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Fulminant cardiac amyloidosis

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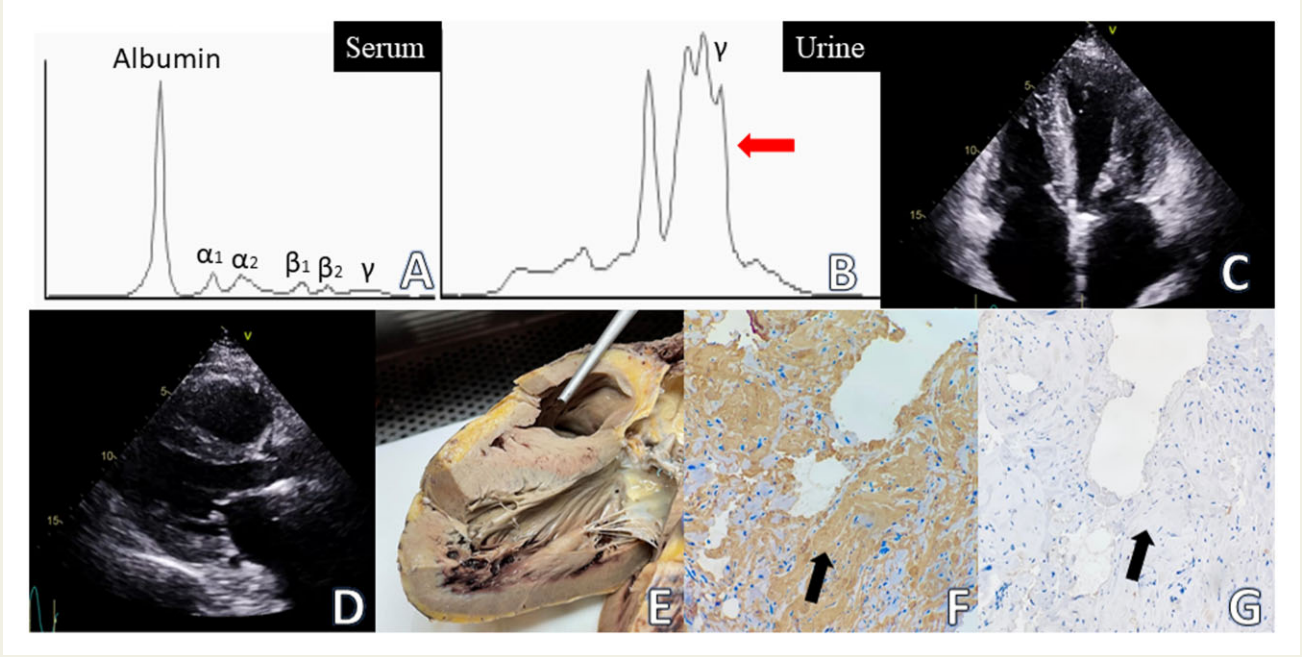
A 56-year-old man presented at the emergency department with dyspnoea. His history included a bioprosthetic aortic valve (Trifecta valve, 21 mm) replacement 4 months prior due to severe aortic stenosis resulting in a first episode of heart failure. Postoperatively, he developed atrial fibrillation, so anticoagulation with Apixaban and rate control strategy with Bisoprolol were started. Left ventricular ejection fraction (LVEF) after surgery was mildly reduced (45%).

On this occasion, he developed cardiogenic shock. His electrocardiogram showed poor r-wave progression and widespread q waves in precordial leads (Supplementary material online, *Figure S1*). His chest X-ray revealed a right-sided pleural effusion. Transthoracic echocardiography showed severely reduced LVEF (10–15%) with biventricular hypertrophy and thickened interatrial septum, with a normally functioning aortic prosthesis (*Panels C* and *D*, *Videos 1* and 2). Transoesophageal echocardiogram ruled out any degree of aortic stenosis or regurgitation. Inotropic support was initiated. The patient suffered a cardiac arrest with electromechanical dissociation. Advanced cardiopulmonary resuscitation manoeuvres were successful and an intra-aortic balloon pump was implanted.

