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Better late than never: assessment of arrhythmogenic cardiomyopathy in an elderly patient

Más vale tarde que nunca: evaluación multimodal de la miocardiopatía arritmogénica en el paciente anciano

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A 74-year-old woman without previous medical history except for the left bundle branch block was admitted for evaluation of recurrent syncopes. During admission, she experienced a sustained self-limited monomorphic ventricular tachycardia together with new syncope. Initial echocardiography displayed moderate biventricular systolic dysfunction. Cardiac magnetic resonance imaging (CMRI) confirmed these findings (Fig. 1) and revealed patchy subepicardial areas of late gadolinium enhancement within the left ventricular inferolateral and apical segments (red arrow) and an aneurysm was found in the right ventricular apex, containing a rounded thrombus (blue arrow) which persisted 10 days after intravenous anticoagulation therapy.

Due to the clinical suspicion of biventricular arrhythmogenic cardiomyopathy with sustained ventricular arrhythmias, cardioverter-defibrillator (ICD) implantation was decided. Subcutaneous approach, initially preferred due to persistent thrombus, was finally dismissed due to predicted high risk of inappropriate therapies in the screening test. Finally, a transvenous ICD was implanted with defibrillation electrode located in the posterior interventricular vein and left bundle branch pacing (Fig. 2) with a significant reduction of paced QRS duration (Fig. 3) and partial recovery of biventricular function during the follow-up. Subsequently, a genetic study was performed confirming a pathogenic variant in the DSG2 gene, which not only explained the phenotype but also allowed familiar cascade screening (Fig. 4).

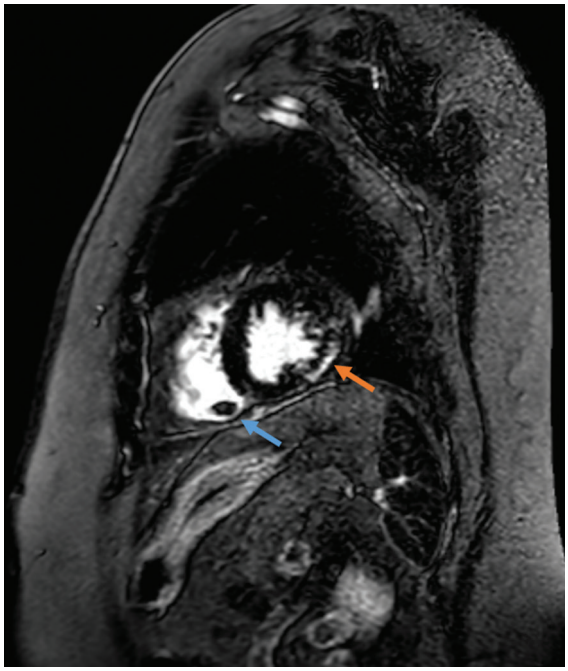

Figure 1. CMRI. Short axis. Inversion-recovery sequence. Late gadolinium enhancement in inferolateral wall (red arrow). Aneurysm in the right ventricle containing a rounded thrombus (blue arrow).

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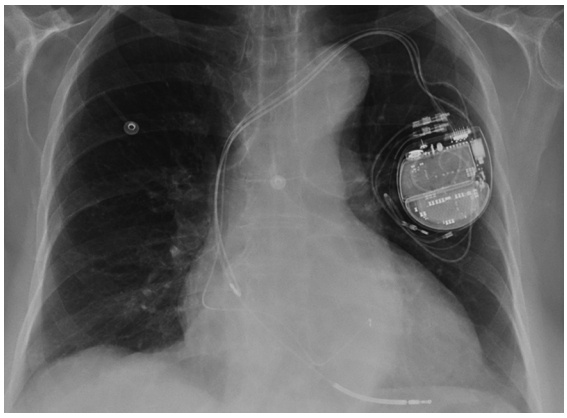

Figure 2. Chest radiography after implantation of ICD.

Arrhythmogenic cardiomyopathy is characterized by replacement of myocardium by fibro-fatty tissue. It is associated with mutations in the genes that encode cardiac desmosomes, crucial proteins for the cardiomyocytes electromechanical connection[1,2]. Although usually diagnosed in youngsters, the greater availability of genetic tests and CMRI help to detect the late phenotypes, with crucial implications for the patient's relatives.

