Journal of Cardiology Cases 19 (2019) 62–65

Contents lists available at ScienceDirect

Journal of Cardiology Cases

journal homepage: www.elsevier.com/locate/jccase

Case Report

Short coupled Torsade de pointes with myocardial injury
A possible sequela of myocarditis

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ARTICLE INFO

Article history:
Received 25 July 2018
Received in revised form 27 September 2018
Accepted 20 October 2018

Keywords:
Short coupled Torsade de pointes
Myocardial injury
Previous myocarditis

ABSTRACT

A 44-year-old woman with previous myocarditis underwent several syncopal events due to self-terminated ventricular fibrillation (VF) with hypokalemia. Electrocardiogram showed a normal QT duration and premature ventricular contraction with a short coupling interval (280 ms), and the R-on-T phenomenon induced Torsade de pointes (TdP) that deteriorated into VF, a condition known as short coupled TdP (ScTdP). Cardiac magnetic resonance imaging showed left ventricular dilatation and diffuse high T2 signal intensity, and endomyocardial biopsy exhibited a failing myocardium. This is a rare case of ScTdP accompanied by a failing myocardium. Some instances of ScTdP may be associated not only with an electrical disorder but also with a structural myocardial disorder.

<**Leaning Objective:** Any cases with short coupled Torsade de pointes (ScTdP) have not reported structural disorders, and ScTdP has been categorized as an idiopathic arrhythmia. This is the first case of ScTdP accompanied by a failing myocardium, which was identified by cardiac magnetic resonance imaging and both light and electron microscopic examination. According to this case, some cases of ScTdP may be associated not only with electrical disorders, but also structural myocardial abnormalities.>

Introduction

Torsade de pointes (TdP) is a known manifestation of long QT syndrome. However, in rare cases TdP occurs with normal QT duration, triggered by premature ventricular contractions (PVCs) with a short coupling interval (<300 ms), a condition known as “short coupled TdP” (ScTdP). Any cases that have involved endomyocardial biopsy in patients with ScTdP have not reported structural disorders [1,2], and ScTdP has therefore been categorized as an idiopathic arrhythmia. This is the first case of ScTdP accompanied by a failing myocardium.

Case report

History of acute myocarditis

A 43-year-old woman with no family history of arrhythmia had several syncopal events with faintness at rest, and she was hospitalized at an outside institution. Electrocardiogram (ECG) showed repeated self-terminated ventricular fibrillation (VF), which disappeared after lidocaine administration. Laboratory tests prior to VF showed elevation of cardiac enzymes [creatine kinase (CK): 1479 U/L; CK-MB: 34 U/L; and troponin I: 0.187 ng/mL, with a normal value of less than 0.1 ng/mL]. Transthoracic echocardiography (TTE) showed diffuse left ventricular (LV) hypocontraction [LV ejection fraction (LVEF): 33%] and pericardial effusion. Coronary artery angiogram (CAG) showed no significant coronary artery stenosis. Based on these results, the patient was diagnosed with acute myocarditis and began amiodarone therapy. After 6? months, TTE showed recovered LV contraction (LVEF: 60%).

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https://doi.org/10.1016/j.jccase.2018.10.004
1878-5409/

