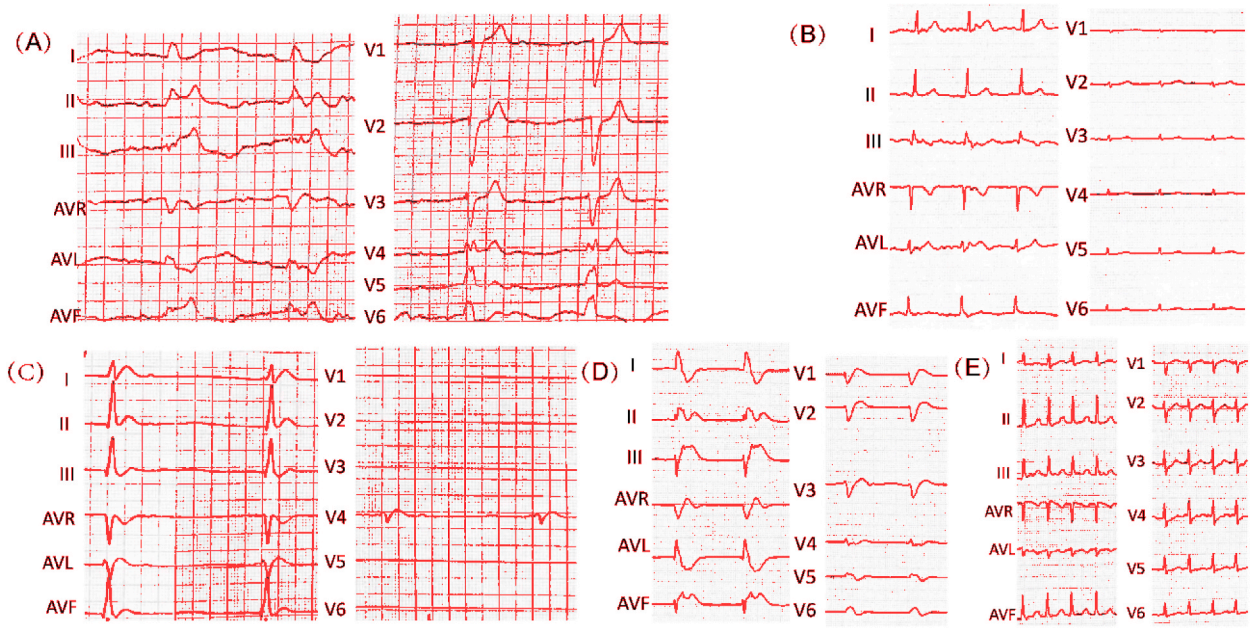

Fig. 1. The patient's ECG changes during two episodes of illness. First episode: (A) Complete atrioventricular block and inferior ST-segment elevation (STE) at home (heart rate 45 bpm); (B) Sinus rhythm recovery after cardiopulmonary resuscitation (CPR), heart rate 73 bpm (precordial leads 1.25 mm/mV). Second episode: (C) Junctional escape rhythm at home (heart rate 32 bpm); (D) Junctional rhythm and inferior STE at the emergency department (heart rate 50 bpm) (precordial leads 5 mm/mV); (E) Sinus rhythm recovery after CPR, heart rate 73 bpm.

