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Case reports

Two simultaneous right ventricular tachycardias in a case of arrhythmogenic right ventricular dysplasia

GIANFRANCO BUJA, BORTOLO MARTINI, ANDREA NAVA

From the Department of Cardiology, University of Padua, Medical School, Padua, Italy

SUMMARY A 51 year old woman with arrhythmogenic right ventricular dysplasia had two types of ventricular tachycardia—(*a*) a regular and sustained tachycardia and with normal frontal plane axis on electrocardiography and (*b*) an irregular non-sustained tachycardia with a leftward frontal plane axis. Changes in the QRS complex were sometimes seen during the sustained ventricular tachycardia. The clinical, electrocardiographic, and electrophysiological data were consistent with the diagnosis of two different and sometimes simultaneous tachycardias originating in the right ventricle.

This case suggests a possible new mechanism for the multiform appearances of the ventricular tachycardia.

Multiple forms of ventricular tachycardias are commonly found in patients with arrhythmogenic right ventricular dysplasia.[1 2] But the simultaneous occurrence of two separate ventricular tachycardias has not been reported before. We report a patient with arrhythmogenic right ventricular dysplasia and unusual multiform sustained and non-sustained ventricular tachycardias, both with a left bundle branch block configuration but normal (+45°) and leftward (−20°) frontal plane axis. The clinical, electrocardiographic, and electrophysiological data support the diagnosis of two different and sometimes simultaneous tachycardias both originating in the right ventricle.

Case report

A 51 year old woman had a three year history of untreated mild hypertension. For many years she had complained of occasional palpitation. The electrocardiogram in sinus rhythm was normal, with inverted T waves in V1 and V2 and flattening or mild inversion in V3, and isolated uniform extrasystoles with a left bundle branch block configuration. In August 1986 she was admitted to the coronary care unit with palpitation and hypotension; the electrocardiogram showed regular, sustained ventricular tachycardia (180/min) and left bundle branch configuration with a normal frontal plane axis (+45°) (fig 1a). Intravenous lignocaine (150 mg) was ineffective but a 150 J electrical shock restored the sinus rhythm.

The chest *x* ray was normal but the M mode and cross sectional echocardiography showed an abnormal trabecular pattern and regional dyskinesia of the inflow and apical segments of the right ventricle with generalised dilatation of the outflow tract. Few hours later brief episodes of non-sustained (4–5 QRS complexes), irregular, repetitive ventricular tachycardia were recorded at a rate of 110–130/min with left bundle branch block configuration and a leftward frontal plane axis (−20°) (fig 1b). On the second day, in the absence of drug treatment, the previous sustained ventricular tachycardia recurred; long electrocardiographic strips sometimes showed aberrations in the QRS complexes (generally in leads II, III, and aVF) that were characterised by gradual variations of the frontal plane axis from +45° to −20°, slight differences in the RR intervals, and fusion complexes (fig 2a). Intravenous flecainide (2 mg/kg) was given slowly and abolished the ventricular tachycardia.

The haemodynamic and angiographic investigation showed a right ventricular end diastolic volume of 100 ml/m^2, an end systolic volume of 68 ml/m^2,

Requests for reprints to Dr Gianfranco Buja, Department of Cardiology, University of Padua, Via Giustiniani 2, 35100 Padova, Italy.