History of the present illness

At age 44?years the patient started taking rikkunshito, a Chinese herbal medicine containing glycyrrhizin acid, for abdominal discomfort. She subsequently had three syncopal events within 3 days and was admitted to our hospital. On admission, her serum potassium level was low (K: 2.7 mEq/L), serum calcium level was in normal range (Ca: 8.7 mg/dL, albumin-corrected calcium level: 8.6 mg/dL) and ECG showed a normal QT duration and PVCs with a short coupling interval (280 ms) (Fig.?1A, B). The patient's PVCs induced the R-on-T phenomenon and she developed TdP, which then deteriorated into VF (Fig.?1C), leading to a diagnosis of ScTdP. The VF resolved after dose escalation of amiodarone and administration of potassium and a β blocker (bisoprolol). Subsequent TTE showed almost normal LV systolic function (LV diastolic diameter: 52 mm; LVEF: 57%) (Fig.?2A,B), but cardiac magnetic resonance imaging (CMR) showed mild LV and right ventricular (RV) dilatation (LV end diastolic volume index: 123 mL/m^2; RV end diastolic volume index: 114 mL/m^2) (Fig.?2C,D) and diffuse high T2 signal intensity with values ranging from 69?to 80 ms on T2 mapping with fine linear late gadolinium enhancement (LGE) in the LV posterior wall (Fig.?2E,F). In addition, she had the late potential on signal-averaged ECG. Challenge tests with pilsicainide (a sodium channel blocker) and epinephrine did not induce ECG changes of Brugada syndrome or long-QT syndrome, respectively. No coronary artery stenosis was observed with CAG. Endomyocardial biopsy from the RV septum showed perinuclear halos with lipofuscin deposits and vacuolar changes of cardiomyocytes (Fig.?3A, B), mild interstitial and perivascular fibrosis (Fig.?3B), and focal fatty deposition (Fig.?3C). Moreover, transmission electron microscopy showed degeneration of myofibrils with proliferation of cardiomyocyte mitochondria (Fig.?3D). Immunohistochemistry (CD3 for T lymphocytes and CD68 for macrophages) revealed no active myocarditis or significant inflammatory

