Obstructive intramural coronary amyloidosis: a distinct phenotype of cardiac amyloidosis that can cause acute heart failure

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A 66-year-old man was admitted to a cardiology unit with diagnosis of congestive heart failure, having complained of worsening asthenia and myalgia for several months before referral. Family history was aspecific. Echocardiography showed a moderately dilated and uniformly hypokinetic left ventricle (end-diastolic diameter 68 mm; ejection fraction 35%); wall thickness was normal. Coronary angiography showed normal epicardial arteries. After temporary improvement during treatment with beta-blockers and ACE-inhibitors, heart failure became severe and left ventricular ejection fraction fell to 20%. Suspected myocarditis prompted a right ventricular endomyocardial biopsy (Panel A), which excluded inflammation but identified amyloid infiltration of small intramural vessels without interstitial involvement. A left ventricular assist device was urgently implanted: histopathological examination of the excised left ventricular apex confirmed obstructive intramural coronary amyloidosis without interstitial deposits and with foci of coagulative necrosis. The patient died a few days later because of gastroenteric haemorrhage (autopsy was not performed) (Panels B–D).

This case documents the existence of isolated intramural coronary obstruction as a peculiar phenotype of cardiac amyloidosis (distinct from the more common amyloidotic cardiomyopathy) (Panels E–I). This rare type of amyloidotic cardiac involvement—which in this patient led to a mistaken clinical diagnosis of myocarditis—must be recognized as one of the possible causes of acute or rapidly progressive heart failure.

Panel A. Endomyocardial biopsy sample showing the small arteriolar vessel (arrow) that led to the diagnosis (H&E staining).
Panel B and C. Electron micrograph of the sample showing specific immunostaining of amyloid fibrils with anti-λ light chain monoclonal antibodies.
Panel D. Echocardiography, 1 day before the implantation of the left ventricular assist device, shows left ventricular dilation. The left ventricular apex removed at implantation displays amyloid exclusively infiltrating small intramural vessels.
Panel E and F. Small intramural arteriolar vessels heavily infiltrated by amyloid deposits [(E) H&E stain and (F) Congo Red stain].
Panel G. Left portion shows hypereosinophilic coagulative necrosis (non-necrotic myocardium is seen on the right); left margin: small vessel infiltrated by amyloid with thrombosis (H&E staining).
Panel H. Polarized light view of Congo Red-stained small intramural arteriolae: there are no amyloid deposits in the myocardial interstitium.
Panel I. Portion of the epicardial apical left anterior descending coronary artery showing the absence of amyloid infiltration.

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