



Autopsy and Case Reports

ISSN: 2236-1960

Hospital Universitário da Universidade de São Paulo

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Autopsy and Case Reports, vol. 7, no. 1, 2017, January-March, pp. 13-15
Hospital Universitário da Universidade de São Paulo

DOI: <https://doi.org/10.4322/acr.2017.007>

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A case of intramural coronary amyloidosis associated with hemodialysis

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Ronny FMH, Kleinman G, Kurtin PJ, Fallon JT. A case of intramural coronary amyloidosis associated with hemodialysis. Autopsy Case Rep [Internet]. 2017;7(1):13-15. <http://dx.doi.org/10.4322/acr.2017.007>

ABSTRACT

Dialysis-related amyloidosis predominantly occurs in osteo-articular structures and dialysis-related amyloid (DRA) substances also deposit in extra-articular tissues. Clinical manifestations of DRA include odynophagia, gastrointestinal hemorrhage, intestinal obstruction, kidney stones, myocardial dysfunction, and subcutaneous tumors. The pathological characteristics of DRA in the heart of hemodialysis patients have rarely been reported. We report the case of a 73-year-old female with a history of cerebral palsy and end-stage renal disease status post two failed renal transplants who had been on hemodialysis for 30 years. The patient was admitted with the working diagnosis of pneumonia. An echocardiography showed markedly reduced biventricular function manifested by low blood pressure with systolic in the 70s and elevated pulmonary artery pressure of 45 mmHg, which did not respond to therapy. Following her demise, the autopsy revealed bilateral pulmonary edema and pleural effusions. There was cardiac amyloid deposition exclusively in the coronary arteries but not in the perimyocytic interstitium. Amyloids were also found in pulmonary and intrarenal arteries and the colon wall. Previous case reports showed that beta 2-microglobulin amyloid deposits in various visceral organs but less frequently in the atrial and/or the ventricular myocardium. In the present case, amyloids in the heart were present in the intramural coronary arteries causing myocardial ischemia and infarction, which was the immediate cause of death.

Keywords

Renal Dialysis; Amyloidosis; Coronary Disease; Autopsy

CASE REPORT

A 73-year-old African American woman was referred to our hospital from her nursing home for complaints of shortness of breath. She had been on hemodialysis for 30 years because of end-stage renal dysfunction due to chronic glomerulonephritis. She had developed dialysis-related amyloid substance deposition in her shoulder and wrist joints and previously underwent bilateral carpal tunnel surgery.

The patient was admitted to hospital with the working diagnosis of healthcare-associated pneumonia and was treated with antibiotics. During her hospital course, she developed low blood pressure and tachycardia. An electrocardiogram showed acute ST segment elevation and atrial fibrillation with ventricular tachycardia. The troponin determination was elevated. An echocardiogram revealed decreased bi-ventricular

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systolic function, which was diffuse; the echo pattern and thickness of the myocardium were within normal limits.

Unfortunately, the patient died, and an autopsy was performed. Pathological findings showed severe amyloid deposition in almost all major organs including the intramural coronary vessels. On gross examination,

the heart showed a recent lateral left ventricular wall infarction, multifocal myocardial replacement fibrosis, and amorphous eosinophilic intimal deposits in the intramural arteries (Figure 1) causing severe stenosis.

Amyloid deposition was exclusively in the coronary arteries but not in the perimycocytic interstitium. Stains for Congo red (Figure 2A) and the amyloid

