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LETTER TO THE EDITOR
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A case report with the progression from atrial fibrillation to complete AV block, heart failure and electrical storm

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When patients initially present with atrial fibrillation along with an enlarged heart and heart failure, followed by atrioventricular block, it's essential to consider genetic factors.[1] Genetic testing can offer crucial diagnostic evidence, aiding in prognosis assessment and the adoption of appropriate treatment strategies.

A 61-year-old male patient was admitted to our hospital on December 18, 2023, complaining of "sudden palpitations over the past 12 years, worsened by chest tightness for the last 9 days". In 2011, he sought treatment at our hospital for sudden palpitations accompanied by left limb weakness, and was diagnosed with "atrial flutter" during an outpatient visit. He was also diagnosed with "cerebral infarction" and was prescribed oral warfarin for anticoagulation and antiarrhythmic drugs to control fast ventricular rate. By August 2020, his cardiac symptoms had worsened, with fatigue and decreased endurance during physical activity. He experienced shortness of breath after walking just 600 meters. An electrocardiogram (ECG) performed at our hospital revealed atrial fibrillation, complete atrioventricular block, and an escape rhythm with a widened QRS complex (Figure 1). Transthoracic echocardiography (TTE) showed overall enlargement of the heart, widening of the pulmonary artery, severely reduced left ventricular function with an estimated ejection fraction of 30%, moderate to severe leakage of the tricuspid valve, mild to moderate leakage of the pulmonary valve, and moderately elevated pressure in the pulmonary artery. On August 19, 2019, he underwent permanent implantation of an dual chamber implantable cardioverter defibrillator (ICD) (Medtronic DDBC3D4). An ventricular electrode was placed in the region of the left bundle branch, with significant fibrosis observed during implantation, and a defibrillator electrode was placed in the apex of the right ventricle.

Postoperative ECG showed a QRS duration of 168 ms, which was 10 ms shorter than before the procedure (Figure 1). The patient had a history of hypertension, chronic hepatitis B infection, and previous cerebral infarction. Following the procedure, he adhered to his prescribed medications and remained free from palpitations and chest tightness for about a year and a half. In March 2022, the patient experienced an episode of ventricular tachycardia, which was resolved with two sequential attempts of anti-tachycardia pacing (ATP) followed by a shock treatment. He subsequently experienced intermittent palpitations and discharges from the ICD, prompting hospitalization to adjust his medication regimen and reprogram the ICD parameters. In May 2022, due to the discovery of ventricular tachycardia due to ICD, amiodarone was added to the patient. On November 9, 2023, at 8:00 PM, he developed chest tightness and received a discharge from the ICD, leading to his admission for further evaluation and management. Due to a significant prolongation of the QT interval and suspicion of torsades de pointes (TdP), amiodarone was immediately discontinued. On December 12, 2023, at 8: 00 PM, he experienced three consecutive ICD discharges, with intervals of 3–5 min. His self-reported heart rate during this episode was 200 beats per minute. Ventricular tachycardia at a frequency of 120-140 beats per minute was subsequently observed on daily ECG monitoring. Another episode of tachycardia occurred on December 18, 2023, at 8:00 PM, with the ECG indicating ventricular tachycardia at a heart rate of 120-140 beats/min. Upon admission, the ECG showed atrial fibrillation, a ventricular pacing rhythm with a QRS duration of 210 ms, and ventricular premature beats (Figure 1). No significant changes were

