Amyloidosis and acute hemorrhage of the kidney, ureter, and bladder

Dany N. Hanna, DO, Jason A. Levy, DO, Jonah S. Marshall, MD
Department of Urology, Hahnemann University Hospital/Drexel University College of Medicine, Philadelphia, Pennsylvania, USA

HANNA DN, LEVY JA, MARSHALL JS. Amyloidosis and acute hemorrhage of the kidney, ureter, and bladder. *Can J Urol* 2017;24(4):8934-8936.

Gross hematuria is a common occurrence in adults. The differential diagnosis is extensive, including: malignancy, trauma, inflammation of the urinary tract, and stones. While, urinary tract amyloidosis represents only a small percentage of causative gross hematuria, it is concerning because of its superficial resemblance to malignant processes. We report the case of an 82-year-old male with concurrent primary amyloidosis of the kidney, ureter and bladder in the setting of acute hemorrhage. Histopathological examination of several biopsied samples confirmed our diagnosis. A nephroureterectomy with bladder cuff was successfully performed without complication along with watchful waiting for the bladder amyloidosis.

Key Words: nephroureterectomy, bladder, kidney, amyloidosis, hemorrhage

Introduction

Amyloidosis refers to a disease process in which insoluble fibrils are deposited into extracellular tissue.[1] Insoluble fibrils originate from soluble precursor proteins. Polypeptide fibrils undergo conformational change at least one point in their life. While this remains an intracellular process, once out in the extracellular environment, these proteins become subject to conformational change from various destabilizing agents.[2,3] Primary amyloidosis develops due to abnormal immune cell disorders such as Waldenström's macroglobulinemia and multiple myeloma. Secondary amyloidosis is a manifestation of chronic inflammatory diseases.[4]

Accepted for publication May 2017

Address correspondence to Dr. Dany N. Hanna, Department of Urology, Hahnemann University Hospital, Broad & Vine Streets, MS 300 – Hospital Administration, Philadelphia, PA 19102 USA

However, primary and localized amyloidosis of the urinary tract is a rare disease, with only approximately 160-200 cases reported in the literature.[5,6] We report the case of an 82-year-old male with concurrent primary amyloidosis of the kidney, ureter, and bladder in the setting of acute hemorrhage.

Case report

An 82-year-old male with a past medical history significant for idiopathic pulmonary fibrosis, chronic obstructive pulmonary disease, prostate cancer status post radiation therapy, and low grade superficial bladder carcinoma presented to the emergency room with complaints of gross hematuria. He admitted to an episode of gross hematuria approximately 1 year prior but did not seek medical attention as the symptoms resolved spontaneously. However, the patient was subsequently admitted to the hospital secondary to urinary clot formation and urinary output obstruction. He was placed on continuous bladder irrigation (CBI) for presumed radiation cystitis. After 2 days of persistent bleeding the patient underwent cystoscopy

which demonstrated diffuse erythema of the bladder wall with active bleeding from the right ureteral orifice. An attempt to pass a guide-wire into the right ureter failed secondary to resistance with a solid mass obstructing the path of the distal ureter. A CT urogram showed hydronephrosis of the right kidney, Figure 1. Retrograde pyelography demonstrated ureteronephrosis of the right distal ureter with a goblet sign lesion. Right ureteroscopy demonstrated a large, solid round mass with a papillary nature. The ureteral wall was inflamed and hyperemic. Gross inspection raised concern for a mass that appeared to be a high grade malignancy and was biopsied. Biopsy specimens were also taken from the bladder, Figure 2.

Histopathological analysis of the specimens demonstrated amyloidosis of the right ureter and bladder. In the bladder, amyloidosis can usually be found along the posterior and lateral walls.[7] Postoperatively, intraurethral hemorrhage continued despite alum administration to the bladder and subsequent amicar bladder irrigation. Bleeding was presumed to come from the ureter and right kidney. A nephrostomy tube was placed and confirmed upper tract hemorrhage. Three days later antegrade ureteroscopy showed a smooth, irregular, solid mass in the renal pelvis without the classic appearance of transitional cell carcinoma. The renal pelvis mass was biopsied and demonstrated amyolidosis.

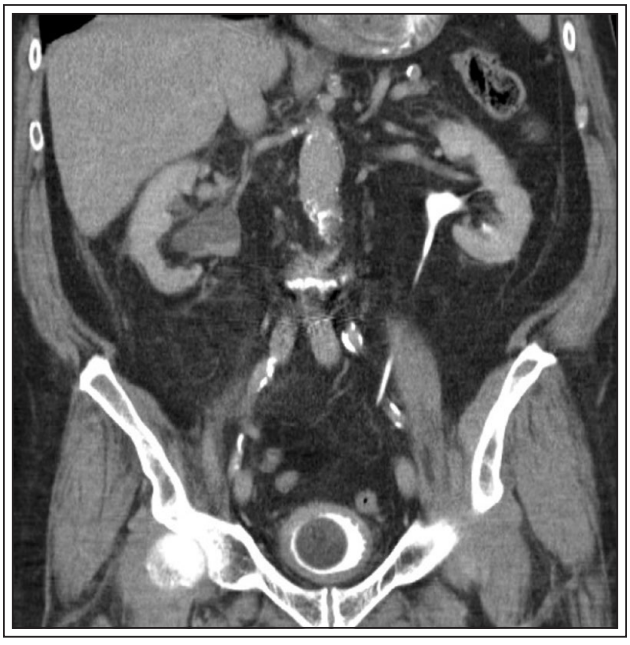

Figure 1. CT of the abdomen and pelvis. A coronal reformatted image from CT of the abdomen and pelvis shows hydronephrosis of the right kidney and adequate drainage of the left. The bladder is decompressed around a foley catheter balloon.

