From wide QRS tachycardia to a diagnostic surprise 🎞

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ABSTRACT

A 29-year old male patient presented with a hemodynamically significant ventricular tachycardia. Despite a comprehensive examination, the correct diagnosis was unfortunately established after two years. We discuss why the correct diagnosis was initially overlooked in physical examination, electrocardiogram, echocardiography and cardiac magnetic resonance imaging, and which findings led the cardiologists to misdiagnose the patient. We have organized this report in a format that the information is presented to a consultant physician by a resident physician to simulate the way such information emerges in the real life as we have encountered. The consultant physician responds as new information is presented, expressing his reasoning with the reader.
(Anadolu Kardiyol Derg 2014; 14: 471-4)

Resident physician: A 29-year old male patient admitted to the emergency service with palpitation and dyspnea. His blood pressure was 80/45 mm Hg and heart rate was 120 beat per minute. Electrocardiogram (ECG) showed wide QRS tachycardia with a left bundle branch block (LBBB)-like pattern (Fig. 1). Emergent direct-current cardioversion was performed due to hemodynamic instability.

Consultant physician: A prompt decision to perform direct-current cardioversion for hemodynamic instability without wasting time for distinguishing ventricular tachycardia (VT) from supraventricular tachycardia with aberrant conduction is the right approach, as in this case. Analysis of the ECG reveals atrioventricular dissociation, Q wave in lead V6 despite LBBB-like pattern, and the interval from QRS onset to S-nadir ≥0.70 milliseconds in lead V1 (1). All these are consistent with VT. LBBB and inferior QRS axis implies that arrhythmogenic focus is around the right ventricular outflow tract (RVOT).

Resident physician: The patient was taken to the intensive care unit. History taking revealed that he suffered from palpitation and exercise-induced dyspnea for six months although he had no known cardiac disease. There was no history of syncope or premature death in his family. Resting ECG showed normal sinus rhythm, right bundle branch block, T wave negativity and epsilon wave-like deflections in leads V2-V3 (Fig. 2). Physical examination was unremarkable except a retrosternal soft holosystolic murmur. Laboratory findings were as follows: The white-cell count was 22100 per cubic millimeter, with 70 percent granulocytes; hemoglobin, 16.2 g/dL; serum creatinine, 1.86 mg/dL; glucose 198 mg/dL; and troponin, 0.43 ng/mL (the upper limit of normal value is 0.06 ng/mL)

Consultant physician: Troponin elevation indicates myocardial injury, and accompanying leukocytosis suggests acute myocarditis in this young patient. Acute coronary syndrome is also probable. However, absence of chest pain and ECG evidence of ischemia, and absence of ST elevation compatible with acute myocarditis make them less likely. Since both diagnoses might still occur in this case, it is required to exclude them with a myocardial perfusion scintigraphy or coronary angiography. Also, at first glance, the findings in ECG can be confused with those of arrhythmogenic right ventricular cardiomyopathy (ARVC) because T wave negativity in leads V2-V3 is a sensitive marker for ARVC. However, this is a normal finding in case of right bundle branch block. The deflections just after the QRS in leads V2-V3 resemble an epsilon wave which is highly specific for ARVC. But one can notice that it does not exist in V1 and those in leads V2-V3 are simply artifact.