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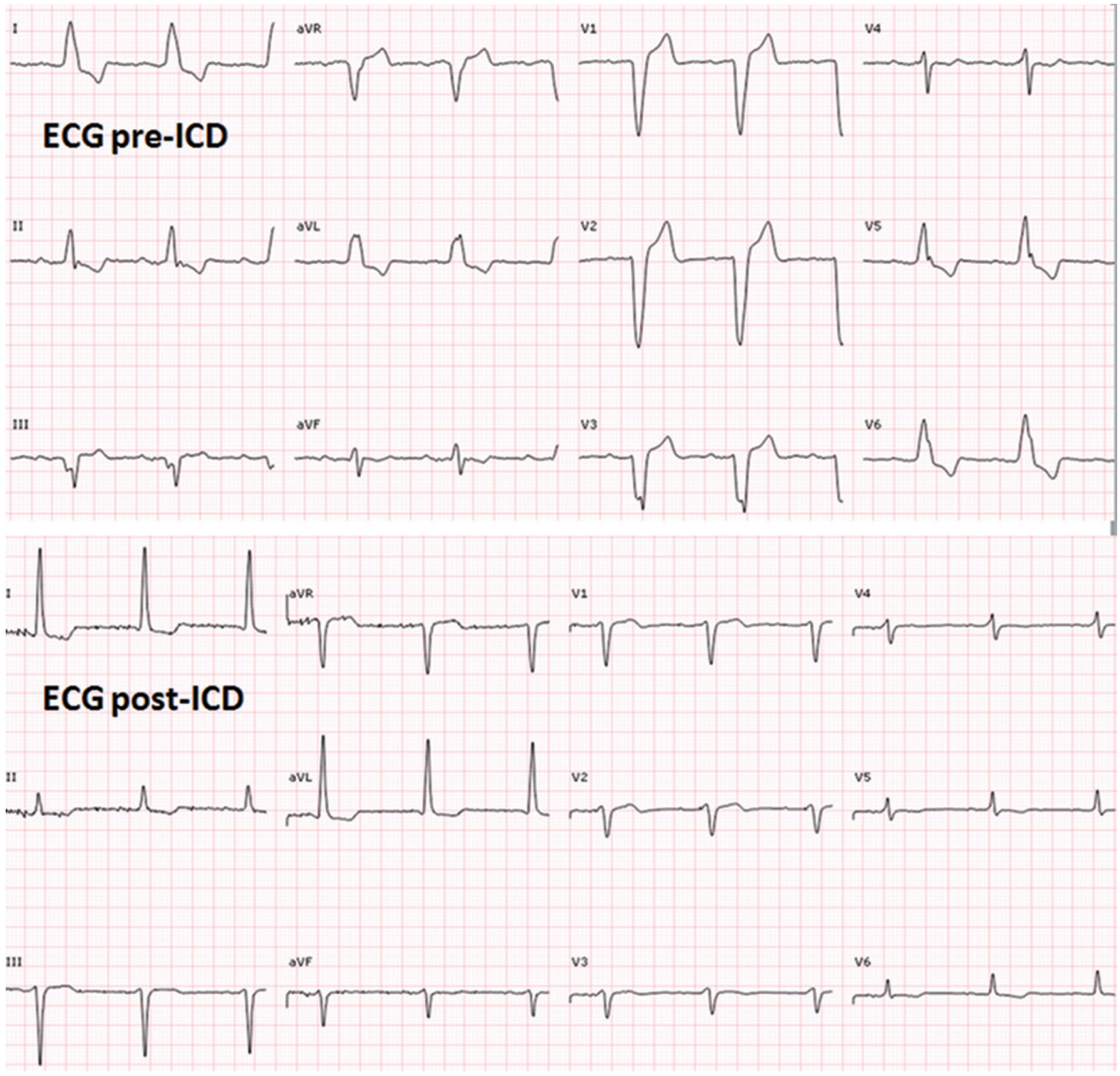

Figure 3. Electrocardiogram before and after ICD implantation with left bundle branch pacing.

