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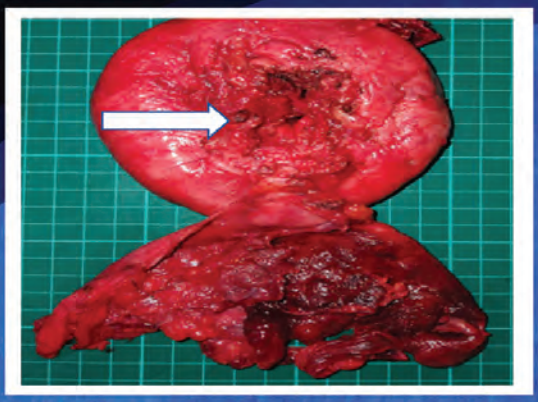
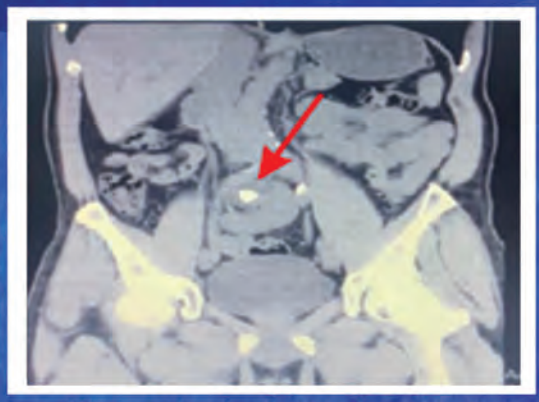


Bladder mass showing rupture point



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ORIGINAL RESEARCH

Minimally Invasive Therapeutic Modalities for Pediatric Urolithiasis: A Single Center Experience from the Philippines

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Introduction: There is a limited data on the incidence of pediatric urolithiasis in the Philippines as well as studies on how it is managed. With the advent of endourology, there is already an evident shift from open stone surgery to minimally invasive surgery as first line in the treatment of pediatric urolithiasis. A tertiary referral center in the country caters to most pediatric urolithiasis cases in the country and offers the different minimally invasive modalities outlined in the latest guidelines. This study aimed to review the different minimally invasive therapeutic modalities for pediatric urolithiasis and its outcomes in the past ten years in a tertiary referral center in the Philippines.
Methods: This retrospective study was conducted through charts review of all pediatric patients (0-18 years) who underwent stone surgery from January 2012 to December 2022. Demographic and clinical data, stone characteristics, types of stone surgery, and outcomes were obtained.
Results: A total of 87 pediatric patients (54 boys, 33 girls) with 105 urolithiases were analyzed. The patients had a mean age of 13 years, with an interquartile range (IQR) spanning from 6 to 18 years. Among them, 62.07% were male, while 37.93% were female. The most prevalent symptom reported was flank pain, affecting 41.38% of the patients, followed by urinary tract infections (UTI) (11.49%) and gross hematuria (8.05%). For the management, 82.86% of the patients underwent minimally invasive stone procedures with extracorporeal shockwave lithotripsy (ESWL) as the most common procedure (44.76%) and ureterolithiases as the most common stone (43.81%). On the other hand, 17.14% of the subjects underwent open stone surgery done mostly on cystolithiases and large ureterolithiases (38.89% each). All patients recovered and were discharged post operatively.
Conclusion: The institution practices minimally invasive approaches in the majority of pediatric patients with urolithiasis. Open stone surgery is reserved for patient with large complex stones and those with anatomic abnormalities.

Key words: children, extracorporeal shockwave lithotripsy, percutaneous nephrolithotomy, ureteroscopy, endourology, open stone surgery

Introduction

The incidence of pediatric urolithiasis has been increasing globally over the last decades.[1-7] Children represent around 2-3% of all cases of urolithiasis with boys in the first decade of age are the most commonly affected and adolescent girls showed the greatest increase in incidence.[1-3] It occurs generally between age 5 and 15 years with marked increase in incidence between 10 and 19 years of age.[4] True incidence of pediatric urolithiasis, however, remains unclear due to shortage of epidemiological studies.[2,4]

Philippines is part of the Afro-Asian stone belt which ranges from Sudan, the Arab Republic of Egypt, Saudi Arabia, the United Arab Emirates, the

Islamic Republic of Iran, Pakistan, India, Myanmar, Thailand, and Indonesia to the Philippines.[1,5] In this area, urolithiasis affects all age groups with prevalence of stone disease ranging from 4-20%.[5] Pediatric urolithiasis remains endemic in low resource countries which may be attributed to hot climate, poor nutrition and diarrheal diseases.[6]

Aside from geographical and socioeconomic factors, most of the children with urolithiasis have an underlying metabolic or infectious etiology.[3,7] Recurrence is high if full metabolic work up is not done to give adequate treatment.[7] Hypercalciuria and hypocitrauria are the most common metabolic disorders noted.[1-2,6-8] Other metabolic abnormalities include hyperuricosuria, and hyperoxaluria as well as stones due to infectious etiology and genitourinary anatomical abnormalities (ureteropelvic junction obstruction and vesicoureteral reflux) provides additional risk factors.[9] Inheritance for autosomal recessive disorders such as primary hyperoxaluria and cystinuria as well as changes in dietary habits such including increase in protein consumption can also be attributed for stone formation.[1,4-5]

Along with the increase in incidence of urolithiasis in the pediatric population there is also a rise in the minimally invasive procedures available for treatment.[9-10] Current internationally-accepted guidelines including guidelines from American Urologic Association (AUA) and European Urologic Association (EAU) include minimally invasive procedures as first-line treatment of both nephrolithiasis and ureterolithiasis in children.[11-12] Minimally invasive procedures include extracorporeal shockwave lithotripsy (ESWL), endourologic procedures including ureteroscopy (URS), retrograde intrarenal surgery (RIRS) and percutaneous nephrolithotomy (PCNL).[11-12] Minimally invasive surgery is the treatment of choice for pediatric stones and it provides success with low complication rates.[10,13-17] Traditional open stone surgery has been reserved for selected complex cases in which minimally invasive modalities are not available or applicable.[14,16-17]

Pediatric urolithiasis has been one of the common problems in a tertiary referral center in the Philippines, however, no data has been published regarding its epidemiology as well as the management and outcomes of the different urologic interventions in the country. The study intends to summarize the demographic and clinical data of pediatric patients (0-18 years if age) with urolithiasis, their stone characteristics and the different available therapeutic modalities the institution has offered over the past ten years, highlighting the minimally invasive approaches for pediatric stone cases and their outcomes.

Methods

This is a single-center, retrospective study from the year 2012 to 2022. All patients age 0 - 18 years admitted in the wards, emergency room and outpatient department at a tertiary government medical center who underwent any of the following procedures below for urolithiasis from January 1, 2012 to December 31, 2022 were included in the study.

- Cystoscopic Procedures
- Ureteroscopy (URS)
- Extracorporeal Shockwave Lithotripsy (ESWL)
- Retrograde Intrarenal Surgery (RIRS)
- Percutaneous or Mini-Percutaneous Nephrolithotomy (PCNL)
- Laparoscopic Surgery
- Open Stone Surgery

All patients who underwent medical treatment and those who underwent any of these procedures for reasons not involving urolithiasis were excluded from the study.

Data were collected through retrospective chart review in the Medical Records Section for admitted/emergency room cases, Operating Room Records for outpatient cases and Endoscopy Unit for ESWL cases. The author reviewed charts of pediatric patients diagnosed with urolithiasis who underwent the different procedures listed above.

To summarize the general and clinical characteristics of the participants, descriptive statistics were employed. Nominal variables were evaluated using frequency and proportion, non-normally distributed interval/ratio variables were assessed using median and interquartile range, and normally distributed interval/ratio variables were evaluated using mean and standard deviation. Shapiro-Wilk test was used to determine normally

distributed interval/ratio variables. 95% confidence intervals were also computed for proportions. Missing variables were neither replaced nor estimated. Data analysis was conducted using R 4.2.2.

Results

A total of 87 pediatric patients with urolithiasis were analyzed (Table 1). The patients in this study had a mean age of 13 years, with an interquartile range (IQR) spanning from 6 to 18 years. Among them, 62.07% were male, while 37.93% were female. The most prevalent symptom reported was flank pain, affecting 41.38% of the patients, followed by urinary tract infections (UTI) (11.49%) and gross hematuria (8.05%). A significant majority of the patients (91.95%) did not have any associated comorbidities, while a minority presented with conditions like acute kidney injury (AKI) (4.6%) secondary from obstructing urolithiasis while others were diagnosed with distal renal tubular acidosis (RTA), global developmental delay, and seizure disorder, each accounting for 1.15% of the total.

Among the 71 patients, the median creatinine level was 0.70 with an interquartile range (IQR) of 0.40-0.95. For the urine culture results, majority of the patients have negative results after 48 to 72 hours (64.58%). Noteworthy bacterial findings from urine cultures included Acinetobacter baumanii, Klebsiella and Enterococcus spp. The remaining patients have insignificant bacterial culture results (20.83%). Almost a third of the total patients were able to obtain stone analysis test with results showing mostly mixed stone types (73.91%) with calcium oxalate as the predominant composition while 26.08% of the other patients each has either calcium oxalate or calcium phosphate stone composition.

The stone characteristics among pediatric patients with urolithiasis were analyzed (Table 2). A total of 105 different stone types based on location were obtained. Some patients had more than one stone type present or more than one stone type were treated in the 10-year period, making the total of 105 types of stone in 87 subjects. The average size of all the stones is 1.4 cm, 1.93 cm for the nephrolithiases, 1.22 cm for the ureterolithiases and 2.7 cm for the cystolithiases. No size was recorded for the urethral calculi.

In terms of location, nephrolithiasis was categorized into four types. Calyceal calculus was the most common and was seen in 14.29% of cases,

Table 1. Demographic and clinical profile of pediatric patients with urolithiasis (n=87).

	Frequency (%); Mean ± SD; Median (IQR)
Age, years	13 (6-18)
Sex	
Male	54 (62.07)
Female	33 (37.93)
Symptoms	
Flank pain	36 (41.38)
UTI	10 (11.49)
Gross hematuria	7 (8.05)
Abdominal pain	6 (6.90)
Dysuria	3 (3.45)
Difficulty urinating	3 (3.45)
Fever	3 (3.45)
Hypogastric pain	2 (2.30)
Vomiting	1 (1.15)
Iliac pain	1 (1.15)
Bipedal edema	1 (1.15)
Lower quadrant pain	1 (1.15)
Comorbidities	
AKI	4 (4.60)
Distal RTA	1 (1.15)
Global developmental delay	1 (1.15)
Seizure disorder	1 (1.15)
Not found	80 (91.95)
Creatinine [n=71]	0.70 (0.40-0.95)
Low	21 (29.58)
Normal	44 (61.97)
High	6 (8.45)
Urine culture (n=48)	
Polymicrobial	4 (8.33)
Acinetobacter baumanii	1 (2.08)
Klebsiella spp	1 (2.08)
Enterococcus spp	1 (2.08)
Negative	31 (64.58)
Insignificant findings	10 (20.83)
Stone analysis (n=23)	
Calcium oxalate	3 (13.04)
Calcium phosphate	3 (13.04)
Mixed stone	17 (73.91)

staghorn calculus in 12.38%, infundibulopelvic calculus in 6.67% and pelvolithiasis in 8.57% respectively. Ureterolithiasis, which is the most common of all the stones, was divided into three regions, showed the proximal third to be the most prevalent location at 20.95%, followed by distal third ureterolithiasis at 11.43%. Cystolithiasis, which includes encrusted double J stents (DJS), was present in 12.38% and lastly, urethral calculus was the least common, observed in 1.90% of the cases. Radiodensity, measured in Hounsfield units (HU), was recorded for 41 samples with the mean radiodensity of 801.30 ± 382.61 for all stones. The mean HU for the nephrolithiasis was 830.56 and 786.40 for the ureterolithiases. For the cystolithiasis, only one HU had been recorded which is 450 and none for urethral calculi. (Table 2).

Among the pediatric patients diagnosed with urolithiasis, various therapeutic procedures were employed to address their condition. Table 3 presents the proportion of urolithiasis cases operated using various therapeutic modalities and the distribution of stone types treated by each modality.

Table 2. Stone characteristics of pediatric patients with urolithiasis.