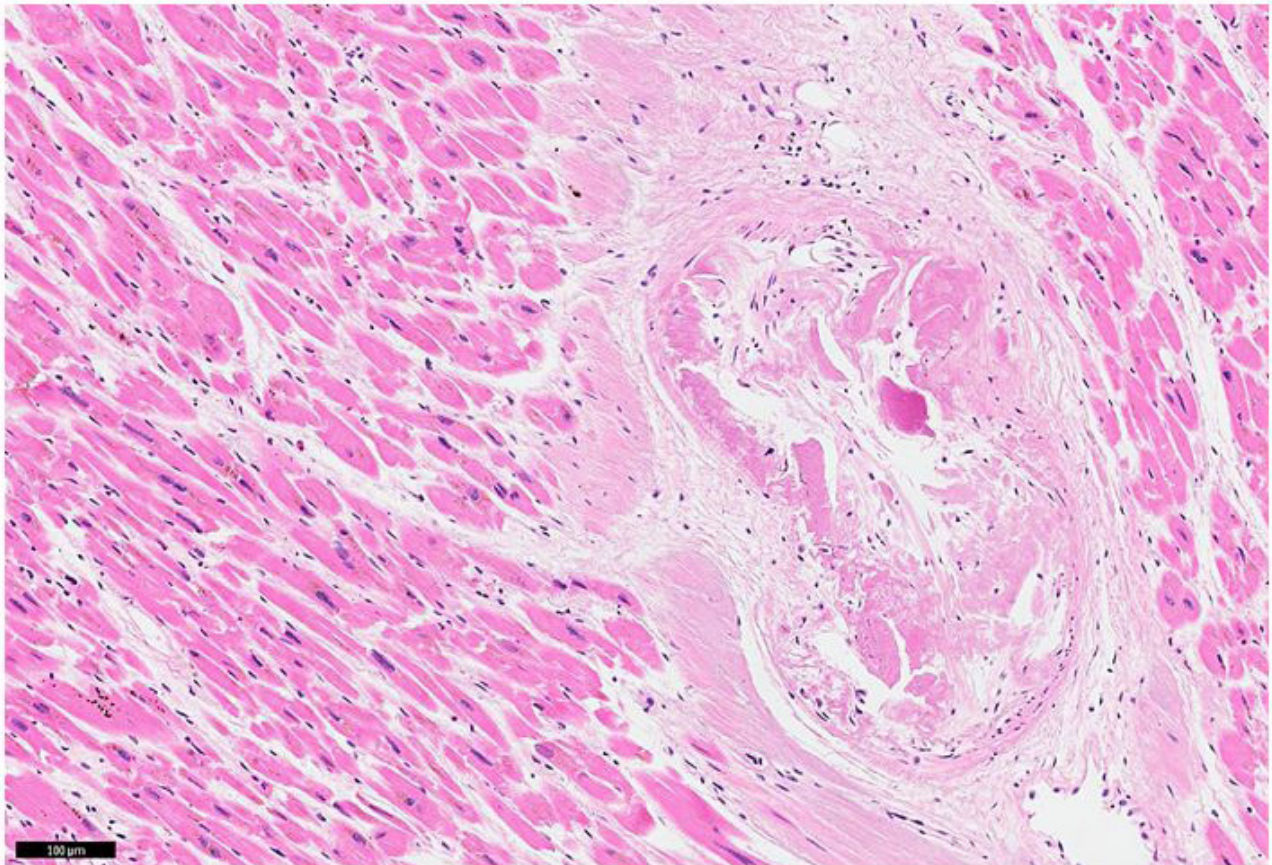


Figure 1. Photomicrograph of the heart showing coronary artery deposition of amyloid filling the lumen (H&E).

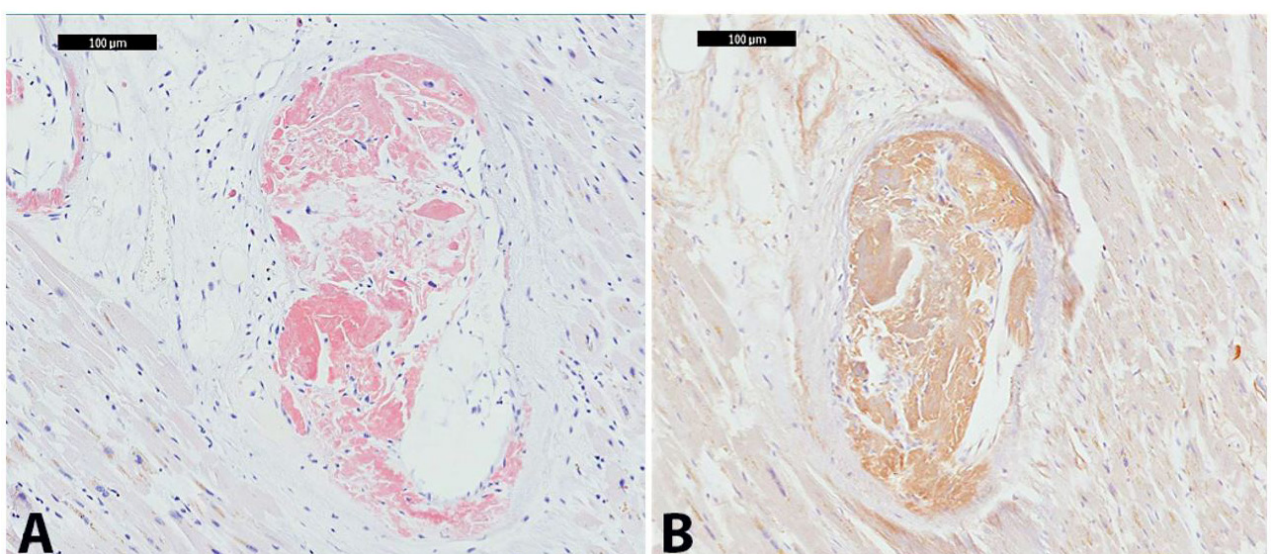


Figure 2. Photomicrograph of the heart showing coronary artery deposition of amyloid. **A** - Congo red; **B** - Amyloid P staining.

P component (Figure 2B) were positive for amyloid deposition in virtually all intramyocardial arteries. Mass spectrometry characterized the amyloids as beta 2-microglobulin, compatible with dialysis-related amyloidosis.

DISCUSSION

Patients with long-term hemodialysis often develop dialysis-related amyloid (DRA), the cause of which is deposition of amyloid fibrils consisting of beta 2-microglobulin.¹⁻³ Previous case reports have shown that beta 2-microglobulin is deposited in various visceral organs and is less frequently involved the heart.³⁻⁵ Especially in the cardiac involvement of DRA, beta 2-microglobulin is deposited in all layers of the atria and/or the ventricle.⁶⁻⁸ In the present case, we found exclusively intramural coronary vessel involvement of DRA at autopsy.

The features of cardiac amyloidosis between primary amyloidosis and DRA are quite different. In patients with hemodialysis-related amyloid substance deposition, even if the echocardiogram does not suggest any cardiac amyloidosis, physicians should consider the possibility of coronary amyloidosis and should check the details of cardiac findings as they may result in cardiac symptoms.

Early amyloidosis without myocardial involvement can produce acute coronary syndrome through the combination of spastic epicardial coronary arteries and obstruction of the intramural coronary arteries. In the management of certain patients with acute coronary syndrome, the possibility of cardiac amyloidosis should be taken into consideration.

Conflict of interest: None

Financial support: None

Submitted on: December 21st, 2016

Accepted on: February 24th, 2017

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