Neth Heart J (2020) 28:295–296
https://doi.org/10.1007/s12471-019-01326-8

Non-sustained ventricular tachycardias, conduction disorders and an impaired left ventricular ejection fraction in a 32-year-old woman

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Published online: 28 August 2019

A 32-year-old woman was referred for cardiological evaluation due to palpitations. She did not have a history of syncope. Her family history was negative for sudden death. Electrocardiography showed a sinus rhythm with a first-degree atrioventricular (AV) block (PR interval 310 ms) and premature ventricular contractions (Fig. 1a). A monomorphic non-sustained ventricular tachycardia was seen on Holter recording. Cardiac MRI showed an impaired left ventricular ejection fraction (LVEF) of 41% with mid-myocardial late enhancement consistent with cardiomyopathy (Fig. 1b). DNA analysis revealed a previously reported pathogenic mutation, c.1130G>A p.(Arg377His) in the *LMNA* gene. The cardiac phenotype associated with mutations in the *LMNA* gene typically includes early-onset AV conduction disorders, tachyarrhythmias, dilated cardiomyopathy, in some cases associated with skeletal myopathy [1, 2]. The presence of non-sustained ventricular tachycardias, LVEF <45% at first evaluation, male sex and non-missense mutations (e.g. ins-del/truncating or mutations affecting splicing) are associated with an increased risk of malignant ventricular arrhythmias in *LMNA* mutation carriers [3].

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Conflict of interest S. Alsters, Y. Polyukhovych, H. Bikker, L. Wong and A.C. Houweling declare that they have no competing interests.

References

1. van Rijsingen IA, Nannenberg EA, Arbustini E, et al. Gender-specific differences in major cardiac events and mortality in lamin A/C mutation carriers. Eur J Heart Fail. 2013;15:376–84.
2. Peretto G, Sala S, Benedetti S, et al. Updated clinical overview on cardiac laminopathies: an electrical and mechanical disease. Nucleus. 2018;9:380–91.
3. van Rijsingen IA, Arbustini E, Elliott PM, et al. Risk factors for malignant ventricular arrhythmias in lamin a/c mutation carriers a European cohort study. J Am Coll Cardiol. 2012;59:493–500.

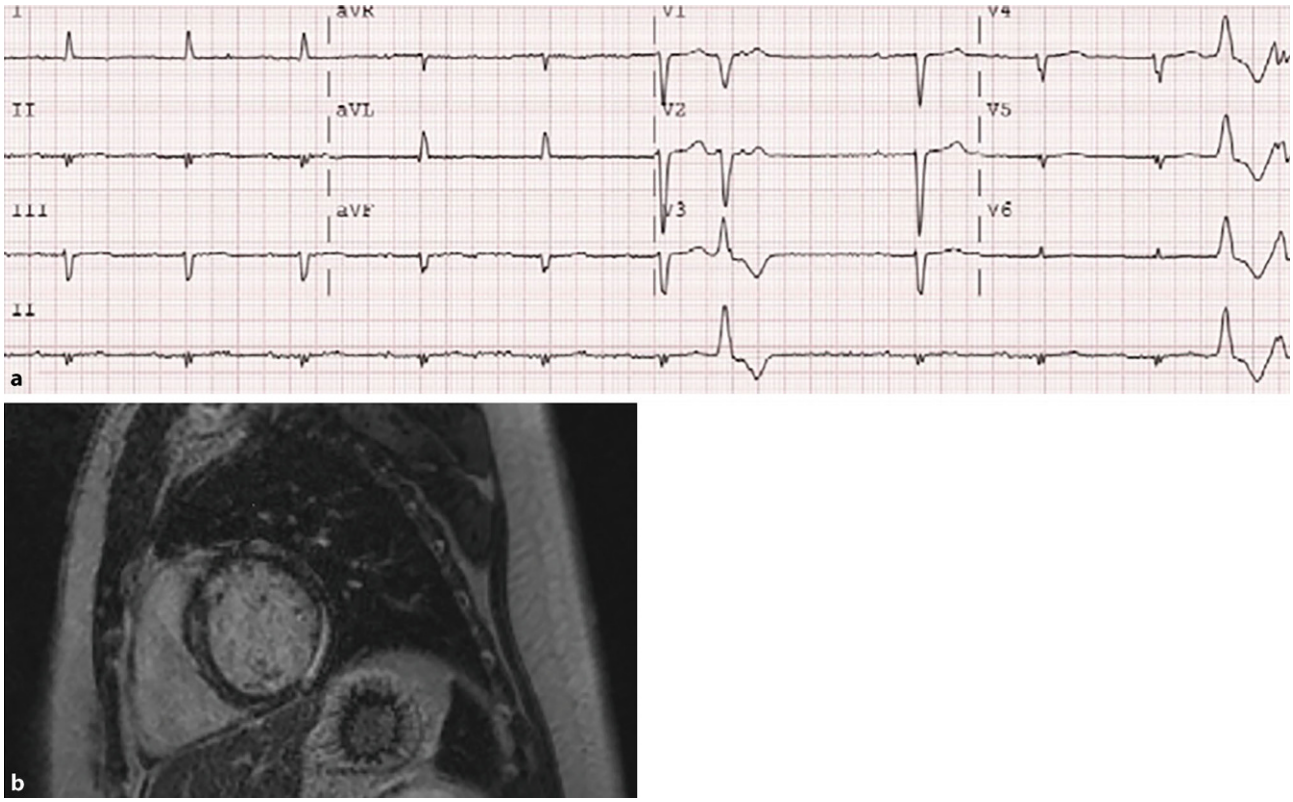

Fig. 1 **a** Electrocardiogram. **b** Cardiac MRI