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RESEARCH ARTICLE

A RARE CASE OF ARRHYTHMOGENIC RIGHT VENTRICULAR DYSPLASIA PRESENTED WITHOUT ARRHYTHMIA AS CONGESTIVE CARDIAC FAILURE

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ABSTRACT

It is a case of ARVD in young patient diagnosed with classical signs and symptoms correlated with ECG and 2D Echo findings, treated accordingly. ARVD can be found in association with diffuse palmoplantar keratoderma, and woolly hair, in an autosomal recessive condition called Naxos disease, because this genetic abnormality can also affect the integrity of the superficial layers of the skin most exposed to pressure stress.

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INTRODUCTION

Arrhythmogenic right ventricular dysplasia (ARVD), or arrhythmogenic right ventricular cardiomyopathy (ARVC), is an inherited heart disease. ARVD is caused by genetic defects of desmosomes. The desmosomes are composed of several proteins, and many of those proteins can have harmful mutations. The disease is a type of nonischemic cardiomyopathy that involves primarily the right ventricle. It is characterized by hypokinetic areas involving the free wall of the right ventricle, with fibro fatty replacement of the right ventricular myocardium, with associated arrhythmias originating in the right ventricle.

CASE HISTORY

A 16 year old boy presented with chief complains of bilateral pitting oedema of pitting type and difficulty in breathing on exersion since last 4 days. There is past history of similar complains twice in last six months for which he has taken some medications from local doctor and symptoms were relieved. No any other past history of any disease, no any habits.

Clinical Examination

On general examination,tempreture –normal,pulse-88/min Blood pressure-96/58 mm of Hg

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Oedema- present

Respiratory system-bilateral fine crepitation present Cardiovascular system- grade 2-3 cardiac systolic murmur auscultated in tricuspid area CNS-patient is conscious, oriented P/A soft non-tender.

Investigations

CBC was normal. S. electrolytes were also unaltered. S. creat was 0.78. RBS – 88 mg% SGPT – 33 U/L. Lipid profile was also unaltered.

Total protiens were 4.2gm%
S. Albumin 1.8mg%.
S. Globulin 2.4mg%
Ratio being 1:1.43.

Urine Routine showed presence of protein in trace amount. On further investigations,

PT was 19.8sec and INR 1.38 some medications from local doctor and Symptoms were

ECG: Epsilon wave in lead V1-V3 with Left Bundle Branch relieved. No any other past history of any disease or no any habits.

ECG: Epsilon wave in lead V1-V3 with Left Bundle Branch Block, Left Axis Deviation.

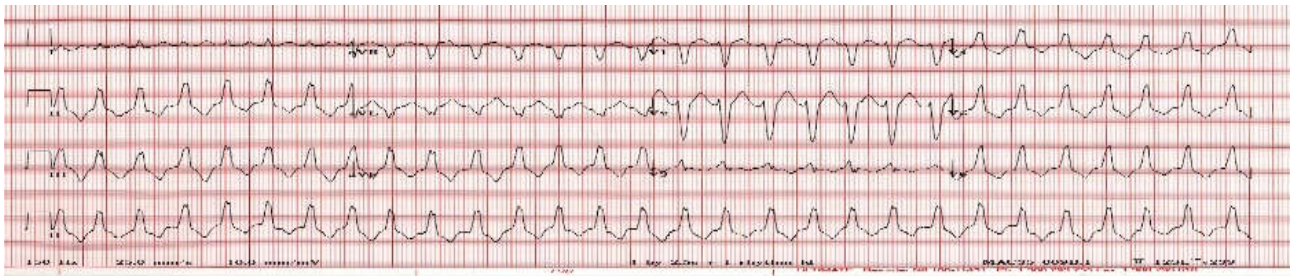

Figure 1. Electrocardiogram of the patient

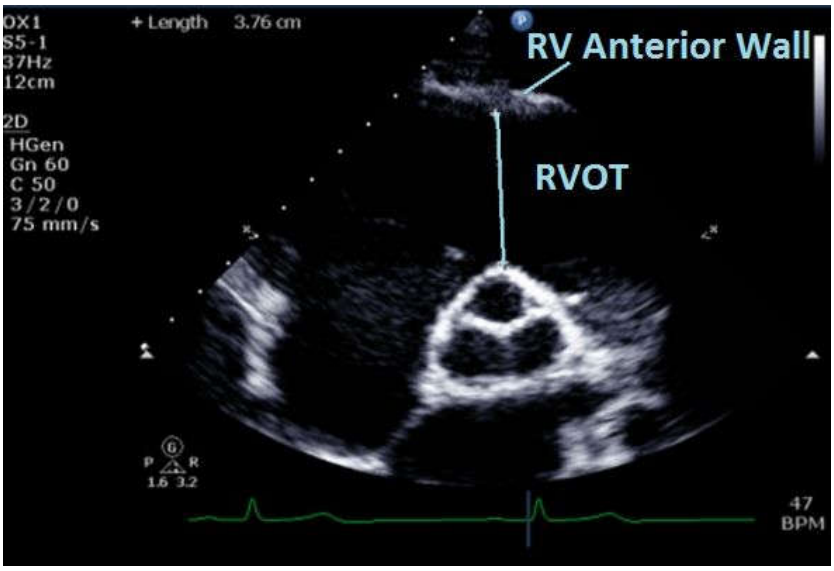

Figure 2. Echo cardiogram of the patient

2D-Echo shows global LV hypokinesia, Biventricular dysfunction, LVEF=30%, Right Ventricular Outflow Tract obstrucytion with severe low pressure TR. IVC collapsing.

Diagnosis: Arrhythmogenic right ventricular dysplasia presented without arrhythmia as congestive cardiac failure.

Treatment

For treating CCF, he was treated with injectable as well as oral diuretics (Furesemide and Spironolactones)

Beta-Blocker and Ramipril were also give n for cardiac re-modelling. Protien suppliments were given.

DISCUSSION

ARVD can be found in association with diffuse palmoplantar keratoderma, and woolly hair, in an autosomal recessive condition called Naxos disease, because this genetic abnormality can also affect the integrity of the superficial layers of the skin most exposed to pressure stress.(1): 513(2) ARVC/D is an important cause of ventricular arrhythmias in children and young adults. It is seen predominantly in males, and 30–50% of cases have a familial distribution. An Intra-cardiac Defibrillator (ICD) is the most effective prevention against sudden cardiac death. Due to the prohibitive cost of ICDs, they are not routinely placed in all individuals with ARVD.

Conclusion

ARVD is arare cause of sudden cardiac death in young patients due to recurrent arrythmia. ARVD may present as a condition without arrythmia and may present as CCF.

REFERENCES

Cox MGPJ, van der Zwaag PA., van der Werf C. *et al.* 2011. "Arrhythmogenic right ventricular dysplasia/ cardiomyopathy" Circulation. 123:2690–700. doi:10.1161/ circulationaha. 110.988287.

Elston, Dirk, 2005. Andrews' Diseases of the Skin: Clinical Dermatology. (10th ed.). Saunders. ISBN 0-7216-2921-0.

Freedberg, *et al.* 2003. Fitzpatrick's Dermatology in General Medicine. (6th ed.).

Fressart V., Guthoit G., Donal E. et al. 2010. "Desmosomal gene analysis in arrhythmogenic right ventricular dysplasia/ cardiomyopathy: spectrum of mutations and clinical impact in practice". Europace. 12: 861–8. doi:10.1093/ europace/euq10.

McGraw-Hill. ISBN 0-07-138076-0.James, William; Berger, Timothy;

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