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Case Report

A Case of Rapidly Progressive Renal Failure with Unearthed Amyloidosis

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Abstract

Amyloidosis-associated kidney disease commonly manifests with chronic glomerular symptoms including heavy proteinuria predominantly albuminuria. Clinical presentation ranges from full-blown nephrotic syndrome, hematuria, and hypertension to renal failure. In India patients with chronic kidney disease are mainly attributed to hypertension and diabetes but an underlying etiology such as amyloidosis needs to be unearthed and shouldn't be ignored as an etiology. We report a case of a 60-year-old man with hypertension and hypothyroidism who presented with frothy urine for several years, b/l pedal edema for 15 days. Over the past 3 months, there was a serial increase in creatinine. As per CKD-EPI equation, the patient was CKD-4. As the patient was suspected to be rapidly progressive renal failure; a renal biopsy was planned. Biopsy reports were suggestive of Amyloidosis. Glomerular, vascular, and tubulointerstitial deposition of amyloid was seen. Based on renal biopsy and IHC staining; the patient's diagnosis was AA-associated secondary renal amyloidosis. Thus in this case renal amyloidosis was an unearthed etiology.

Introduction

Amyloidosis-associated kidney disease commonly manifests with chronic glomerular symptoms including heavy proteinuria predominantly albuminuria. Clinical presentation ranges from full-blown nephrotic syndrome, hematuria, and hypertension to renal failure [1-3]. In India patients with chronic kidney disease are mainly attributed to hypertension and diabetes but an underlying etiology such as amyloidosis needs to be unearthed and shouldn't be ignored as an etiology, especially in cases of rapidly progressive renal failure.

Case presentation

We report a case of a 60-year-old man with hypertension and hypothyroidism who presented with frothy urine for several years, b/l pedal edema for 15 days. Over the past 3 months, there has been a serial increase in creatinine. As per CKD-EPI equation, the patient was CKD-4. As the patient was suspected to be rapidly progressive renal failure; a renal biopsy was planned. Biopsy reports were suggestive of the following findings, vascular and tubulointerstitial deposition of amyloid was seen. IHC for SAA (Serum Amyloid Associated) protein shows intense (3+) positivity along glomerular sites of amyloid deposition. DIF showed 2+ segmental entrapment of IgM and 1+ segmental entrapment of C3. A characteristic apple green birefringence was seen after the application of a congo red stain under polarized light. There was amyloid deposition in the mesangial area, capillary loops, and weakly in the interstitium. Based on renal biopsy and IHC staining; the patient's diagnosis was AA-associated secondary renal amyloidosis; likely to be related to chronic inflammatory disease. Thus in this case renal amyloidosis was an unearthed etiology.

Discussion

Secondary Renal AA Amyloidosis (RAAA) presents with proteinuria and/or nephrotic syndrome and progresses to End-stage Renal Disease (ESRD) insidiously. However, some patients with secondary amyloidosis show a more rapid renal disease progression than the usual course. A study done in Turkey by Celebi ZK, et al. named "Rapidly Progressive Renal Failure in AA Amyloidosis: A New Clinical and Histopathological Entity for an Old Disease" concluded that higher amyloid deposition and severe inflammation revealed in kidney biopsy of secondary RAAA cases can be risk factors for rapidly progressive renal failure [2].

A case report by Mir TH, et al. has admitted that COVID-19-triggered AA amyloidosis can cause rapidly progressive renal failure and it's prudent to follow severe COVID-19 survivors. Many chronic viral infections like hepatitis B, C, and HIV have been associated with AA amyloidosis [4].

A case report by Anupama YJ, et al. named "Rapidly progressive glomerulonephritis in a patient with renal amyloidosis: Case report and review of the literature" concluded that crescentic glomerulonephritis associated with renal amyloidosis is a rare occurrence but it's important to suspect this possibility when the patient has rapid worsening of renal functions in the background of renal amyloidosis [5].

Crosthwaite A, et al. have reported a case of 67 years 67-year-old man with a background of IgGκ [kappa] multiple myeloma and primary amyloidosis who developed deranged renal functions. A renal biopsy was done which revealed crescentic glomerulonephritis and amyloid deposition. This patient progressed to end-stage renal disease. Thus here AL amyloidosis was an underlying cause [6].

Muñiz-Pacios L, et al. reported a case of a 64-year-old male who had a loss of 17 kg, asthaenia, anorexia and anemia. A gastroscopy was performed along with a biopsy which revealed a neoplastic proliferation with a solid pattern suggestive of GIST. AA amyloid deposits were observed in biopsies of the gastric mucosa and the tumor. The patient subsequently showed lower limb edema and diarrhea. The lab showed: hemoglobin 8.7 g/dl, serum creatinine 1.3 mg/dl, albumin 1.36 g/dl, proteinuria 5.1 g/day and sediment with 4-6 red blood cells/high power field. Given the poor condition of the patient, renal biopsy was not performed and it was assumed that amyloidosis previously observed in gastrointestinal biopsies was responsible for the nephrotic syndrome and diarrhea. The renal function continued to deteriorate and finally, he was initiated on hemodialysis in this case; secondary amyloidosis because of GIST was accountable for rapidly progressive renal failure [7].

Costa Mylene, et al. reported a case of a 79-year-old woman; a known case of Sjogren syndrome who presented with rapidly progressive renal failure and complaints of asthenia, anorexia, and generalized edema. She presented with acute renal failure (creatinine was 6.0 mg/dl; it had been 0.99 mg/dl 1 year before admission and 1.67 mg/dl 1 month previously). Clinical results showed the patient met the diagnostic criteria for primary SS, and neoplastic hematological disease was excluded. Renal biopsy revealed Congo red-positive glomerular deposits thus proving diagnosis of AA amyloidosis [8].

Kukuy Olga L, et al. conducted a study named "Amyloid storm: acute kidney injury and massive proteinuria, rapidly progressing to end-stage kidney disease in AA amyloidosis of familial Mediterranean fever". They described an acute condition, termed 'amyloid storm', manifesting with a rapid (≤ 2 weeks) increase in serum creatinine and urine protein, which has never been characterized in FMF amyloidosis [9].

Conclusion

In a case of rapidly progressing renal failure; amyloidosis could be an underlying etiology of clinical deterioration. Cases where there is proteinuria and deranged kidney function test; amyloidosis can be included as a differential. Although it is a well-known complication of chronic inflammatory disease only a few studies have evaluated the natural history, prognostic markers, and treatment of AA amyloidosis. Sometimes etiologies of secondary amyloidosis such as tuberculosis, rheumatic disease, and neoplasm may remain underinvestigated or underdiagnosed; unless clinically significant findings are present. With multiple case reports; amyloidosis causing rapidly progressing renal failure is always a possibility. The role of biopsy can never be underestimated.

Ethical declaration

This case report was planned after obtaining informed patient consent (in the language the patient understands) for the publication of the data and with his free will to participate in the case report.

The identity of the patient is confidential

Institutional review board no. was not assigned as it was a case report.

This article has been reviewed by all the authors.

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