A Rare Clinical Case of Symptomatic Ventricular Tachycardia caused by Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC)

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INTRODUCTION

- Arrhythmogenic right ventricular cardiomyopathy (ARVC) is a clinical entity characterized by ventricular arrhythmias of RV origin and ventricular pathology wherein there is fibrous or fibro-fatty replacement of the myocardium leading to initial regional wall motion abnormalities then progressing to global RV dilation with myocardial thinning throughout the process.(1)
- ARVC is a inherited cardiopathy that affects roughly one third of first degree relatives as shown in studies looking at common mutations involved in the ARVC. Siblings had a three-fold-increased risk of ARVD/C diagnosis compared with parents and children. There is both an autosomal dominant and recessive form, with the dominant being more common (3)
- Prevalence is estimated at 1 in 2000 to 1 in 5000(2)
- Presentation rarely occurs prior to puberty and is most common between the ages of 10-50, with a mean age of 30 upon diagnosis. Some 20% of patients are diagnosed after 50 years old.Symptoms include palpitations(67%), syncope(32%), atypical chest pain(27%), dyspnea(11%), and, rarely, sudden cardiac death(6%) (4)
- These patients are at high risk for ventricular arrythmias. The most common arrythmia is a sustained or nonsustained monomorphic VT that originates in the RV and therefore has a left bundle branch block (LBBB) pattern
- Supraventricular tachyarrhythmia can be seen in up to 25% of patients with ARVC(6). ARVC can also present with sudden cardiac death and has been implicated in the premature death of athletes

OVERVIEW OF THE CASE

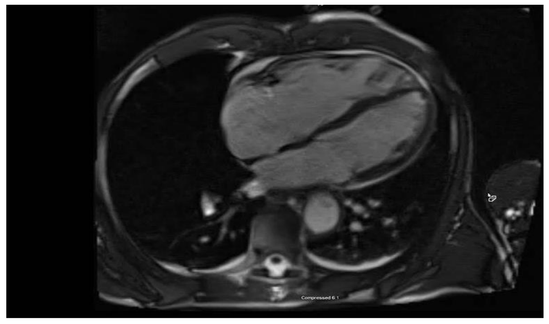
- The case in question is regarding a elderly male who developed monomorphic Ventricular tachycardia(MMVT) and was found to have ARVC
- ARVC suggested by Transthoracic Echocardiogram, confirmed via Cardiac MRI
- Patient diagnosed with Ventricular tachycardia likely as a result of ARVC
- This condition predisposes the patient to future arrythmias, Therefore a ICD was installed during his stay to prevent future sudden cardiac death
- This case was explored due to the abnormally late onset of ARVC in this patient, a unclear family history and his symptomatic presentation. All these characteristics are things clinicians should be aware of when considering this condition

OBJECTIVES

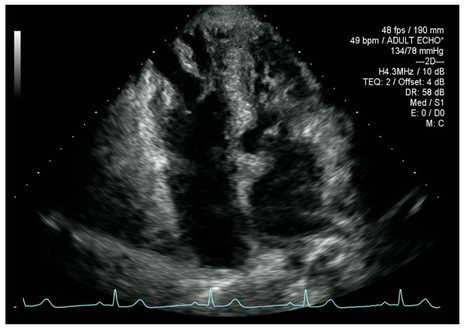
- Recognize the signs and symptoms of ARVC. Why does it warrant clinical suspicion?
- Learn about the diagnosis and interventions for ARVC
- Recognize the utility in considering ARVC as a possible differential for cardiac arrythmias

CLINICAL COURSE

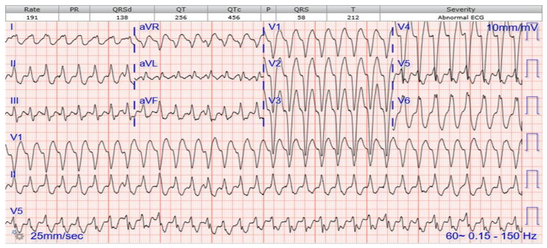
- Patient is a 74 year old male with a medical history of hyperlipidemia, hypertension, a stable abdominal aortic aneurysm, hypothyroidism, and syncope likely falsely attributed to a vasovagal etiology with Raynaud's who presented with tachycardia, palpitations with associated headache, lightheadedness, nausea.
- The patient' initial EKG showed monomorphic ventricular tachycardia.). patient became hypotensive so a DCCV at 120J was performed and the patient converted to NSR
- A EKG performed shortly after showed ST depressions in the lateral leads suggestive of ischemia, and the patient was transferred to Good Sam. A initial troponin was 0.027, a repeat troponin prior to transfer(multiple hours later due to transport delay) was 0.300. A EKG done prior to transfer showed normalization of the prior ST depressions. The patient was started on amiodarone and heparin prior to transfer on the advice of the cardiologist. On arrival the patient admitted to discontinuing his medications including lisinopril, atorvastatin, aspirin, and levothyroxine.
- The patient's ROS was positive for fatigue present for months, shortness of breath, palpitations and mild generalized weakness. His vital signs upon arrival were stable with borderline bradycardia. On physical exam the patient was a thin appearing male elderly male
- A echocardiogram revealed a normal LV with a EF of 55-60%, a moderately dilated RV with moderately depressed RV systolic function with the RV appearing more dilated toward the apex with significant trabeculation suggestive of ARVC.
- The EKG also demonstrated criteria for ARVC specifically the Inverted T waves in V1-V2, Slurred S upstroke seen and the presence of a initial ventricular arrythmia
- Clinically there was a suspicion for ARVC however other causes could not be ruled out. Uhl's anomaly, Brugada syndrome, Sarcoidosis, ischemic cardiomyopathy was considered
- The MRI showed a indexed end diastolic volume of 129 mL/sq m and RV EF of 40%. Also seen was right ventricular hypokinesia which was most prominent at the free wall towards the base and it had right ventricular wall thinning and hyper trabeculation. There also appeared to be delayed hyperenhancement of the free wall towards the base suggesting fibrosis.
- A Left heart catheterization was then performed and came back with no evidence of blockages. It was then decided that the patient would benefit from a dual chamber ICD was placed for secondary prevention. Patient was discharged with follow up in place.