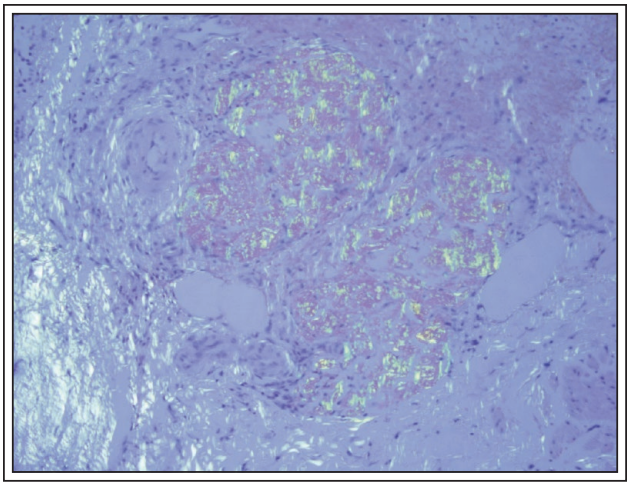

Figure 2. Bladder biopsy. A bladder specimen viewed under polarized light shows classic apple green birefringence with Congo-red stain.

The patient was ultimately diagnosed with amyloidosis of the kidney, ureter, and bladder with gross hemorrhage; which, could not be controlled with conservative measures. Bleeding from the kidney continued despite despite aminocaproic acid irrigation through a nephroureteral stent. The patient was ultimately transfused with three units of packed red blood cells. Definitive treatment via a right robotic-assisted laparoscopic nephroureterectomy with bladder cuff was performed. Final pathologic specimens confirmed the diagnosis of primary amyloidosis of the kidney and ureter. Postoperatively he was stable with some persistent gross hematuria. After several days of CBI the hematuria resolved. The patient was admitted to rehabilitation, where he remained for almost 3 weeks due to multiple comorbidities. Prior to discharge, the patient developed acute onset hematuria and was taken to the OR for clot evacuation and fulguration of bleeding lesions. After discharge the patient has remained disease free 1 year. Our intention was to perform cystoscopy with retrograde pyelogram every 3 months for the first 2 years, every 6 months for years 3-5, and annually for years 5-10.

Discussion

While primary amyloidosis in the urinary system is rare, simultaneous amyloidosis of the kidney, ureter and bladder is an exceptionally rare diagnosis.

Amyloidosis is a generic term used to describe the deposition of insoluble proteins in extracellular tissue

leading to a potentially pathologic state.[1] While it is thought that the amyloid proteins themselves do not have malignant potential, they become pathologic through their mass-effect capabilities.[8] Differentiating this disorder from malignant processes is imperative, especially in the setting of poor symptoms. Diagnosis cannot be definitively made with radiographic imaging alone. Instead, a biopsied tissue sample with Congo red staining is necessary for definitive diagnosis. The presence of amyloid fibrils will classically demonstrate an apple-green birefringence under polarized light.[9]

Once identified, a decision on disposition of the amyloidosis must be made. Treatment options revolve around several factors: size and location of amyloid deposition, surgical urgency, and comorbidities. Patients can be treated conservatively with intravesical dimethylsulfoxide or oral colchicine. Resection of smaller lesions can be performed using endocautery. For dramatic disease, removal of organs may be warranted. Although patients may respond well to nephroureterectomy, some may needlessly undergo the procedure because of its resemblance to malignancy.[10] Given our patient's obstruction, acute and persistent hemorrhage, and location in the renal pelvis and distal ureter, a decision was made to perform a nephroureterectomy. In our patient, it was found in the dome and right lateral wall. We decided to observe the bladder for further progression.

Conclusion

Concurrent primary amyloidosis of the kidney, ureter and bladder is an exceptionally rare presentation. Diagnostic work up must be exhaustive as its gross and radiographic appearance mimics urothelial malignancy. While amyloid proteins themselves may not have malignant potential, they appear to alter physiologic function through mass-effect. When necessary, surgical procedures such as nephroureterectomy may be indicated. □

References

1. Kumar V, Abbas AK, Aster JC, Fausto N. Robbins & Cotran Pathologic Basis of Disease, 8e. Saunders, 2009.
2. Bellotti V, Nuvolone M, Giorgetti S et al. The workings of the amyloid diseases. *Ann Med* 2007;39(3):200-207.
3. Eisenberg D, Jucker M. The amyloid state of proteins in human diseases. *Cell* 2012;148(6):1188-1203.
4. Lachmann HJ, Goodman HJ, Gilbertson JA et al. Natural history and outcome in systemic AA amyloidosis. *N Engl J Med* 2007;356(23):2361-2371.
5. Kobayashi T, Roberts J, Levine J, Degrado J. Primary bladder amyloidosis. *Intern Med* 2014;53(21):2511-2513.
6. Tolofari S, Ansari A, Knight RJ. A rare case of hematuria; primary amyloidosis of the bladder neck. *Urol Case Rep* 2016;10:48-50.
7. Auge BK, Haluszka MM. Primary amyloidosis of the bladder. *J Urol* 2000;163(6):1867-1868.
8. Chitale S, Morsey M, Peat D, Webb R. Amyloidosis of lower genitourinary tract: a review. *EAU-EBU Update Ser 5* 2007;70-76.
9. Ding X, Yan X, Ma X et al. Localized amyloidosis of the ureter: a case report and literature review. *Can Urol Assoc J* 2013;7(11-12): E764-E767.
10. Mark IR, Goodlad J, Lloyd-Davies RW. Localized amyloidosis of the genito-urinary tract. *J R Soc Med* 1995;88(6):320-324.