

Non-amyloidotic light-chain deposition cardiomyopathy

Clinical Case Portal

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Case Report

Light-chain deposition in myocardium usually manifests as amyloidotic restrictive cardiomyopathy. In rare cases paraproteinemia can deposit as a non-fibrillary infiltrate that does not show typical tintorial attributes of amyloidosis but resembles its clinical features. Clinical suspicion and correct diagnosis are important since early treatment of underlying cell plasma dyscrasia can reverse cardiac dysfunction. We present images from the case of a 47-year-old woman with fatal non-amyloidotic light-chain deposition cardiomyopathy that developed left atrial mechanical dysfunction while in sinus rhythm complicated with systemic arterial embolization.

Patient history prior to current observation :

A 47-year-old woman sustained a 8-month history of heart failure. She had been managed in an outpatient clinic. At symptoms onset, she underwent transthoracic echocardiography that revealed "concentric hypertrophy and normal ventricular function". Initial diagnosis of non-obstructive hypertrophic cardiomyopathy was made, but small doses of propranolol first, and verapamil later resulted in significant worsening of symptoms. Diuretics and vasodilators were started, but heart failure symptoms progressed. She was then referred to our hospital outpatient clinic for evaluation.

Clinical findings on admission, evolution and outcome :

Outpatient work-up in our hospital:

· **ECG** (fig. 1): Sinus rhythm, nonspecific intraventricular conduction delay and repolarization

alterations.

- **Chest X-ray** (fig. 2): Cardiomegalia, mild bilateral pleural effusion, pulmonary vascular congestion and interstitial oedema.
- **Blood tests**: Haemogram, coagulation, serum creatinine, thyroid hormones, ferritin and angiotensin-converting enzyme were normal. Erythrocyte sedimentation rate was high at 80 mm/h. Serum electrophoresis showed a monoclonal peak of lambda light-chains, while levels of IgG, IgA and IgM were slightly decreased (fig. 3).
- **Urine analysis**: Proteinuria (2.1 g/dL). Monoclonal lambda light-chain peak of 3.5 mg/dL (Bence-Jones Proteinuria).
- **Transthoracic echocardiography**: Concentric (relative wall thickness 0.99) increment of left ventricular mass (113 g/m²). Left ventricular ejection fraction 58%. (fig. 4; fig. 5; fig. 6). Note that fractional shortening (fig. 4 and fig. 5) is well preserved, but longitudinal shortening assessed in fig. 6 is reduced. Left atrial dimensions were normal. Valves had no significant dysfunction. Mild pericardial and left pleural effusions were found. Mitral inflow (fig. 7 showed short E wave deceleration time (120 ms) and a hardly visible atrial contribution. Pulmonary vein flow (fig. 8) showed a marked prevalence of the diastolic component and the absence of retrograde A wave. Both Doppler patterns were consistent with restrictive physiology and left atrial mechanical dysfunction. Pulsed tissue Doppler interrogation of mitral annulus at both its septal and lateral points disclosed E' wave velocity of 6 cm/s, pointing towards restrictive cardiomyopathy rather than constrictive pericarditis.
- Rectal mucose biopsy was negative for Congo red staining.

Discussion

Evolution and outcome

Still awaiting results of the above mentioned tests, our patient was admitted due to acute lower limb ischaemia. ECG showed sinus rhythm. Ischaemia improved on heparin, but heart failure worsened despite intensive therapy and our patient died on the fifth day from admission. **Autopsy** found deposition of extracellular homogeneous eosinophilic material negative for Congo red stain and positive for lambda light-chain immunostaining in myocardium (fig. 9; fig. 10), pancreas, spleen and kidney. Histology did not reveal plasma cell disease in bone marrow or elsewhere. **Mural thrombi** were found in both atria. Recent infarction of **embolic** pattern were noted in left kidney and spleen.

Conclusion

Although cardiac amyloidosis is the characteristic expression of paraproteinemia deposition in myocardium, non-amyloidotic light-chain deposition cardiomyopathy should be considered in the differential diagnosis of restrictive cardiomyopathy.

- Its recognition is important since reversibility has been reported after successful treatment of underlying plasma cell dyscrasia.
- Amyloidosis shows typical staining with Congo red. Diagnosis of non-amyloidotic light-chain deposition is confirmed by paraproteinemia demonstration, absence of Congo red staining and positive immunostaining for immunoglobulin light chains in biopsy specimens.
- Nonamyloidotic light-chain deposition cardiomyopathy has been reported to mimic clinical and echocardiographic features of cardiac amyloidosis, including atrial electromechanical dissociation (fig. 1; fig. 7; fig. 8) and embolic events.
- Whether anticoagulation should be instituted in case of left atrial mechanical dysfunction despite electrocardiographic sinus rhythm is debatable. Echocardiography has a role in left atrial electromechanical dissociation diagnosis.

References

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Fig. 1 :

[Non-amyloidotic light-chain cardiomyopathy ECG Non-specific QRS-T abnormalities](#)



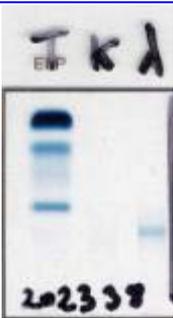
Fig. 2 :

[Non-amyloidotic light-chain cardiomyopathy Chest X-ray film: heart failure](#)



Fig. 3 :

[Serum immunoelectrophoresis: lambda light-chain monoclonal band](#)



Video 1 :

[Non-amyloidotic light-chain cardiomyopathy TTE PLAX](#)



Video 2 :

[Non-amyloidotic light-chain cardiomyopathy TTE 2D PSAX](#)



Video 3 :

[Non-amyloidotic light-chain cardiomyopathy TTE 2D AP4C](#)



Fig. 4 :

[Non-amyloidotic light-chain cardiomyopathy TTE PWD Restrictive mitral inflow](#)

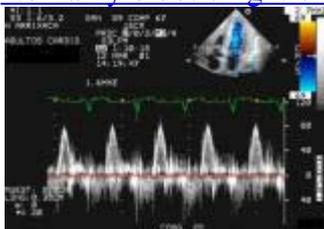


Fig. 5 :

[Non-amyloidotic light-chain cardiomyopathy TTE Pulmonary vein flow restrictive pattern](#)

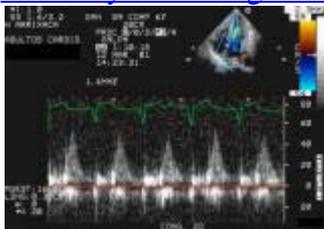


Fig. 6 :

[Non-amyloidotic light-chain cardiomyopathy Histology Myocardium Congo red staining No signs of amyloidosis](#)

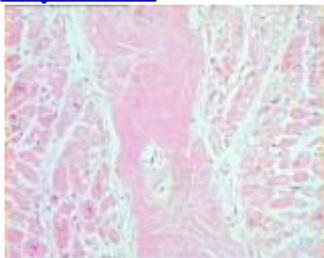


Fig. 7 :

[Non-amyloidotic light-chain cardiomyopathy Histology lambda light-chain immunostaining](#)