	Frequency (%); Mean ± SD; Median (IQR)
Size, cm [n=92]	1.40 (0.80-2.10)
Nephrolithiasis [n=45]	1.93
Ureterolithiasis [n=41]	1.22
Cystolithiasis [n=6]	2.7
Location	[n=105]
Nephrolithiasis [n=44]	
Staghorn calculus	13 (12.38)
Calyceal calculus	15 (14.29)
Infundibulopelvic calculus	7 (6.67)
Pelvolithiasis	9 (8.57)
Uteropelvic junction calculus	0
Ureterolithiasis [n=46]	
Proximal third	22 (20.95)
Middle third	5 (4.76)
Distal third	12 (11.43)
Uterovesical junction calculus	7 (6.67)
Cystolithiasis (including encrusted DJS) [n=13]	13 (12.38)
Urethral calculus [n=2]	2 (1.90)
Radiodensity, HU [n=41]	801.30 ± 382.61
Nephrolithiasis [n=18]	830.56
Ureterolithiasis [n=22]	786.40
Cystolithiasis [n=1]	450

For percutaneous nephrolithotomy (PCNL) which was done to 19 cases (includes 5 mini-PCNL), the predominant stone type addressed was staghorn calculus, constituting 52.63% of all PCNL cases. This was followed by pelvolithiasis at 26.32% and infundibulopelvic calculus at 21.05%. Extracorporeal shockwave lithotripsy (ESWL), conducted in 47 cases, was most used for treating calyceal calculus and the proximal third ureterolithiasis, each comprising 29.79% and 25.53%. Ureterovesical junction calculus and distal third ureterolithiasis made up 12.77% each of the ESWL cases. Middle third ureterolithiasis and infundibulopelvic calculus were the least common for ESWL, at 6.38% and 4.26%. Ureteroscopy, performed in 13 cases, was primarily employed for the proximal third ureterolithiasis, accounting for almost half of the ureteroscopy cases (46.15%). The distal and middle third ureterolithiases each made up 30.77% and 23.08%, respectively. For cystoscopy (including cystolithotripsy), conducted in 8 cases, the primary indication was cystolithiasis (including 4 encrusted DJS), accounting for 75% of the cases while urethral calculus was accounted for 25% of the cystoscopy cases. Lastly, open stone surgery, performed in 18 cases (17.14%), was most frequently used for cystolithiasis (including 1 encrusted DJS) with 38.89%. Proximal third ureterolithiasis and staghorn calculus each made up 22.22% and 16.67% of the open surgery cases. Calyceal calculus and ureterovesical junction calculus were the least common, both at 5.56%.

All 87 patients achieved recovery, denoting a recovery rate of 100% with a confidence interval (CI) of 95.77% to 100%. Similarly, the discharge rate for these patients was also 100% (CI: 95.77%-100%), indicating that all patients were successfully discharged post-treatment. (Table 4)

On the aspect of post-operative complications or conditions seen on Table 4, post operative fever/ febrile episodes were the most frequently observed symptom, presenting in 17.24% of patients (CI: 10.74%-26.52%). Fever may be attributed to different causes since most of these patients have negative blood and urine cultures post operatively

Table 3. Proportion of cases operated using the different modalities.

	Percutaneous nephrolithotomy (n=19)	**Extracorporeal shockwave lithotripsy (n=47)**	**Ureteroscopy (n=13)**	**Cystoscopy (n=8)**	**Open stone surgery (n=18)**
	Frequency (%, 95% CI)				
Nephrolithiasis					
Staghorn calculus	10 (52.63, 31.71-72.67)	0	0	0	3 (16.67, 5.84-39.22)
Calyceal calculus	0	14 (29.79, 18.65-43.98)	0	0	1 (5.56, 0.99-25.76)
Infundibulopelvic calculus	4 (21.05, 8.51-43.33)	3 (6.38, 2.19-17.16)	0	0	0
Pelvolithiasis	5 (26.32, 11.81-48.79)	4 (8.51, 3.36-19.93)	0	0	0
Uteropelvic junction calculus	0	0	0	0	0
Ureterolithiasis					
Proximal third	0	12 (25.53, 15.25-39.51)	6 (46.15, 23.21-70.86)	0	4 (22.22, 9-45.21)
Middle third	0	2 (4.26, 1.17-14.25)	3 (23.08, 8.18-50.26)	0	0
Distal third	0	6 (12.77, 5.98-25.17)	4 (30.77, 12.68-57.63)	0	2 (11.11, 3.10-32.80)
Uterovesical junction calculus	0	6 (12.77, 5.98-25.17)	0	0	1 (5.56, 0.99-25.76)
Cystolithiasis (including encrusted DJS)	0	0	0	6 (75, 40.93-92.85)	7 (38.89, 20.31-61.38)
Urethral calculus	0	0	0	2 (25, 7.15-59.07)	0

Table 4. Clinical outcomes of pediatric patients with urolithiasis.

	Frequency (%, 95% CI)
Recovered	87 (100, 95.77-100)
Discharged	87 (100, 95.77-100)
Fever/ febrile episodes	15 (17.24, 10.74-26.52)
Fluid collection	1 (1.15, 0.20-6.23)

upon investigation. Fluid collection, which could be an indicator of potential complications such as urinoma, was observed in one patient (1.15%) (CI: 0.20%-6.23%). This patient underwent ultrasound guided aspiration of the fluid collection likely urinoma and was discharged unremarkably.

Discussion

A total of 87 patients with 105 stone types (based on location) who underwent different stone procedures in a 10-year period were done and supervised mainly by three pediatric urology consultants in the institution. About two thirds of the subjects were males and one third were females with a mean age of 13 years. This is supported in other studies in which showed higher urolithiasis occurrence in males than in females with studies citing a M/F ratio of 1.3/1 to as high as 3.1/1.[1-2,7,18-19] Most common presentations were flank pain (41.38%) and recurrent urinary tract infection (11.49%). Symptomatology varies per age group, usually infants present with crying, irritability and vomiting in 40% of the cases while the classic renal colic and hematuria were more common in older age groups since ureterolithiases were more common in adolescents.[2,10,20] Majority of the patients have negative stone cultures which is a requirement for some of the minimally invasive procedure such as PCNL. Full metabolic workup, though it is recommended for stone disease in this age group, is not available in the institution. In the available data, only few subjects have co-morbidities and only 26.44% of the patients submitted for stone analysis. For the patients who submitted for stone analysis, 73.91% were mixed stones with calcium oxalate as the primary component. This is comparable to other retrospective studies done in other countries in which calcium oxalate is the most common stone component in the pediatric population.[6,16,19] Children with anatomic abnormalities, urinary tract infections and metabolic disturbances are known to have higher risk for stone recurrence.[21-22] A study on the metabolic abnormalities in pediatric Filipino patients is suggested.

Majority of the stones were managed via minimally invasive approach (82.86%) with ESWL as the most common procedure (44.76%). The average size of ureterolithiasis was 1.22 cm in largest diameter while nephrolithiasis has an average size of 1.93 cm. Both have an average HU of <1000. All of these features render both stone types amenable for minimally invasive approaches. With a high success rates in adults, minimally invasive procedures rapidly became the standard treatment for children with urolithiasis and since children have higher stone recurrence rates, minimally invasive procedures were favored in this population type instead of repeated open stone procedures.[9,17,21] With refinement in the technique, it offers short operative time, less exposure to anesthesia, excellent stone-free rates with minimal morbidity.[17,23]

No laparoscopic stone surgery was done in the institution in the 10-year period and its role has yet to be maximized by the institution for the years to come. Laparoscopic procedures will probably be used in patients with large stone burden in which it can reproduce what an open stone surgery can do using minimal access.[24-25] It has been proven to be safe and feasible in the pediatric population with high success and minimal complications.[24-25] No robot-assisted surgeries were also done in the institution as the institution has yet to acquire one, though, the indications are almost the same as for the laparoscopic surgery. Retrograde Intrarenal Surgery, on the other hand, was only introduced in the institution later in the year 2022. RIRS has been proven safe and effective in the pediatric population and is an emerging option for stones <2 cm.[26-27]

Only 17.14% underwent open stone surgery with the majority done on cystolithiasis and ureterolithiasis (38.89% each). Cystolithiases have a large average diameter of 2.7 cm which was deemed appropriate for an open stone surgery. On the other hand, open stone surgeries done on ureterolithiasis were mostly on those larger than 2 cm. Cystolithiases are endemic to some developing countries attributed to low protein diet, use of goat milk and dehydration.[28] For the management of lower tract calculi, minimally invasive approach such as laser cystolithotripsy is advisable for small stones (<1 cm) to avoid iatrogenic urethral stricture especially in the pediatric male population.[29] Percutaneous lithotripsy or open cystolithotomy is preferred for large bladder calculi.[23,29] For urethral calculi, cystoscopic procedures such as

laser lithotripsy or pushback of the calculus for posterior urethral calculus (to be followed by laser cystolithotripsy or cystolithotomy to extract or fragment the urethral stone) or manual extraction for anterior urethral calculus can be done.[30] For open stone surgeries involving nephrolithiasis, mostly were done on large staghorn calculi more than 4 cm and all of these were mostly done during the first four years of the study where minimally invasive approach such as PCNL is only starting to be practiced in the institution.

All patients were discharged stable after the procedures whether be an open or a minimally invasive procedure. Fifteen patients had fever post operatively (10 underwent minimally invasive approach while 5 underwent open stone surgery) which resolved with ambulation, hydration and for some, IV antibiotics , while 1 patient had fluid collection seen on ultrasound after an open stone procedure (extended open pyelolithotomy) which was managed via percutaneous aspiration and delayed removal of double J stent. Studies suggest indications for an open stone surgery include anatomic abnormalities (diverticular stones, presence of ureteropelvic junction obstruction), complex large stone burden which is not expected to have good results with minimally invasive approaches and repeated endourologic failure.[14,16-17]

Conclusion

The tertiary referral center in the Philippines caters to a number pediatric urology cases and referrals yearly including urolithiasis. Due to the advancement in technology and availability of specialized instruments, there is a rise of minimally invasive therapeutic modalities as standard approach to pediatric urolithiasis which are proven to be safe and effective in the pediatric population. The institution already practices minimally invasive approaches in the majority of the pediatric stone cases and open stone surgery is only reserved for selected cases such as large complex stones and those with concurrent anatomic abnormalities. There is still room for other minimally invasive advancements such as laparoscopic and robot assisted surgeries in the future.

Conflict of Interest

The authors declare no conflicts of interest in writing the study.

Ethical Considerations

The study was conducted after approval by the Institutional Review Board.

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ORIGINAL RESEARCH

The Intraoperative Anatomic Difference Between the Use of a Standard Cystoscope when Compared to Standard Operating Microscope as an Innovative Approach of Performing a Subinguinal Varicocelectomy with Intraoperative Vascular Doppler: Preliminary Result of a Novel Technique

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Objective: The varicocoele exist in approximately 35-40% of primary male factor infertility while two to 10 percent of cases presents with pain.[1] Most surgeons favor subinguinal microscopic varicocoelectomy because it offers superior improvement in semen parameters and reproductive outcome with the least complication rate. This study aimed to show an innovative surgical technique in the management of men with varicoceles.

Methods: Subinguinal varicocelectomies were performed by a single surgeon on all patients starting with a standard cystoscope stabilized by a customized mechanical holding system attached to the operating bed. All the presumed vascular channels, vas deferens and lymphatics were isolated and marked with vascular loops and surgical ties. After all the presumed vessels were tagged, the standard operating microscope was brought to the operative field and full microsurgical dissection was carried out.

Results: Ten varicocelectomies were performed on six men with a mean age of 30.5 years. 13 arteries, 84 veins, and 20 lymphatics were identified by the cystoscope while 18 arteries, 93 veins, and 29 lymphatics were identified by the standard operating microscope. Comparing the two modalities, 72%, 90%, and 69% of the arteries, veins, and lymphatics, respectively, were correctly identified by the cystoscope when compared to the latter.

Conclusion: Subinguinal varicocelectomy using a standard cystoscope could be offered as an alternative surgical approach in men with varicoceles as it can identify veins comparable with that of the standard operating microscope. In addition, a standard cystoscope can also identify, to some degree, lymphatics and arteries during surgical dissection. This innovative surgical technique can serve as a valuable option in the treatment of men with varicoceles.

Key words: varicocelectomy, microscope, cystoscope, veins, arteries, lymphatics

Introduction

Varicocele exists in approximately 15% of the general male population.[1] It is considered as the most common etiology of primary and secondary male factor infertility in 35% and 80% of cases, respectively.[2,3] However, its main pathophysiology remains largely undiscovered. When clinical palpable varicocele coexists with impaired semen quality, surgical repair may

potentially restore spermatogenesis and fertility. Both the European Association of Urology (EAU) and American Association of Urology (AUA) recommended surgical varicocelectomy in infertile men with palpable varicoceles and abnormal semen parameters.[4,5] Meanwhile, about 2% to 10% of patients with varicoceles present with pain and up to 80% of them will have improvement and/or resolution of their symptoms after varicocelectomy.[6]

To date, there have been limited randomized, controlled, prospective clinical studies that compare various techniques to determine the gold standard for the treatment of varicocele in infertile men. Consequently, the best surgical approach for varicocelectomy is still unknown. However, most surgeons favor subinguinal microsurgery.[7,8] In a randomized clinical trial comparing the different varicocelectomy techniques, subinguinal microsurgical varicocelectomy provided the best outcome in terms of semen parameters improvement and reproductive outcome with the least complication and recurrence rate.[9] Nearly half (43.2%) of clinicians (n= 574) are routinely using operating microscope according to a global practice survey conducted from 59 countries.[10] In a local survey among Filipino urologists, only 4.58% of respondents use microscope in performing varicocelectomy. This is mainly due to a lack of exposure to microsurgery training and due to a limited number of hospitals equipped with standard operating microscope thus limiting the practice of microsurgical varicocelectomy.