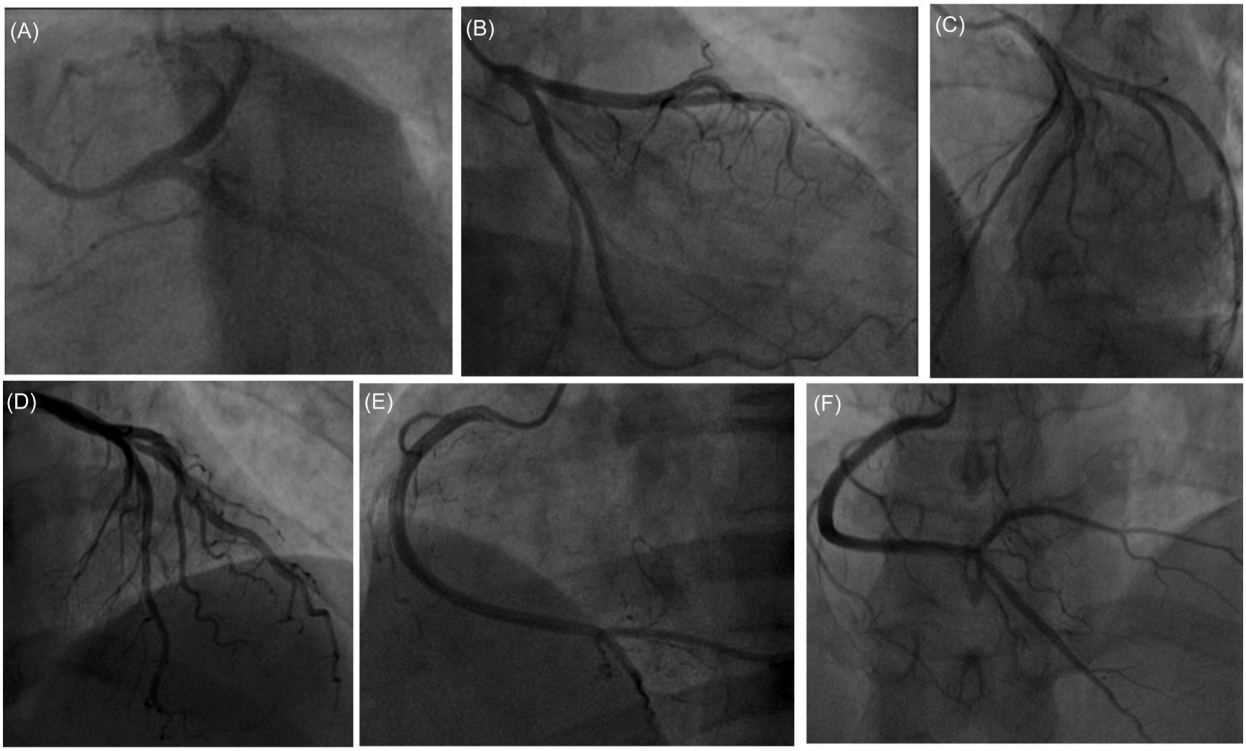

Fig. 2. Coronary angiography with different projection angles revealed normal left (A–D) and right coronary arteries(*E*-F).

symptoms improved gradually. On day seven, she had another episode of CAVB due to coronary spasm that progressed to hemodynamic collapse (Fig. 3A). During this period, the ECG showed progressive widening and notching of the R wave. The patient received immediate cardiopulmonary resuscitation, intravenous adrenaline and atropine, and correction of acid-base imbalance. After 30 minutes, ventricular fibrillation occurred on the ECG, which converted to CAVB after one unsynchronized electrical defibrillation, and then reverted to sinus rhythm and normal tissue perfusion (Fig. 3B). The patient experienced four more episodes of similar ECG changes at 12, 18, 72, and 84 hours after the onset. Each episode was accompanied by a sudden bradycardia, and a rapid transition from sinus rhythm to CAVB or escape rhythm. Blood pressure and peripheral oxygen saturation were unmeasurable, and Kussmaul breathing was observed. Each episode was promptly managed with cardiopulmonary resuscitation, intravenous adrenaline and atropine, electrical defibrillation as needed, and advanced life support to correct acid-base imbalance. She experienced several adverse