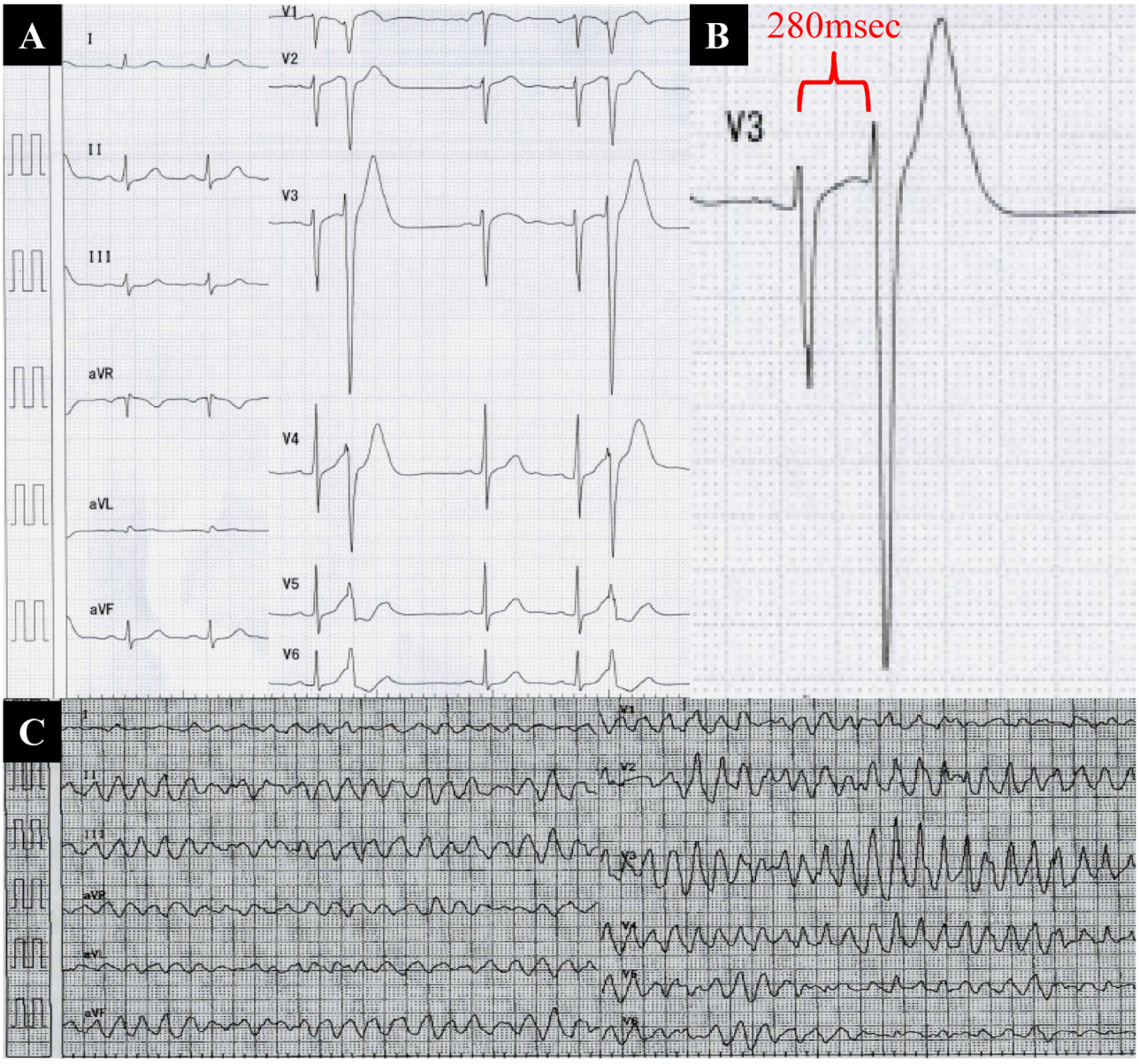

Fig. 1. PVCs triggering ScTdP, and subsequent VF. (A, B) PVCs with a short coupling interval time of 280 ms. (C) VF induced by ScTdP.
PVC, premature ventricular contraction; ScTdP, short coupled Torsade de pointes; VF, ventricular fibrillation.

