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A PECULIAR CASE OF NEPHROTIC STORM WITH ACUTE KIDNEY INJURY PRESENTING AS AMYLOIDOSIS IN A YOUNG FEMALE

General Medicine

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ABSTRACT

Renal amyloidosis, particularly secondary amyloidosis (AA amyloidosis), is a rare glomerular disease, typically arises in the context of chronic inflammatory conditions or infections. We present a unique and novel case of renal amyloidosis in a previously healthy 30-year-old female, demonstrating an acute kidney injury (AKI) complicated by nephrotic syndrome. The patient developed rapidly progressive renal failure without classic systemic manifestations of amyloidosis, challenging initial diagnostic approaches. Renal amyloidosis, a complication of chronic inflammatory or infectious diseases, often presents as nephrotic syndrome and progressive renal dysfunction. The kidney is the most commonly affected organ in amyloidosis, and a high index of suspicion is necessary for early diagnosis. This case highlights the rare association of intramuscular injections leading to localized chronic inflammation and subsequent amyloid deposition in a young, otherwise healthy individual.

KEYWORDS

Amyloidosis, Nephrotic Syndrome, Acute Kidney Injury (AKI), Renal Amyloidosis, Secondary Amyloidosis, Proteinuria, Arteriovenous Fistula (AVF).

INTRODUCTION

Amyloidosis refers to a group of disorders characterized by the extracellular deposition of amyloid proteins in tissues, leading to organ dysfunction[1]. Renal amyloidosis is most commonly seen in the form of nephrotic syndrome, characterized by massive proteinuria, hypoalbuminemia, edema, and progressive renal dysfunction. Secondary amyloidosis (AA amyloidosis), typically associated with chronic inflammatory diseases, is one of the most common types of renal amyloidosis[2,3]. In this report, we describe an unusual case of secondary amyloidosis in a 30-year-old female without classic systemic manifestations or chronic inflammatory diseases, highlighting the need for increased awareness and diagnostic diligence in such cases.

Case Study

A 30-year-old female, mother of two children, with no pre-existing medical conditions, presented with a history of rapidly progressing pedal edema, which evolved into anasarca over 6-7 days. Her serum creatinine levels rose from 2.6 mg/dL to 9.6 mg/dL within this period. Additionally, she developed oliguria, which progressed to anuria. Upon detailed history elaboration, the patient revealed history of a gluteal abscess, which had been present for one month. This abscess had been surgically drained at an outside primary facility. Given the clinical presentation, sepsis secondary to the abscess was suspected, and empirical antibiotics were started. Haemodialysis was undertaken every alternate day to manage the volume overload associated with worsening renal failure.

Day	On Admission	4th	7th HD +	12th HD+	15th HD+	17th
S. Creatinine	4.7	8.3	9.6	8.0	4.8	3.8
WBC	13500	14000	14500	10000	11000	10000

Laboratory investigations showed an elevated serum creatinine level, initially 4.7 mg/dL, which increased to 9.7 mg/dL within a week. Urine routine showed albumin (+2), and the urine protein-to-creatinine ratio was markedly elevated at 22,270 mg/g. Hemoglobin was 9.3 g/dL, white blood cell count was 13,500/µL, and platelet count was 3.04 lakhs/µL. Complement levels were notable for low C3 (47 mg/dL) and normal C4 (13.94 mg/dL). Ultrasound of the abdomen and pelvis showed bulky kidneys with preserved corticomedullary differentiation. The patient was also negative for any significant infections on pus culture and sensitivity. Considering the low complement levels, an autoimmune cause for glomerulonephritis was initially suspected. Her ANA was negative. However, despite negative cultures for infection, the patient's condition continued to deteriorate.

Renal Biopsy was performed, which revealed 14 glomeruli showing global sclerosis with organized deposits in each glomerulus which were weakly PAS-positive and strongly positive for Congo red staining(Fig 1), confirming the diagnosis of secondary amyloidosis. The tubules showed features of acute tubular injury, including focal flattening of tubular epithelium and necrotic debris, suggesting acute kidney injury secondary to amyloid deposits.