The main purpose of this study is to present an innovative approach of performing varicocelectomy using a standard cystoscope connected to a camera system that is more readily available and more affordable that may possibly serve as a valuable alternative option to a more expensive, less available standard operating microscope. In addition, it demonstrates the intraoperative anatomic difference during surgical dissection using a standard cystoscope compared to a standard operating microscope.

Methods

This study is a descriptive comparative observational study which was conducted in 2021-2022. The study protocol (REC-2020-61) was submitted to and approved by the Ethics Review Board of the hospital. All patients were provided with written informed consent.

Patients

A total enumeration of patients with varicoceles who met the inclusion/exclusion criteria. Inclusion Criteria (All Criteria): >18 years old, clinically-palpable varicoceles with abnormal semen parameters, intractable scrotal pain; Exclusion Criteria (At least one): Subclinical varicocele, recurrent varicocele, concomitant cryptorchidism) were included in the study. This study was conducted in a one-year time period from the time of approval in 2021.

Instruments

Cystoscope and a Camera System

A standard cystoscope (straight forward telescope 0°, diameter 4 mm, length 30 cm, autoclavable, fiber optic light transmission incorporated) was utilized in this study. A French 21 sheath connected to a telescope bridge with 1 lockable channel housed the cystoscope. The cystoscope was connected to the camera head of the existing laparoscopy tower with a 1080p resolution (Figure 1a). The video captured was transmitted in a full high-definition monitor. A fiber optic light cable (diameter 4.8 mm, length 300cm) with straight connector was attached to the light source. A customized autoclavable mechanical holding system mounted on the operating room bed rail held the cystoscope vertically (Figure 1b). This consists of a mechanical holding arm with the adjustable articulating stand and extension rod with clamp jaw to reach adjustment of the articulated stand with lateral clamp for height and angle adjustment of the articulated stand. This allowed the cystoscope to be optimally positioned thereby minimizing any interference during the surgical procedure. A clamping cylinder boarded the cystoscope and it allowed vertical movement and rotation of the cystoscope. An actual set up of the subinguinal varicocelectomy using a standard cystoscope is shown in Figure 1c.

Figure 1. (a) Cystoscope connected to the camera head of the existing laparoscopy tower with a 1080p resolution, (b) Customized autoclavable mechanical holding system attached on the operating room bed rail, (c) whole set up of subinguinal varicocelectomy using a standard cystoscope.

Techniques

The subinguinal microsurgical varicocelectomy technique was carried out to all patients who consented to be part of the study by a single surgeon. Approximately 2-cm incision over the external inguinal ring was made. The spermatic cord was identified and encircled with penrose drain or secured with a clamp. The standard cystoscope was utilized initially in all cases. All the presumed vascular channels, vas deferens and lymphatics were isolated and marked with vascular loops and surgical ties (white vascular loop: vas deferens, blue vascular loop: artery, black tie: veins and white tie: lymphatics). After all the presumed vessels were tagged, the standard operating microscope (Zeiss TIVATO 700 with advanced surgical visualization of 1080p or 4K camera and a large external monitor) was brought to the operative field (Figure 2).

A full microsurgical dissection was done, and appropriate tags were applied if there were additional structures identified. An intraoperative vascular doppler (8 MHZ Surgical Doppler System) was used to confirm the preservation of the arteries. To conclude the surgical technique, all cremasteric veins, external spermatic veins and internal spermatic veins were ligated while the vasal vein, arteries and lymphatics were preserved. Thereafter, approximation of the subcutaneous tissues and skin closure using subcuticular technique followed.

Figure 2. Standard operating microscope with 1080p or 4K camera used during the procedure.

Results

A total of six subjects with a mean age of 30.5 years were included in the study. Four of them had bilateral varicoceles while two had unilateral varicoceles which was equivalent to 10 varicocelectomies performed. All subjects had clinically significant grade 2-3 varicoceles. Sixty seven percent (67%) of the cases consulted due to male factor infertility while the remaining 33% was due to intractable pain.

Using the standard cystoscope, 13 arteries, 84 veins, and 20 lymphatics were identified. Upon use of the standard operation microscope, 18 arteries, 93 veins, and 29 lymphatics were identified. When comparing both findings, 72% of arteries, 90.3% of veins, and 69% of lymphatics were correctly identified by using the standard cystoscope.

Discussion

Varicocele is the abnormal dilatation and tortuosity of the pampiniform venous plexuses within the spermatic cord.[11] Testicular hyperthermia is a known pathologic reason causing the detrimental effect of varicocele on reproductive outcome. Although other primary proposed hypotheses were presented such as venous pressure, testicular blood flow, hormonal imbalance, toxic substances, and reactive oxygen species (ROS).[12] Response of the testis to heat stress, hypoxia and inflammation subsequently produce excessive amount of ROS.[13] This also inactivates enzymes and proteins necessary for spermatogenesis. Men with varicoceles can also present with scrotal pain. The probable mechanisms for this include compression of the surrounding neural fibers by the dilated venous complex, elevated testicular temperature, increased venous pressure, hypoxia, oxidative stress, hormonal imbalances, and the reflux of toxic metabolites of adrenal or renal origin.[6]

An ideal varicocelectomy procedure should have optimal results with minimal complications, such as varicocele recurrence or persistence, hydrocoele formation and testicular artery injury.[14] In a study conducted by Liu X et al (2013), injury of the spermatic artery remains 24% (12/50) in the loupe-assisted procedure, and 45.7% (32/70) using traditional open surgery.[15] The incidence of accidental testicular artery ligation in microsurgical varicocelectomy is approximately 1% after evaluating 2,102 cases of microsurgical varicocelectomy that was confirmed intraoperatively by observation of pulsatile twitching of the ligated vessel stump under 25x magnification.[16] In the present study using a standard cystoscope, the authors did not encounter any incidence of spermatic artery injury, and the identification rate for arteries was 72.22% (13/18). Only 9.67% (84/93) of spermatic veins were missed using the standard cystoscope which is lower than 22.9% (43/188) of spermatic veins missed using a magnifying loupe in a study conducted by Hao Zhang et al.[17] The study yielded a comparable result with the use of operating microscope in terms of identification of spermatic veins. According to a study by Richter et al (2001), hydrocoele formation is the most encountered complication at varicocelectomy by urologists and andrologists, reported in 40.4% of patients, in spite of the use of some form of optical magnification by 70% of surgeons.[18] Testicular edema, hydrocoele formation and reduced testicular function were reported to be due to the division of lymphatics at varicocelectomy.[19] In the present study, 68.97% (20/29) of lymphatics were correctly identified by the standard cystoscope. However, there was one structure identified as vein in the standard cystoscope but it turned out to be lymphatic vessel under the standard operating microscope. There was also one lymphatic vessel that was to be incorrectly ligated with a vein under the standard cystoscope but was identified separately under the standard operating microscope yielding a 6.89% (2/29) incorrect identification of lymphatics under standard cystoscope which is lower compared to a 14.3% (18/126) of incorrect identification in the loupe-assisted varicocelectomy conducted by Hao Zhang et al.[17] Present study findings showed that concomitant use of intraoperative vascular doppler during microsurgical varicocelectomy increased the accuracy of preservation of arteries although there was no added benefit in identification in the initial result. Comparing it to a study by Cocuzza et al (2010), they found out that the use of intraoperative vascular doppler allowed more arterial branches to be preserved, and more internal spermatic veins to be likely ligated.[21]

This study has its own limitations. Although all the varicocelectomies were performed by a single surgeon, the authors had a limited number of cases. In addition, the microsurgical technique using the standard operating microscope was always performed after the standard cystoscope approach. As a result, the accurate number of vessels including arteries, veins, and lymphatics that a standard operating microscope alone could identify could not be reported. The study only had 1 set of patients for comparison. A proper randomized controlled trial comparing the two approaches would be a better study design for this hypothesis. The need for customized mechanical holding system, video system and the ability to adjust or adapt the hand and eye coordination while looking at the monitor while performing the microsurgery are also important considerations in doing this innovative approach.

Overall, no difficulty was experienced with the use of standard cystoscope connected to a camera system held on an autoclavable mechanical holding system. The study will be further conducted to include more subjects and with separate randomized groups to enhance the power of the study.

Conclusion

Subinguinal varicocelectomy using a standard cystoscope could be offered as an alternative surgical approach in men with varicoceles as it can identify veins comparable with that of the standard operating microscope. In addition, a standard cystoscope can also identify, to some degree, lymphatics and arteries during surgical dissection. This innovative surgical technique can serve as a valuable option in the treatment of men with varicoceles.

Author Disclosure Statement

The present study is in no way related to any company or individual but solely an interest of innovative surgical management for varicoceles.

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ORIGINAL RESEARCH

Cancer Detection Rate of MRI Fusion-targeted and Systematic Prostate Biopsy Based on Urologist-performed MRI Reading and Contouring in a Government Tertiary Hospital

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Introduction: Prostate cancer is a significant health problem worldwide. Transrectal ultrasound guided biopsy has limitations in the detection of clinically significant disease, hence, new imaging including multiparametric MRI and MRI targeted biopsy is developed. In most centers, reading and contouring of the prostate and identification of significant lesions on MRI are performed by radiologists. In this institution, these steps are performed by a urologist.
Objective: To determine the clinically significant cancer detection rate in patients undergoing MRI fusion-targeted and random systematic prostate biopsy where MRI PIRADS scoring, identification of lesions and contouring are performed by a trained urologist in a Philippine tertiary hospital.
Methods: This is a cross-sectional study of patients who underwent MRI fusion prostate biopsy in the Philippine General Hospital (PGH) from June 2021 to June 2023. Clinically significant cancer (csCancer) detection rates were calculated for MRI fusion prostate biopsy, random systematic prostate biopsy, and PIRADS scoring. Concordance was also determined between PIRADS scores and histopathological results.
Results: Forty six (46) patients who underwent MRI fusion biopsy in PGH were included in the study, representing a total of 90 lesions identified by urologists using mpMRA with PIRADS scores of at least 3. Of the patients, 13 (14.4%) were diagnosed with csCancer, while a large proportion was diagnosed with benign prostatic tissue. The csCancer detection rate of MRI fusion biopsy was 28.3% (13/46) and 8.7% (4/46) for random biopsy. The csCancer detection rate was 11.1%, 14.6%, and 36.4% for PIRADS 3, 4, and 5, respectively.
Conclusion: The detection rate of clinically significant prostate cancer using MRI fusion-targeted prostate biopsy based on urologist-performed MRI reading and contouring was superior to random systematic approach. The positive predictive value of PIRADS scores when interpreted by urologists was lower compared to reported values in the literature and did not show concordance. This may reflect lowered thresholds for labeling prostate lesions as suspicious in urologists.

Key words: Prostate cancer, Cancer detection, MRI fusion prostate biopsy, PI-RADS

Introduction

Prostate cancer is a significant health problem worldwide, with an estimated 1.4 million cases and 375,000 deaths in 2020.[1] It is the second most frequent cancer diagnosis made. In the Philippines, prostate cancer is the third most common cancer among men, with an estimated 8,297 new cases and

3,283 deaths in 2020.[2] The incidence of prostate cancer in the Philippines has been increasing in recent years, likely due to the aging population and increased awareness and screening.[3]

Traditionally, the diagnosis of prostate cancer has relied on transrectal ultrasound-guided (TRUS) biopsy. However, TRUS biopsy has limitations in the accurate detection and characterization of prostate cancer, particularly in the detection of clinically significant disease.[4] This has led to the development of new imaging techniques, including multiparametric magnetic resonance imaging (mpMRI) and MRI-targeted biopsy. These have shown promise in the accurate detection and characterization of clinically-significant prostate cancer, defined as Gleason score ≥ 3+4.

MRI fusion prostate biopsy involves a reading and contouring of the prostate and suspicious lesions on MRI images and targeted biopsy of identified lesions. At the Philippine General Hospital, these steps are all performed by a urologist trained in an advanced course in reading of prostate MRI (MRI PRO at the Monash University). Additionally, random systematic sampling of the prostate is also performed to increase the rate of detecting cancer. MRI-targeted biopsy has shown excellent diagnostic performance in detecting clinically significant prostate cancer.[5] However, some studies have revealed that MRI-targeted prostate biopsy can miss the detection of clinically significant prostate cancer in 7-20% of cases that were detected by random systematic sampling.[6,7] However, differences in cancer detection rates between targeted and systematic biopsies in Filipino patients have not been reported.

PIRADS is a standardized system for the interpretation and reporting of prostate mpMRI, which is based on the likelihood of clinically significant prostate cancer.[8] PIRADS V2.1 scores range from 1 to 5, with a score of 1 indicating very low suspicion of clinically significant prostate cancer, while a score of 5 indicates very high suspicion. At the Philippine General Hospital, patients with PIRADS score of at least 3 are advised to undergo prostate biopsy. A meta-analysis of studies reported that the cancer detection rate or positive predictive values of PIRADS 1, 2, 3, 4, and 5 were 6%, 9%, 16%, 59%, and 85%, respectively.[9] In a study done in Filipinos that compared PIRADS scores with transperineal prostate sector biopsy, the cancer detection rate for PIRADS 3, 4, and 5 were 3.3%, 58.7%, and 94.7%, respectively.[10] A recent study in Filipinos showed fair concordance between PIRADS and Gleason scores when mpMRI are read and contoured by a radiologist, with cancer detection rates of 27.3%, 42.4%, 65.3%, and 83.8% for PIRADS 2, 3, 4, and 5, respectively. However, the predictive performance of PIRADS scores read by urologists in detecting clinically significant prostate cancer on MRI fusion-targeted biopsy in Filipinos has not been reported.