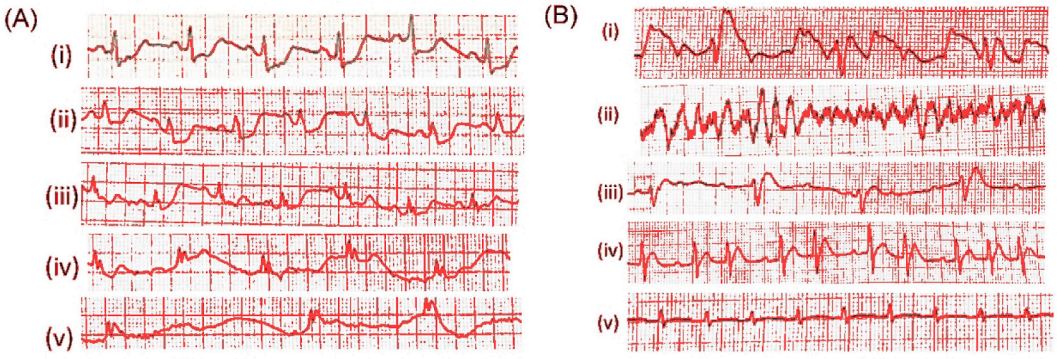

Fig. 3. The patient had another life-threatening arrhythmia due to coronary spasm on day seven. (A) ECG (lead II) before cardiopulmonary resuscitation (CPR): (i) Sinus rhythm, 90 bpm (8:23 a.m.); (ii) Sinus rhythm with broad R waves (lambda wave), 94 bpm (8:24 a.m.); (iii) Sinus rhythm with broader and notched R waves, 94 bpm (8:25 a.m.); (iv) High-grade atrioventricular block (AVB), 68 bpm (8:26 a.m.); (v) Complete AVB, 34 bpm, no blood pressure (8:26 a.m., 33 seconds after iv-chart). Chest compression, adrenaline and atropine given. (B) ECG after CPR: (i) Lambda waves still present after 30 minutes of compression, with ST-segment elevation resembling nonischemic "action potential" or unusual "tombstone" (8:54 a.m.); (ii) Ventricular fibrillation (8:54 a.m.); (iii) Complete AVB after 150 J defibrillation (8:54 a.m.); (iv) First-degree AVB with atrial ectopy (8:55 a.m.); (v) Sinus tachycardia, 140 bpm (8:57 a.m.).

outcomes following cardiopulmonary resuscitation, such as cerebral edema, hyperthermia, gastrointestinal bleeding, hospital-acquired infection, and multiple organ failure. However, her vital signs improved gradually with supportive care. Her consciousness improved from coma to vegetative state, and enteral nutrition was resumed. On day 27, she underwent tracheostomy and was extubated successfully.

Her family history revealed that her uncle and two sisters had sudden cardiac death at a young age. To rule out inherited cardiac disorders (e.g., cardiac ion channelopathies), whole exome sequencing was performed for the patient, her husband and son, her brother and his son, and no abnormalities were detected. Therefore, the cause of cardiac arrest was attributed to CAVB secondary to coronary spasm. She received an ICD on day 66 and was discharged on postoperative day eight. She remained in a vegetative state and in good physical condition during the 32-month follow-up, with no episodes of syncope or shock. The timeline of the case is shown in Fig. 4.

3. Discussion

Coronary spasm with CAVB is a rare and life-threatening cardiovascular complication that can lead to cardiac arrest or sudden cardiac death [4]. The pathogenesis is unclear, but it may involve coronary spasm-induced myocardial ischemia, infarction, fibrosis, autonomic imbalance, and genetic factors [4]. The incidence and prognosis depend on the severity, duration, location, extent, frequency, and triggers of coronary spasm. The incidence of complete or high-degree atrioventricular block due to coronary spasm ranges from 1 % to 5 %, with the right coronary artery being the most frequently affected (60 %–80 %). The mortality rate of this condition is high (10 %–20 %) [4].

The diagnosis of coronary spasm with CAVB is based on clinical presentation, ECG, and coronary angiography. The main clinical features are chest pain, syncope, or shock. A 24-h Holter monitoring can capture the ECG changes during episodes of coronary spasm, such as ST-segment elevation, T-wave inversion, and CAVB. In our case, the Holter monitoring during the second admission allowed us to observe the entire process of CAVB due to coronary spasm. The rapid and recurrent ECG changes made it unlikely that coronary thrombolysis or plaque rupture was involved. The ST-segment elevation in the inferior leads suggested right coronary artery involvement. We also recorded an important ECG sign - the “lambda wave” in the limb lead of the ECG monitoring during the attack. The lambda wave is an abnormal ventricular depolarization and repolarization wave that indicates a high risk of sudden cardiac death [2].

