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CASE REPORT

Arrhythmogenic right ventricular cardiomyopathy and left atrial tachycardia: a case report

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Abstract Little is known about atrial arrhythmias in arrhythmogenic right ventricular cardiomyopathy (ARVC). A 46-year-old man with definite ARVC presented with palpitations and exertional dyspnoea. Electrocardiogram showed a supraventricular tachycardia. Despite no prior cardiac surgery or atrial fibrillation ablation, electrophysiological study revealed a left atrial (LA) re-entrant circuit characterized by a slow fractionated potential bounded by two areas of double potentials giving a figure-of-eight pattern on activation map. Located on the LA roof within a zone of low bipolar voltages, this unusual substrate can be associated with a primitive atrial myopathy in ARVC.

Keywords ARVC • Supraventricular tachycardia • Left atrial re-entrant tachycardia • Low voltage area • Atrial scar • Case report

Learning points

- Atrial arrhythmias are common in patients with arrhythmogenic right ventricular cardiomyopathy (ARVC).
- Left atrial re-entrant arrhythmia may be associated with a primitive atrial myopathy due to ARVC.

Introduction

Arrhythmogenic right ventricular cardiomyopathy (ARVC) is an inherited cardiomyopathy characterized by a progressive loss of myocytes with fibrofatty replacement.[1] Originally described as a right ventricular (RV) disease, ARVC belongs to a broad disease spectrum that includes left dominant and biventricular forms.[2,3] Some evidence suggests that ARVC may directly involve atrial chambers[4–6] but the thin-walled nature of the human atrial myocardium limits conventional imaging accuracy to assess fibrofatty infiltration. Furthermore, most of human autopsy series focused on ventricular assessment.[2,3]

Case report

A 46-year-old Caucasian gentleman presented with a 2 months history of palpitations and exertional dyspnoea. On physical examination, he had a heart rate of 75 beats/min, a blood pressure of 115/80 mmHg, and a temperature of 36.9 °C. Cardiac auscultation revealed normal first and second heart sounds. No signs of congestive heart failure were noted. Electrocardiogram (ECG) showed a supraventricular arrhythmia with positive P-waves in inferior and V1 leads but negative P-waves in lead I (*Figure 1*).

His past medical history was significant for paroxysmal atrial fibrillation and typical flutter treated by cavo-tricuspid isthmus ablation. Class I obesity (body mass index of 34 kg/m^2) was his sole cardiovascular risk factor.

He was classified as having definite ARVC according to the 2010 revised Task Force Criteria[1] at the age of 41 through familial screening. Three major and one minor criteria were found out: ARVC confirmed in a first-degree relative, inverted T-waves in precordial leads, lateral RV dyskinesia with RV ejection fraction <40% by magnetic resonance imaging (MRI) and late potentials on signal averaged ECG.

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Timeline

Five years prior to presentation	Medical history significant for arrhythmogenic right ventricular cardiomyopathy (ARVC)
Two months prior to presentation	Onset of palpitations and exertional dyspnoea
Initial presentation	Physical examination showed a heart rate of 75 beats/min, a blood pressure of 115/80 mmHg, no signs of congestive heart failure Electrocardiogram revealed a supraventricular arrhythmia with positive P-waves in inferior and V1 leads
Seven months after initial presentation	Left atrial (LA) re-entrant tachycardia diagnosed on electrophysiological study Figure-of-eight pattern on the LA activation map with an unusual substrate located in the roof of the left atrium Successful radiofrequency catheter ablation
Follow-up (29 months after catheter ablation)	Patient free from arrythmia

Clinical course of ARVC was marked by a myocarditis mimicking an acute coronary syndrome. Coronary arteriograms were normal. Cardiac MRI showed a subepicardial enhancement in the postero-lateral wall of the left ventricle (LV). Subsequent cardiac MRI showed no gadolinium enhancement in the same region.

Last cardiac examination revealed an asymptomatic patient with normal LV volume, mass, and ejection fraction. Left atrium was mildly dilated (surface 28 cm^2, volume 70 mL). Right ventricular function was altered (34% ejection fraction on MRI) with a normal end-diastolic volume (71 mL/m^2). His medications included nadolol 80 mg daily and warfarin. He had no implantable cardioverter defibrillator.