Reports of cancer detection rates of diagnostic procedures in Filipino patients with suspected prostate cancer are very few. Moreover, the diagnostic performance of these tests may vary across institutions and readers. Previous studies have demonstrated that contouring inter-reader discrepancies occur among radiologists and between radiologists and urologists.[11,12] There is a need to determine the diagnostic value of MRI fusion targeted biopsy, random systematic biopsy, and PIRADS performed by urologists at the Philippine General Hospital in order to make better clinical decisions from the results and also to prevent subjecting patients to unnecessary procedures which are not without risks.

Methods

This was a cross-sectional study of patients who have undergone MRI fusion prostate biopsy in the Philippine General Hospital from June 2021 to June 2023. This study was conducted among patients who have undergone MRI fusion prostate biopsy in the Philippine General Hospital from June 2021 to June 2023 where the MRI was read and interpreted by and the contouring and identification of lesions for biopsy by an advanced-prostate-MRI-reading-trained urologist (MRI PRO at Monash University),

Inclusion criteria

1. Patients who underwent MRI fusion prostate biopsy in the Philippine General Hospital from June 2021 to June 2023

2. MRI was read and interpreted by an advanced prostate MRI reading-trained urologist, and the contouring and identification of lesions for the targeted biopsy were performed by an advanced prostate MRI reading-trained urologist

Exclusion criteria

1. Cases with significant missing data (>50% of required information missing)
2. MRI was read and interpreted by a radiologist only
3. Contouring and identification of the lesions for the targeted biopsy was performed by a radiologist

The records and operations log of the Division of Urology were reviewed and searched for patients who have undergone MRI fusion prostate biopsy in the Philippine General Hospital from June 2021 to June 2023. Cases were screened and assessed for inclusion in the study based on the preset criteria. The PIRADS scores of the lesions, as interpreted by the trained-urologist, and the Surgical Pathology results were obtained and recorded from the patients' medical records.

The data collected included the following:

- Patient age
- PSA prior to surgery (in ng/mL) - latest level of prostate specific antigen (in ng/mL) taken at the nearest date prior to MRI-fusion prostate biopsy
- PIRADS Score – most recent PIRADS score of each identified lesion from preoperative multiparametric prostate MRI,as identified by a trained urologist
- Location and Zone of Identified Lesions on mpMRI – location and zone of the identified lesions in the multiparametric MRI
- Size and Number of Identified Lesions on mpMRI – greatest diameter of each of lesions identified and the number in the multiparametric prostate MRI results
- Prostate Size on mpMRI- prostate size determined on preoperative mpMRI as measured by the trained urologist
- Gleason Score – Gleason grading system score (expressed as Grade Group per International Society of Urologic Pathology) for each lesion
- Complete histopathological report - histopathological report for each prostate biopsy core sample, including presence of prostate cancer, cancer histology, Gleason score and other pathological findings (e.g., inflammation, hypertrophy, atrophy)

Continuous variables (age, size of lesion and prostate, PSA) were expressed as mean and standard deviation and categorical variables (location and zone of lesion, Gleason score, other histopathological findings) were expressed as counts and frequencies. The total number of clinically significant prostate cancer cases were the number of patients with Gleason score $\geq$ 3+4 upon either MRI fusion-targeted or random systematic prostate biopsy. Clinically significant cancer (csCancer) detection rate was calculated as follows:

$$\text{csCancer detection rate (targeted)} = \frac{\text{No. of patients w/ clinically significant prostate cancer (targeted)}}{\text{Total no. of patients biopsied}}$$

$$\text{csCancer detection rate (random systematic)} = \frac{\text{No. of patients w/ clinically significant prostate cancer (systematic)}}{\text{Total no. of patients biopsied}}$$

The performance of PIRADS read by urologists in predicting clinically significant prostate cancer was evaluated by comparing the most recent PIRADS score of each identified lesion on mpMRI to the histopathology result. The corresponding positive predictive value for PIRADS scores 3, 4, and 5 were calculated as:

$$\text{csCancer detection rate (PIRADS score)} = \frac{\text{No. of clinically significant prostate cancer lesions for a PIRADS score}}{\text{Total no. of lesions for a PIRADS score}}$$

This can also be referred to as the positive predictive value of each PIRADS score.

Kendall's τ_B (tau-b) was determined to evaluate whether there was congruence between PIRADS scores and Gleason scores because these outcomes were ordinal variables. For tests that showed statistical significance (p-value < 0.05), the relationship between PIRADS and Gleason scores was interpreted as insignificant, fair, moderate, good, and strong for Kendall's τ_B 0–0.20, 0.21–0.40, 0.41–0.60, 0.61–0.80, and 0.80–1, respectively.

The study protocol was approved by the University of the Philippines Research Ethics Board (UPM-REB) and was implemented in accordance to the principles of Good Clinical Practice and the Declaration of Helsinki.

Results

Between June 2021 to June 2023, forty six (46) patients underwent MRI fusion prostate biopsy in the Philippine General Hospital where MRI reading and contouring was performed by urologists. Their clinical, imaging, and pathological profiles are summarized in Table 1. The mean age of patients was 66.5 years, with median pre-biopsy PSA of 12.63 ng/mL and median prostate size of 43 grams. A total of 90 lesions were identified using mpMRI, with median lesion size of 11 mm. Patients were fairly distributed across PIRADS scores 3, 4, and 5 when considering the highest PIRADS score. At a lesion-level, more lesions were classified as PIRADS 4. Lesions were more frequently located in the midgland or apex and in the transition zone.

Of the lesions identified, 21 were histopathologically diagnosed as prostate cancer, with 4 lesions classified with Gleason score of 6 (3+3) and 17 lesions with a Gleason score of at least 7 (3+4). Thus, there were 13 patients diagnosed with csCancer (14.4%) and 2 non-csCancer. A majority of patients were histopathologically diagnosed with benign prostatic tissue.

The csCancer detection rate of MRI fusion-targeted prostate biopsy was 28.3% (13/46), while for random systematic prostate biopsy, the rate was 8.7% (4/46). It should be noted that all patients that were found to have csCancer were detected by MRI fusion-targeted prostate biopsy.

Comparison of urologist-interpreted PIRADS scores with MRI fusion prostate biopsy results

Table 1. Clinical, imaging, and pathological profile of patients who underwent MRI fusion prostate biopsy in the Philippine General Hospital from June 2021 to June 2023.

Parameter	Total (%)
Number of patients	46
Mean age in years (SD)	66.5 (10.2)
Median pre-biopsy PSA in ng/mL (range)	12.63 (4.2-185)
Median prostate size in grams (range)	43 (16-120.4)
Number of prostate lesions	90
Median lesion size in mm (range)	11 (2.6-36)
Patient-level PIRADS score (highest PIRADS)	
3	15 (32.6)
4	14 (30.4)
5	17 (37.0)
Lesion-level PIRADS score (highest PIRADS)	
3	27 (30.0)
4	41 (45.6)
5	22 (24.4)
Location of lesion	
Base	20 (22.2)
Midgland	30 (33.3)
Apex	31 (34.4)
Base to Midgland	2 (2.2)
Midglant to Apex	4 (4.4)
Base to Apex	3 (3.3)
Zone of lesion	
Central	5 (5.6)
Transition	44 (48.9)
Peripheral	37 (41.1)
Transition and Peripheral	4 (4.4)
Gleason scores of targeted lesions	
3+3=6	4
3+4=7	8
4+3=7	4
4+4=8	3
4+5=9	0
5+4=9	2
Diagnosis	
Clinically-significant prostate cancer	13 (14.4)
Non-clinically-significant prostate cancer	2 (2.2)
Atypical small acinar proliferation	1 (1.1)
Benign prostatic tissue	26 (28.9)
Benign prostatic tissue with chronic prostatitis	2 (2.2)
Prostatitis	2 (2.2)

Table 2. Comparison of urologist-interpreted PIRADS scores with MRI fusion prostate biopsy results.

PIRADS Score	Clinically significant prostate cancer on MRI fusion prostate biopsy pathology results			csCancer detection rate (PIRADS score)
	Positive	Negative	Total	
PIRADS 3	3	24	27	11.1%
PIRADS 4	6	35	41	14.6%
PIRADS 5	8	14	22	36.4%

are shown in Table 2. The csCancer detection rate (or positive predictive value) was 11.1%, 14.6%, and 36.4% for PIRADS 3, 4, and 5, respectively. Concordance between these measures was nominally fair, based on Kendall's τ_B of 0.221. However, this was not statistically significant (p=.16108).

Discussion

Detection of clinically significant disease is an important step in the diagnosis of prostate cancer. In addition to TRUS biopsy, PIRADS using mpMRI, MRI fusion targeted biopsy, and systematic random biopsy are options for diagnosing prostate cancer. While MRI modalities are typically performed by radiologists, urologists contour and interpret prostate MRI images in some centers, such as the Philippine General Hospital.

In the current study, MRI fusion-targeted prostate biopsy was able to detect all csCancer cases, showing better diagnostic performance than systematic biopsies. However, among targeted biopsies performed, only 28.3% were truly clinically significant. Moreover, the positive predictive values of PIRADS scores were lower compared to previous reports in the Filipino population.[13] These data suggest possible false positives in the labeling of prostate lesions as suspicious and assigning PIRADS scores when performed by urologists. This could reflect lower thresholds for targeted biopsies in urologists. More clinical experience and close follow-up of patients may be helpful in providing feedback to urologists who perform contouring and interpretation of prostate MRI.

Previous work showed mismatch between segmentations performed by urologists and radiologists.[12] Agreement between readings was positively correlated with lesion size, suggesting that this variable could be an important factor to consider when performing targeted biopsy. However, agreement among radiologists was also moderate, indicating challenges in performing these diagnostic procedures.

This study has several limitations, notably the small sample size and lack of comparison with radiologist readings. Future studies may include more patients as the institution gains more experience with performing targeted biopsies.

Conclusion

The detection rate of clinically-significant prostate cancer using MRI fusion-targeted prostate biopsy based on urologist-performed MRI reading and contouring was 28.3%, which was superior to random systematic approach. The positive predictive value when interpreted by urologists was 11.1%, 14.6%, and 36.4% for PIRADS 3, 4, and 5, respectively, but did not show concordance. This may reflect lowered thresholds for identifying prostate lesions as suspicious in urologists.

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CASE SERIES

En bloc Prostatectomy Combined with Abdominoperineal Resection on Locally Advanced Rectal Adenocarcinoma Involving the Prostate: A Case Series

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The genitourinary tract is known to be infiltrated in 3–10% of cases of advanced rectal adenocarcinoma. It is usually managed with total pelvic exenteration with urinary diversion. Complications are encountered because of this diversion. This case series aims to present four cases of locally advanced rectal adenocarcinoma involving the prostate who underwent en bloc prostatectomy in Jose Reyes Memorial Medical Center.

Methods: This case series consists of review of records of hospital charts, documentation of specimens from the database of the department.

Results: The study consists of four (4) male patients in their 50s presenting with primary rectal adenocarcinoma with invasion to the prostate, bladder or seminal vesicle. Two out of 4 patients had shorter hospital days (6 days) while 2 patients spent 14 and 28 days. Two out of 4 patients have anastomotic leakage, 1 had nosocomial pneumonia and no one had urinary tract infection, postoperatively. All 4 patients had a histopathologic diagnosis of rectal adenocarcinoma with a tumor size of 3.5 to 7.0 cm. Two patients are positive for lymphovascular space invasion while 1 patient has a positive line of resection margin. Two patients had neoadjuvant chemotherapy and radiotherapy while 1 patient had 1 adjuvant chemotherapy and radiotherapy. One patient was lost to follow-up.

Conclusion: En bloc prostatectomy combined with abdominoperineal resection to treat locally advanced rectal adenocarcinoma provides good local control with the risk of having less postoperative complications. Since there is only one stoma, there is less chance of infection and better quality of life. Complete resection of the tumor can be obtain but can also cause urologic morbidity even after chemotherapy and radiotherapy.