Coronary angiography can also help in diagnosing coronary spasm, as it can reveal the location and extent of the vasospasm, as well as the presence or absence of atherosclerotic plaques [5]. Moreover, acetylcholine can be used to induce or rule out coronary vasospasm. In our case, her coronary angiography during the first admission showed no significant atherosclerosis. Therefore, coronary vasospasm was diagnosed based on her clinical presentation, normal coronary angiography, and the typical ECG changes during the episode. She did not undergo coronary angiography during the second admission per her family’s wishes. Considering the genetic predisposition to coronary spasm and the history of sudden death in several family members at a young age, we performed whole exome sequencing for the patient and her close relatives. Importantly, we ruled out any known genetic missense mutations, and confirmed the diagnosis of idiopathic coronary vasospasm.

The treatment of coronary spasm with CAVB involves medical therapy, temporary pacemaker, and ICD [2,3,6–8]. Medical therapy mainly includes calcium channel blockers (CCB) and nitrates, which can prevent and relieve coronary spasm and reduce the risk of CAVB [6]. However, the optimal duration of these drugs is unclear. Percutaneous coronary interventions, which can improve coronary blood flow and reduce ischemia, are not effective for coronary spasm without significant plaques. Temporary pacemaker can be used as a short-term measure for patients who do not respond to drugs or have life-threatening symptoms such as syncope or shock. ICD can prevent death from coronary vasospasm with CAVB, especially when associated with life-threatening arrhythmias [9–11]. This

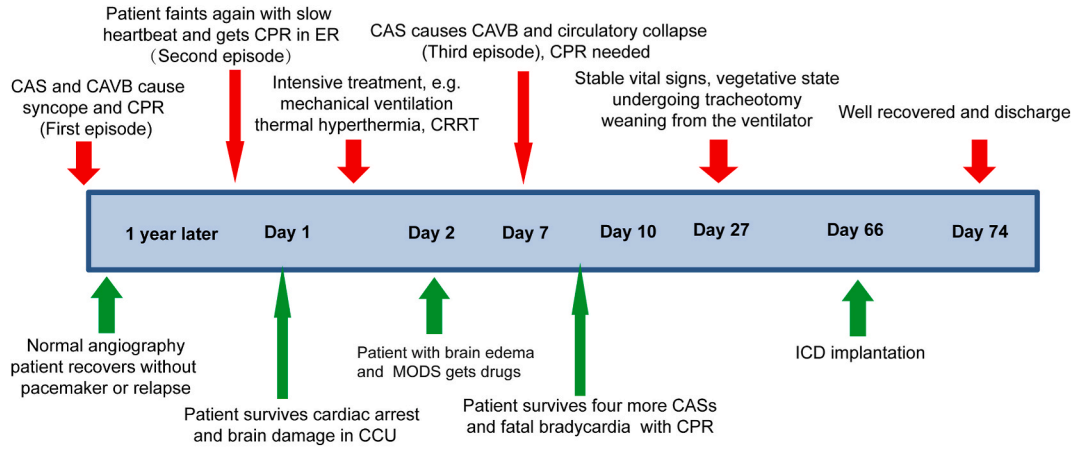

Fig. 4. Timeline of the case. CAS coronary artery spasm, CAVB complete atrioventricular block, CCU coronary care unit, CPR cardiopulmonary resuscitation, CRRT continuous renal replacement therapy, ICD Implantable cardioverter defibrillator.

efficacy has been confirmed by several clinical trials and observational studies. However, there are no guidelines for ICD implantation in primary prevention for coronary vasospasm despite its potential to cause lethal arrhythmias. In secondary prevention, contemporary guidelines recommend ICD implantation in these patients, if they are already on optimal medical therapy or if medical therapy is not tolerated (Class IIa) [12]. The indication for ICD should be restricted to life-threatening situations, which are defined by the following criteria: i) ventricular fibrillation or sustained ventricular tachycardia; ii) syncope or cardiac arrest; iii) high-degree atrioventricular block or sinoatrial block; iv) left ventricular ejection fraction less than 35 %; v) familial history of sudden cardiac death or inherited arrhythmia syndrome [10]. Therefore, we suggest that the risks and benefits of each patient should be carefully assessed, and an individualized decision should be made. Considering the recommendations, our patient would have benefited from an ICD implantation after the first episode in terms of survival and quality of life.