Owing to the persistence of symptoms, an electrophysiological study was proposed. Left atrial (LA) activation mapping was achieved during tachycardia and revealed a presumably re-entrant circuit. The activation map depicted in *Figure 2* was characterized by a combination of a slow fractionated potential (purple tag) bounded by two areas of double potentials (blue tags) giving a figure-of-eight pattern. This substrate was located in the roof of the left atrium, mid-distance between the appendage, and the right superior pulmonary vein (PV) ostium. This area was also characterized by low bipolar voltages (*Figure 2*, Panel C). Radiofrequency current application at the site of fractionation (purple tag on *Figure 2*), resulted in abrupt termination of the tachycardia, which was not inducible thereafter.

Over a follow-up of 29 months, the patient was free from any arrhythmia.

Discussion

The reported prevalence of atrial arrhythmias in patients with ARVC varies between 4 and 42%.[1,2] Camm *et al.*,[2] described a 14.1% rate with atrial fibrillation being the most common (80%), followed by atrial flutter (31%) and other SVTs (23%). Left atrial re-entrant tachycardia is an uncommon arrhythmia mostly describe in patients with prior cardiac surgery or catheter ablation of atrial fibrillation. Atrial incision, gap in circumferential ablation lines around PVs and/or in additional lines serve as the ideal substrate for focal or macro-reentrant atrial tachycardias.[3]

Desmosomal proteins are present throughout the cardiac system including the atrial myocardium.[2] Some evidence suggests that ARVC related fibrofatty lesions may involve atrial chambers. Deteriorated

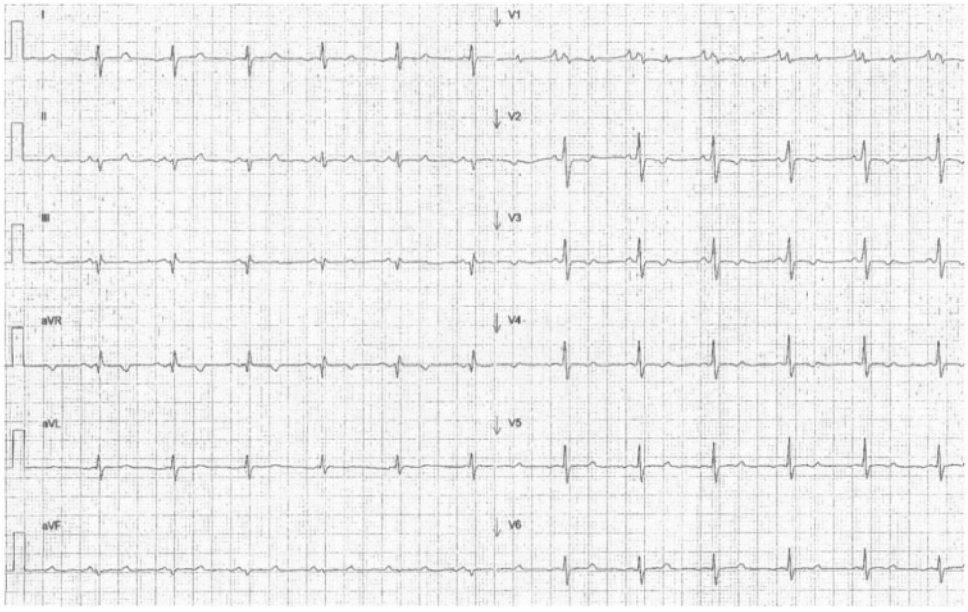

Figure 1 Twelve-lead electrocardiogram showing an atrial arrhythmia with positive P-waves in inferior leads and V1, negative P-waves in I. Regular 2: 1 ventricular response.

