Abstract

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The serial accumulator

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Background

A 74 year old lady came to our with non-productive cough, sloping edema and worsening dyspnoea for about 10 days, she had already performed a chest CT scan at home which showed slight bilateral baseline fluid. She decided to go to the ED where they found a mild normocytic normochromic anemia (Hb 11.4 g/dL), Troponin I 148 ng/L, BNP 2554 pg/mL, and LDH 330 U/L. The troponin curve performed with dosage at three and ten hours was not significant due to ischemic in progress. At the anamnestic interview, evidence of arterial hypertension, chronic ischemic heart disease, recent PMK implant (approximately 20 days earlier for the onset of symptoms for BAV II° Mobitz 2), and previous HCV infection subjected to eradicating antiviral therapy. On admission, the patient was dyspnoeic but hemodynamic ally stable. In the light of the suggestive picture of congestive heart failure, intravenous diuretic therapy was set up with furosemide and potassium canrenoate with consequent benefit on respiratory dynamics. On the fifth day of hospitalization, in order to monitor cardiac function, it was decided to perform a new echocardiogram, which showed severe concentric hypertrophy of the left ventricle with preserved volume, moderate mitral regurgitation and sclerocalcified aortic valve, concluding for probable storage disease. Approximately three days after the last echocardiographic check-up, the patient reported acute precordial pain, the ECG was not suggestive of ACS but there was a rapid increase in the Troponin I value (34624 pg/mL) and consequently the patient went to UTIC to perform coronary angiography, which was negative for coronary lesions. In consideration of the suspicion of cardiac infiltrative pathology, it was decided to perform:

- serum immunofixation for and lambda chains, which later proved positive for the monoclonal lambda component;
- bone scan with HMDP, tested negative (Score 0 by Perugini);
- end myocardial biopsy, which confirmed the presence of a picture compatible with storage disease, specifically attributable to ‘‘cardiac amyloidosis with my cytolysis’’;
- Immuno-electromicroscopy, with evidence of an ultra-structural picture compatible with AL Amyloidosis (lambda light chains).

Once the acute symptoms were controlled and the patient stabilized, the decision was made to discharge the patient with an indication to continue the therapeutic process at a specialist reference center [1-3].

Discussion

Cardiac amyloidosis is a severe and progressive infiltrative disease caused by hereditary or acquired physiological abnormalities that lead to the accumulation of amyloid fibrils in the heart. More than 98% of cardiac amyloidosis is caused by just two types of proteins: immunoglobulin light chains and transthyretin. Amyloidosis AL has an annual incidence of approximately 1 per 100,000 people in the United States.

Typically, cardiac amyloidosis manifests itself with a large number of extra cardiac signs and symptoms, defined as "red flags", which can guide the diagnostic suspicion.

These include proteinuria, macroglossia, bruising of the skin, and a history of carpal tunnel syndrome. But there are also cardiac red flags, such as the presence of heart failure with a level of NT-proBNP disproportionate to the echocardiographic evidence, right heart failure in the presence of normal valvular and ventricular function, persistently elevated troponin levels, a QRS voltage too low or an early conduction disturbance.

However, even in the absence of "red flags", a storage disease must always be suspected in patients with multiple ischemic events but with unaffected coronary, and in acute decompensated patients. The therapeutic approaches are distinguished in those aimed at treating the comorbidities deriving from the disease,

such as heart failure, arrhythmias, conduction disturbances, thromboembolism, aortic stenosis, and actual disease-modifying treatments which are able to act on different stages of production of amyloid fibrils, from formation to assembly [4-6].

References

1 Nils de Marneffe (2022) Cardiac amyloidosis: a review of the literature. Acta Cardiol 77: 683-692.

2 Maurer MS (2017) Common questions encountered in the diagnosis and management of Cardiac Amyloidosis. Circulation 135: 1357-1377.

3 Dubrey SW (2011) Amyloid diseases of the heart: assessment, diagnosis, and referral. Heart 97: 75-84.

4 Writing Committee (2023) ACC Expert consensus decision pathway on comprehensive multidisciplinary care for the patient with Cardiac Amyloidosis: A report of the American College of Cardiology solution set oversight committee. J Am Coll Cardiol 81: 1076-1126.

5 Palladini G (2020) Daratumumab plus CyBorD for patients with newly diagnosed AL Amyloidosis: safety run-in results of Andromeda. Blood 136: 71-80.

6 Gillmore JD (2016) No biopsy Diagnosis of Cardiac Transthyretin Amyloidosis. Circulation 133: 2404-2412.