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A Case of Intramural Coronary Amyloidosis Associated With Hemodialysis

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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 perimyocytic interstitium. Stains for Congo red (Figure 2A) and the amyloid

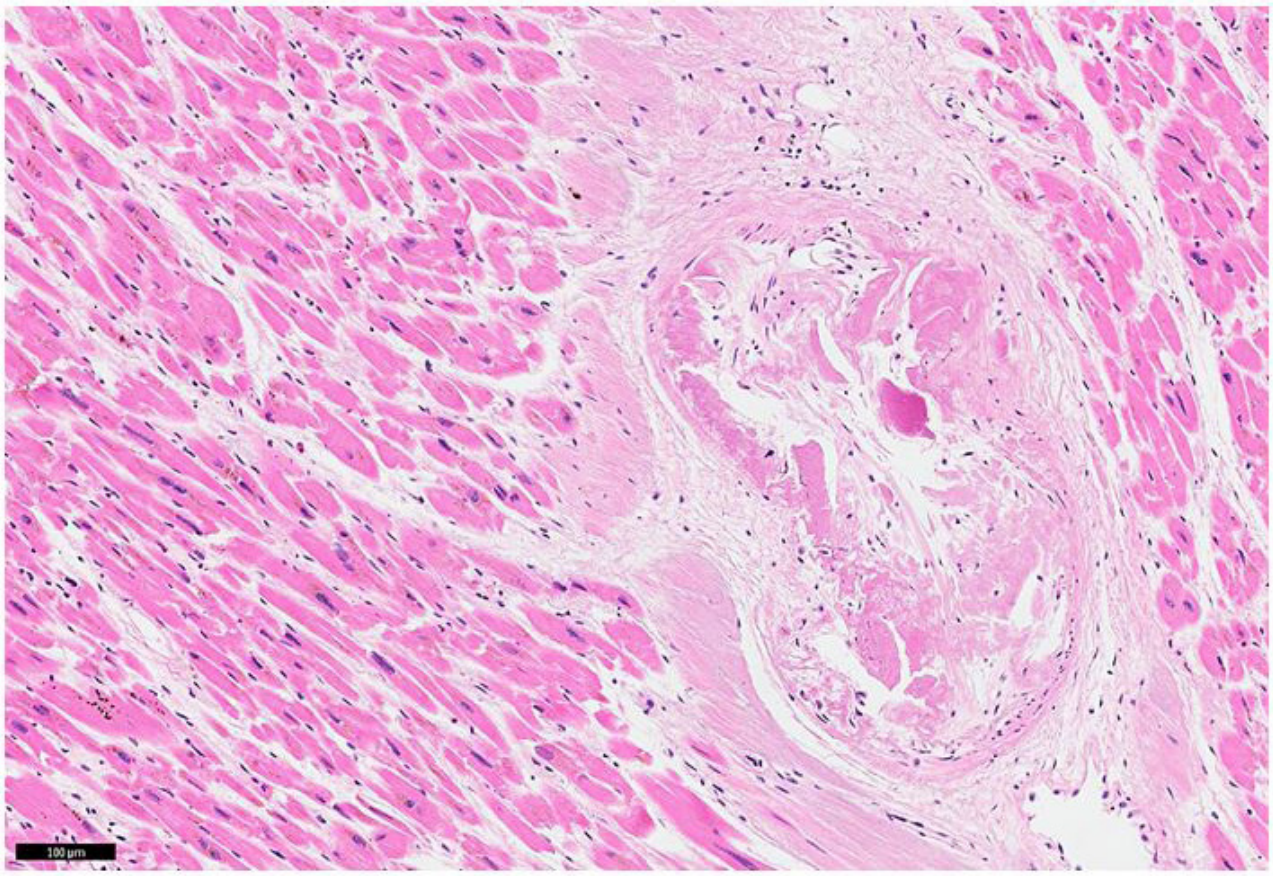

Figure 1. Photomicrograph of the heart showing coronary artery deposition of amyloid filling the lumen (H&E).