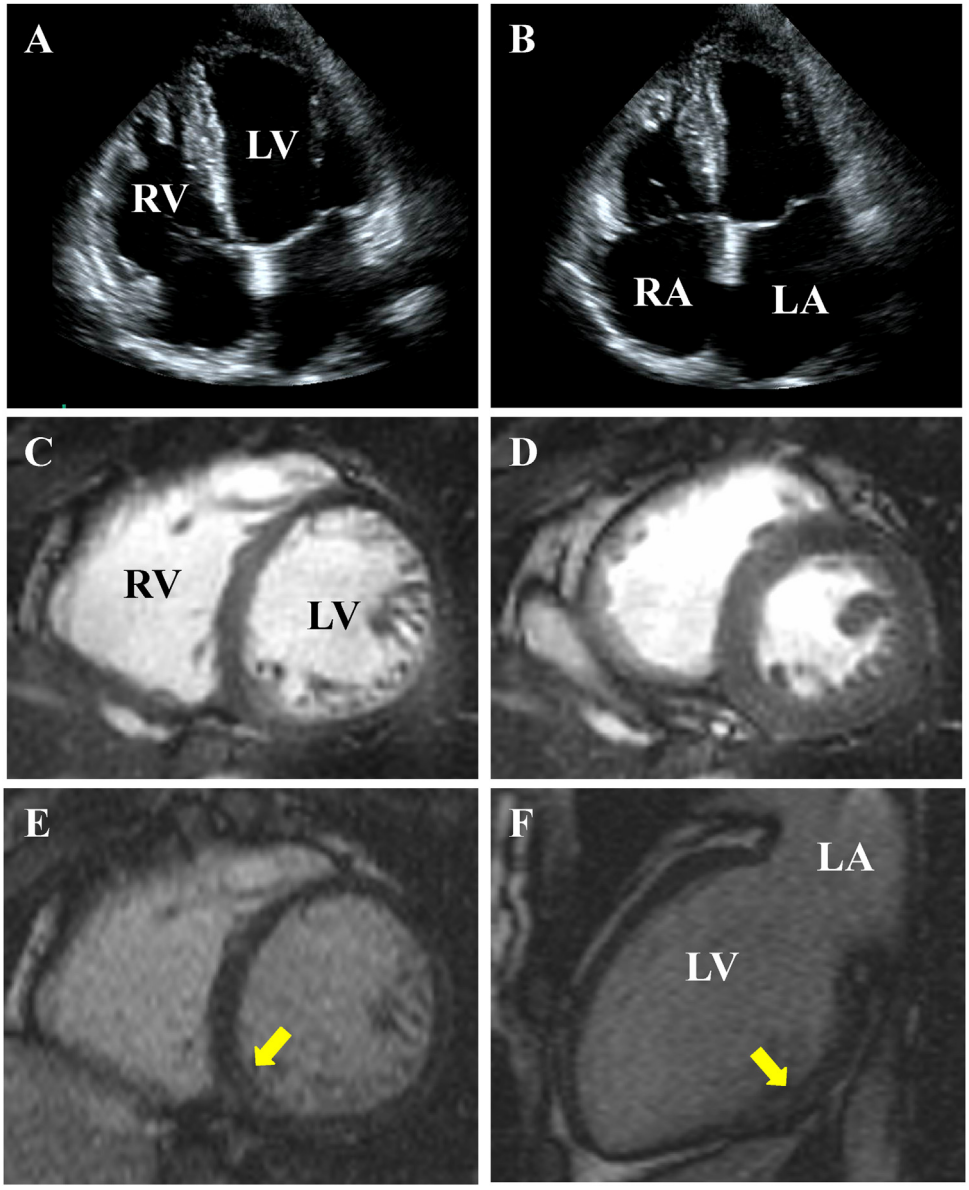

Fig. 2. TTE and CMR findings. TTE with an apical four-chamber view in the end-diastolic phase (A) and the end-systolic phase (B) show left ventricular dilatation and mildly reduced contraction. CMR with a cine image obtained with a coronal view in the end-diastolic phase (C) and the end-systolic phase (D) show mild dilatation of the LV and RV. Delayed enhancement CMR with a coronal view (E) and vertical long-axis view (F) show fine linear late gadolinium enhancement (yellow arrow).
CMR, cardiac magnetic resonance imaging; LA, left atrium; LV, left ventricular; RA, right atrium; RV, right ventricular; TTE, transthoracic echocardiography.

cell infiltration, and no findings of conditions such as amyloidosis and sarcoidosis that can lead to secondary cardiomyopathy. These histologic abnormalities were not specific, but suggested a failing myocardium, and therefore we considered the possibility of underlying cardiomyopathy or alternations in myocarditis. Based on these findings, we concluded that the patient's ScTdP was associated with post-inflammatory myocardial injury. Genetic testing showed no mutations in KCNQ1, KCNH2, SCN5A, or RYR2

To prevent sudden cardiac death, we performed catheter ablation and implanted an implantable cardioverter-defibrillator (ICD). An electrophysiological study (EPS) showed PVCs originating in the RV apex (RVA) with a pre-potential (possibly a Purkinje potential). We performed catheter ablation targeting the initial PVCs, and the number of PVCs was significantly reduced. Since the ICD implantation, the patient has not required appropriate ICD discharge for more than 3?years.

Discussion

To the best of our knowledge, this case is the first case of ScTdP accompanied by a failing myocardium. ScTdP has a poor prognosis [3], but is not well understood due to its low incidence. Several previous studies found that electrical disorders were involved in the pathogenesis of ScTdP. Leenhardt et?al. showed that as many as 64% of patients with ScTdP had PVCs originating in the RVA [3].? Furthermore, Kusano et?al. revealed that in a ScTdP patient, low potassium levels shortened repolarization only in the RVA and not in other lesions, including the LV apex [4].?Haissaguerre et?al. demonstrated that a short coupling time was a specific characteristic of PVCs originating in the Purkinje system [5].?In addition, previous cases with ScTdP that have performed various examinations such as endomyocardial biopsies from the RVA had no structural disorders [1,2].?Based on the above findings, it is

