Biventricular arrhythmogenic dysplasia: How CMR fills the diagnostic void of the Task Force Criteria

Kirtivardhan Vashistha, Jian Hu, Mahathi Indaram, Jayshiv Badlani, Sarumathi Thangavel, Abhas Khurana, Kartikeya Kashyap, Robert Biederman, Victor Farah, Vinh Nguyen

[1] Department of Medicine; [2]Division of **Cardiovascular Medicine**, Allegheny Health Network, Pittsburgh, PA

Introduction

- ARVD/C is a genetic heart condition usually seen in young men characterized by rhythm abnormalities due to fibrofatty replacement of myocardium.
- Reports have shown left ventricle (LV) involvement as the disease progresses but its prevalence still remains low.
- Existing diagnostic criteria include considerations of family history, EKG changes, echocardiogram or MRI findings, as well as histologic tissue characterization of the myocardium.
- Current criteria does not include MRI tissue characterization.

Case Presentation

- A 20 year-old male athlete with a history of eosinophilic esophagitis presented with palpitations.
- EKG showed RBBB pattern. TTE showed RV hypertrophy and dilation. Cardiac MRI (CMR) was pursued for morphologic and tissue characterization of the RV. Make font as big as possible but no smaller that 36pt.
- CMR showed a dysmorphic RV with right to left septal concavity deformation, free wall akinesis with microaneurysmal formation, RV end diastolic volume >110 ml/m2, and RVEF <40%.
- Late gadolinium enhancement (LGE) showed biventricular enhancement involving the entire RV free wall as well as the subepicardium of LV apical cap with basal extension to mid ventricular septum.
- Microaneurysm was also present at the LV apex.

Discussion

- Per 2010 Revised Task Force Criteria, the diagnosis is based on structural, functional and electrophysiological abnormalities which reflect histological changes[1].
- CMR can be used to meet structural criteria, based on RV shape, size, and function but tissue characterization is not included[2].

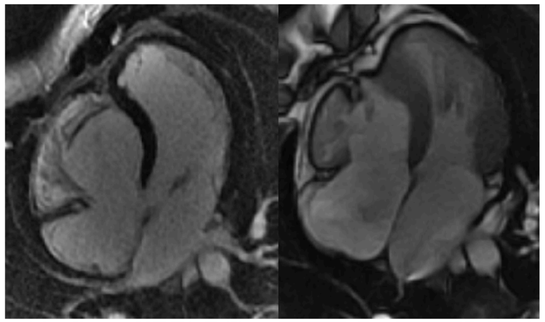

Fig a) RV Late gadolinium enhancement
b) RV micro aneurysm
c) LV late gadolinium enhancement
d) LV micro aneurysm

- Although our patient met the CMR ARVD criteria for structural alterations, it is the presence of the biventricular LV LGE pattern that is atypical for arrhythmogenic LV cardiomyopathy1,3.
- With growing focus on the disease due to advances in tissue characterization by cardiac imaging, the condition has broadened to include left dominant and biventricular subtypes2.
- The existing criteria synthesized 10 years ago lacks consideration of noninvasive tissue findings described in our case, which contributes to it's under recognition1.
- Further revision of the task criteria to include LGE and updated CMR sequences such as T1/T2 mapping and fat suppression sequences may provide diagnostic guidance to fulfill nonimaging criteria to establish the diagnosis.

Conclusion

- The 2010 Task Force Criteria does not recognize the robust tissue characterization of cardiac MRI, leading to under recognition of the disease.
- We advocate for an emphasis on CMR tissue characterization and/or revision to the outdated Task Force Criteria.

References

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