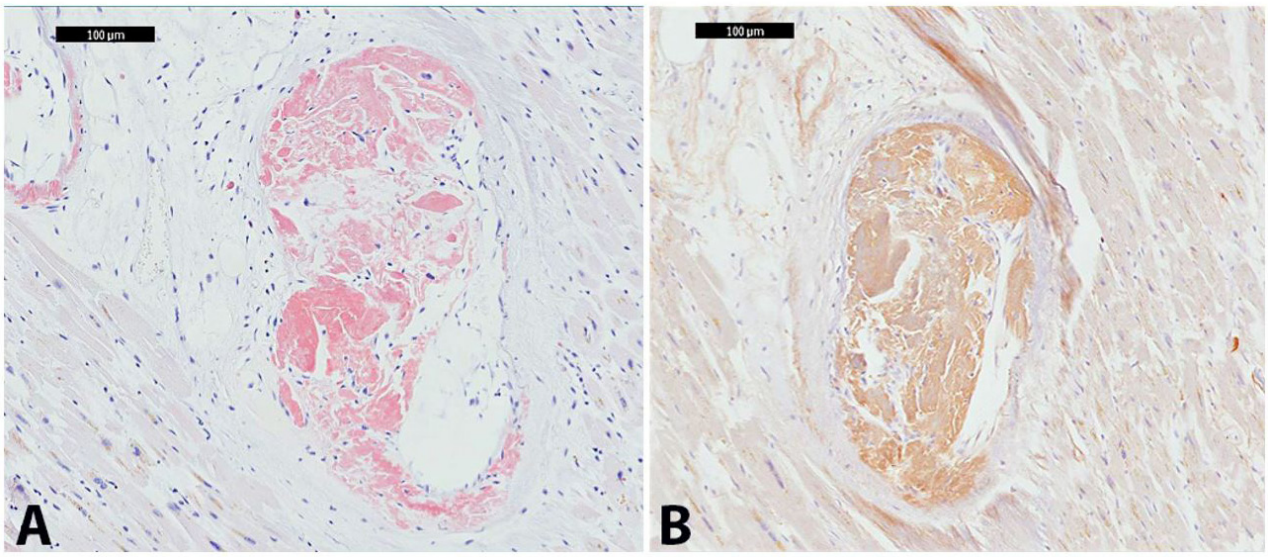

Figure 2. Photomicrograph of the heart showing coronary artery deposition of amyloid. **A** - Congo red; **B** - Amyloid P staining.

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.

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REFERENCES

1. Shirahama T, Skinner M, Cohen AS, et al. Histochemical and immunohistochemical characterization of amyloid associated with chronic hemodialysis as beta-2-microglobulin. Lab Invest. 1985;53(6):705-9. PMid:2415776.
2. Gal R, Korzets A, Schwartz A, Rath-Wolfson L, Gafter U. Systemic distribution of beta-2-microglobulin-derived amyloidosis in patients who undergo long-term hemodialysis. Arch Pathol Lab Med. 1994;118(7):718-21. PMid:8024407.
3. Vrana JA, Gamez JD, Madden BJ, Theis JD, Bergen HR 3rd, Dogan A. Classification of amyloidosis by laser microdissection and mass spectrometry-based proteomic analysis in clinical biopsy specimens. Blood. 2009;114(24):4957-9. PMid:19797517. http://dx.doi.org/10.1182/blood-2009-07-230722.
4. Choi H, Heller D, Picken MM, Sidhu GS, Kahn T. Infarction of intestine with massive amyloid deposition in two patients on long-term hemodialysis. Gastroenterology. 1989;96(1):230-4. PMid:2909422. http://dx.doi.org/10.1016/0016-5085(89)90785-3.
5. Ogawa H, Saito A, Hirabayashi N, Hara K. Amyloid deposition in systemic organs in long-term hemodialysis patients. Clin Nephrol. 1987;28(4):199-204. PMid:3319310.
6. Campistol JM, Argilés A. Dialysis-related amyloidosis: Visceral involvement and protein constituents. Nephrol Dial Transplant. 1996;11(Suppl 3):142-5. PMid:8840330. http://dx.doi.org/10.1093/ndt/11.supp3.142.
7. Takayama F, Miyazaki S, Morita T, Hirasawa Y, Niwa T. Dialysis-related amyloidosis of the heart in long-term hemodialysis patients. Kidney Int. 2001;59:S172-6. PMid:11169005. http://dx.doi.org/10.1046/j.1523-1755.2001.59780172.x.
8. Masuda M, Ishimura E, Ochi A, et al. Serum β2-microglobulin correlates positively with left ventricular hypertrophy in long-term hemodialysis patients. Nephron Clin Pract. 2014;128(1-2):101-6. PMid:25376242. http://dx.doi.org/10.1159/000365447.