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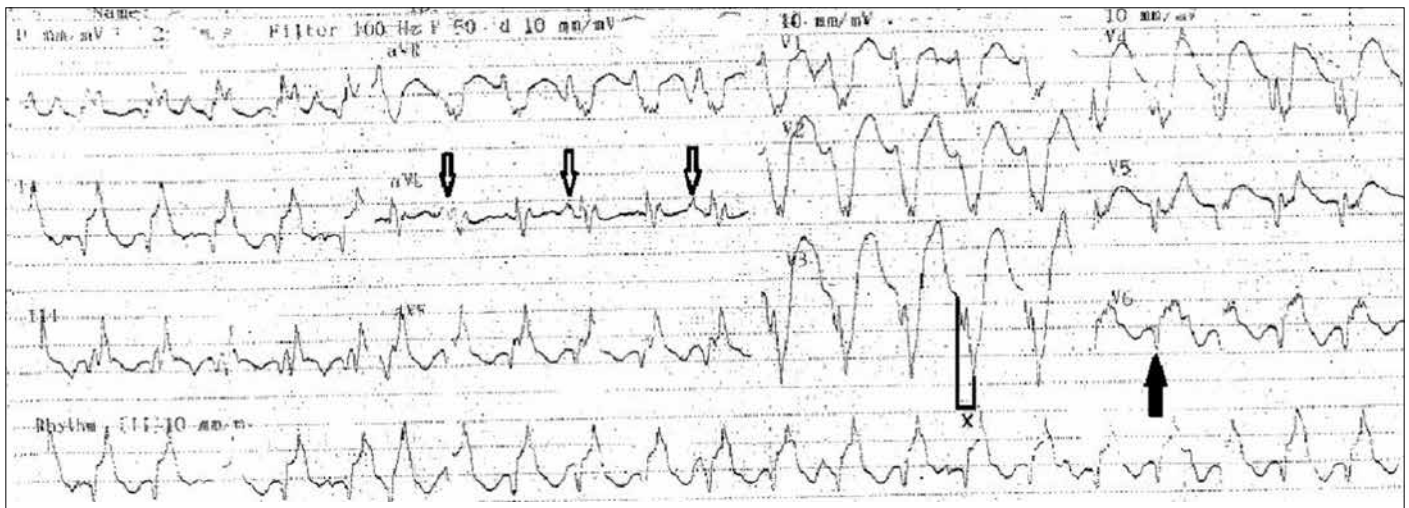
Figure 1. Wide QRS tachycardia with a left bundle branch block-like pattern. Atrio-ventricular dissociation (*blank arrows*), Q wave in lead V6 despite left bundle branch block-like pattern (*filled arrow*), and the interval from QRS onset to S-nadir (X) ≥0.70 milliseconds indicates ventricular tachycardia
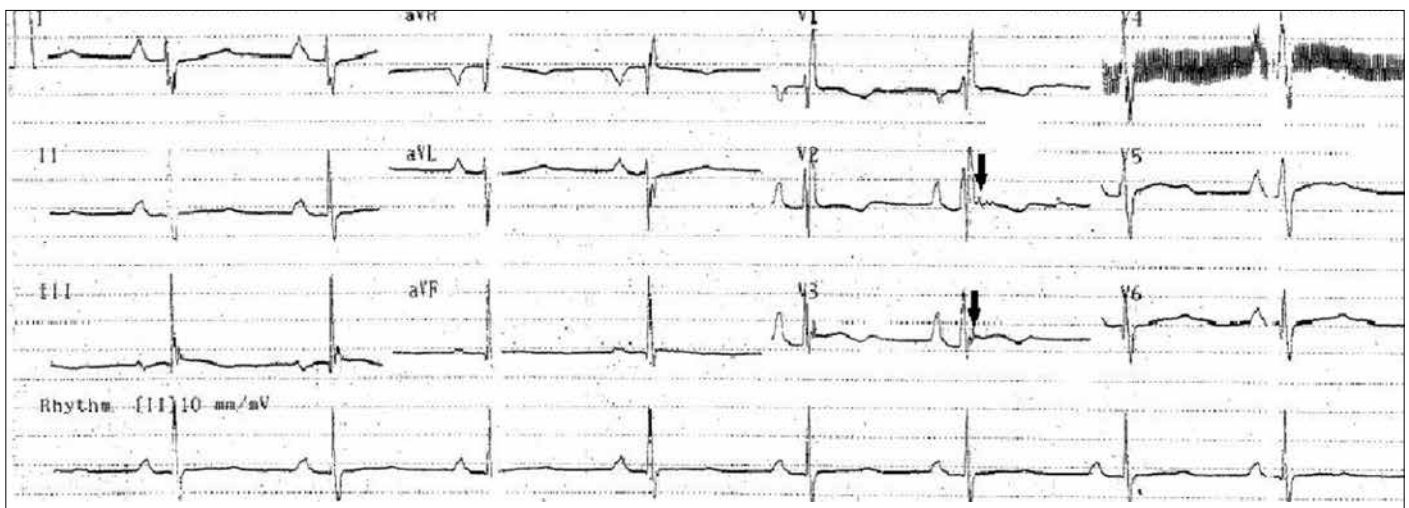

Figure 2. Normal sinus rhythm with right bundle branch block and P pulmonale. *Filled arrows* indicate epsilon wave-like deflections in leads V2-V3

Resident physician: During follow-up, hemodynamically significant VT recurred and direct current cardioversion was successfully performed again. Intravenous amiodarone was administered. Then coronary angiography was undertaken and revealed slow flow in the left anterior descending artery. The left circumflex and right coronary arteries were normal.

Consultant physician: Although slow flow in the left anterior descending artery makes one consider an ischemic etiology, this is not the case because RVOT is supplied by right coronary artery which is known to be normal in this patient. Accordingly, acute coronary syndrome is ruled out. At this stage, transthoracic echocardiography (TTE) would be of a great help.

