CASE REPORT

The nightmare ECG of a spontaneously aborted sudden cardiac death

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We present the case of a spontaneously aborted sudden cardiac death related to an high-rate ventricular tachycardia in an healthy man with bileaflet mitral valve prolapse. An effective treatment of the patient, with complete suppression of ventricular ectopic activity, was achieved only combining medical therapy, ICD and catheter ablation, thus overcoming the limitations of each approach.

KEYWORDS

catheter ablation, mitral valve prolapse, sudden cardiac death, ventricular arrhythmia, ventricular tachycardia

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A 55-year-old caucasian man without relevant personal or familiar pathological history experienced syncope while driving car. Nonsustained repetitive runs of a high-rate monomorphic wide-complexes irregular tachycardia were reported by the emergency service. Medical treatment with amiodarone, magnesium sulfate, lidocaine, and bisoprolol had been initiated and had resulted effective, in the acute setting, in suppressing the arrhythmia. At admission in our Coronary Care Unit, the patient was stable, did not complain any ongoing symptom and his physical examination revealed only a mild systolic murmur. Laboratory tests and baseline electrocardiography (ECG) were unremarkable. Echocardiography showed a floppy mitral valve with bileaflet prolapse and mild-to-moderate insufficiency; left ventricular (LV) ejection fraction was normal, as all other findings were. Coronary angiography excluded pathological alterations in epicardial arteries.

The ECG taken by the emergency service was finally retrieved (Figure 1a) and showed nonsustained repetitive runs of monomorphic ventricular tachycardia (VT), with irregular cycles (minimum: 160 ms, 375 bpm). A supraventricular aberrant arrhythmia could be ruled out, because (i) QRS complexes present right-bundle-branch-block-like morphology and right axis deviation, with an aspect (see lead V_1) incompatible with aberrancy, and with all morphology-based algorithms concordant for VT; (ii) some beats with different morphology could represent fusions (or, less likely,

ventricular ectopies (VE) with another origin), confirming atrio-ventricular dissociation; (iii) a single sinus beat is followed by a short-coupled VE, that is identical to VT beats, falls on top of the T wave and triggers another VT run; (iv) such sinus beat makes unlikely the hypothesis of atrial fibrillation runs; (v) there is no sign of ventricular pre-excitation in the baseline ECG (Figure 1b), that could justify an hypothesis of atrial fibrillation with rapid accessory pathway conduction. Analyzing the surface vectors, the arrhythmic focus could be expected to be placed posteriorly in the LV, in a mid-to-basal location.

During hospitalization, the same nonsustained VTs recurred, despite medical therapy. Symptomatic bradycardia and hypotension during attempts of bisoprolol up-titration required to anticipate the dual-chamber implantable cardioverter defibrillator (ICD) implantation, to allow atrial pacing. Bisoprolol up-titration, even with amiodarone and lidocaine, was not effective in preventing other presyncopal nonsustained VTs, that were only responsive to intravenous midazolam. Two days after ICD implantation, an arrhythmic storm occurred (14 appropriate ICD shocks in the VF-zone).

The patient underwent catheter ablation. ECG features of the arrhythmia, suggesting its likely origin, and the echocardiographic findings of bileaflet mitral valve prolapse (Figure 1c) oriented to consider LV mapping as priority. Through transaortic approach, VE were mapped in the LV using Biosense Webster CARTO3 system. The earliest activation during VE was found between the

FIGURE 1 Panel (a) electrocardiogram taken by the emergency service. Panel (b) electrocardiogram at admission in our Coronary Care Unit, under medical therapy. Panel (c) transthoracic echocardiography frames showing bileaflet mitral valve prolapse

posterior papillary muscle insertion and the mitral annulus, within the posterior LV wall, where low-voltage, fragmented potentials were observed (Figures 2 and 3), with completely negative deflection of the unipolar signal on the mapping catheter. Radiofrequency ablation was performed at those sites and acutely completely suppressed ventricular arrhythmias, even during isoprenaline challenge. Drugs were stopped and nonsustained VTs, with irregular (generally longer) cycles and the same morphology, were observed (Figure 4a). Amiodarone and bisoprolol were reinitiated and finally effectively suppressed the VE (Figure 4b). At discharge, even an exercise testing was negative. At the 4-month follow-up visit, no arrhythmic episodes have been detected by the ICD, and no

symptoms complained by the patient. The patient refused a magnetic resonance imaging scan.