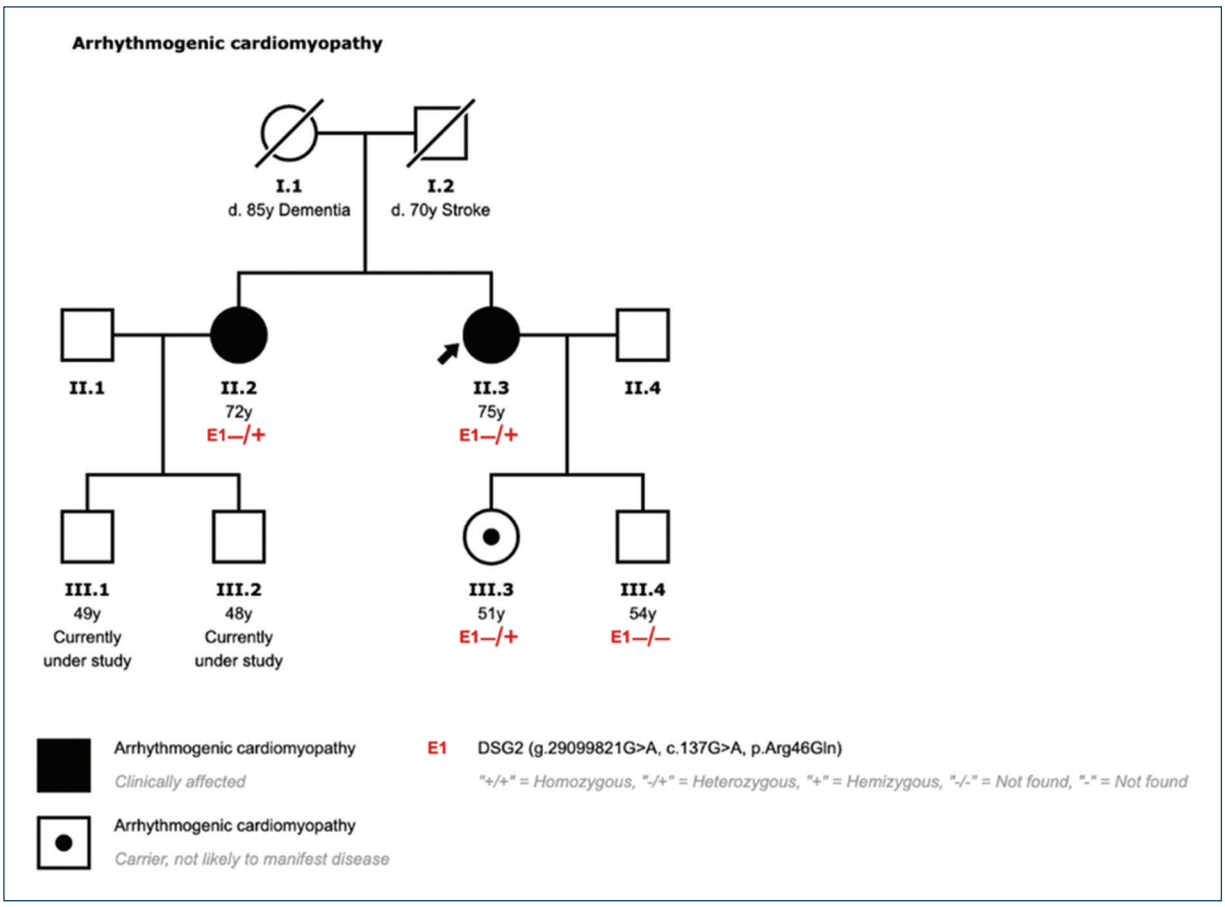

Figure 4. Family tree of the patient.

Conflicts of interest

All authors declare no pertinent conflicts of interest for the present study.

Ethical disclosures

Protection of human and animal subjects. The authors declare that the procedures followed were in accordance with the regulations of the relevant clinical research ethics committee and with those of the Code of Ethics of the World Medical Association (Declaration of Helsinki).

Confidentiality of data. The authors declare that they have followed the protocols of their work center on the publication of patient data.

Right to privacy and informed consent. The authors have obtained the written informed consent of the patients or subjects mentioned in the article. The corresponding author is in possession of this document.

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