Acquired Hemophilia presenting as Gross hematuria following Kidney Stone – A case report and review of the literature

Max Schmidt-Bowman 1, Lael Reinstatler 2, Eric P. Raffin 2, Joseph E. Yared 2, John D. Seigne 2, Einar F. Sverrisson 2

1 Geisel School of Medicine at Dartmouth, Hanover, NH; 2 Department of Surgery, Section of Urology, Dartmouth Hitchcock Medical Center, Lebanon, NH

ABSTRACT

A rare condition in itself, acquired hemophilia A, seldom presents as isolated gross hematuria. It is a serious condition with a high mortality rate and thus clinical suspicion followed by prompt diagnosis is imperative (1). In fact, only 8 cases of such presentation of this condition have been reported thus far in the literature. Of these, none describe the initial presentation of hematuria with the inciting event of a kidney stone. We present a case of a 67-year-old man with signs and symptoms of nephrolithiasis accompanied by profuse hematuria, who was subsequently found to have developed expression of factor VIII inhibitor leading to acquired hemophilia A.

CASE REPORT

A 67-year-old man was transferred to our hospital, a large academic medical center, for work-up of persistent gross hematuria. The patient first presented to his local hospital eight days prior with the complaint of gross hematuria and left flank pain. A computed tomography (CT) scan was obtained, revealing an obstructing 2-3 mm left ureteropelvic junction (UPJ) stone and associated hydronephrosis. Physical exam also showed a fever of 102°F. Urologic evaluation was not available so he was subsequently transferred to a different outside hospital for management, where he was admitted to the medical intensive care unit (MICU) and taken to the operating room (OR) for ureteral stent placement. A retrograde pyelogram was normal without filling defects, however urine from his left ureteral orifice was noted to be bloody and there was a hydronephrotic drip reported. A postoperative abdominal and pelvic CT with and without contrast showed clot in the left collecting system and hydronephrosis with a 3 mm lower pole stone. His hematuria persisted, though his vital signs and he-
Acquired hemophilia presenting as gross hematuria is not altogether unheard of, as demonstrated by two case reports in the literature. Shander et al. describe an 82-year-old male who presented with 8 days of gross hematuria. On further work up, he was determined to have acquired hemophilia A with subsequent spontaneous hemorrhage of a previously identified renal cyst (5). Similarly, Hosier et al. report the case of a 54-year-old female who presented with gross hematuria and was later diagnosed with acquired hemophilia A. In this case, however, the inciting cause of bleeding was not identified and it was assumed to be secondary to spontaneous hemorrhage (6). A third case describes gross hematuria in a 60-year-old male with congenital hemophilia A rather than acquired hemophilia A, however the case is notable because the genitourinary hemorrhage was so profound that a cystectomy with ileal conduit was eventually required (7). Urolithiasis has not previously been reported as the inciting cause of hematuria in acquired hemophilia A, as was the case in the patient described above.
Various agents have been used to acutely treat this disorder with success mostly related to the severity of the bleed. Patients with low auto- antibody titers may be treated with human factor VIII concentrate and DDAVP (1-deamino-8-D-arginine vasopressin) whereas patients with high titers are usually treated with aPCC (FEIBA) (8). In their review of fourteen patients with acquired hemophilia over 8 years, Holme et al. describe their success using FEIBA, NovoSeven, corticosteroids, and/or cyclophosphamide. Two of their patients had no response to treatment, six had a partial response, and the remainder were successfully treated (1). Importantly, they noted that some of the cohort required procedures and these patients were effectively treated with FEIBA peri-operatively without any bleeding complications. In a larger retrospective review, Lak et al described their experience with 34 patients diagnosed with acquired hemophilia A. Over 79% of their patients experienced complete response to treatment. The median time of treatment was 4 months. Seven of their patients required surgical procedures and all were effectively treated without any intraoperative bleeding complications. Two of their patients died, one from bleeding and one from their underlying disease (2).

CONCLUSION

Acquired hemophilia A is a rare bleeding disorder that may present as gross hematuria, with or without an inciting factor such as urolithiasis. This diagnosis should be suspected in the setting of an isolated prolonged PTT. Prompt recognition and treatment can be lifesaving.

CONFLICT OF INTEREST

None declared.

REFERENCES

1. Afshar-Oromieh A, Avtzi E, Giesel FL, Holland-Letz T, Linhart HG, Eder M, et al. The diagnostic value of PET/CT imaging with the (68)Ga-labelled PSMA ligand HBED-CC in the diagnosis of recurrent prostate cancer. Eur J Nucl Med Mol Imaging. 2015;42:197-209.

2. Afshar-Oromieh A, Haberkorn U, Schlemmer HP, Fenchel M, Eder M, Eisenhut M, et al. Comparison of PET/CT and PET/ MRI hybrid systems using a 68Ga-labelled PSMA ligand for the diagnosis of recurrent prostate cancer: initial experience. Eur J Nucl Med Mol Imaging. 2014;41:887-97.

3. Palier CJ, Antonarakis ES. Management of biochemically recurrent prostate cancer after local therapy: evolving standards of care and new directions. Clin Adv Hematol Oncol. 2013;11:14-23.

4. Freedland SJ, Humphreys EB, Mangold LA, Eisenberger M, Dorey FJ, Walsh PC, et al. Risk of prostate cancer-specific mortality following biochemical recurrence after radical prostatectomy. JAMA. 2005;294:433-9.

5. Mussi TC, Garcia RG, Queiroz MR, Lemos GC, Baroni RH. Prostate cancer detection using multiparametric 3 - tesla MRI and fusion biopsy: preliminar results. Int Braz J Urol. 2016;42:897-905.

6. Lee DJ, Recabal P, Sjoberg DD, Thong A, Lee JK, Eastham JA, et al. Comparative Effectiveness of Targeted Prostate Biopsy Using Magnetic Resonance Imaging Ultrasound Fusion Software and Visual Targeting: a Prospective Study. J Urol. 2016;196:697-702.

7. Mariotti GC, Costa DN, Pedrosa I, Falsarella PM, Martins T, Roehrborn CG, et al. Magnetic resonance/transrectal ultrasound fusion biopsy of the prostate compared to systematic 12-core biopsy for the diagnosis and characterization of prostate cancer: multi-institutional retrospective analysis of 389 patients. Urol Oncol. 2016;34:416.e9-416.

8. Muthigi A, George AK, Sidana A, Kongnyuy M, Simon R, Moreno V, et al. Missing the Mark: Prostate Cancer Upgrading by Systematic Biopsy over Magnetic Resonance Imaging/Transrectal Ultrasound Fusion Biopsy. J Urol. 2017;197:327-334.

Correspondence address:
Lael Reinstatler, MD
Department of Surgery, Section of Urology
Dartmouth Hitchcock Medical Center
1 Medical Center Drive
Lebanon, NH 03756
Fax: +603 650-4985
E-mail: lael.s.reinstatler@hitchcock.org