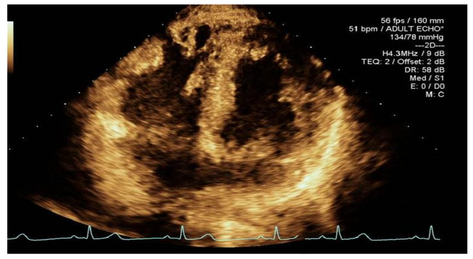
The RV is much larger (most anterior structure) than the LV. This is abnormal. Also visible are numerous trabeculations are suspicious for ARVC. The RV free wall near the apex is also quite thin



RV on the left-hand side is, again, enlarged and shows trabeculations in the apex.



Ventricular Tachycardia



Enlarged RV free wall and trabeculations

DISCUSSION

- Diagnostic criteria include major and minor criteria. For diagnosis you need 2 major or 1 major and 2 minor, or 4 criteria from different categories.
- The categories were Echo, MRI, RV angiography, Tissue characterization of the wall, EKG findings, Arrythmias and Family history.

What is the utility of being aware of this condition?

- It is an important cause of sudden cardiac death in young people and athletes, commonly presents during exercise(8) and has a strong familial component.
- Patients often display dizziness, palpitations, and syncope but can have atypical chest pain as well so its important to include it in a differential. Also important to realize up to 40% of patients are asymptomatic.
- The ventricular arrythmias that this condition can present with can often be confused with Brugada syndrome, Dilated cardiomyopathy with RV involvement and RVOT tachycardia

How is ARVC diagnosed? What interventions are available? How are these patients screened and what surveillance is recommended?

- Diagnosis is challenging as there is no one test that definitively diagnoses ARVC. A EKG may show Inverted T waves in the right precordial leads V1 to V3, epsilon waves(reproducible low amplitude signals between end of QRS complex to onset of T wave) in the right precordial leads V1 to V3 (1 and 2).
- Echocardiography may show a hypokinetic RV, a very thin RV free wall, or an enlarged RV.
- Another option is a cardiac MRI which may show regional RV akinesis, dyskinesia and can show a transmural diffuse bright signal in the RV wall suggesting myocardial thinning.
- Finally another option is a endomyocardial biopsy which can show fibrous replacement of the RV free wall however its sensitivity is low.
- For ARVC management the goal is to prevent sudden cardiac death and decrease the chance that it could occur.
- Pharmacologically the patient could receive a beta blocker for arrythmia suppression and Sotalol has been found most beneficial, it is also recommended the patient receive Warfarin as anticoagulation to prevent thrombus formation due to a decreased ejection fraction and dyskinesis(reference 9).
- Another more invasive option is radiofrequency catheter ablation which is used to treat refractory or continued ventricular tachycardia (10)
- A ICD is the most effective at preventing sudden cardiac death and is often called for in this condition.
- Cardiac transplant surgery can also be considered if the arrythmia is uncontrollable with the above interventions or failure to manage ventricular failure (11).
- All first degree relatives of a patient with ARVC should be screened. Screening would include a echocardiogram, EKG, a cardiac MRI and exercise stress test
- Patients with ARVC need lifelong clinical follow up to periodically evaluate new onset or worsening symptoms, progression of functional/morphological ventricular abnormalities and ventricular arrhythmias. Surveillance should include a resting 12 lead EKG, echocardiogram, 4 hour Holter monitoring and a exercise stress test every 1-2 years depending on age, symptoms and disease severity.

CONCLUSION

- This case was interesting due to the presenting symptoms with no cardiac history and no clear family history known by the patient.
- ARVC often presents at a younger age and it was abnormal to see it present in a older individual who hadn't had any prior cardiac workup for ARVC.
- Cases of ARVC have a strong familial component so screening of family members will be important.
- Physicians should keep ARVC on the differential especially in patients with a family history of SCD and new ventricular arrythmias with no prior documented history of cardiac issues.
- If suspected the patient should receive a workup including EKG, echocardiogram and possibly a cardiac MRI depending on the clinical picture.

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