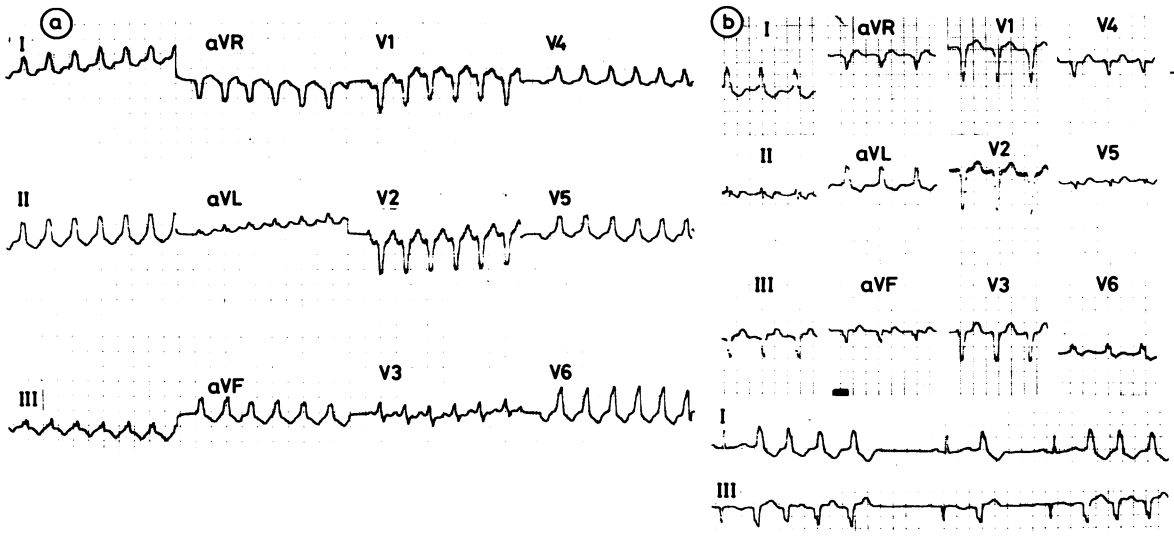

Fig 1 *(a) Electrocardiogram showing regular sustained ventricular tachycardia at a rate of 180 beats/min with left bundle branch configuration and normal axis (+45°). (b) Electrocardiogram showing irregular non-sustained repetitive ventricular tachycardia at a rate of 110–130 beats/min with left bundle branch configuration and a leftward frontal plane axis of −20°. The electrocardiographic strip of I and III shows the pattern of the ventricular tachycardia.*

and an ejection fraction of 32%; structural alterations of the trabecular zone, hypokinesia of the inferior and apical wall, bulging of the inflow tract, and infundibular dilatation were also present. The left ventricle was normal.

At electrophysiological study the basal intervals were normal (PA = 35 ms, AH = 100 ms, HV = 40 ms). Delayed and fractioned electrical potentials were recorded from the right ventricular outflow tract. Clinical non-sustained ventricular tachycardia with a leftward frontal plane axis appeared spontaneously, and a complete catheter electrode endocardial mapping, performed in eight different sites in the right ventricle, showed that electrical activation started in the right ventricular apex (fig 2b). Furthermore, sustained ventricular tachycardia with a normal frontal plane axis was easily induced by right ventricular extrastimuli during ventricular drive at 600 ms with coupling intervals for S1–S2 of 280 ms to the right ventricular refractory period (220 ms). Accurate catheter electrode endocardial mapping showed that electrical activation started in the right ventricular outflow tract (fig 2b). In addition, during the induced clinical sustained ventricular tachycardia, the previous alteration of the QRS complexes appeared spontaneously, and catheter electrode endocardial mapping showed a simultaneous early electrical activation in both the right ventricular apex and outflow tract (fig 2b). Right ventricular overdrive at a cycle length of 300 ms or a bolus of intravenous flecainide restored the sinus rhythm during sustained tachycardia but not during non-sustained ventricular tachycardia, which reappeared after the infusion. Also, after flecainide right ventricular incremental pacing—with two or three premature stimuli during three different ventricular drives, and bursts, delivered to either the right ventricular apex or right ventricular outflow tract, did not induce the sustained ventricular tachycardia. Oral flecainide (200 mg/day) was given but at a follow up on 24 hour Holter monitoring sporadic episodes of the non-sustained ventricular tachycardia persisted. Treatment with amiodarone (1000 mg/week) was successful and 12 months later she was free of symptoms and arrhythmias.

Discussion

This patient with arrhythmogenic right ventricular dysplasia that affected the apex and the inflow and the outflow segments of the right ventricle may be described as having pleomorphic ventricular tachycardia.[1] But she was unusual in having two types of ventricular tachycardia: (*a*) a sustained ventricular tachycardia with normal frontal plane axis and (*b*) an irregular, non-sustained, repetitive ventricular tachycardia with a leftward frontal plane axis that was not corrected by intravenous flecainide. Electrocardiographic strips recorded during the sustained ventricular tachycardia sometimes showed a gradual shift of the QRS axis from normal to left axis

