

Successful Surgical Treatment of an Electrical Storm in a Patient With Atypical Noncompaction Cardiomyopathy

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A 34-year-old male patient with known atypical, predominantly right ventricular noncompaction cardiomyopathy¹ was admitted to our department because of repeated ventricular tachycardias and implantable cardioverter defibrillator shocks (Figure A). An extracorporeal life support system was implanted because of hemodynamic instability, and an electrophysiological study was performed. Epicardial access was achieved by subxiphoid puncture. Endo- and epicardial electro-anatomical voltage mapping (Ensite NavX, St Jude) revealed earliest activation in the area of the noncompaction myocardium (Figure B). Ablation was attempted with radiofrequency (irrigated tip, max 40 Watt) from epi- and endocardial access. However, the patient was still inducible in programmed ventricular stimulation, and during monitoring at the intensive care unit, sustained ventricular arrhythmias occurred. Stepwise, antiarrhythmic treatment including amiodarone, sotalol, and finally ranolazin was used to establish a stable sinus rhythm. However, after >48 hours with stable sinus rhythm, again sustained ventricular tachycardia occurred, and therefore, the patient was allocated to open surgery. Intraoperative epicardial ablation points (see arrows) marked the suspected origin of the arrhythmias (Figure B and C). Accordingly, the arrhythmogenic substrate in the right ventricular apex was completely

removed. Histology and immunohistology showed severe fibrosis in Masson Trichrome stain and infiltration of CD68+ macrophages as arrhythmogenic substrate (Figure D and E). After excision of the noncompaction myocardium, the patient was in stable sinus rhythm. Antiarrhythmic medication was terminated beside amiodarone and β -blocker, and the patient was discharged. Six months after discharge with amiodarone and β -blocker, right ventricular stimulation via the implantable cardioverter defibrillator revealed no ventricular arrhythmia, and thus, antiarrhythmic medication was terminated. The patient was free from heart failure symptoms. Our case emphasizes the importance of an interdisciplinary heart team approach for patients with complex cardiomyopathies.

Disclosures

None.

References

1. Towbin JA, Lorts A, Jefferies JL. Left ventricular non-compaction cardiomyopathy. *Lancet*. 2015;386:813–825. doi: 10.1016/S0140-6736(14)61282-4.

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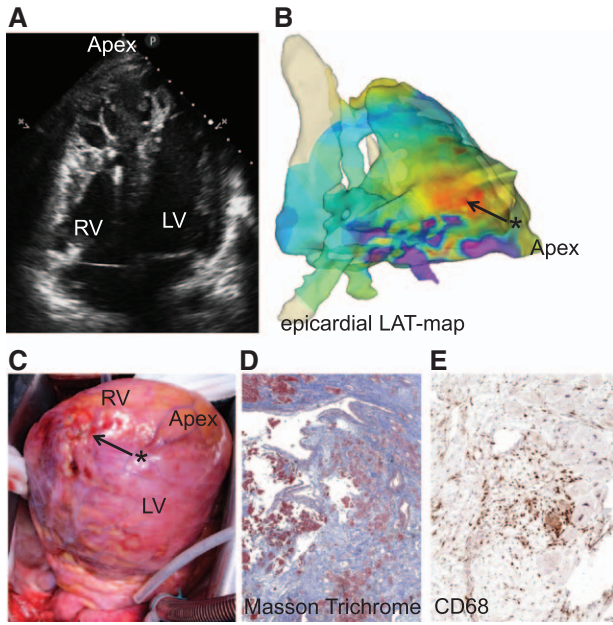


Figure. **A**, Echocardiographic 4-chamber view showing asymmetrical noncompaction cardiomyopathy. **B**, Epicardial electroanatomical map showing local activation time (LAT) during tachycardia. *Ablation site. **C**, Intraoperative situs. *Ablation site. **D** and **E**, Histology and immunohistology of excised myocardium show wide areas of fibrosis (Masson trichrome) and severe infiltration of CD68+ macrophages. LAT indicates local activation time; LV, left ventricle; and RV, right ventricle.

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