Scholars Journal of Medical Case Reports

Abbreviated Key Title: Sch J Med Case Rep ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online) Journal homepage: https://saspublishers.com/journal/sjmcr/home

Electric Findings of the Left Dominant Arrhythmogenic Dysplasia: Features Need to Be Considered

Soumia Saidi^{1*}, Amel Halfaoui², Karima Bentahar¹, Nashwan Ghanem²

¹Department of Cardiology B, Ibn Sina Hospital, University Mohamed V, Rabat, Morocco ²Department of Cardiology Simone Veil Hospital, Eaubonne, France

*Corresponding author: Saidi Soumia DOI: 10.36347/sjmcr.2019.v07i06.002 | Received: 05.06.2019 | Accepted: 15.06.2019 | Published: 30.06.2019

Abstract Case Report

The combined RV / LV involvement in the arrhythmogenic ventricular dysplasia is very rare but it is linked to a darker prognosis when dominant in the left ventricle. We report a case of a 42 -year-old male diagnosed with Left dominant dysplasia. The purpose of our case report is to shed light on the electrical aspects of left dominant dysplasia. **Keywords:** Left Dominant, arrhythmogenic, Dysplasia, Electrocardiogram.

Copyright @ 2019: This is an open-access article distributed under the terms of the Creative Commons Attribution license which permits unrestricted use, distribution, and reproduction in any medium for non-commercial use (NonCommercial, or CC-BY-NC) provided the original author and source are credited.

INTRODUCTION

The combined RV / LV involvement in the arrhythmogenic ventricular dysplasia is very rare but it is linked to a darker prognosis when dominant in the left ventricle, hence the interest of implantation of defibrillator in primary prevention in these patients.

CASE REPORT

A 42-year-old athlete without cardiovascular risk factors, personal or familial history of cardiac disease, he was admitted to the Emergency Department (ED) for syncope with head trauma during exercise. The Clinical examination was normal .The 12-lead electrocardiogram showed sinus rhythm with negative T waves in precordial V4-V5 and inferior leads, QRS fragmentation in aVL and epsilon waves in II, III, and aVF. On telemetry there were many polymorphic ESVs, with couplets.



Fig-1: The 12-lead electrocardiogram showed sinus rhythm withT-wave inversion in inferolateral leads and none demonstrated T-wave inversion in V1-V3

The complementary tests excluded any other probable diagnosis of syncope with a negative Tilt test, a normal brain CT scan, and a normal basal routine investigations. A coronary angiography was performed, that does not show any obstruction or stenosis

The echocardiography showed global hypokinesia of the left ventricle with a dilated and hyperkinetic right ventricle. The Magnetic resonance imaging showed that showed a LV global hypokinesia with an apical dyskinesia, systolic dysfunction of LV of 41%; a very dilated RV (RVTD at 180ml / m2), and RVEF at 31% with free wall extensive dyskinesia. The late enhancement was extensive, affecting the lateral, septal posterior wall and the apex without obvious enhancement of the RV.



Fig-2: Cardiac magnetic resonance imaging of the patient reveals late gadolinium enhancement (left to right: 4 chamber, 2 chamber and midventricular view), mainly within the epicardial and midmyocardial layers of the left ventricle (LV), compatible with fibrofatty infiltration involving primarily the apex, lateral, septal posterior wall

The patient was treated with beta blockers with single-chamber cardioverter-defibrillator implantation.

DISCUSSION

Left-dominant arrhythmogenic cardiomyopathy or isolated arrhythmogenic LV cardiomyopathy is now a recognized entity, however, its diagnosis criteria still remains unclear in comparaison with the right sided dysplasia [1]. The purpose of our case report is to shed light on the electrical aspects of left dominant dysplasia.