Resident physician: TTE results were as follows: normal left ventricular function with an ejection fraction of 60 to 65%, normal function of aortic and mitral valves; normal aortic and pulmonary blood flow velocities, dilated right atrium, dilated right ventricle (RV) with severe systolic dysfunction and excessive trabeculation, moderate tricuspid valve regurgitation (TR), and systolic pulmonary artery pressure of 70 to 80 mm Hg. Atrial septal defect was excluded by performing saline contrast injection. Echocardiographer considered ARVC or isolated RV noncompaction as the most likely diagnoses and recommended cardiac magnetic resonance imaging for the differential diagnosis. Later in the follow-up, blood tests were as follows: The white-cell count was 14500 per cubic millimeter; serum creatinine, 1.24 mg/dL; glucose, 98 mg/dL; troponin, 7 ng/mL; and normal thyroid function tests.

Consultant physician: Acute myocarditis is not supposed to present with a normal left ventricle and dilated RV. Idiopathic RVOT tachycardia is also excluded because of the presence of structural heart disease. Neither ARVC nor isolated RV non-
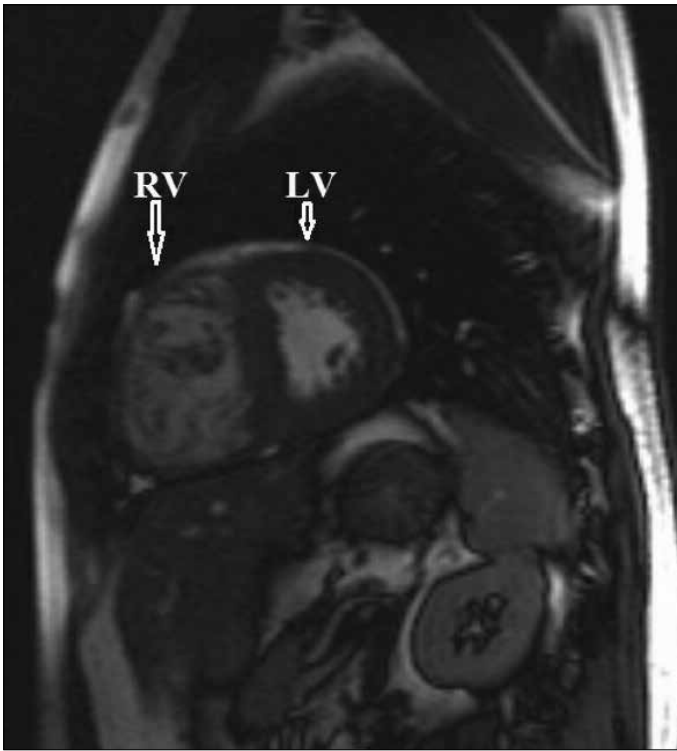
Figure 3. Magnetic resonance imaging shows thin-walled dilated right ventricle with excessive trabeculation
LV - left ventricle; RV - right ventricle

compaction alone is compatible with troponin elevation. However, troponin elevation might be caused by subendocardial ischemia/injury due to hypotension related to VT. Isolated RV noncompaction without LV involvement is such a rare condition that there have been only few case reports (2, 3). It is even debatable to adopt it as a distinct entity since increased trabeculation may occur secondary to severe ventricular dilatation. On the other hand, severe pulmonary hypertension from any cause might be the reason for all these findings. Cardiac magnetic resonance imaging would be helpful for the differential diagnosis.

Resident physician: Cardiac magnetic resonance imaging showed right heart dilatation with excessive trabeculation and thin RV wall (Fig. 3, Video 1). However, no specific diagnosis was able to be established. Putting together all the findings, ARVC was established as the most likely final diagnosis. The patient was prescribed oral beta blocker and amiodarone and was implanted with an implantable cardioverter-defibrilator (ICD). He was discharged in a stable condition after 11 days.

Consultant physician: One of the major criteria for ARVC suggested by the Task Force is global RV dilatation (4). This finding was shown in this case but other possible findings such as RV wall aneurysm/dyskinesia were not reported. Also, pulmonary hypertension cannot be explained by ARVC and appears to have been ignored in this case.

Resident physician: In the next two years, the patient had multiple admissions for recurrent VT. Anti-arrhythmic drugs (amiodarone, metoprolol, sotalol, propafenone, or combinations of these drugs) were not helpful to prevent attacks. Catheter ablation was attempted in two different sessions and was not successful. All these made clinicians consider the possibility of an additional condition responsible for the refractoriness to treatment. Accordingly, a reappraisal of the case was initiated. First, TTE was repeated by an experienced echocardiographer. The result of TTE was very surprising: The patient had severe congenital pulmonary valve stenosis. Despite poor echogenic image quality, the echocardiographer succeeded to acquire a systolic gradient of almost 100 mm Hg which was hardly perceptible. Pathologic findings were in fact related to the severe PS, not to the ARVC. Therefore, pulmonary balloon valvuloplasty was performed and a 20 mm Hg gradient was successfully achieved. ICD was left in place in case of VT recurrence in the future.

