

Echocardiographic findings of biopsy proven systemic and cardiac amyloidosis

Clinical Case Portal

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Case Report

We describe a case of systemic amyloidosis with cardiac involvement diagnosed by transthoracic echocardiography and histologic study.

Patient history prior to current observation :

A 62-year-old caucasian woman was evaluated at our institution because of dizziness. She has been previously hospitalized in medical department where systemic amyloidosis with renal and liver involvement was diagnosed. For severe renal impairment the patient was in chronic dialytic treatment.

Her medical history was remarkable for systemic hypertension, monoclonal gammopathy and thyroidectomy for papillary carcinoma.

Clinical findings on admission, evolution and outcome :

Physical examination: Blood Pressure = 140/80 mmHg, Heart Rate = 87/min regular, 3/6 apical systolic murmur, clear lungs, no peripheral oedema, hepatomegaly or jugular veins' distention.

ECG: Sinus rhythm at 87 bpm with low voltages in peripheral leads and pseudo-infarction pattern in precordial leads (fig. 1).

Transthoracic echocardiography showed nondilated ventricles with wall thickening and "granular sparkling" appearance of the myocardium (fig. 2, fig. 3, fig. 4), preserved left ventricular global systolic function (fig. 5), diastolic dysfunction with a restrictive pattern (fig. 6). Aortic and mitral leaflets appeared slightly thickened (fig. 5) and color-Doppler showed mild aortic and mitral regurgitation. Atrial chamber appeared enlarged with no evidence of atrial septum thickening (fig. 5).

Immunohistochemical stainings: myocardial (fig. 7), renal (fig. 8), bone marrow (fig. 9) and colon (fig. 10) amyloid deposition.

Conclusion

Our case is representative of a typical case of systemic amyloidosis with cardiac involvement. The diagnosis is important for prognosis and therapy. However, this diagnosis can be difficult because the clinical presentation may mimic other infiltrative cardiomyopathies or storage disorders, as well as hypertrophic cardiomyopathy. Even in absence of histological confirmation of the diagnosis, there is evidence that a combination of echocardiographic and ECG features (low voltage ECG and increased interventricular septal thickness), in patients with clinical suspicion of cardiac amyloidosis, could allow a correct diagnosis with a high positive predictive value (1).

References

Rahman JE et al. Noninvasive Diagnosis of Biopsy-Proven Cardiac Amyloidosis. J Am Coll Cardiol 2004; 43:410-5.

Fig. 1 :
[Cardiac amyloidosis_Electrocardiogram](#)

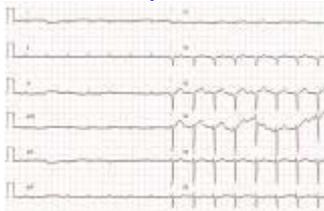
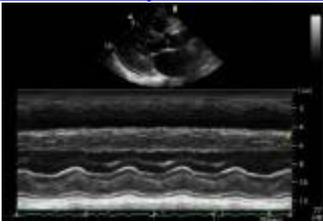


Fig. 2 :

Cardiac amyloidosis Transthoracic left ventricle M-Mode



Video 1 :

Cardiac amyloidosis Transthoracic short-axis view



Video 2 :

Cardiac amyloidosis Transthoracic apical four-chamber view



Fig. 3 :

Cardiac amyloidosis Transmitral Doppler flow and Tissue Doppler Imaging

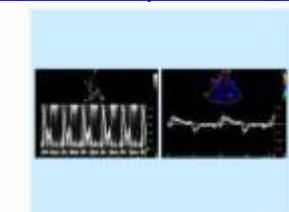


Fig. 4 :

Cardiac amyloidosis Endomyocardial biopsy

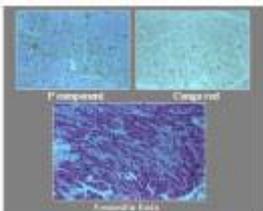


Fig. 5 :

Cardiac amyloidosis Renal byopsy

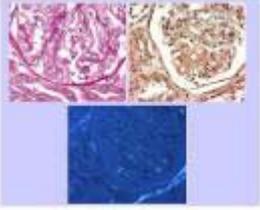


Fig. 6 :
Cardiac amyloidosis Bone marrow biopsy

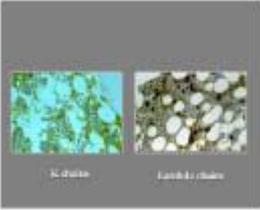


Fig. 7 :
Cardiac amyloidosis Colon biopsy

