A rare presentation of alkaptonuria: Extensive prostatic calculi with highlight of stones found in a unique paraprostatic urethral diverticulum

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ABSTRACT

INTRODUCTION: We present a case of Alkaptonuria (AKU) presented with severe lower urinary tract symptoms (LUTS) secondary to extensive prostatic calculi deposited in the para prostatic diverticulum. Prostatic calculi are seen in chronic prostatitis, chronic pelvic pain syndrome and benign prostate hyperplasia; however, in patients with AKU, prostatic calculi and/or calcifications are more extensive. A para prostatic diverticulum is a rare entity in males; however, it should be considered in patients with AKU based on this case report.

DIAGNOSIS, THERAPEUTIC INTERVENTIONS, AND OUTCOMES: A patient with AKU presented with extensive prostatic calculi that were deposited in para prostatic diverticulum, urethra and urinary bladder. The stones were successfully managed endoscopically.

DISCUSSION: Paraprostatic diverticula or urethral diverticula are rare entities. They may be congenital or acquired due to recurrent Urinary Tract Infections (UTIs) or distal urethral obstruction. The distal obstruction of the urethra secondary to stones was the most likely etiology of the paraprostatic diverticulum in our case. The possible mechanisms behind stone formation in our case were chronic stasis and urinary infection within a urethral diverticulum proximal to the urethral obstruction.

CONCLUSION: This case sums up a rare case of AKU and paraprostatic diverticular stones; in addition, it highlights the role endoscopic management of prostatolithiasis in AKU patient.

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1. Introduction

Alkaptonuria (AKU) is rare inherited genetic disorder with late complications such as arthritis and ochronosis [1]. Patients with AKU excretes alkapton (homogentisic acid) in the urine due to the congenital lack of the enzyme: homogentisate 1,2-dioxygenase, which mediates the essential step in the catabolism of phenylalanine and tyrosine [1]. When the urine excretion of homogentisic acid is very high, urine turns dark upon standing or alkalinization as a result of forming polymerized products of Alkapton. The presence of Alkapton in the urine has a high lithogenic effect [1].

PKU has a very low prevalence (1:100,000–250,000) in most ethnic groups [2]. The prevalence of AKU is unknown in Jordan; however, in a study done in 2009 in a single village in southern region of Jordan, forty cases with AKU were identified among 17 screened families with history of AKU, the study concluded that prevalence of AKU among Jordanian is likely to be higher than the worldwide rates due to high rates of consanguineous marriages [3].

Prostatic calculi are common, with 99% of asymptomatic adult men noted to have some degree of prostatic calcification at the time of autopsy, regardless of age [4].

Although prostatic calculi are seen in chronic prostatitis, chronic pelvic pain syndrome and benign prostate hyperplasia, none of these have prostatic calculi or calcification as extensive as in AKU [5].

We present a case of severe lower urinary tract symptoms (LUTS) caused by extensive prostatic calculi that were deposited in a para prostatic diverticulum, the urethra and the urinary bladder.

2. Case presentation

A known case of hypertension, asthma and AKU presented to Urology clinic complaining of severe LUTS: obstructive symptoms including poor stream, straining, hesitancy, and incomplete emptying of urinary bladder, in addition to irritative symptoms including frequency, urgency and nocturia. These symptoms
started 2 months prior to his current presentation with a history of passing stones during urination and acute urinary retention that was relieved by suprapubic catheterization and cystolitholapaxy for his urinary bladder stones. He also reports blackish discoloration of his urine and history of recurrent urinary tract infections (UTIs).

On examination, a urethral stone was felt in the shaft of the penis during palpation, bladder was not full. By digital rectal examination, the prostate was hard with indentation mostly representing palpable stones in the prostatic urethra and para prostatic diverticulum. Other extra genitourinary manifestations due to AKU disease were apparent; including the presence of thoracic lumbar back pain for the past 3 years with an incidental finding of advanced changes of AKU seen in the spine (Fig. 1A, KUB), short stature, and black discoloration of the nose and ear cartilages.

Laboratory investigations revealed a normal complete blood count, kidney function tests, serum parathyroid hormone, uric acid, calcium, and prostate specific antigen (PSA). The only positive finding was the presence of red blood cells and white blood cells in his urine analysis; however, his urine culture results came back negative.

A Kidney, Ureter and Bladder plain film (KUB) and unenhanced urinary tract computed tomography (CT) were done with results as shown in (Fig. 1A) and (Fig. 1B + C) respectively.