Key words: En bloc prostatectomy, abdominoperineal resection, rectal adenocarcinoma

Introduction

Colorectal cancer (CRC) was noted to be the third most common cancer in 2015. In 2018, Asia had the highest proportions of both incident (51.8%) and mortality (52.4%) CRC cases (all genders and ages) per 100,000 population in the world.[1]

In 2022, an estimated 106,180 cases of colon cancer and 44,850 cases of rectal cancer will be diagnosed in the US, and a total of 52,580 people will die from these cancers.[2]

During surgery for colorectal carcinoma, an infiltration of urological organs is found in 5–10% of patients and may rise to 50% when considering patients with locally advanced tumors (T4) at

presentation. Organs adherent to an advanced colorectal carcinoma must be resected to ensure en bloc removal of the mass. Due to the close anatomical relationship of colorectum and urological organs, the latter are at high risk for extirpation during surgery for advanced colorectal carcinoma.[3]

When there is bladder or the prostate infiltration, anterior pelvic exenteration is the best treatment that achieves the largest percentage of tumor free margins and the best 5-year survival. There are few cases of prostatic infiltration wherein the bladder can still be preserved and prostatectomy and abdominoperineal block resection are performed.[4] The study aims to determine the free resection margin, urologic complications, histopathologic findings and local recurrence on patients with locally advanced colorectal cancer who underwent en bloc prostatectomy and to discuss the management and outcomes of patients with locally advanced colorectal cancer.

The Cases

Case 1

Y.E., a 50 year-old male with no bowel movement for three days and occasional bilious vomiting. He is a known case of rectal adenocarcinoma stage IIC (pT4bN0M0) with invasion to prostate and left seminal vesicle. He underwent neoadjuvant short course of radiotherapy for 5 cycles and had 4 cycles of neoadjuvant FOLFOX. Patient underwent abdominoperineal resection en bloc resection, seminal vesicle and coccygectomy, en bloc prostatectomy after 4 months. Intraoperatively, noted a rectal mass 3 cm from anal verge with invasion of the prostate and seminal vesicle (Figure 1). Four days postoperatively, patient noted have increasing creatinine levels secondary to ischemic acute tubular necrosis. Cystogram was done 2 weeks postoperatively and noted extravasation of contrast and indwelling foley catheter was maintained. Repeat whole abdominal CT scan revealed fistulous tract on the posterior bladder wall and retroperitoneal space. Patient had hospital acquired pneumonia, 2 weeks postoperatively. Patient had percutaneous tube nephrostomy insertion, bilateral one month postoperatively. Patient was discharged after 28 days and had adjuvant chemotherapy for 4 cycles and no urinary symptoms on follow-up.

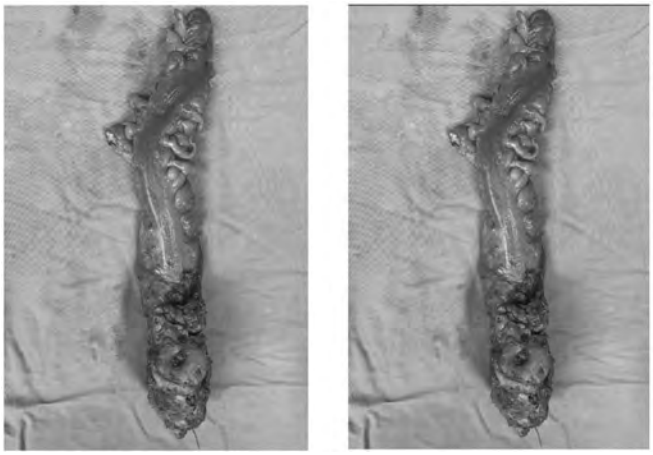

Figure 1. Rectal mass 3 cm from anal verge with invasion of the prostate and seminal vesicle.

Case 2

R.A., a 50-year old male with 1 year history of difficulty in defecation and anal pain associated with anorexia and weight loss. He consulted and subsequently underwent proctosigmoidoscopy with biopsy and colonoscopy which showed rectal adenocarcinoma. Patient underwent sigmoid loop colostomy followed by neoadjuvant chemoradiotherapy for 5 cycles with partial response. On follow-up after 8 months, pelvic MRI showed an eccentric short-segment, heterogeneously-enhancing, nodular wall thickening seen in right lateral aspect of the mucosal/submucosal layer of the lower rectum which is approximately 2.6 cm away from the anal verge. There are few prominent subcentimeter inguinal lymph nodes seen bilaterally. Prostate gland is not enlarged with a weight of 17.5 grams. Patient was referred intraoperatively because of a noted constricting mass at the middle rectum adherent to the prostate (Figure 2). Abdominoperineal resection with en bloc prostatectomy was done. Patient was sent home after 6 days and maintained on indwelling foley catheter for 2 weeks. Cystogram was done prior to removal of IFC, and noted no extravasation of contrast. Patient was has no urinary symptoms on follow-up.

Figure 2. Patient 2 was referred intraoperatively and noted a constricting mass at the middle rectum adherent to the prostate.

Case 3

P.D., a 52 year-old male came in due to prolapsed anal mass and rectal bleeding. Colonoscopy biopsy was done which showed rectal carcinoma with invasion to the bladder. Abdominoperineal resection with en bloc prostatectomy was done with intraoperative findings of rectal mass on the right anterolateral, 1 cm from anal verge up to midrectum with invasion to prostate gland (Figure 3). Patient was sent home after 6 days with unremarkable hospital stay. Patient was advised radiotherapy but was lost to follow-up. Noted leakage after 3 months postoperatively but resolved spontaneously after 1 year.

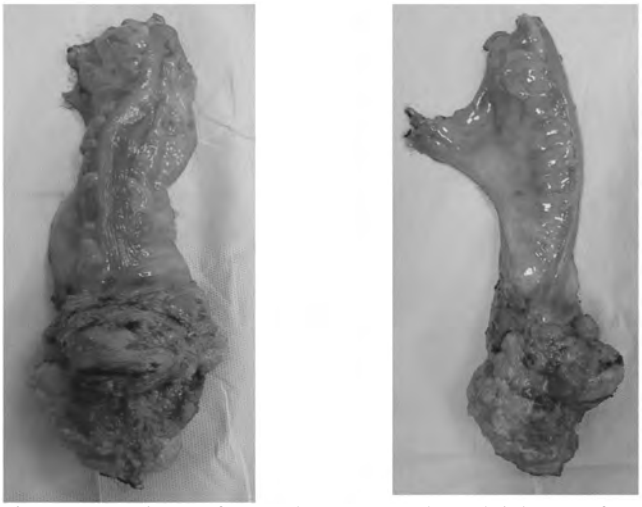

Figure 3. Specimen of a rectal mass anterolateral right 1 cm from anal verge up to midrectum with invasion to prostate gland.

Case 4

S.R., a 57-year-old male who came in due to six months history of hematochezia. Work-up was done which showed rectal adenocarcinoma with prostate involvement. Patient underwent abdominopelvic resection with en bloc prostatectomy and noted an annular, circumferential constricting mass with perianal and prostate involvement. Cystogram was done after 14 days and noted no extravasation of contrast. Patient was sent home after 14 days with no complications and no urinary symptoms. Patient had 13 cycles of adjuvant radiotherapy and 6 cycles of adjuvant chemotherapy.

Result

The study consists of four (4) male patients on their 50s presenting with primary rectal adenocarcinoma with invasion to the prostate, bladder or seminal vesicle (Table 1).

Table 1. Patient demographics.

Patient No.	Age	Tumor	Diagnosis
1	50	Primary	Rectal adenocarcinoma with invasion to the prostate and left seminal vesicle
2	50	Primary	Rectal adenocarcinoma with prostate invasion
3	52	Primary	Rectal carcinoma with invasion to the bladder
4	57	Primary	Rectal adenocarcinoma with prostate involvement

Postoperatively, 2 out of 4 patients have anastomotic leakage, 1 had nosocomial pneumonia, no one had urinary tract infection (Table 2). Patient number 3 noted leakage 3 months postoperatively. Two (2) out of 4 patients had short hospital days (6 days) while 2 patients spent 14 and 28 days.

Table 2. Postoperative complications.

Patient No.	Anastomotic Leakage	Urinary Infection	Hospital acquired Pneumonia	Days in the hospital
1	Yes	No	Yes	28 days
2	No	No	No	6 days
3	Yes	No	No	6 days
4	No	No	No	14 days

All 4 patients had a histopathologic diagnosis of rectal adenocarcinoma with a tumor size of 3.5 to 7.0 cm. One patient had no residual tumor. Two (2) patients are positive for lymphovascular space invasion while 1 patient has a positive line of resection margin (Table 3).

Two patients had neoadjuvant chemotherapy and radiotherapy while 1 patient had 1 adjuvant chemotherapy and radiotherapy. One patient was lost to follow-up.

One patient had liver metastasis on follow-up after 1 year, while 1 patient expired after 1 year post surgery.

Table 3. Histopathology.

Patient No.	Diagnosis	Tumor size	Lymphovascular Invasion	Margin	AJCC classification
1	Rectal adenocarcinoma, extending up to the prostate	3.5 cm	Not demonstrated	Circumferential resection margin, with the nearest measuring 0.2 cm	pT4bN0M0
2	Rectal adenocarcinoma	No residual tumor	Not demonstrated	Not applicable	
3	Rectal adenocarcinoma, well-differentiated, rectum	4.0 cm	Positive for lymphovascular space invasion	Negative all lines of resection	pT3N1aMx
4	Adenocarcinoma, well-differentiated, invading up to the peritoneum, rectum and extending up to the skin of the anus.	7.0 cm	Positive for lymphovascular space invasion	Positive for malignant cells: - Prostate -Lines of resection, prostate - 5 out of 16 (5/16) isolated paracolic lymph nodes	pT4bN2aMx

Table 4. Other management.

Patient No.	Radiotherapy	Chemotherapy
1	neoadjuvant short course of radiotherapy for 5 cycles	1. neoadjuvant chemotherapy x 4 cycles (FOLFOX) 2. adjuvant chemotherapy x 4 cycles
2	neoadjuvant radiotherapy x 5 cycles with partial response	neoadjuvant chemotherapy x 5 cycles with partial response
3	Lost to follow-up	Lost to follow-up
4	13 cycles of adjuvant radiotherapy	6 cycles of adjuvant chemotherapy

Table 5. Follow-up.

Patient No.	Recurrence	Metastasis
1	Yes	Yes (liver)
2	No	No
3	Lost to follow-up	Lost to follow-up
4	N/A	N/A

Discussion

Total pelvic exenteration is the standard approach in treating patients with locally advanced rectal adenocarcinoma with a staging of ≥ T3, involving the seminal vesicles, trigone of the urinary bladder and prostate.[5] It involves en bloc removal of the rectum, urinary bladder, distal ureters, and reproductive organs with curative intent, with negative surgical margins.[6] R0 margin is important for long-term survival.[7]

The creation of the ileal conduit stoma is important and if not done properly, can jeopardize the postoperative recovery by the constant leak of urine around an ill-fitting appliance.[8] This procedure results in double stomas and compromises quality of life. TPE results in chronic morbidity often secondary to urinary diversion.

Majority of recurrences will occur within 2 years after surgery and 90% by 5 years. If the patient exceeds 5 years after surgery without recurrence, the chances of developing a recurrence later become unlikely.[9] Recurrent disease detection based on history and physical examination alone varies from 15 to 40%.[10]

All of the patients in the studies are in their 50s who underwent abdominoperineal resection with en bloc prostatectomy. Ureteral injury and urinary leak after urinary diversion are the most common causes of morbidity and mortality in patients who have undergone TPE.[5]

Postoperatively, all four patients had cystogram after 2 weeks and patient number 1 noted extravasation of contrast which is managed conservatively with indwelling foley catheter. 1 patient had leakage 3 months postoperatively which resolved spontaneously. Only 1 patient had hospital acquired pneumonia. No patient had urinary tract infection.

In terms of being cost-effective, 2 patients had shorter hospital stay compared to the other 2 patients. Two (2) out of 4 patients had short hospital days (6 days) while 2 patients spent 14 and 28 days.

If adequate surgical margins can be achieved, without a total cystectomy, local and distal failure rates will not be lessen by cystectomy and urinary diversion.[5]

The tumor size ranges from of 3.5 to 7.0 cm and 1 patient had no residual tumor. Two (2) patients are positive for lymphovascular space invasion while 1 patient has a positive line of resection margin. One patient had liver metastasis follow-up after 1 year. One patient was advised adjuvant chemotherapy and radiotherapy but was lost to follow-up after surgery and that same patient expired after 1 year.

Conclusion

En bloc prostatectomy combined with abdominoperineal resection to treat locally advanced rectal adenocarcinoma provides good local control with the risk of having less postoperative complications. However, certain conditions like nosocomial infection will cause longer hospital stay. Since there is only one stoma, there is less chance of infection and better quality of life. Complete resection of the tumor can be obtained but can also cause urologic morbidity even after chemotherapy and radiotherapy. Even after the surgery, follow-up is important for the continuation of management because locally advanced rectal carcinoma requires a multi-disciplinary team in its treatment process.

Disclosure

No conflict of interest because the funding for this study came from the primary investigator.