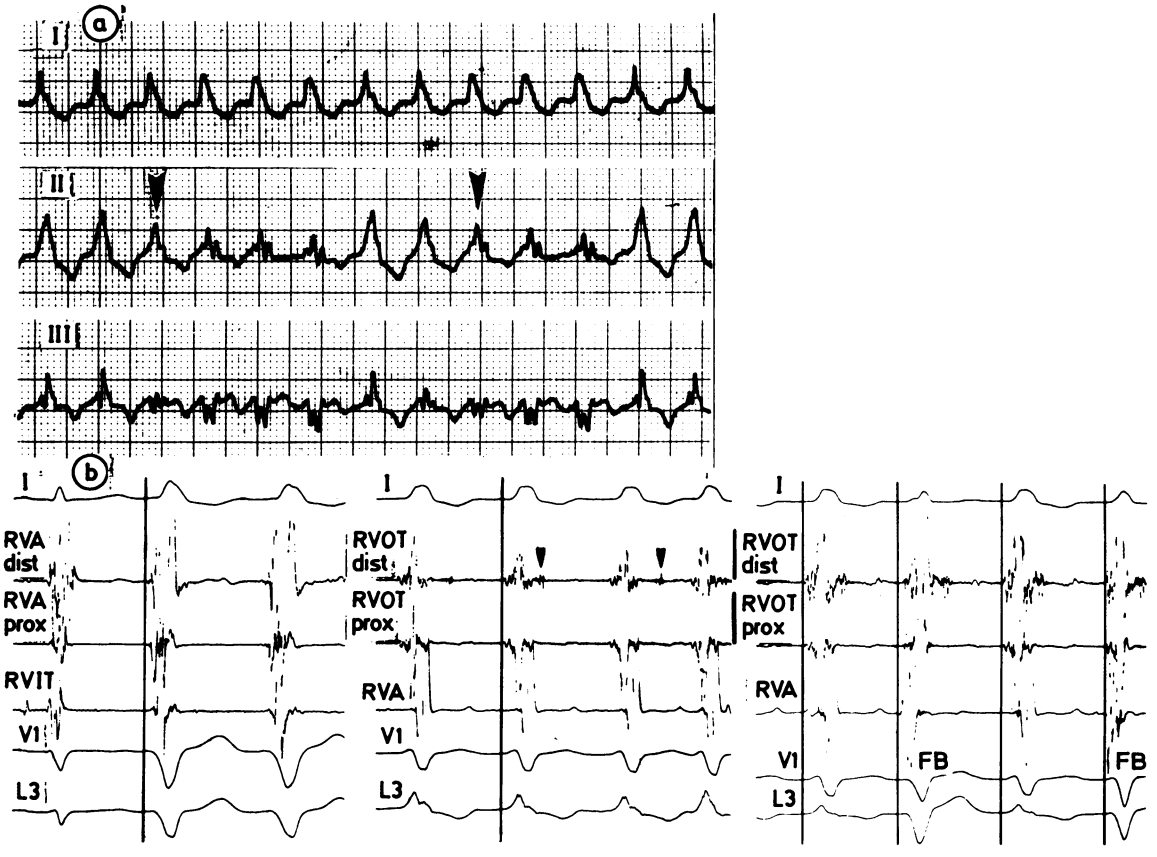

Fig 2 *(a) Electrocardiographic trace of leads I–III during the spontaneous sustained ventricular tachycardia. Note the shift of frontal plane axis, the slight change in RR intervals, and the presence of fusion beats (arrows). (b) Spontaneous non-sustained ventricular tachycardia (left panel), the clinical sustained ventricular tachycardia induced by ventricular stimulation (middle panel), and the spontaneous alteration of the QRS complex (see fig 2a) recorded during the sustained ventricular tachycardia (right panel). Top to bottom: lead I, electrograms from the right ventricular apex (RVA distal and proximal electrodes), the right ventricular inflow tract (RVIT), the right ventricular outflow tract (RVOT distal and proximal electrodes), and VI and III. The earliest electrical activation was recorded in the RVA (left panel) and in the RVOT (middle panel). In the right panel, during the pleomorphism, the earliest activation changes were recorded in the RVOT in the first and the third beat, in the RVA (nearly simultaneously) in the second beat, and simultaneously (RVA and RVOT) in the fourth. FB, fusion beat. Late fractioned electrical potentials were recorded in RVOT (arrows). The vertical lines mark the onset of the earliest endocardial electrograms. Note that in each panel the electrograms precede the onset of the surface QRS complex. Paper speed 100 mm/s.*

deviation, slightly irregular RR intervals, and fusion beats.

Our case resembles a case reported by Strasberg *et al* who found ventricular tachycardia with right bundle branch block and right and left axis deviation in opposite directions[3]. In the absence of electrophysiological data they suggested that their patient had two separate foci or a common focus, within or close to the left bundle branch, with different exit pathways and different electrophysiological properties. In our case the clinical and electrocardiographic behaviour and the response to the electrical stimulation and to flecainide strongly support the presence of two different electrophysiological mechanisms. Moreover, the results of endocardial catheter electrode mapping clearly showed that the two tachycardias had separate origins.

Another unusual feature of our case was the occurrence of alterations in the QRS complexes during sustained ventricular tachycardia. Electrophysiological mapping of the right ventricle showed the simultaneous onset of ventricular activation in

both the apex and outflow tract, and this is further evidence that the two different tachycardias occurred together. As far as we know, mapping has never shown this feature before. Nevertheless, we cannot exclude the possibility of a single site of origin with different routes of impulse propagation and the simultaneous arrival of impulses at the apex and outflow tract. Our case shows that there can be widespread involvement of the right ventricle even when the heart silhouette is normal and there are no important clinical signs; this suggests a possible new mechanism for the multiform appearances of the ventricular tachycardia.[4]

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