Case report

Isolated left ventricular non-compaction in association with ventricular tachycardia

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KEY WORDS

Cardiomyopathy
Non-compaction of left ventricle
Ventricular tachycardia

A B S T R A C T

A 32-year-old young male was found to have non-sustained, repetitive, monomorphic ventricular tachycardia of right bundle branch morphology during routine pre-anaesthetic evaluation for orthopaedic surgery. Echocardiography and left ventricular angiogram were suggestive of isolated non-compaction of left ventricular apex with systolic dysfunction. He was successfully managed with anti-arrhythmic drugs and had an uneventful 9-month follow-up. The index case is an unusual association of asymptomatic, non-sustained ventricular tachycardia with isolated ventricular non-compaction.

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Introduction

Isolated ventricular non-compaction (IVNC) is a rare cardiomyopathy characterised by the loss of compaction of myocardial fibre meshwork during intrauterine life. This is possibly due to an arrest of intrauterine compaction of the myocardial fibres in the absence of any other structural heart disease.¹ It is characterised by the presence of deep intertrabecular recesses in the hypertrophied, frequently hypokinetic segment of ventricular myocardium.^{2,3} The clinical manifestation includes triad of ventricular systolic dysfunction, thromboembolism and malignant tachy-arrhythmias.⁴ We hereby report a case of IVNC of left ventricle (LV) presented with non-sustained ventricular tachycardia (VT), which was effectively treated with anti-arrhythmic drugs.

Case report

A 32-year-old male, manual labourer by occupation was referred to our outpatient clinic in July 2009, for his electrocardiogram (ECG) evaluation prior to right knee joint

arthroscopy for osteo-chondritis. He did not have any cardiac symptoms like palpitation, dyspnoea, angina, or syncope. He was non-smoker, normotensive, and non-diabetic. There was no family history of sudden cardiac death in any of the family members. General physical examination revealed irregular, normal volume radial pulse. Rest of his general and systemic examination was within normal limits. The ECG revealed repetitive runs of non-sustained, monomorphic VT of right bundle branch block (RBBB) morphology (Figures 1A and B). Echocardiogram features of LV apical IVNC as characterised by hyper-trabeculation and inter-trabecular recesses and >2:1 ratio of non-compacted to compacted layer of left ventricular wall,³ was present as evident in Figure 2. There was no clot at left ventricular apex. Coronary angiography revealed normal epicardial coronaries. Left ventriculogram revealed spongi-form appearance of apex and diaphragmatic surface (Figure 3) suggestive of localised non-compaction of LV as described by us in our previous publication.⁵ The LV ejection fraction was 30%. The pulmonary artery pressure was 25/10 mmHg (mean 15 mmHg). A diagnosis of IVNC-LV type with systolic dysfunction and haemodynamically stable non-sustained, monomorphic VT of LV origin was made. His tachycardia could be controlled with amiodarone 400mg twice a day tablets (Figure 1C). There were no runs of VT on 24-hour Holter monitoring on day 10 of admission. He was discharged on tablets—amiodarone 200mg, ramipril 5mg, and aspirin

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Figure 1 (A, B) Electrocardiogram showing repetitive runs of non-sustained ventricular tachycardia of right bundle branch block morphology. (C) Tachycardia controlled with the administration of amiodarone and sotalol.

150mg per day. An implantable cardioverter defibrillator (ICD) could not be implanted because of financial constraint. He is doing well and had an uneventful 9-month follow-up. The ECG did not show any runs of VT or ventricular premature complexes, echocardiogram showed the unchanged LV ejection fraction of 30%.

Discussion

Out of the clinical triad of IVNC,⁴ the index case had two manifestations in the form of LV systolic dysfunction and

non-sustained VT. It is uncommon to encounter IVNC as an underlying structural abnormality in a patient with VT.⁶ Nevertheless, ventricular arrhythmia is the frequent complication in IVNC patients on long-term follow-up.^{7,8} The incidence is variably reported as 4.6% in 238 IVNC patients at 6 months of follow-up by Fazio Ge et al⁷ and 41% in 34 IVNC patients at 44 months of follow-up by Oechslin et al.⁸ The precise mechanism of ventricular arrhythmia is not clear. A relatively decreased perfusion and ischaemia-related fibrosis at sub-endocardial non-compacted area has been postulated as the cause of electrical inhomogeneity and micro-reentry resulting into ventricular tachycardia.^{6,9} Steffel et al. have

Figure 2 Echocardiogram in modified apical four chamber view with anterior tilt shows non-compaction of left ventricular apex. LV: left ventricle, RA: right atrial, RV: right ventricle.

Figure 3 Left ventricular angiogram in right anterior oblique 30° showing the spongiform appearance of apex and diaphragmatic surface.

tried to risk stratify these cases by VT induction during electro-physiological studies.¹⁰ The medical treatment of VT includes beta-blockers and anti-arrhythmic drugs like amiodarone, which was effective in the index case. There are reports of ICD⁸/cardiac resynchronisation therapy implantation,¹¹ radiofrequency ablation,⁶ and even cardiac transplantation^{8,12} in IVNC cases with malignant ventricular tachy-arrhythmias. The long-term prognosis of these patients is variable depending on the New York Heart Association (NYHA) functional class, LV systolic dysfunction, presence of malignant ventricular arrhythmias, and cerebral thromboembolism.^{4,10}

Nevertheless, our case had an uneventful 9-month follow-up on medical treatment only.

In conclusion, IVNC may be the underlying cause for ventricular tachycardia in a young individual, evaluated for idiopathic VT. A careful echocardiographic evaluation can diagnose the underlying unclassified cardiomyopathy. As the long-term prognosis is variable depending upon the NYHA functional class, LV systolic dysfunction, and associated arrhythmias, the management should be individualised depending upon the risk stratification.

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