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CASE REPORT

Ruptured Urachal Mucinous Cystic Tumor of Low Malignant Potential: A Case Report and Review of Literature

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Neoplasms of the urachus are uncommon, consisting of only 0.17% of all bladder malignancy. Mucinous cystic tumor of low malignant potential (MCLTMP) is a rare subtype with only 26 cases reported in the literature as of 2023. These tumors may present in a variety of ways such as hematuria, mucusuria, lower abdominal pain and irritative lower urinary tract symptoms. This is a case of 43-year-old female presenting at the emergency room for abdominal pain initially managed as a case of ovarian new growth in complication who underwent exploratory laparotomy, adhesiolysis, bilateral salpingectomy, partial cystectomy enbloc removal of urachal with anterior peritonectomy and excision of umbilicus. Histopathologic examination revealed mucinous cystic tumor of low malignant potential (MCLTMP) of the urachus. It is important to consider the possibility of a ruptured urachal cyst in a female patient who presents with hypogastric abdominal pain. A transabdominal and transvaginal ultrasound may lead to an incorrect diagnosis. In such cases where the patient presented with an acute abdomen, knowledge that a ruptured urachal cyst maybe a differential for such masses would lead to a strategic laparotomy incision aimed at a possible en-bloc removal of the umbilicus while maintaining the connections of the possible urachal mass to the urinary bladder.

Key words: neoplasms of the urachus, urachal tumors, mucinous cystic tumor of low malignancy potential (MCLTMP)

Introduction

Mucinous cystic tumor of low malignant potential (MCLTMP) is a rare subtype of the neoplasms of the urachus. As of 2023, there are only 26 case studies reported in the literature.[1] The case presented as a unique addition to the existing literature as it is the first documented instance of a urachal MCTLMP presenting as acute abdomen with a palpable right lower quadrant mass.

The Case

The patient is a 43-year-old female who presented with a 3-week history of mild hypogastric pain and enlarging abdomen. There were no reported lower urinary tract symptoms or bowel movement changes. She was prescribed pain medications that only temporarily relieved her symptoms. Two weeks after initial treatment, she was rushed to the emergency room due to severe hypogastric abdominal pain. She presented with

a 7cm x 5cm x 5cm palpable right lower quadrant mass with direct and rebound tenderness. Whole abdominal ultrasound revealed a 11.6cm x 10.8cm x 6.8cm ruptured unilocular cyst anterior to the uterus more to the right which was interpreted as a ruptured right adnexal mass probably benign by subjective assessment and IOTA-Adnexa with a 6.25% risk of malignancy (Figure 1A). The right adnexal mass has mixed level echo fluid and multiple echogenic foci within with a point of rupture at the lateral pole measuring 2 cm. It has a capsule measuring 0.4 cm. On Doppler studies, the right adnexal mass shows absent vascularity. (Figure 1B) Transvaginal ultrasound was also done with note of an anteverted uterus with regular contour, cervix with a homogeneous stroma and distinct endocervical canal as well as a uniform and hyperchogenic endometrium. (Figure 1C) She was brought to the operating room with an initial plan of exploratory laparotomy, possible unilateral salpingectomy right. Intraoperatively, a ruptured cystic mass just beneath and adherent to the anterior abdominal wall was noted measuring 13cm x 13cm with egress of yellowish mucinous fluid (Figure 2). The mass had a thick capsule with papillae within. (Figure 4) Upon further exploration, the mass was confluent with the anterior bladder dome (Figure 3). Urology service was then called to assess intraoperatively wherein a suspicion of a large urachal mass was entertained. Since the connections to the umbilicus have already been severed, the surgery proceeded first with a partial cystectomy and en-bloc excision of mass and anterior peritoneum. The umbilicus and the remaining attachments where then excised after. On further evaluation, the patient had normal uterus, normal bilateral adnexae and bowels. The patient had no untoward events post-op and was sent home voiding freely.

Gross pathological examination of the excised mass complex consists of a 25cm x 13.0cm x 6.0cm solitary, tan-brown mass with an attached anterior peritoneum. The external surface of the cystic mass had a grey to tan-brown (13.0cm x 13.0cm x 0.5cm) with dull external surface, located abutting the nearest bladder margin of resection. An oriented rupture point was present on the superior surface of the cystic mass. The internal surface was dark brown and rough with wall thickness measuring

Figure 1A. Holoabdominal ultrasound showing the anatomical relation of the mass, bladder and uterus. The mass is anterior to the uterus more to the right. (UP PGH 2023).

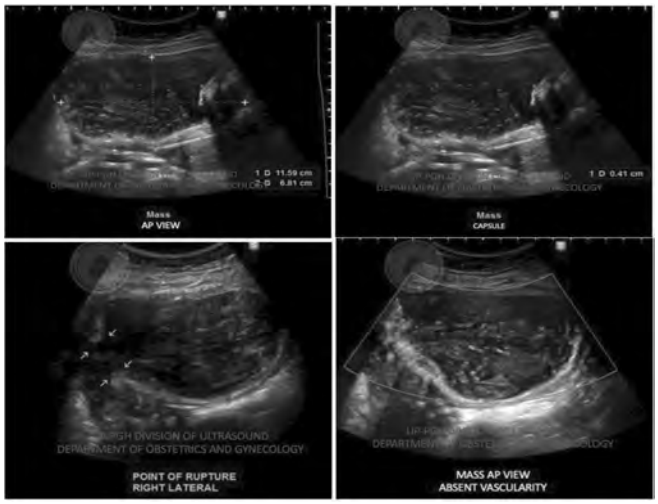

Figure 1B. Holoabdominal ultrasound shows right adnexal mass as a unilocular cyst measuring 11.6cm x 10.8cm x 6.8cm (volume: 445.5 cc) with mixed level echo fluid and multiple echogenic foci within. There are no solid areas and papillary excrescences seen. The capsule measures 0.4 cm. There is a point of rupture measuring 2.0 cm at the right lateral pole of the mass with the contents egressing through the defect. On Power Doppler, the right adnexal mass shows absent vascularity (UP PGH, 2023).

0.3 cm. The cystic mass did not grossly involve the muscularis of attached bladder dome. The oriented anterior peritoneum was grey to dark brown, irregular sheet of tissue (10.0cm x 10.0cm x 1.0cm). The attached bladder dome (6.0cm x 4.0cm x 1.5cm) had a tan-brown, smooth internal surface. The excised umbilicus consisted of a solitary, cream to tan brown, cylindrical, soft to doughy tissue

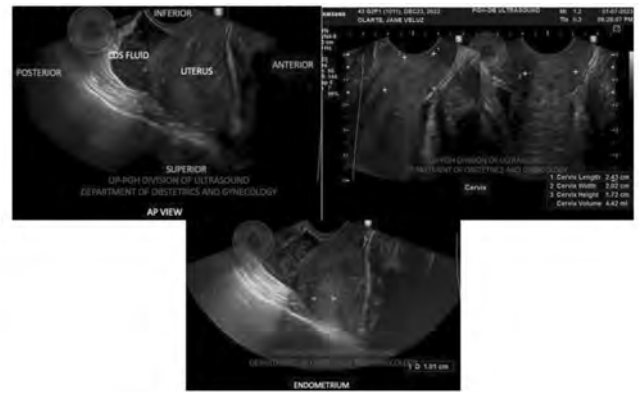

Figure 1C. Transvaginal ultrasound shows The uterus is anteverted with regular contour and homogeneous echopattern measuring 6.9cm x 6.2cm x 3.2cm. The cervix measures 2.4cm x 2.0cm x 1.7cm with homogeneous stroma and distinct endocervical canal. The endometrium is uniform, hyperchogenic, measuring 1.0cm. The endometrial midline is not defined. The endometrial-myometrial junction is regular.

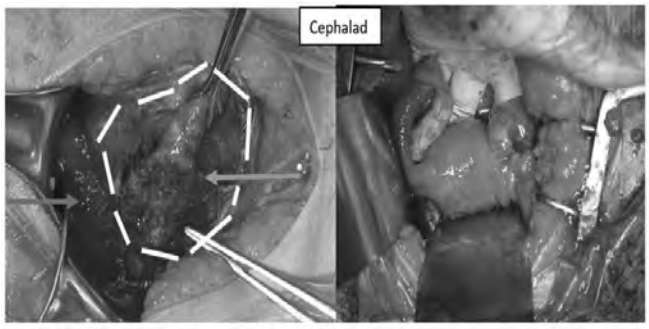

Figure 2. Left 15cm x 10cm cystic mass with egress of jelly-like material (blue arrow) from the point of rupture (Green arrow) Dashed lines show outline of the cystic mass Right. The uterus and bilateral adnexae appear grossly normal.

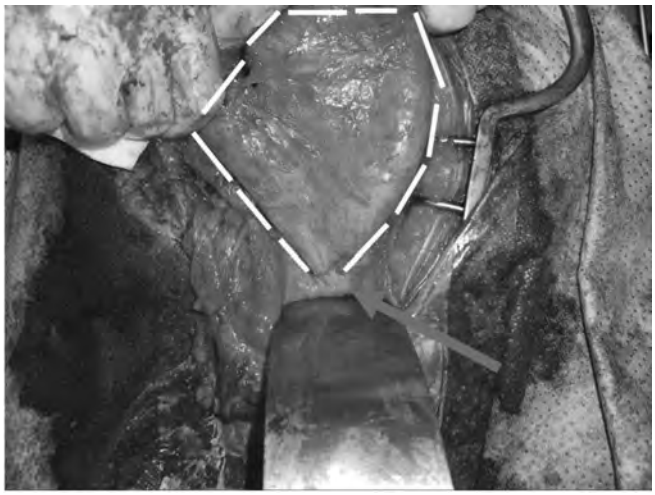

Figure 3. 10cm x 15cm mass (white dashed lines) reflected superiorly with confluence to the dome of the urinary bladder (blue arrow).

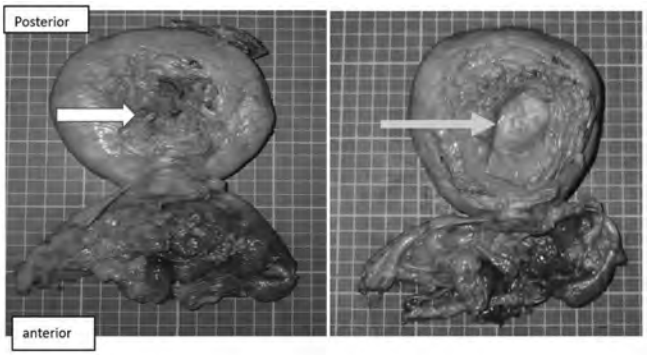

Figure 4. Left The excised mass complex consisted of a 25cm x 13.0cm x 6.0cm solitary, tan-brown bladder mass showing rupture point (White arrow) with an attached anterior peritoneum (Dark arrow). Right- The excised mass complex with attached urinary bladder dome (arrow) UP-PGH, 2023

measuring 4.0cm x 3.0cm x 1.5cm and an attached umbilical skin measuring 2.6cm x 2.0cm x 0.3cm. (Figure 4)

Histologic features of the cystic mass lining show the one to three-layer stratification of the cuboidal to low columnar glandular-type epithelium. Each individual cell has scant eosinophilic cytoplasm, hyperchromatic nuclei with moderate atypia and no definite mitoses. Consistent with the mucinous subtype of the tumor, pools of mucin are prominently seen as the abundant pale eosinophilic stringy material (Figure 5)

The final histopathological diagnosis was urachal mucinous cystic tumor of low malignant potential (MCTLMP). The excised umbilicus and fallopian tubes had benign findings.

A post-operative chest CT scan revealed pulmonary tuberculosis with bronchiectactic and fibrotic changes. No evidence of metastasis was noted. The patient did not receive additional treatment.

At 3 months post-op, she underwent surveillance cystoscopy which showed a smooth urethra and smooth bladder walls with no noted intravesical masses or abnormal mucosal surface. She is on regular follow-up at the outpatient department wherein she is scheduled for periodic imaging with abdominal CT scan with IV contrast.

Discussion

The urachus is a midline tubular structure which is an embryologic remnant of the allantois.