Bileaflet mitral valve prolapse syndrome has been recognized as an underestimated cause of sudden cardiac death in otherwise healthy subjects (Sriram et al., 2013; Syed et al., 2016); its structural hallmark has been identified as the fibrosis of papillary muscles and infero-basal LV free wall, reflecting the myocardial stretch by the prolapsing leaflets and elongated chordae (Basso et al., 2015).

Our patient was successfully treated only combining medical therapy, ICD, and ablation, thus overcoming the limitations of each single approach. A pivotal role was played by the index ECG, that resulted crucial both for diagnosis and for treatment.

FIGURE 2 Top: fluoroscopic views showing catheters placement during radiofrequency applications. Bottom: surface and intracavitary tracings during mapping of ventricular tachycardia on the posterior left ventricular wall, showing early low-voltage, fragmented potentials (arrow) on the distal dipole of the roving catheter, with far-field atrial potential (asterisk) on the proximal dipole, indicating proximity to the mitral annulus. Abbreviations: ABL, ablation/roving catheter; AP, antero-posterior projection; CS, coronary sinus catheter; d, distal dipole; ICD, defibrillation catheter; L 9-10, proximal coronary sinus dipole; L 1-2, distal coronary sinus dipole; LAO, left anterior oblique projection; p, proximal dipole; RA, right atrial appendage catheter; RAO, right anterior oblique projection; RV, right ventricular diagnostic catheter

FIGURE 3 Right: postero-anterior view of the activation map of the left ventricle, obtained with Biosense Webster CARTO3 system, showing the earliest activations, during ectopies, located in a region between the posterior papillary muscle insertion and the mitral annulus, within the posterior left ventricular wall (bigger white spot; in the color-coded scale, red indicates the earliest activation, while purple indicates the less early activation, relative to the apex of the ectopic QRS in V_4). Left: surface electrocardiogram and intracavitary tracings from the roving catheter, showing low-voltage, fragmented potentials on the distal dipole (MAP 1-2), with completely negative deflection of the unipolar signal (MAP 1), 32 ms earlier than the onset of the surface signal. Abbreviations: MAP, ablation/roving catheter; PA, postero-anterior view

FIGURE 4 Panel (a) electrocardiogram taken 2 days after the ablation procedure, showing recurrence of the ectopic activity, in form of isolated beats and irregular long cycle "tachycardia"; note some degrees of fusion between ventricular ectopies and sinus beats. Panel (b) electrocardiogram at discharge, showing absence of ectopic activity

CONFLICT OF INTEREST

None.

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REFERENCES

- Basso, C., Perazzolo Marra, M., Rizzo, S., De Lazzari, M., Giorgi, B., Cipriani, A., ... Iliceto, S. (2015). Arrhythmic mitral valve prolapse and sudden cardiac death. *Circulation*, 132, 556–566. <https://doi.org/10.1161/CIRCULATIONAHA.115.016291>
- Sriram, C. S., Syed, F. F., Ferguson, M. E., Johnson, J. N., Enriquez-Sarano, M., Cetta, F., ... Ackerman, M. J. (2013). Malignant bileaflet mitral valve prolapse syndrome in patients with otherwise idiopathic

out-of-hospital cardiac arrest. *Journal of the American College of Cardiology*, 62, 222–230. <https://doi.org/10.1016/j.jacc.2013.02.060>

Syed, F. F., Ackerman, M. J., McLeod, C. J., Kapa, S., Mulpuru, S. K., Sriram, C. S., ... Noseworthy, P. A. (2016). Sites of successful ventricular fibrillation ablation in bileaflet mitral valve prolapse syndrome. *Circulation Arrhythmia and Electrophysiology*, 9, e004005. <https://doi.org/10.1161/CIRCEP.116.004005>

How to cite this article: Coluccia G, Punzi R, Polini S, La Rosa C, Rillo M. The nightmare ECG of a spontaneously aborted sudden cardiac death. *Ann Noninvasive Electrocardiol*. 2019;24:e12547. <https://doi.org/10.1111/anec.12547>