Spontaneous Massive Bladder Hematoma in the Late Period After Kidney Transplantation in Patients with AA Amyloidosis: Two Cases

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ABSTRACT Massive bladder hematoma can occur following a trauma, neoplastic states, or as a longterm side effect of a pelvic irradiation therapy. It is reported, albeit rarely, in the patients with amyloidosis which can affect bladder and can manifest itself as hematoma. However, it hasn’t been yet reported in kidney transplant recipients. A 31 year-old male with chronic glomerulonephritis received kidney transplantation. He developed proteinuria and frequent episodes of lung infection because of bronchiectasis after seven years from transplantation. Duodenal biopsies showed AA-amyloidosis. Macroscopic hematuria appeared, and cystoscopy revealed hematoma and perforation of bladder wall. A 56 year-old male with secondary amyloidosis due to familial Mediterranean fever had kidney transplantation. After two years, cystoscopy was performed because of macroscopic hematuria and revealed large hematoma that completely filled the bladder. To our knowledge, these are the very first cases with AA amyloidosis who developed spontaneous bladder hematoma in kidney transplant recipients.

Keywords: Amyloidosis; hematoma; urinary bladder; familial Mediterranean fever; kidney transplantation

Systemic amyloidosis which is caused by the deposition of circulating misfolded proteins can occur due to plasma cell abnormalities [light chain (AL) or heavy chain (AH) amyloidosis], chronic inflammatory diseases (deposition of serum amyloid A, AA amyloidosis), in the presence of genetic mutations (deposition of transthyretin, ATTR amyloidosis) and in some other rare conditions. Massive bladder hematoma can occur following a trauma, neoplastic states, or as a longterm side effect of a pelvic irradiation therapy. It is also reported, albeit rarely, in patients with systemic primary (AL) or secondary (AA) amyloidosis which can affect the bladder and can manifest itself as macroscopic hematuria, bladder hematoma, or bladder perforation. We presented two cases of spontaneous massive bladder hematoma in the late period after the transplantation in kidney recipients with systemic AA amyloidosis.
were not any abnormality about coagulation parameters. Later on, he was admitted to emergency department with dizziness, lower urinary tract symptoms and fever. While the diagnostic work-up was still ongoing, he had sudden cardiac arrest, was resuscitated and admitted to intensive care unit with the diagnosis of urinary sepsis. Despite all efforts, he died because of urinary sepsis and heart failure.

The legal representatives of both of the patients have given their informed consents for these case reports.

**DISCUSSION**

Amyloid deposition may occur in the condition of an abnormal protein presence like light chains, in association with excess exposure to a normal protein like serum amyloid A (SAA) and for undiscovered reasons increased with age.\(^1\) The clinical presentation of amyloidoses depends upon the affected organs that include mainly the kidney, heart, nervous system, liver, and gastrointestinal and urinary tracts. Secondary amyloidosis occurs as a consequence of long-standing inflammatory diseases that lead to overproduction of serum amyloid A in the liver. Rheumatoid arthritis, familial Mediterranean fever (FMF), ankylosing spondylitis and Chron’s disease are commonly associated with AA amyloidosis.\(^4\)

Secondary amyloidosis tends to accumulate within the vascular structures which cause amyloid angiopathy with increased fragility in blood vessels and impaired vasoconstriction which may explain why secondary amyloidosis of bladder has a high potential of massive hematuria and why renal biopsy in amyloidosis is known to be a hazardous procedure because of increased risks of bleeding.\(^5,6\) Although it is seen rarely, bladder amyloidosis mainly presents itself with painless macroscopic hematuria. Additionally, secondary bladder amyloidosis has a lower incidence than primary bladder amyloidosis.\(^7\)

In renal transplant recipients, macroscopic hematuria occurs with the prevalence of 12% after transplantation mostly because of infections, surgical complications, malignancies, graft rejections, recurrences of primary disease and calculus formation.\(^8\) Although there is a case of secondary bladder amy-
loidosis that presented itself with macroscopic hematuria in an FMF patient treated with renal transplantation, bladder hematoma or perforation without an obvious reason like trauma, clotting disorder, history of radiation or surgery is an extreme case and, to our knowledge, these are the very first cases with AA amyloidosis in renal transplant recipients. Although we had no chance to confirm bladder amyloidosis by biopsy, in both of these cases, there weren’t any other reasons about the patients to be prone to bleeding except from amyloidosis.

The control of underlying inflammatory disease is the main strategy for the treatment. Amyloidosis resulting from bronchiectasis has become rarer with the development of antimicrobial agents, but in immunosuppressive patients like renal transplantation recipients, it can be a problematic situation just like in our first case. Continuous colchicine use is recommended to prevent the involvement of other organs, even for end-stage renal disease patients with FMF, so its importance is greater in renal transplantation to prevent graft kidney amyloidosis.4

**CONCLUSION**

We presented two cases of AA amyloidosis involving bladder caused hematoma and perforation, one of which being after renal transplantation. Both patients died because of the systemic involvements of amyloidosis. We believe that, in kidney recipients with systemic amyloidosis, bladder hematoma should be considered as an adverse prognostic factor.

**REFERENCES**

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