The long-term outcomes of ICD for secondary prevention of coronary spasm-induced cardiac arrest remain unclear [13]. Some studies have reported that the first three months after the index event are the most critical period for cardiovascular events [14]. Smoking cessation and CCB therapy are the main prognostic protective factors. Other factors that have been associated with poor prognosis are advanced age, elevated C-reactive protein, multivessel spasm, significant atherosclerotic lesion, and reduced ejection fraction [14]. This patient lacks these risk factors and would likely fall into the low-risk group for recurrence. She took CCB for about one month after the first episode of coronary spasm. However, she experienced a recurrence after one year, indicating that the risk period for adverse cardiovascular events may extend beyond three months, particularly in the absence of continuous CCB therapy.

Whether ICD implantation is appropriate in coronary spasm patients with aborted sudden cardiac arrest, even before assessing the response to medical therapy, is not well known and requires further studies. It has been proposed to use the result of the acetylcholine provocation test under optimal medical therapy in resuscitated CAS patients as a criterion for secondary ICD implantation [15]. Recently, several cases of coronary spasm leading to hemodynamic compromise and recurrent syncope, bradycardia (e.g., ventricular standstill, complete atrioventricular block, etc.) following brief cardiopulmonary resuscitation have been described in the literature [2,3,9]. These cases showed no obstructive coronary artery disease on angiography and had a favorable outcome after ICD implantation and 1–2 months of follow-up without further episodes. A recent multicenter European retrospective cohort study assessed the clinical outcomes of such patients under different therapeutic strategies and evaluated the necessity of ICD placement in conjunction with optimal medical therapy [11]. The results indicate that ICD implantation was performed in nearly 90 % of patients. About a quarter of patients experienced appropriate ICD shocks during a median follow-up of 59 months. These findings, along with our own case, suggest that patients with coronary spasm and aborted sudden cardiac death may still have arrhythmic events in a considerable period. Owing to some genetic vulnerability, and ICD implantation may be a preferable treatment option for the patients experiencing with life-threatening arrhythmias due to coronary spasm. This case report adds to the scarce literature on this rare condition, although it does not confirm coronary spasm by acetylcholine provocation testing. It also presents some distinctive features: 1) We recorded the ECG variations during the event with fine time resolution. 2) Our patient did not receive ICD implantation after the first event of aborted sudden cardiac death and had another out-of-hospital cardiac arrest one year later, resulting in irreversible hypoxic brain injury. This unfavorable outcome highlights the high recurrence risk and poor prognosis of patients with arrhythmias secondary to coronary spasm, who are not adequately treated with medical therapy. During the almost 3-year follow-up after ICD implantation, the patient had no more syncope or shock episodes. We think that the combination of ICD and medications, such as CCB, was effective in preventing further episodes of syncope or shock due to coronary spasm and CAVB. The ICD could provide backup pacing and defibrillation in case of life-threatening arrhythmias, while the drugs could reduce the frequency and severity of coronary spasm. We do not know if the ICD prevented any fatal arrhythmias in this period, but we believe that ICD implantation improved her chances of survival.

4. Conclusion

Patients with coronary vasospasm and life-threatening complications, such as complete atrioventricular block, may benefit from ICD implantation. This intervention may improve their long-term survival and quality of life outcomes.

Ethics statement

This study was conducted in accordance with the Declaration of Helsinki (as revised in 2013). Written informed consent was obtained from the patient's family.

Funding

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

Data availability statement

The raw data supporting the conclusions of this article will be made available by the authors, without undue reservation.

CRediT authorship contribution statement

Wei Gao: Writing – review & editing, Writing – original draft, Methodology, Investigation, Data curation, Conceptualization.

Yingwu Liu: Writing – review & editing, Supervision, Investigation, Formal analysis, Data curation. **Tong Li:** Writing – review & editing, Resources, Formal analysis, Conceptualization.

Declaration of Competing interest

The authors declare that there is no conflict of interests regarding the publication of this article.

