Fulminant cardiac amyloidosis

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Short title: Fulminant cardiac amyloidosis

Short abstract: A 56-year-old man developed cardiogenic shock. Echocardiography showed biventricular thickening with systolic dysfunction. Diagnosis of AL Cardiac Amyloidosis was confirmed by pathology and the patient died from severe complications.

Disclosure: The Author(s) declare(s) that there is no conflict of interest

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Figure 1:

Supplementary figure 1:
Clinical message:

A 56-year-old man presented at the emergency department with dyspnea. His history included a bioprosthetic aortic valve (Trifecta valve, 21 mm) replacement four 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 figure 1). 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 S1 and S2). Transesophageal 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 maneuvers 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 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-labeled 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 Hematoxylin-eosin staining revealed acellular and amorphous material deposits with Congo Red positivity, despite treatment with potassium permanganate. Immunohistochemical 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).
Consent statement: 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.
Supplementary figure 1.
12. Supplemental files (i.e. movies, videos etc do not include patient details)

Video S1.mp4
12. Supplemental files (i.e movies, videos etc do not include patient details)

Video S2.mp4