In the following hours, a biventricular assist device (Levitronix Centrimag) was implanted. Considering the deterioration of the patient despite the treatment of aortic stenosis and the echocardiographic findings described above, diagnosis of an infiltrative



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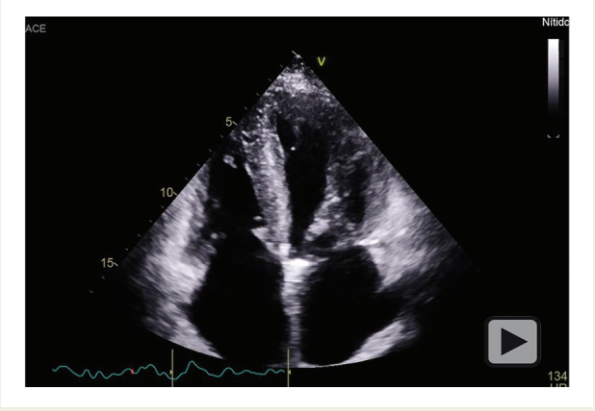
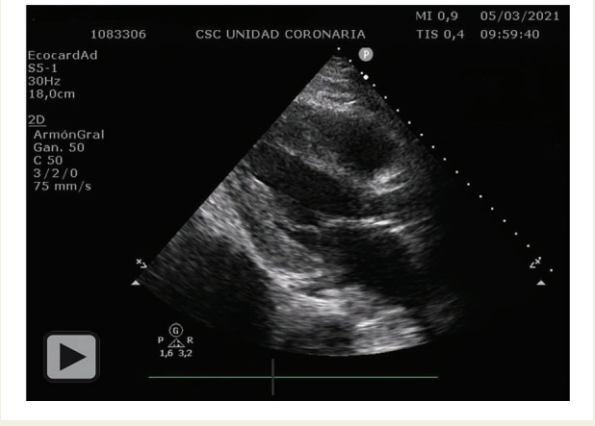


Video 1 Transthoracic echocardiography showing severe reduced Left ventricular ejection fraction with biventricular hypertrophy and thickened interauricular septum.



Video 2 Transthoracic echocardiography showing biventricular hypertrophy.

cardiomyopathy was suspected and serum and urine protein electrophoresis were performed. Cardiac magnetic resonance would be essential in the diagnosis of an infiltrative cardiomyopathy and a bone scintigraphy with 99m technetium-labelled bisphosphonates could confirm TTR cardiac amyloidosis, but the instability of the patient did not allow us to perform either of the two tests. A monoclonal component of kappa light chains was detected in the urine (*Panel B*, red arrow) but not in the serum sample, which was normal (*Panel A*). An endomyocardial biopsy with haematoxylin-eosin staining revealed acellular and amorphous material deposits with Congo Red positivity, despite treatment with potassium permanganate. Immuno histochemical staining confirmed the diagnosis of AL amyloidosis with a high expression of kappa light chains (*Panel F*, black arrow) and restriction of lambda light chains (*Panel G*, black arrow). Targeted treatment with corticosteroids, Bortezomib and Cyclophosphamide was started. Notwithstanding, the patient suffered from device cannulae anastomosis bleeding, requiring four surgical reinterventions. The balance between device-related thrombosis and bleeding complications became challenging, since the patient started with uncontrolled gastrointestinal bleeding related to duodenal ulcers. These complications eventually caused the death of the patient. Marked biventricular hypertrophy was confirmed at autopsy (*Panel E*).

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

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Consent: The authors confirm that witnessed verbal consent for submission and publication of this case report including images and associated text has been obtained from the patients detailed in this case report. The patient has since become deceased and has no surviving next of kin. This has been discussed with the editors.

Conflict of interest: None declared.