Comment

If the correct diagnosis would have been established, pulmonary balloon valvuloplasty would be performed at the first place. This in turn might have provided a decrease in RV pressure and RV wall ischemia. VT could be controlled, and accordingly, it would be a better choice not to implant ICD initially. A watchful waiting under the anti-arrhythmic treatment would suffice. On the other hand, ICD implantation could still be required, because re-entry foci of myocardial fibrosis related to longstanding pressure overload at the late stages of RV failure might still predispose the patient to VT despite intraventricular pressure decrease provided by pulmonary balloon valvuloplasty.

It is worth to discuss about the possible reasons why the correct diagnosis might have been overlooked by each diagnostic method:

Physical examination

Physical findings consistent with RV failure and a retrosternal holosystolic murmur were actually determined at the beginning. However, the murmur was mistakenly attributed only to that of TR. The auscultatory findings of the accompanying TR was prominent and overshadowed the more subtle signs of severely stenotic pulmonary valves with reduced motion.

Electrocardiogram

VT with a LBBB-like pattern and inferior axis indicated the focus originating from the RVOT. Idiopathic RVOT tachycardia and ARVC were then considered as the most likely diagnoses which initiated a chain of bias. Moreover, artifacts at leads V2-V3 in resting ECG were misinterpreted as an epsilon wave.

Transthoracic echocardiography

The high clinical probability of ARVC written in the request form for TTE might have made the first echocardiographer focus on primarily whether there were findings compatible with ARVC

without giving adequate interest for pulmonary stenosis. Poor image quality might have inhibited to detect the exact flow velocity across the pulmonary valve as well. Underestimation of jet velocity by malalignment of continuous wave Doppler beam, or using pulsed wave Doppler and sampling the pulse volume proximal to the stenotic site could be the other possible explanations. A high TR velocity indicating increased RV systolic pressure was also misinterpreted as an indicator of pulmonary hypertension which, in this fashion, should have not been diagnosed in case of pulmonary stenosis. Absence of RV hypertrophy could be explained by late presentation of the case since RV dilatation and decreased wall thickness dominated the picture at this late stage.

Cardiac magnetic resonance imaging

Global RV dilatation seen in this case could be regarded as one of the major findings of ARVC. It has high sensitivity but low specificity. Thin RV wall could be considered compatible with
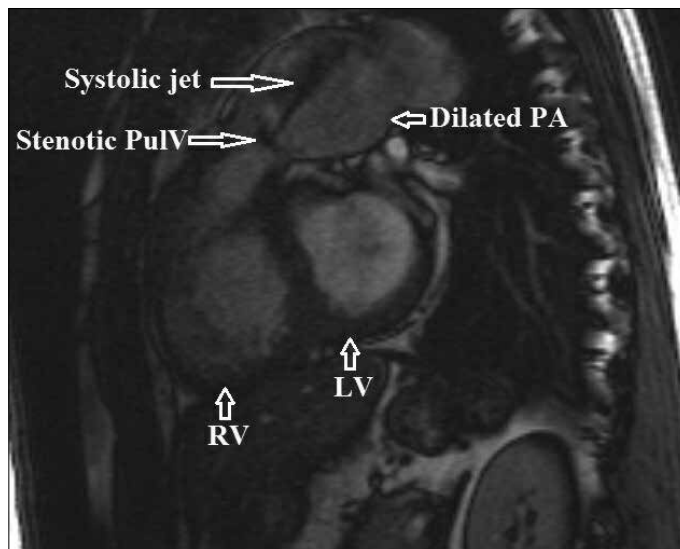

Figure 4. Magnetic resonance imaging of the right ventricular outflow tract clearly demonstrates ejection flow of the stenotic pulmonary valve and post-stenotic pulmonary artery dilatation
LV - left ventricle; PA - pulmonary artery; PulV - pulmonary valve; RV - right ventricle

ARVC as well. However, it is likely that both ECG and TTE results suggesting ARVC might cause a bias for the radiologist. Accordingly, too much focus on the presence or absence of ARVC findings might have made him overlook the PS which was in fact clear to ascertain by cine-magnetic resonance imaging of the RVOT (Fig. 4, Video 2).

Conclusion

This report shows detrimental effect of bias for overreliance to medical equipment. To avoid bias in performing or interpreting any procedure, it is important to look for any other possibilities *in a standard manner*. Each pathologic finding (e.g., so-called pulmonary hypertension as in this case) should also be explained by a cogent reason.

Conflict of Interest: none.

Video 1. Cine-magnetic resonance imaging shows thin-walled dilated right ventricle with excessive trabeculation
Video 2. Cine-magnetic resonance imaging of the right ventricular outflow tract clearly demonstrates ejection flow of the stenotic pulmonary valve and post-stenotic pulmonary artery dilatation

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