Two operative sessions were done to completely clear his stones in the urethra, paraprostatic diverticulum and bladder. During the first operation, cystoscopy revealed multiple urethral, urinary bladder, and left para prostatic diverticulum black stones. His urethral stones were pushed back up to urinary bladder and cystolitholapaxy were used to eliminate urinary bladder stones. A 3-way urethral catheter was inserted into urinary bladder. Another unenhanced urinary tract CT (Fig. 2A + B) and pelvic magnetic resonance imaging (MRI) (Fig. 2C + D) were taken postoperatively.

Few days after his first operation, the patient developed pulmonary embolism and started on an anticoagulant (enoxaparin), then the patient course was complicated by clot retention, so suprapubic catheter was inserted and patient was kept on continuous irrigation.

Repeat cystoscopy revealed urethral and left paraprostatic diverticulum with multiple black stones that were then fragmented using a pneumatic lithotripter, and evacuated by inserting a 3-ways Foley’s catheter. An unenhanced urinary tract CT (Fig. 3) was taken for comparison with the his baseline CT prior to operation (Fig. 1B + C).

The weight of the stones was 78 g, and chemical calculus analysis showed black stones with a composition of 20% uric acids and 80% calcium oxalate.
After few days from the last cystoscopy procedure, Foley’s catheter was removed followed by suprapubic catheter removal, after that, patient was passing urine with good stream and discharged home. The patient has been visiting outpatient clinic with a regular follow up visits without any bothersome urinary symptoms.

3. Discussion

Prostatolithiasis in AKU is of two types; true prostatolithiasis with deposition of calculi in the prostatic gland and false prostatolithiasis with the location of calculi in the pars prostatica urethra [6]. In our case the false prostatolithiasis type was found. Paraprostatic diverticula or urethral diverticula are rare entities and may be congenital or acquired due to recurrent UTI or distal urethral obstruction that causes outpouching of the urethral wall as a true diverticulum [7]. The paraprostatic diverticulum in our case was secondary to the distal obstruction of the urethra by stones. MRI plays an important role in the diagnosis of urethral diverticulum [8]; however, the stones are not easily identified on MRI.

Most of the urethral calculi are reported in developing countries, owing to the higher prevalence of bladder calculi, having migrated to the posterior urethra [9]. Rarely a calculus may form primarily within the urethra when a stricture is present, or it may form in a pouch or a diverticulum that opens into the urethra [10].

The possible mechanism behind stone formation in our case is chronic stasis and urinary infection within a urethral diverticulum and proximal to a urethral obstruction that lead to the formation of urethral diverticular stones [11,12].

Extensive prostatic calculi of variable sizes with stones in the para prostatic diverticulum are very rare in routine clinical practice and literature. Most of them were treated by surgery. We report a case of extensive and variable sized stones in the para prostatic diverticulum, urethra and urinary bladder in a patient with AKU that were successfully treated via an endoscopic approach. Calculi were fragmented with the pneumatic lithotripter and by pushing the stones back to the bladder to fragment the stones per a cystolitholapaxy approach via a transurethral route. The patient was stone-free as illustrated in the non-contrast postoperative urinary tract CT (Fig. 3) when compared with his pre-op CT (Fig. 1B + C).

This case also highlights the role for endoscopic management of prostatolithiasis in AKU patient. Our work has been reported in line with the SCARE criteria [13].

Conflicts of interest

The authors declare that no conflicts of interest exist.

Funding

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Ethical approval

Since this is just a case report and patient’s confidentiality was assured, there was no need for ethical approval based on our institution regulations.

Consent

“Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request”.

Author contribution

Husam MF. Masoud1: He was a part in the primary inpatient managing urology team, also participated in data collection, data analysis and interpretation, and writing the paper.

Hussam H. Alhawari: He was consulted as an endocrinologist to help with inpatient management in regards to Alkaptonuria. He also participated in writing and reviewing the paper.

Nosihab T. Alryalat: She was consulted as a radiologist.

Muayyad M. Murshidi: Participated in data collection, data analysis and interpretation, and writing the paper.

Mujalli M. Murshidi: He was the consultant urologist within the inpatient managing urology team, also participated in data collection, data analysis and interpretation, and writing the paper.

Registration of research studies

Not registered since this is just a case report.

Guarantor

All above authors accept full responsibility for the work and/or the conduct of the study, had access to the data, and controlled the decision to publish.

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