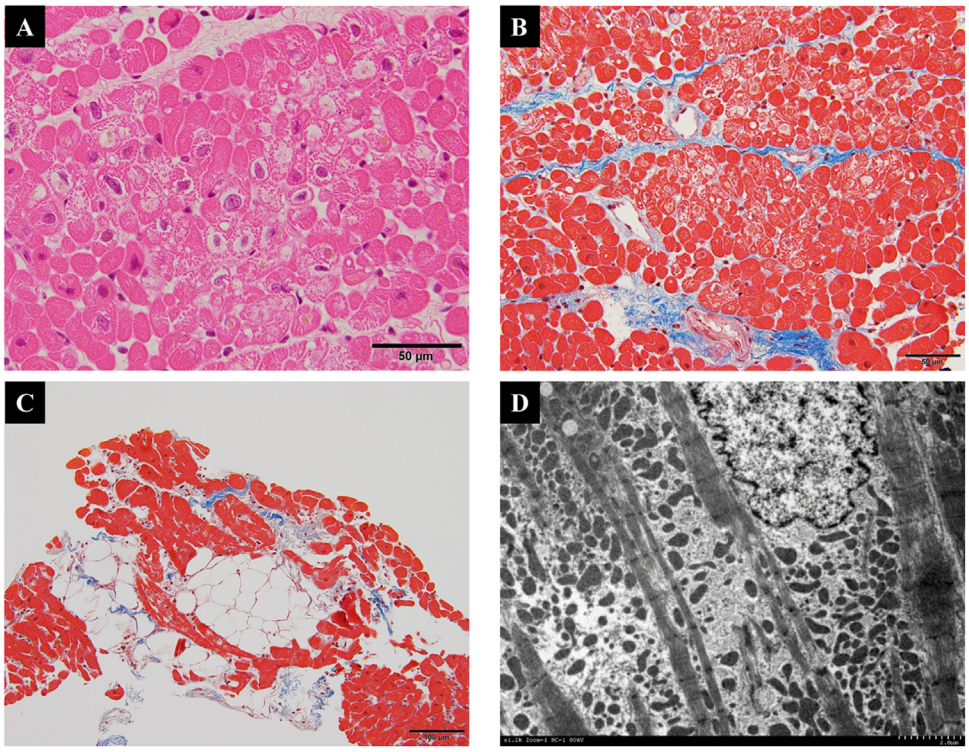

Fig. 3. Histological findings on myocardial biopsy of the right ventricle. (A,B) Perinuclear halo with lipofuscin deposits and vacuolation of myocardial cytoplasm (A: 10×, HE; B: 20×, Masson's trichrome). (C) Interstitial and perivascular fibrosis, and focal fatty deposition (10×, Masson's trichrome). (D) Transmission electron microscopy showed degeneration of myofibrils with proliferation of cardiomyocyte mitochondria.
HE, hematoxylin and eosin stain.

considered that short coupled PVCs can be caused by electrical disorders related to Purkinje fibers at the RVA, mediated by low potassium levels. In fact, our case of ScTdP also demonstrated hypopotassemia and short coupled PVCs originating in the RVA.

The novel finding in our case is that ScTdP occurred in the context of a failing myocardium. The CMR and both light and electron microscopic examination of endomyocardial biopsy samples identified myocardial injury that was probably related to previous myocarditis. Diffuse high T2 signal intensity on CMR indicated diffuse myocardial edema and damage. Thus, myocardial damage may have caused the initial PVCs in the RVA. Interestingly, LGE on CMR, which showed more significant myocardial fibrosis compared with other areas, was located in the LV posterior wall. It could be considered that myocardial fibrosis in the LV posterior wall related as a substrate in maintaining the arrhythmia. Given these findings, cardiac damage in our case might have played a role not only in the initiation of PVCs but also maintaining the arrhythmia.

The optimal management of ScTdP is still unknown. Implantation of an ICD is the only effective therapy for preventing sudden cardiac death. A short report stated that the calcium blocker verapamil was recommended for ScTdP prevention [3].?However, calcium blockers are not recommended as routine therapies for structural heart disease [6].?β-blockers are commonly effective for TdP prevention, and therefore may be a reasonable alternative to calcium blockers for ScTdP patients with a failing myocardium.

Little is known about ScTdP, so future studies are needed, including those evaluating the relationship between ScTdP and the failing myocardium.

Conclusion

Some cases of ScTdP may be associated not only with electrical disorders but also structural myocardial abnormalities.

Conflict of interest

None.

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