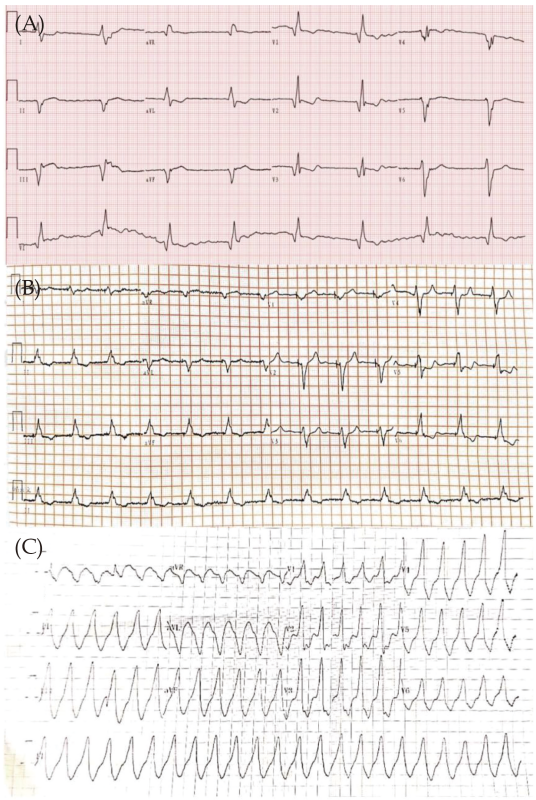

Figure 1 Electrocardiogram before and after ICD implantation. (A): Pre-ICD implantation ECG showing atrial fibrillation with third-degree atrioventricular block. The ventricular rhythm exhibits right bundle branch block characteristics, with a QRS duration of 178 ms. (B): Post-ICD implantation ECG, with the atrial lead placed in the inflow tract of the interventricular septum. The paced QRS duration is 168 ms, reflecting a 10 ms reduction compared to the preoperative measurement. (C): ECG during a ventricular tachycardia episode, showing a ventricular rate of 157 beats/min.

noted on cardiac ultrasound. Genetic testing revealed a variation in the LMNA gene (Figure 2). The patient's family history revealed that his elder sister exhibited similar clinical manifestations and underwent permanent pacemaker implantation due to heart enlargement and complete atrioventricular block. The primary admission diagnoses included arrhythmias (atrial fibrillation, complete atrioventricular block, ventricular tachycardia), sympathetic electrical storm following ICD implantation, dilated cardiomyopathy with Grade III cardiac function (NYHA scale), unstable angina due to coronary heart disease, very high-risk hypertension, and a history of previous cerebral infarction. The patient received intensive treatment for heart failure, continued antiplatelet therapy, anticoagulation, statin therapy, and other treatments for coronary heart disease, as well as anti-

Sample	Detection of genes	Position	Variability situation	Variant type
The patient	LMNA	chr1: 156106717	c.1386_1390del	heterozygosis

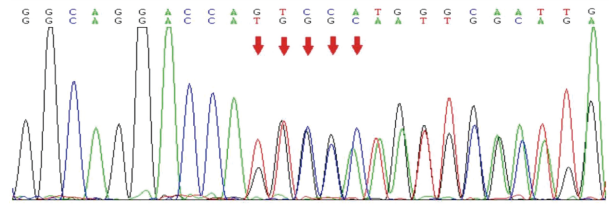

Figure 2 Gene second-generation sequencing report. A deletion in the LMNA gene, located at position c.1386_1390, is indicated by the red arrow in the figure.

symptomatic excitability management including antianxiety, sedation, and Antagonistic sympathetic drugs. During follow-up, the patient continued to experience intermittent palpitations but did not require further ICD shock therapy.

The patient was diagnosed with atrial flutter, atrial fibrillation, and stroke despite having a low stroke risk score, indicating significant left atrial fibrosis. Unfortunately, there was no reliable evaluation method available at that time. Subsequent implantation of an incompatible MRI ICD failed to provide improvement in cardiac magnetic examination. However, based on the observed difficulty in multiple insertions of the 3830 electrode and poor electrode parameters during ICD implantation,[1] it can be inferred that there is evident ventricular fibrosis. As the disease progressed, the entire heart gradually enlarged, left ventricular ejection fraction decreased, and the patient developed third-degree atrioventricular block. Considering the similar situation in the patient's sister, a possibility of gene mutation was considered. Genetic testing was conducted to identify mutations in the LMNA gene.[2]