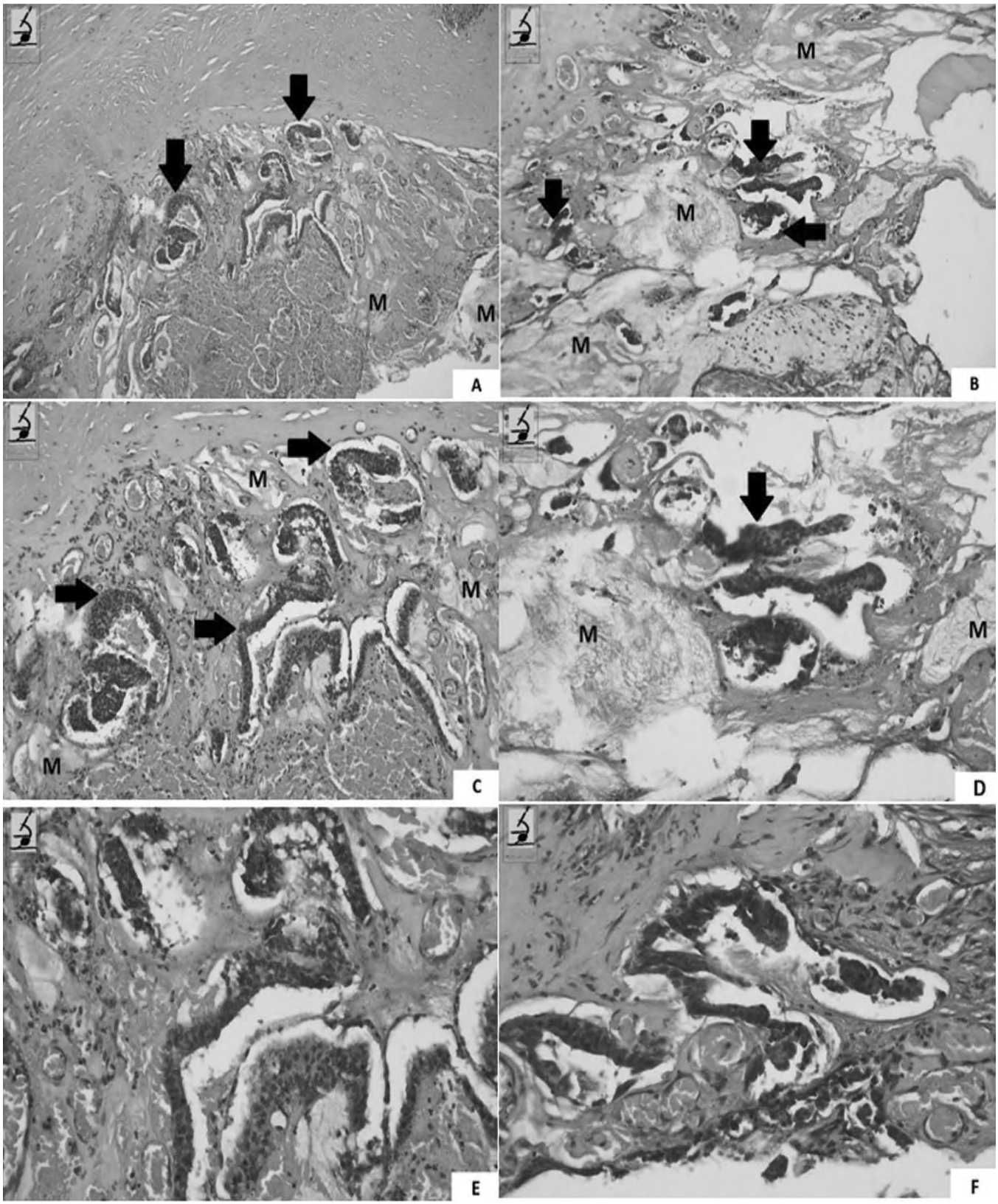

Figure 5. A. and B. (H&E 40X) Internal surface of the cystic mass lined by glandular structures (arrows) in pools of mucin (M). **C**. and **D**. (H&E 100X) Closer magnification of the glands (arrows) and mucin (M). **E**. and **F**. (H&E 400X) High magnification of the glands highlighting its tufted to pseudopapillary architecture, stratification and atypia.

This tubular structure usually involutes before birth or in early infancy forming the median umbilical ligament. However, 32% of adults are reported to have remnant urachus resulting in various clinical presentations such as a patent urachus (most common), umibilical-urachal sinus, vesicourachal diverticulum and urachal cysts.[2] Histologically, the persistent urachus lumen is lined with urothelial epithelium or columnar glandular epithelium. Its walls are formed by smooth muscle in continuation with muscularis propria of the bladder. The exact etiology is not fully understood, but it is believed that the malignant conversion of the columnar or glandular metaplastic epithelium of the urachus may result in the urachal carcinoma.[3]

Neoplasms of the urachus are extremely rare causes of bladder malignancy representing 0.17% of all bladder cancers.[1] There are only a few urachal glandular tumors that are considered completely non-malignant. Villous adenomas and mucinous cystadenomas are examples of completely benign tumors. Villous adenomas present with prominent villopapillary structure, lined by dysplastic mucinous epithelium. Mucinous cystadenomas on the other hand, shows unilocular or multilocular cystic spaces, lined by a single layer of non-dysplastic cuboidal or columnar epithelium.[2]

The mucinous cystic neoplasms are further classified according to the 2016 WHO classification into three entities: Cystadenoma, Mucinous Cystic Tumor of Low Malignant Potential (MCTLMP) and mucinous cyst adenocarcinoma. As of 2023, there are only 48 reported cases of mucinous urachal neoplasms and only 26 cases of mucinous cystic tumor of low malignant potential.[1] The presented case is the first one reported by the institution.

Typically, these tumors manifest with non-specific symptoms like hematuria, mucusuria, urinary frequency, and vague lower abdominal pain. In contrast, some patients exhibited no tumor-associated symptoms, with the tumor being discovered either through palpation or incidentally on imaging. Only two of the reported cases presented with abdominal pain, and merely three cases involved a palpable mass. The case presented is the first documented instance of a urachal MCTLMP presenting as acute abdomen with a palpable right lower quadrant mass. This case stands out in its presentation, thereby enriching the authors' understanding of the variability in clinical manifestations of urachal MCTLMP and highlighting the importance of considering it in differential diagnoses for similar presentations. In a recent review of clinicopathologic characteristics of MCTLMP cases in 2021, it was reported that the incidence is equal between men and women with age ranging from the second to eight decade of life.

The work-up of abdominal masses is one of the highlights of the case. Due to the unusual presentation as well as the failure to recognize the urachal cyst during the initial work-up prior to presenting as an acute abdomen, no cross-sectional imaging like CT scan or MRI was done. Even during transvaginal and abdominal ultrasound done at the ER setting, there was no suspicion of the mass being of urachal in origin. Generally, a midline location is an important feature that helps distinguish an ovarian mass from an urachal mass or bladder mass. Ultrasound may show a mixed echo between the bladder dome and abdominal wall on the umbilical level. However, in the present case, the transvaginal and abdominal ultrasound showed the mass was more to the right and anterior to the uterus. While ultrasonography can demonstrate the tumor as a complex midline mass with a supravesical location, a multiplanar CT or MRI can better differentiate the tumor's anatomic relationship to the nearby structures effectively. Ultrasound is also subject to the operator's technical ability and patient's body habitus.[4]

Abdominal CT scan could reveal a heterogenous, lobulated hypodense mass extending from umbilicus to dome of the urinary bladder wall. Lesions usually show no enhancement after contrast infusion but have internal septa or calcifications of their walls. MRI provides better imaging for the diagnosing the urachus due to its ability to capture detailed images in both coronal and sagittal views, which suit the slanted orientation of the urachus. MRI is particularly useful for showing the spread of disease within the local area or even to other parts of the body. Urachal tumors can appear in various forms—solid, cystic, or both, often containing a jelly-like substance known as mucin. Another common feature of urachal carcinoma is the presence of psammomatous calcifications; fine, round calcifications which are found in most of these cases. Thus, when there is uncertainty on

the location of a pelvic mass, using MRI could be of significant benefit.[5]

First-line and definitive treatment remains to be mass excision, urachectomy or urachectomy combined with partial cystectomy.[6] However, this case was initially assessed as a ruptured ovarian neoplasm, a recognized gynecological emergency, the planning for a possible urachectomy was not considered. An important factor of the case was the failure to identify the urachal cyst during initial evaluation, as discussed earlier. During exploratory laparotomy, the mass was densely adherent to the anterior peritoneum, which hindered further peritoneal dissection and the identification of the bladder. This finding was crucial, as it significantly deviates from the typical presentation of ovarian neoplasms. Even in cases of large ovarian neoplasms, such adherence to the anterior abdominal wall is uncommon. This case illustrates the importance of maintaining a broad differential diagnosis, especially when encountering atypical presentations during surgical procedures. The possible connections of the urachal cyst and the umbilicus was probably dissected during the laparotomy or during further adhesiolysis.

In the literature, there is no reported recurrence or post-operative morbidities noted after mass surgical excision. The prognosis for MTLCP remains good with only 1 mortality described as "death from other causes" and the rest were free of recurrence or metastasis for up to 84 months of follow-up.[1] In contrast to this excellent prognosis, invasive urachal adenocarcinoma has a five-year survival rate 45%.[5] Due to limited data, there is still possibility for further complications, growth, malignant transformation for MCTLMP. Therefore, it is suggested that after surgical excision, periodic imaging and cystoscopy should be done to monitor for recurrence. Cystoscopy and periodic imaging would contribute to the medical literature and increase overall understanding of these neoplasms.[1]

Conclusion

Mucinous cystic tumor of low malignant potential (MCLTMP) is a rare subtype of the neoplasms of the urachus which carries an excellent prognosis. Although rare, the clinician must be familiar with its clinical features and anatomical considerations. This case illustrates the importance of maintaining a broad differential diagnosis, especially when encountering atypical presentations during surgical procedures. Surgical excision, adequate resection and regular follow-up are required to avoid missing the potential malignant component and prevent devastating prognosis.

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CASE REPORT

Non-reducible Inguinoscrotal Hernia of the Urinary Bladder as a Direct Component of a Pantaloon Hernia: A Case Report and Review of Literature

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Pantaloon hernias are a subset of abdominal hernias wherein both indirect and direct components of inguinal hernias are present on one side. Inguinoscrotal hernia of the urinary bladder is a rare condition involved in less than 4% of inguinal hernias and more so if they are part of a direct component of a pantaloon hernia. This is a case of a 65-year-old male who presented a non-reducible right inguinoscrotal mass. Inguinoscrotal herniation of the urinary bladder was preoperatively diagnosed through imaging. Intra-operatively a pantaloon hernia was noted with the herniated urinary bladder as the direct component and omentum as the indirect component. The patient underwent cystourethroscopy, inguinal exploration right, and mesh hernioplasty. The patient had an unremarkable post-operative course and was discharged with improved conditions. Awareness of this clinical condition will aid in the diagnosis, and proper management and prevent post-operative complications.

Key words: pantaloon hernia, urinary bladder hernia, cystocoele, hernioplasty

Introduction

An inguinal hernia is an opening in the myofascial plain of the oblique and transversalis muscles that can allow for herniation of intraabdominal or extraperitoneal organs. These groin hernias can be divided into indirect, direct, and femoral based on location.[1] Pantaloon hernia or saddlebag hernia is characterized by the presence of both indirect and direct inguinal hernias at the same laterality. The incidence of pantaloon hernia in males is 5.6% in males and approximately 1.8% in females.[2] Inguinoscrotal hernia of the urinary bladder was first described by Levine in 1951 as a scrotal cystocoele is a rare event that is common among men. It occurs in about 1- 4% of inguinal hernias in the general population.[3]

According to Baruchu (2018), patients with urinary bladder hernia are diagnosed incidentally and develop without a specific symptom but generally present inguinal pain or swelling associated with voiding or lower urinary tract symptoms (LUTS). Less than 7% of inguinal bladder hernias are diagnosed prior to surgery, 83% are diagnosed intraoperatively, and 16% are diagnosed postoperatively due to complications.[3]

Here is a case of a total urinary bladder hernia as a direct component of a pantaloon hernia diagnosed pre-operatively. This paper aimed to highlight the rarity of the case as well as the

importance of complete history taking, physical examination, and diagnostic workup in order to prevent postoperative complications common in this case.

The Case

The patient in this case is a 65-year-old, Filipino male, a known diabetic, with a BMI of 30.3 who came with a one-week history of notable non-reducible bulging right inguinoscrotal mass which was associated with urinary frequency, straining, and feeling of incomplete bladder emptying. The patient had a one-year history of lower urinary tract symptoms predominantly urinary frequency and nocturia. He was noted to have an enlarged prostate and was treated with alpha-adrenergic blockers and 5-alpha reductase inhibitors which offered relief of symptoms. The patient had a family history of hypertension and diabetes mellitus on both maternal and paternal sides. Past medical history revealed a 4-year history of diabetes mellitus and gouty arthritis with no prior surgeries.

A pertinent physical examination revealed a right inguinal mass extending toward the right scrotum. The mass was non-reducible, non-tender, soft, and cystic in character. The right and left testis were palpable and unremarkable. Rectal examination revealed a smooth, rubbery, non-tender prostate, approximately 30 grams. The initial consideration at that time was a non-reducible indirect right inguinal hernia.

Laboratory studies were within normal limits except for an elevated HbA1c of 7.2% (4.3-6.4%). Urinalysis demonstrated bacteriuria (3027/HPF), microcytic hematuria (42/HPF), and pyuria (42/HPF) with a normal PSA of 4ng/dL.

Plain computed tomography (CT) imaging (Figures 1 & 2) showed a widened right inguinal canal and herniation of the urinary bladder into the right inguinoscrotal region. The kidneys were noted to be normal in size with the collecting system not dilated. A static cystogram (Figure 3) was also performed which showed an outlined urinary bladder with a significant portion herniating down towards the right inguinoscrotal region.

The patient was scheduled for, cystourethroscopy possible TURP, and right inguinal exploration. On cystoscopy, the median lobes of the prostate

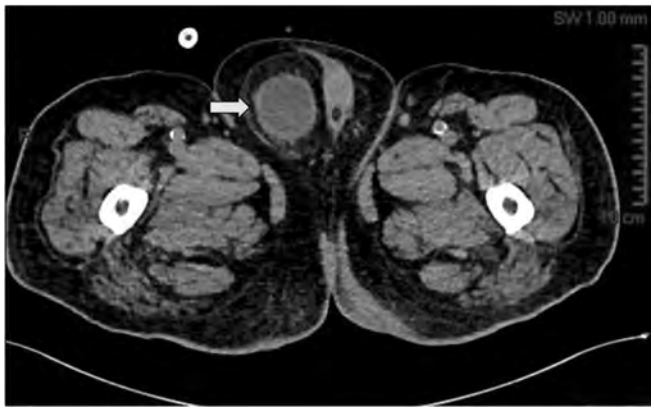

Figure 1. Axial CT scan of the abdomen and pelvis showing the herniation of the urinary bladder into the right inguinoscrotal area (arrow).