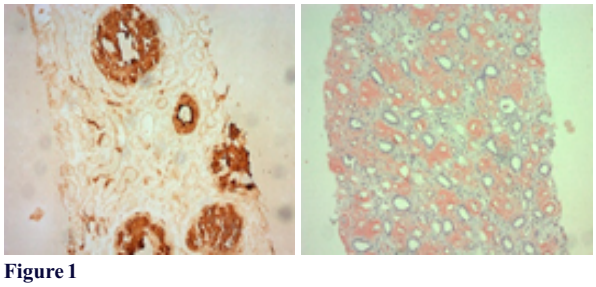

Figure 1

Upon further questioning, the patient recalled receiving an intramuscular injection for back pain at the site of the gluteal abscess before the abscess developed. This event raised the possibility of chronic local inflammation as the underlying trigger for amyloid deposition.

DISCUSSION

Amyloidosis is a rare cause of nephrotic syndrome and AKI, often presenting in the setting of chronic infections, inflammatory diseases, or familial conditions such as Familial Mediterranean Fever (FMF)[3]. In secondary amyloidosis (AA amyloidosis), the deposition of serum amyloid A (SAA) protein occurs as a consequence of prolonged inflammatory stimuli. The kidney is the most common organ affected, and renal involvement often leads to nephrotic syndrome, AKI, or chronic kidney disease[4].

Our patient, a young woman with no prior chronic inflammatory diseases, presented with a rapid onset of nephrotic syndrome and AKI. Interestingly, she did not exhibit the typical systemic signs of amyloidosis such as macroglossia, periorbital purpura, or joint pains. The potential trigger in this case appeared to be localized chronic inflammation resulting from an intramuscular injection, which is a rarely recognized etiology for secondary amyloidosis.

Amyloid deposits can involve the glomeruli, tubules, interstitium, and vasculature of the kidney[5]. In this case, the glomeruli showed amyloid deposits with typical Congo red staining. Renal biopsy remains the gold standard for diagnosing renal amyloidosis, and Congo red

staining is critical for detecting amyloid fibrils.

Secondary amyloidosis, particularly when associated with chronic infections or inflammatory conditions, can progress rapidly to end-stage renal disease (ESRD)[6]. Management of the underlying condition is essential to prevent further amyloid deposition. In our patient, upon the diagnosis of amyloidosis, wound debridement, antibiotics and haemodialysis was continued for 2-3 weeks. She however remained anuric upon follow up and was therefore advised for AVF construction and required maintenance haemodialysis thrice weekly .She has no transplant prospects.

CONCLUSION

This case highlights the rare presentation of renal amyloidosis in a young, previously healthy individual without systemic inflammatory disease. The case underscores the importance of considering amyloidosis in the differential diagnosis of nephrotic syndrome and AKI, particularly when no obvious cause for renal dysfunction is evident. The diagnosis of amyloidosis should prompt further investigation to identify potential underlying triggers such as chronic inflammation or infection.

REFERENCES

1. Tan SY, Pepys MB. Amyloidosis. Histopathology. 1994 Nov;25(5):403-14. doi: 10.1111/j.1365-2559.1994.tb00001.x. PMID: 7868080.
2. Gurung, Reena et al. Renal Amyloidosis: Presentation, Diagnosis, and Management. The American Journal of Medicine, Volume 135, S38 - S43
3. Siligato R, Gembillo G, Calabrese V, Conti G, Santoro D. Amyloidosis and Glomerular Diseases in Familial Mediterranean Fever. Medicina (Kaunas). 2021 Oct 1;57(10):1049. doi: 10.3390/medicina57101049. PMID: 34684086; PMCID: PMC8541210.
4. Feitosa VA, Neves PDMM, Jorge LB, Noronha IL, Onuchic LF. Renal amyloidosis: a new time for a complete diagnosis. Braz J Med Biol Res. 2022 Oct 3;55:e12284. doi: 10.1590/1414-431X2022e12284. PMID: 36197414; PMCID: PMC9529046
5. Khalighi MA, Dean Wallace W, Palma-Diaz MF. Amyloid nephropathy. Clin Kidney J. 2014 Apr;7(2):97-106. doi: 10.1093/ckj/sfu021. Epub 2014 Mar 13. PMID: 25852856; PMCID: PMC4377792..
6. Scarpioni R, Ricardi M, Albertazzi V. Secondary amyloidosis in autoinflammatory diseases and the role of inflammation in renal damage. World J Nephrol. 2016 Jan 6;5(1):66-75. doi: 10.5527/wjn.v5.i1.66. PMID: 26788465; PMCID: PMC4707170.