Our reported patient had a negative T waves in inferior and lateral leads with epsilon waves in II, III, and aVF; suggesting a prevalent LV disease and an absent of RV involvement

However our review of the literature found a few publications that has reported similar aspects but not yet well indentified, the negative T waves in inferior and lateral leads were described in almost all reported cases [2-8]. The other two criteria: epsilon wave in inferior leads and the QRS fragmentation in a VL were reported only once. Carlos Galva *et al.* [5]. has reported the epsilon wave in inferior leads when the LV is involved , and Ardan M. *et al.* [6]. Has reported the QRS fragmentation in a VL.

The diagnosis of arrhythmogenic LV cardiomyopathy poses a great challenge and these findings remain unclear and not yet recognized by any consensus or established criteria for this rare entity which is probably underestimated.



Fig-3: Epsilon waves in II, III, and aVF (red arrows, Panel A and B) and QRS fragmentation in aVL(red arrows, Panel C)

CONCLUSION

Arrhythmogenic right ventricular dysplasia is an important cause of sudden cardiac death in young adults, but the prognosis gets worse with left ventricle involvement or dominance , which requires a well defined diagnostic criteria of this rare entity.

Figures

Figure 1: The 12-lead electrocardiogram showed sinus rhythm withT-wave inversion in inferolateral leads and and none demonstrated T-wave inversion in V1-V3

Figure 2: epsilon waves in II, III, and aVF (red arrows, Panel A and B) and QRS fragmentation in aVL(red arrows, Panel C)

Figure 3: Cardiac magnetic resonance imaging of the patient reveals late gadolinium enhancement (left to right: 4 chamber , 2 chamber and midventricular view), mainly within the epicardial and midmyocardial layers of the left ventricle (LV), compatible with fibrofatty infiltration involving primarily the apex , lateral, septal posterior wall.

REFERENCES

- 1. Jacoby D, McKenna WJ. Genetics of inherited cardiomyopathy. European heart journal. 2011 Aug 2;33(3):296-304.
- Navarro-Manchón J, Fernández E, Igual B, Asimaki A, Syrris P, Osca J, Salvador A, Zorio E. Left dominant arrhythmogenic cardiomyopathy caused by a novel nonsense mutation in desmoplakin. Revista Española de Cardiología (English Edition). 2011 Jun 1;64(6):530-4.
- Bazoukis G, Letsas KP, Xia Y, Tse G, Li KH. A novel desmin mutation causing severe left ventricular arrhythmogenic cardiomyopathy/dysplasia. Journal of thoracic disease. 2018 Sep;10(Suppl 26):S3100.
- Smaldone C, Pieroni M, Pelargonio G, Dello Russo A, Palmieri V, Bianco M, Gentile M, Crea F, Bellocci F, Zeppilli P. Left-dominant arrhythmogenic cardiomyopathy. Circulation: Arrhythmia and Electrophysiology. 2011 Aug;4(4):e29-32.
- Galvão Braga C, Silva P, Salgado A, Magalhães S, Themudo R. Isolated left ventricular arrhythmogenic dysplasia. European Heart Journal–Cardiovascular Imaging. 2014 Feb 20;15(8):907-.
- Saguner AM, Buchmann B, Wyler D, Manka R, Gotschy A, Medeiros-Domingo A, Brunckhorst C, Duru F, Mayer KA. Arrhythmogenic left ventricular cardiomyopathy: suspected by cardiac magnetic resonance imaging, confirmed by identification of a novel plakophilin-2 variant. Circulation. 2015 Aug 11;132(6):e38-40.
- 7. Ionescu AA, Chivulescu M, Nicula A, Popescu BA, Ginghina C, Jurcut R. A case of

arrhythmogenic cardiomyopathy–not only a right ventricular disease. Romanian Journal of Cardiology. 2016 Dec 1;26(4).

 Miles C, Finocchiaro G, Papadakis M, Gray B, Westaby J, Ensam B, Basu J, Parry-Williams G, Papatheodorou E, Paterson C, Malhotra A. Sudden Death and Left Ventricular Involvement in Arrhythmogenic Cardiomyopathy. Circulation. 2019 Jan 31.

© 2019 Scholars Journal of Medical Case Reports | Published by SAS Publishers, India