Acknowledgements

None.

References

[1] G.M. Cioffi, A. Gasperetti, G. Tersalvi, M. Schiavone, P. Compagnucci, F.B. Sozzi, et al., Etiology and device therapy in complete atrioventricular block in pediatric and young adult population: contemporary review and new perspectives, J. Cardiovasc. Electrophysiol. 32 (11) (2021) 3082–3094.
[2] G. Wang, N. Zhao, C. Zhang, S. Zhong, X. Li, Lambda-like ST-segment elevation in acute myocardial infarction triggered by coronary spasm may be a new risk predictor for lethal ventricular arrhythmia: a case report, Medicine 97 (49) (2018), e13561.
[3] A. Kojima, T. Shirayama, J. Shiraishi, T. Sawada, Subcutaneous implantable cardioverter-defibrillator was inappropriate for use in a patient with aborted sudden cardiac death due to coronary spastic angina: a case report, European heart journal Case reports 4 (6) (2020) 1–5.
[4] J. Jewulski, S. Khanal, K. Dahal, Coronary vasospasm: a narrative review, World J. Cardiol. 13 (9) (2021) 456–463.
[5] S. Stern, A. Bayes de Luna, Coronary artery spasm: a 2009 update, Circulation 119 (18) (2009) 2531–2534.
[6] B. Gao, Z. Zhang, J. Qian, C. Cao, X. Hua, M. Chu, et al., The use of calcium channel blockers in the treatment of coronary spasm and atrioventricular block, Cell Biochem. Biophys. 72 (2) (2015) 527–531.
[7] S. Sueda, T. Shinohara, N. Takahashi, J. Shite, T. Shoji, M. Akao, et al., Questionnaire in patients with aborted sudden cardiac death due to coronary spasm in Japan, Heart Ves. 35 (12) (2020) 1640–1649.
[8] S. Sueda, H. Kohno, Optimal medications and appropriate implantable cardioverter-defibrillator shocks in aborted sudden cardiac death due to coronary spasm. Internal medicine (tokyo, Japan) 57 (10) (2018) 1361–1369.
[9] A. Guevara, M. Patel, D. DeLurgio, Recurrent syncope and ventricular arrhythmias induced by coronary artery spasm. Cardiovascular revascularization medicine : including molecular interventions 40s (2022) 225–230.
[10] D. Adepoju, J.A.L. Smith, S.J. Leslie, Life-threatening coronary vasospasm treated by implantable cardioverter defibrillator: the warning signs, Case reports in cardiology 2022 (2022), 4504028.
[11] M. Rodríguez-Mañero, T. Oloriz, J.B. le Polain de Waroux, H. Burri, B. Kreidieh, C. de Asmundis, et al., Long-term prognosis of patients with life-threatening ventricular arrhythmias induced by coronary artery spasm. Europace : European pacing, arrhythmias, and cardiac electrophysiology : journal of the working groups on cardiac pacing, arrhythmias, and cardiac cellular electrophysiology of, the European Society of Cardiology 20 (5) (2018) 851–858.
[12] S. Riaz, V. Raj, S. Shah, Refractory vasospastic angina and sudden cardiac arrest: is implantable cardioverter defibrillator indicated and is it always protective? Cureus 12 (8) (2020), e9613.
[13] A. Ueno, A. Kawabe, T. Sugiyama, M. Ishikawa, A. Uema, M. Shimoyama, et al., Out-of-hospital cardiac arrest related to coronary arterial spasm in three elderly patients with no obstructive coronary artery disease, Journal of cardiology cases 16 (5) (2017) 154–157.
[14] A. Matta, F. Bouisset, T. Lhermusier, F. Campelo-Parada, M. Elbaz, D. Carrié, et al., Coronary artery spasm: new insights, J. Intervent. Cardiol. 2020 (2020), 5894586.
[15] S. Sueda, T. Sakaue, T. Okura, Spasm provocation tests under medication may help decide on medical or mechanical therapy in patients with aborted sudden cardiac death due to coronary spasm, Intern. Med. (Tokyo) 59 (11) (2020) 1351–1359.