Acute Ventricular Tachycardia in Young Man with Arrhythmogenic Right Ventricular Cardiomyopathy

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BACKGROUND

- Arrhythmogenic right ventricular cardiomyopathy (ARVC) is a rare heart muscle disease is characterized by fibrofatty replacement of the myocardium leading to electrical instability and ventricular arrhythmias and increasing risk of sudden death.
- It is a autosomal dominant genetic disorder of the myocardium can cause ventricular tachycardia and sudden cardiac death in young people and athletes.
- The clinical hallmark of the disease is ventricular arrhythmias, arising predominantly from the right ventricle.
- Many patients with ARVC are asymptomatic. Presenting symptoms include palpitations, syncope, chest pain, dyspnea, sudden cardiac death, and heart failure.
- The diagnosis of ARVC is difficult and is based on ECG abnormalities, arrhythmias, structural abnormalities and tissue characteristics, as well as family history and genetics.
- In this case report, we discuss a young patient with known ARVC mutation but who remained asymptomatic until his presentation in the ED.

CASE DESCRIPTION

- 21-year-old male presented to the ED in ventricular tachycardia
  - –EMS gave him 80 mg lidocaine intravenously
  - –BP on arrival was stable, alert, mentating well
- In the ED, patient experienced intermittent ventricular tachycardia (Fig 1, 2).
- Magnesium and potassium repleted in the emergency department, all other labs are unremarkable.
- Admitted and started on metoprolol as well as amiodarone
- Subcutaneous ICD implanted, discharged home in stable conditions.

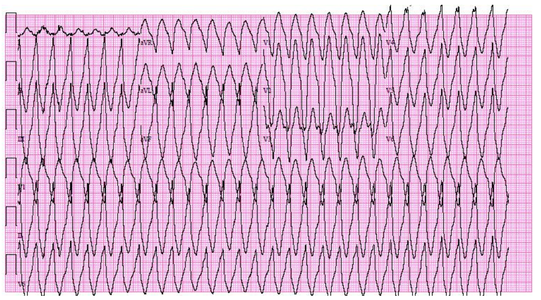

Figure 1. Ventricular tachycardia

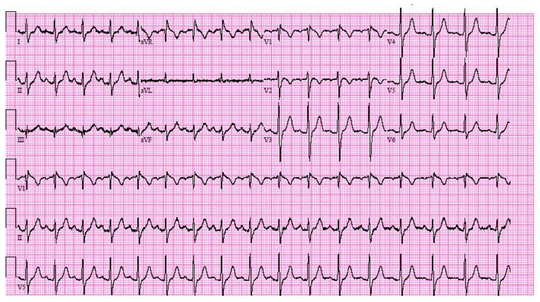

Figure 2. Return of sinus rhythm

DISCUSSION

- ARVC is one of the many genetic structural cardiac disease that can cause clinically significant events important for emergency physicians to recognize including tachyarrhythmia, syncope and sudden cardiac death.
- Scarring of the myocardial tissue leads to right ventricular dilation, and ventricular arrythmias originate from the right ventricle.
- Patient can present in wide complex tachycardias, as well as syncope, palpitations, dizziness.
- After initial stabilization and resuscitation, other essential parts of management include electrolyte repletion, anti-arrythmic medications, rate control with beta blockade, and ultimately admission for ICD for definitive management.

CONCLUSIONS

- ARVC is a genetic structural myocardial disease that causes RV dilation and arrhythmias (Fig 3).
- Diagnosis is made through a combination of patient history, clinical exam, ECG, cardiac imaging and genetic testing.
- ARVC can present in the ED in wide array of presentations, including wide complex tachycardias.
- After initial stabilization and resuscitation, the patient should be admitted for monitoring and definitive management with an ICD.

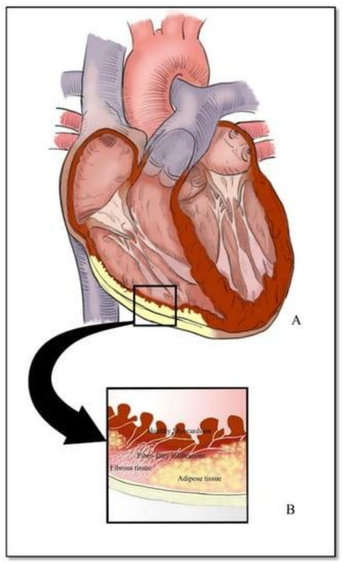

Figure 3. Representation of the white-yellow discoloration of the myocardium on macroscopic examination (**A**) and fibrous-adipose tissue deposition proceeding from the epicardial region towards the subendocardial portion of the myocardium (**B**).