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Case Report

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"Amyloid Storm" As a Rare Etiology of Acute Kidney Injury: Case Series and Review of Literature

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Introduction

"Amyloid storm" is a recently defined hyper inflammatory condition that causes acute kidney injury. Criteria of "amyloid storm are: Increase of creatinine and proteinuria levels at least two levels and increase of CRP values to at least ten times compared to highest normal level in less than two weeks [1]. Even though it was first reported in patients with AA amyloidosis secondary to Familial Mediterranean Fever (FMF), it has also been described in patients with clinical conditions other than FMF [2]. We aimed to describe the clinical features of our patients who presented with amyloid storm.

Cases

The clinical features of our 3 amyloid storm patients are summarized in Table 1.

	Patient 1	Patient 2	Patient 3
Age	33	47	63
Sex	Male	Male	Female
Creatinine at presentation	7.03 mg/dl	12.66 mg/dl	7.99 mg/dl
Proteinuria at presentation	12868 mg/day	5568 mg/day	10766 mg/day
CRP at presentation	78.2 mg/l	209 mg/l	206.3 mg/l
Procalcitonin at presentation	0.56 µg/l	1.81 µg/l	0.53 µg/l
Basal creatinine	0.94 mg/dl	1 mg/dl	1.12 mg/dl
Basal proteinuria	2+ (not quantified)	342 mg/day	1+ (not quantified)
Condition that triggered amyloid storm	Pneumonia	Pneumonia	Lymphadenitis in the right inguinal lymph node
Renal biopsy	AA amyloidosis in glomerular mesangium, tubulus basement memebrane, interstitium and vessel wall	AA amyloidosis in glomerular, mesangium, walls of arterioles and arteries	AA amyloidosis in mesangial nodules, arterioles and capillary walls
Other organs with biopsy proven AA amyloidosis	Stomach, duodenum and colon	-	Bone marrow
Pre-existing rheumatic disease	None	Ankylosing spondylitis	None
Treatment for the rheumatic disease	-	Sulfasalazine, NSAIDs	-
MEFV mutation status	Negative	Negative	Negative
Final diagnosis	Idiopathic AA amiloidosis	AA amyloidosis secondary to ankylosing spondylitis	AA amyloidosis secondary tosystemic inflammatory disease (Unspecified granulomatous disease)
Treatment initiated	Colchicine (0.5 mg/day) + Canakinumab (150 mg /month)	Colchicine (0.5 mg/day) + Adalimumab (40 mg/ 2 weeks)	Colchicine (0.5 mg/day)*
Current status (in terms of survival and renal function)	Alive, continues hemodialysis three times a week, preparing for renal transplantation	Alive, continues hemodialysis three times a week, preparing for renal transplantation	Exitus due to septic shock while on hemodialysis three times a week
Final CRP	5.6 mg/l	6.9 mg/l	269.2 mg/l*
*Patient 3 succumbed to septic shock before Anakinra could be initiated			

Table 1: Clinical features of the patients with amyloid storm.

Discussion

Amyloidosis constitutes the etiology of chronic kidney disease in 0.83 % of Turkish patients undergoing routine hemodialysis [3]. Recently, clinicians have begun to encounter patients that present with acute kidney injury and are subsequently diagnosed with amyloidosis, which was defined as "amyloid storm" [1].

FMF amyloidosis patients with "amyloid storm" were younger and associated with higher risk for progression to end stage renal disease (ESRD) and mortality [1]. Two of our patients were younger than 50. All of our patients progressed to ESRD. After one-year follow-up, one patient died due to septic shock.

Cases of amyloid storm were also identified in patients without FMF. Bektaş and colleagues reported 9 patients fulfilling the definition of "amyloid storm". Five patients had FMF. Two patients had ankylosing spondylitis, 1 patient had amyloidosis secondary to non-FMF auto inflammatory disease, 1 patient had idiopathic AA amyloidosis [2]. None of our 3 patients had clinical symptoms of FMF and MEFV mutations of our patients were negative. Patient 2 has ankylosing spondylitis. Patient 1 and Patient 3 did not have a rheumatological diagnosis. Patient 1 did not have any other findings, therefore we considered him to have "idiopathic" AA amyloidosis. Patient 3 did not have a rheumatological diagnosis. However, her past medical history was notable for the lung biopsy performed 6 months ago, which revealed granulomatous inflammation which was negative for tuberculosis. Additionally, during admission, findings suggestive of systemic granulomatous disease (dermopathic lymphadenopathy with histiocytic infiltration in the inguinal lymph node biopsy and reactive plasmacytosis in the bone marrow biopsy) were detected. However, patient died before a definite diagnosis could be reached.

The most common trigger for amyloid storm was infections [1,2]. Similar to the literature all of our patients presented with infections.

Treatment of the amyloid storm is supportive [1]. All of our patients received supportive treatment and colchicine. Patient 1 and patient 2 received biologic agents (canakinumab and adalimumab respectively) and even though we planned to treat patient 3 with anakinra, her clinical condition deteriorated and she died. CRP of the first two patients decreased to close to normal levels after initialisation of biological agents.

In conclusion, we present three more non-FMF patients that have clinical and renal histopathological findings compatible with amyloid storm triggered by infections and all became hemodialysis dependant.

Informed Consent: Informed consent was obtained from the two patients that are still alive and from the family of the third patient who passed away.

Conflict of Interest: None

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