Meanwhile, besides LMNA cardiomyopathy, the association of atrioventricular block and ventricular arrhythmias should raise suspicion of myocarditis or cardiac amyloidosis. Myocarditis can present as trioventricular block and ventricular arrhythmias, but its course is usually acute or subacute, with elevated troponin, diffuse or subepicardial patterns of delayed gadolinium enhancement (LGE) in cardiac magnetic resonance (CMR), and precursors to viral infection. In this patient, there were no elevated markers of acute inflammation, other tests did not show signs of active myocarditis. Cardiac amyloidosis, especially transthyroxin protein amyloidosis (ATTR-CM), which can manifest as atrioventricular block, ventricular arrhythmias and heart failure.[3] However, typical signs such as left ventricular wall

thickening with low-voltage ECG, and longitudinal strain "apical retention" pattern were not observed in this patient. In addition, the ventricular arrhythmias of amyloidosis was more associated with atrial enlargement or microcirculation ischemia than with deep septal fibrosis.[4] Studies indicate that among genetic causes of DCM, LMNA mutations are the second most common and follow an autosomal dominant inheritance pattern.[5] LMNA mutations account for 33% of cases in patients with both DCM and conduction disorders, with approximately 85% of affected individuals exhibiting cardiac involvement.[6] The penetrance of LMNA-associated DCM is high, with nearly all patients over 60 years old developing cardiac manifestations.[7] Myocardial dysfunction and arrhythmias resulting from LMNA mutations are collectively termed LMNA cardiomyopathy. Its primary manifestations include DCM and conduction abnormalities, which may present independently or in combination. LMNA cardiomyopathy can appear as isolated ventricular dilation, arrhythmogenic cardiomyopathy, reduced systolic function without ventricular dilation, or classic DCM.[6,8] Compared to other forms of non-ischemic DCM, LMNA cardiomyopathy progresses more rapidly, often without significant ventricular chamber enlargement or wall thinning, and is associated with a higher incidence of malignant arrhythmias and the need for heart transplantation. In this patient, cardiac dysfunction progressed slowly, whereas arrhythmias worsened rapidly, with evident bundle branch block and recurrent ventricular tachyarrhythmic storms. Left bundle branch pacing initially improved QRS duration following ICD implantation, but over time, QRS duration widened, and QT interval prolongation ensued with bundle branch involvement. Although ICD upgrade to CRT-D was considered,[9] the patient exhibited a QRS duration of 210 ms with bundle branch block, and given the potential for non-response, optimization of pharmacological therapy was prioritized.

The patient presented with permanent atrial fibrillation, heart failure, and intermittent episodes of ventricular tachycardia, meeting the indications for ICD implantation. Given the patient's high pacing needs, the defibrillation electrode was placed at the right ventricular apex to optimize defibrillation efficiency. However, pacing at this site could potentially disrupt cardiac synchronization, so a two-chamber pacemaker was chosen. The other ventricular electrode was positioned in the left bundle branch region to improve synchronization of ventricular pacing. It is crucial to deactivate the automatic mode switch when setting the tachycardia detection function, as this prevents the misinterpretation of ventricular tachycardia as delayed supraventricular tachycardia, which could result in unnecessary discharges from both atrial and ventricular interfaces. The patient experienced pre-syncope symptoms without discharge, and the ICD mistakenly identified supraventricular tachycardia, leading to delayed discharge. Additionally, most slow ventricular tachycardia episodes occurred at a rate of 120-140 beats/min, and ATP treatment occasionally accelerated the ventricular tachycardia, triggering ICD shocks. The management of slow ventricular tachycardia focused on sedation, reducing sympathetic activity, and using antiarrhythmic medications. However, due to significant QT interval prolongation, amiodarone use was restricted. Propranolol, which blocks sympathetic excitability both peripherally and centrally, provided more effective antiarrhythmic therapy. In cases where slight anxiety or fatigue triggers slow ventricular tachycardia,[10] sympathetic ganglion block could be considered as an optional therapy to reduce sympathetic activity and decrease the frequency of electrical storms.[11] For radiofrequency ablation of ventricular tachycardia, the patient's polymorphic episodes and potential extensive myocardial fibrosis with multiple reentry circuits suggest that complete elimination of ventricular tachycardia may not be achievable.

Several promising drugs are currently under development, including MEK1/2 inhibitors (PD98059 and smeitinib),[12] INK inhibitors (SP600125 and Calbiochem), small molecule inhibitors (ARRY-371797), AKT/mTOR pathway regulator (Sirolimus), calcium sensitizer SCH-00013, and a derivative of diazone. These drugs show potential for mitigating the effects of LMNA gene mutations related to Lamin-altered gene therapy.[13,14] Progress has also been made in antisense oligonucleotide therapies targeting exon 11, which may be effective for LMNA cardiomyopathy caused by exon 11 missense mutations.[15] Gene editing therapy using adenovirus delivery has successfully corrected LMNA gene mutations in multiple organ cells in mice, leading to rejuvenated phenotypes and extended lifespans.[14] Previous studies have shown that heart transplantation not only improves cardiac function but also reduces the occurrence of malignant arrhythmias. Therefore, heart transplantation remains a viable option for eligible patients.