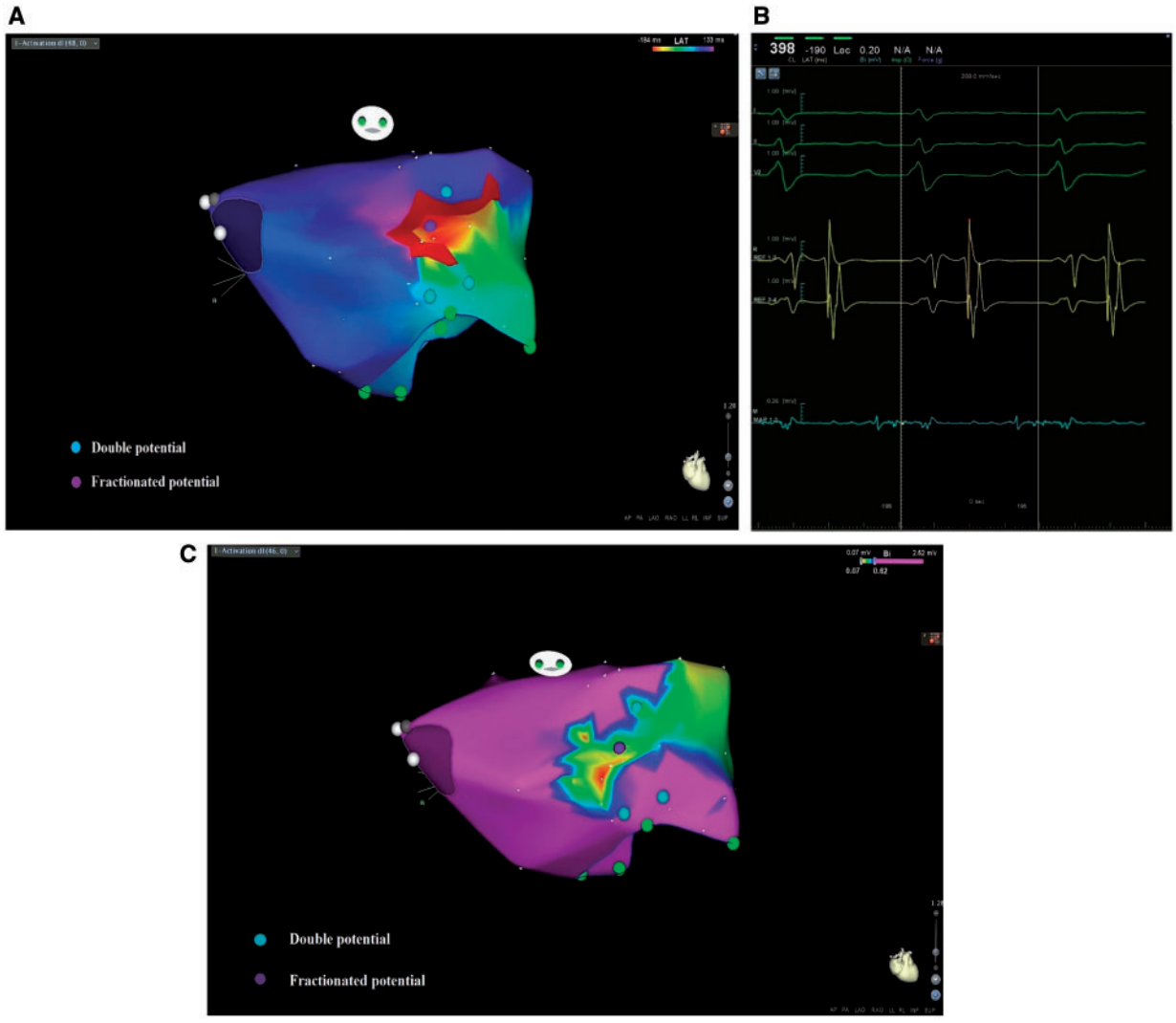

Figure 2 Panel (*A*) Three-dimensional activation map showing a figure-of-eight re-entry with slow conduction (purple tag) double potentials (blue tags) located between the base of the left atrial appendage and the right superior pulmonary vein. Panel (*B*) Surface electrocardiogram (green), intracardiac electrograms from the coronary sinus (yellow), and fractionation recorded by the mapping catheter (light blue) at the site indicated by a purple tag on the map. Panel (*C*) Bipolar voltage map, purple colour indicate > 0.62 mV.

atrial electrical activation (prolonged P-wave duration and abnormal P-wave morphology) has been observed in patients with ARVC compared to age- and sex-matched healthy control subjects.[4] Presence of fatty tissue was found within the sino-atrial node in two patients with ARVC[5] and LA fibrofatty myocardial replacement was identified in 9 of 12 boxers with ARVC.[6] Unfortunately, the thin-walled nature of the human LA myocardium makes atrial biopsy unwise and limits conventional imaging accuracy to assess fibrofatty infiltration. Furthermore, most of the human autopsy series focused on ventricular assessment.[2]

Conclusion

We reported a 46-year-old man with ARVC and LA re-entrant tachycardia. Located at the roof of the LA chamber, the substrate appeared within an unusual zone of low bipolar voltages. Adding to a growing body of evidence suggesting atrial fibrofatty myocardial replacement in ARVC, we hypothesize that the observed low voltage area and related LA arrhythmia can be associated with a primitive atrial myopathy in ARVC.

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.

Author contributions: C.K. and Z.S. were involved in compilation of data and writing of this manuscript. F.B. and D.L. were involved in the management of the case.

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