LMNA cardiomyopathy progresses rapidly, often

presenting as DCM with heart failure and arrhythmias such as atrial fibrillation and atrioventricular block. Early optimization of heart failure treatment and ICD implantation can reduce the risk of sudden cardiac death. In cases of recurrent electrical storms, the association of atrial arrhythmias, complete heart block, and heart failure with family history of conduction system disease should raise a suspicion of LMNA cardiomyopathy, indicating genetic testing. Ultimately, heart transplantation may be necessary for some patients.

DISCLOSURE

Conflict of Interests

None.

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REFERENCES

[1] Zeppenfeld K, Tfelt-Hansen J, de Riva M, *et al.* 2022 ESC Guidelines for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death. *Eur Heart J* 2022; 43: 3997–4126.
[2] Topriceanu CC, Alfarih M, Hughes AD, *et al.* The atrial and ventricular myocardial proteome of end-stage lamin heart disease. *Acta Myol* 2023; 42: 43–52.
[3] Buttà C, Zappia L, Laterra G, Roberto M. Diagnostic and prognostic role of electrocardiogram in acute myocarditis: A comprehensive review. *Ann Noninvasive Electrocardiol* 2020; 25: e12726.
[4] Lehtonen J, Uusitalo V, Pöyhönen P, *et al.* Cardiac sarcoidosis: phenotypes, diagnosis, treatment, and prognosis. *Eur Heart J* 2023; 44: 1495–1510.
[5] Lazarte J, Jurgens SJ, Choi SH, *et al.* LMNA variants and risk of adult-onset cardiac disease. *J Am Coll Cardiol* 2022; 80: 50–59.
[6] Hasselberg NE, Haland TF, Saberniak J, *et al.* Lamin A/C cardiomyopathy: young onset, high penetrance, and frequent need for heart transplantation. *Eur Heart J* 2018; 39: 853–860.
[7] Ferradini V, Cosma J, Romeo F, *et al.* Clinical features of LMNA-related cardiomyopathy in 18 patients and characterization of two novel variants. *J Clin Med* 2021; 10: 5075.
[8] Wang Y, Dobreva G. Epigenetics in *LMNA*-Related Cardiomyopathy. *Cells* 2023; 12: 783.
[9] Sidhu K, Castrini AI, Parikh V, *et al.* The response to cardiac resynchronization therapy in LMNA cardiomyopathy. *Eur J Heart Fail* 2022; 24: 685–693.
[10] Savastano S, Baldi E, Compagnoni S, *et al.* Electrical storm treatment by percutaneous stellate ganglion block: the STAR study. *Eur Heart J* 2024; 45: 823–833.
[11] Savastano S, Dusi V, Baldi E, *et al.* Anatomical-based percutaneous left stellate ganglion block in patients with drug-refractory electrical storm and structural heart disease: a single-centre case series. *Europace* 2021; 23: 581–586.
[12] Judge DP, Lakdawala NK, Taylor MRG, *et al.* Long-term efficacy and safety of ARRY-371797 (PF-07265803) in patients with lamin A/C-related dilated cardiomyopathy. *Am J Cardiol* 2022; 183: 93–98.
[13] Sayed N, Liu C, Ameen M, *et al.* Clinical trial in a dish using iPSCs shows lovastatin improves endothelial dysfunction and cellular cross-talk in LMNA cardiomyopathy. *Sci Transl Med* 2020; 12: eaax9276.
[14] Shemer Y, Mekies LN, Ben Jehuda R, *et al.* Investigating *LMNA*-related dilated cardiomyopathy using human induced pluripotent stem cell-derived cardiomyocytes. *Int J Mol Sci* 2021; 22: 7874.
[15] Lee JM, Nobumori C, Tu Y, *et al.* Modulation of LMNA splicing as a strategy to treat prelamin A diseases. *J Clin Invest* 2016; 126: 1592–1602.

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