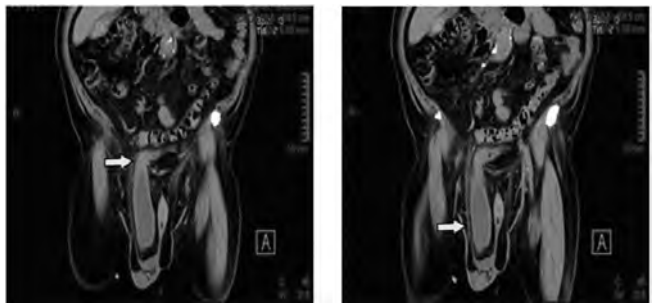

Figure 2. Coronal CT scan of the abdomen and pelvis showing the herniation of the urinary bladder into the inguinoscrotal area (arrows).

Figure 3. Cystogram showing a contrast-filled urinary bladder herniated towards the right inguinoscrotal area.

were not enlarged, bilateral slit-like orifices were identified, and mild trabeculations were noted. Upon inguinal exploration, an indirect inguinal hernia containing the omentum was identified (Figure 4A). As the dissection proceeded medially, the herniated urinary bladder was noted (Figure 4B) as it passed medially to the external inguinal ring and enters the scrotum together with the herniated omentum.

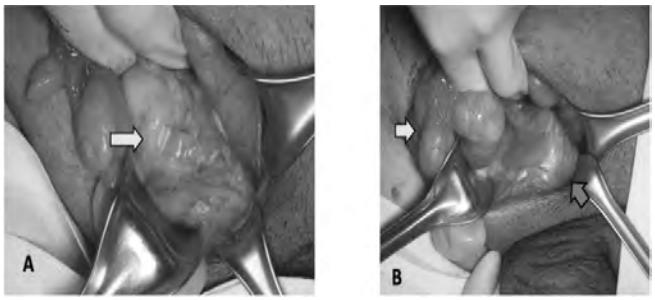

Figure 4. Inguinal Exploration: **A**. Indirect inguinal hernia containing the omentum (arrow). **B**. Pantaloon hernia; Indirect component containing omentum (arrow) and directly herniating urinary bladder (dark arrow) entering the right scrotum.

Careful blunt and sharp dissection was done. The herniated portion of the urinary bladder was completely withdrawn from the scrotum (Figure 5A) and reduced (Figure 5B). The indirect component of the pantaloon hernia was carefully separated from the spermatic cord and contents were reduced. Figure 6 shows the hernial sac after the reduction of the omentum. A Lichtenstein repair was done to decrease the risk of recurrence.

A post-operative cystogram was done (Figure 7) showing the reduced urinary bladder in the pelvis with a smooth mucosal outline, no filling defects, and no leak.

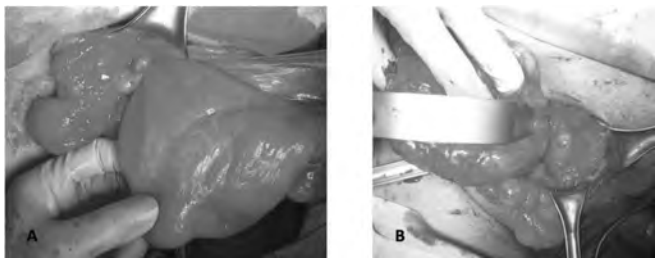

Figure 5. Urinary Bladder Hernia: **A**. Portion of the urinary bladder completely withdrawn from the right scrotum. **B**. Post-reduction image of the herniated urinary bladder.

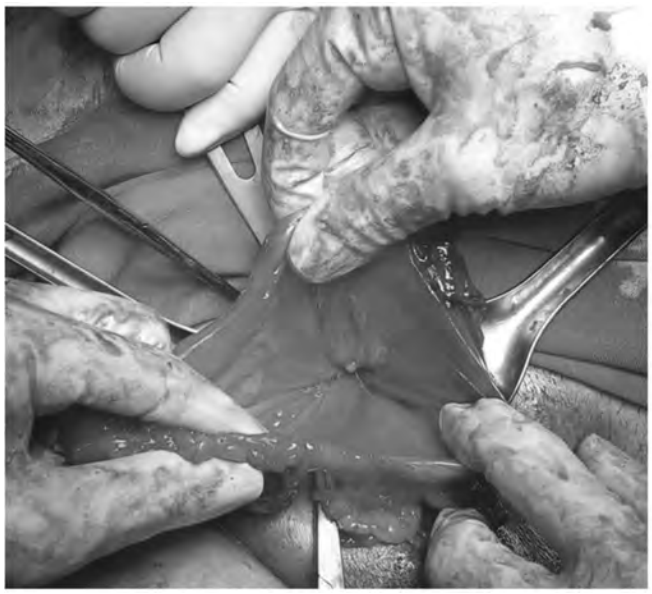

Figure 6. The hernial sac after reduction of its contents.

Figure 7. Cystogram showing the reduced contrast-filled urinary bladder in its normal pelvic location with smooth mucosal lining and no leak.

The postoperative course was uneventful, and the patient was discharged on the 2nd postoperative day.

Discussion

Abdominal wall hernias mostly occur in the groin. These groin hernias can be divided into

indirect, direct, and femoral based on location.[1] The most common subtype of groin hernia in men and women is indirect inguinal hernia.[5] Pantaloon hernia is a subtype that is characterized by the presence of both indirect and direct inguinal hernias at the same laterality. Urinary bladder involvement occurs in 1-4% of inguinal hernias in the general population.[3] The etiology may be related to bladder outlet obstruction due to an enlarged prostate, chronically distended bladder, decreased bladder tone, pericystitis, adhesion of the bladder wall to the perivesical fat, obesity, and weakness of the pelvic floor muscles in combination with a weak abdominal wall.[6,8]

A systematic review done by Branchu et at.[3] reported that herniation of the urinary bladder into the inguinoscrotal area mainly occurred in a male patient who was overweight and over the age of 50 years old. Although some patients with inguinoscrotal bladder hernia may be asymptomatic, many would report having inguinal swelling, lower urinary tract symptoms, pain, and reduction of inguinal mass after voiding.[1]

It has been reported that less than 7% of bladder hernias are diagnosed preoperatively, while 16% of those are diagnosed postoperatively due to complications.[8]

The most common radiologic modalities that may be used as an adjunct to history and physical examination would include ultrasonography, computed tomography (CT), and magnetic resonance imaging (MRI)[5] as well as voiding cystourethrography which is considered the best diagnostic modality for inguinoscrotal bladder hernia.[6,7] High-resolution ultrasound with 5-, 7.5, or 10-MHz transducers allows better anatomic depiction and higher sensitivity for the detection of scrotal abnormalities.[7] CT scans and MRIs provide static images that are able to delineate groin anatomy, detect groin hernias, and exclude potentially confounding diagnoses[5] as well as help in surgical planning.

Oruç et al.[9] found that 11.2% of the hernias in a review of 190 cases were associated with urologic malignancies including bladder carcinoma and prostate carcinoma. Hence, it is imperative that inguinoscrotal bladder hernias be diagnosed preoperatively so that appropriate surgical and medical plans are made.

The presented case was diagnosed preoperatively, Hence a careful and well-planned approach was done. The possibility of having morbid postoperative complications was greatly reduced because necessary steps were taken.

Surgical repair of hernias can be performed open, laparoscopic, or with robotic assistance[5] however open surgical repair is the preferred treatment.[6] The surgical approach depends on the surgeons' preference and the patient's status and condition.[6] The standard of treatment for inguinoscrotal bladder hernia is either reduction or resection of the herniated bladder.[7] Indications for bladder resection include bladder damage during hernioplasty, necrosis of the bladder neck, bladder tumors, bladder diverticulum, and hernia neck of less than 5 mm in diameter[6] which were not present in this case.

Conclusion

Inguinal hernia is one of the most common disease entities that general practitioners, surgeons, and urologists may encounter. However, being aware of other disease entities that may accompany or present similar to inguinal hernias must always be kept in mind. A detailed history and physical examination of all patients are paramount to having an accurate diagnosis. Inguinoscrotal bladder hernias should be diagnosed preoperatively as much as possible to prevent complications. General surgeons and urologists alike must be aware that this kind of rare condition exists.

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CASE REPORT

Leaving No Stone Unturned: A Case Report of Squamous Cell Carcinoma of the Kidney Associated with a Staghorn Calculus

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Squamous cell carcinoma (SCC) of the kidney is a rare malignancy and has a poor prognosis because it is usually detected and presents at an advanced stage of the disease. Due to lack of studies regarding its clinical course and its radiologic features, it is usually not detected and presents as an incidental finding in histopathologic results. This type of malignancy more often is related to Renal stones secondary to chronic infection, inflammation, and irritation. A 52-year-old Filipino woman was referred to this institution due to flank pain and constant weight loss. The authors administered antibiotics then performed simple nephrectomy on her. A pathologic examination revealed Squamous Cell Carcinoma with Sarcomatoid differentiation. Four months after the operation, patient was readmitted due to lowback pain and generalized weakness which led to a suspicion of a possibility of Bone Metastasis. Patient was offered further workups such as whole abdominal CT scan with contrast and other palliative treatments however she refused and opted to be discharged despite medical advice. Patient then died 2 weeks after being home against medical advice.

Key words: squamous cell carcinoma, staghorn calculus

Introduction

Literature provides that Renal cell carcinomas, especially arising from the renal cortex, accounts for about 80% to 85% of all primary renal neoplasms.[1] The most frequent type of renal cell carcinoma is the transitional cell carcinoma, which constitutes approximately 8% of the total incidence. Less frequently, there are parenchymal epithelial tumors, which include pathologies such as oncocytomas, collecting duct tumors, angiomyolipomas, and renal sarcomas.[2]

Renal cell carcinomas are predominantly seen in men aged 50 to 70. On a global scale, they exhibit varying degrees of incidence. In the United States alone, there are approximately 63,000 fresh diagnoses annually, leading to nearly 14,000 fatalities. It was noted that most of the increases in cases in the US observed since the 1980s have been diagnosed at an early stage of the disease.[1,3] In the Philippines, a total of 2,384 new cases of kidney cancer were recorded in 2020 alone and amassing 1,229 cancer deaths. Accordingly, it was concluded that renal cell carcinoma is the most common type of renal malignancy in the country.[4]

The screening of renal cell carcinoma typically includes the following: 1) urine test with possibility of hematuria; 2) hematological parameters indicative of anemia or high serum calcium levels; and 3) a renal and bladder ultrasound showing a solid mass or a complex cyst, which can have septations and nodules.[2,5] As to imaging, a plain and contrast enhanced CT scan of the kidneys, ureters,

and bladder are staples in its diagnosis. Through these imaging modalities, renal cell carcinoma will demonstrate significant enhancements, which can even reach greater than 20 Hounsfield units (HU) after contrast. The imaging modalities can also detect lymphadenopathy and invasion to the renal vein or inferior vena cava or invasion to the adjacent organs, greatly assisting in the approach to management. Moreover, the aforementioned use of the CT scan allows for the detection of metastases, including the extent of involvement among the bones of the abdomen and pelvic regions. Accordingly, if distant metastases are suspected, a whole-body CT scan is still warranted.

Rare cases of renal cell carcinoma also include squamous cell carcinoma of the kidney. Even as it is rare, it is still vital to explore this as a potential differential diagnosis, especially when a renal mass coincides with persistent renal calculi. Chronic irritation, inflammation, and infections that facilitate squamous metaplasia of the renal pelvis epithelium, are known factors that can contribute to the said diagnosis. Thus, cases of chronic pyelonephritis, which is a state of chronic inflammation and chronic irritation of the renal parenchyma through stones are significant considerations needed to be explored when considering a diagnosis of squamous cell carcinoma.[6]

This case report presents a Filipino patient who was diagnosed with stage IV (T1bN0M1) squamous cell carcinoma of the kidney associated with a single staghorn calculus. The paper focuses on the perspective of the patient's management, as well as the comparison of the unique features of this case contrasted against similar cases reported in peer-reviewed literature.

The Case

The patient is a 52-year-old female who came to the hospital with a chief complaint of flank pain and constant weight loss, which started and persisted about three months prior to her hospital consultation. It was also noted that the patient did not present with symptoms of hematuria. Upon being given the requisite ancillaries and laboratory evaluations, the patient was diagnosed to have a non-functioning kidney, secondary to Obstructive Nephropathy, which was also secondary to a left staghorn calculus, hence the patient was subsequently scheduled for a simple nephrectomy of the left kidney.

The operation was conducted without any intraoperative complications.

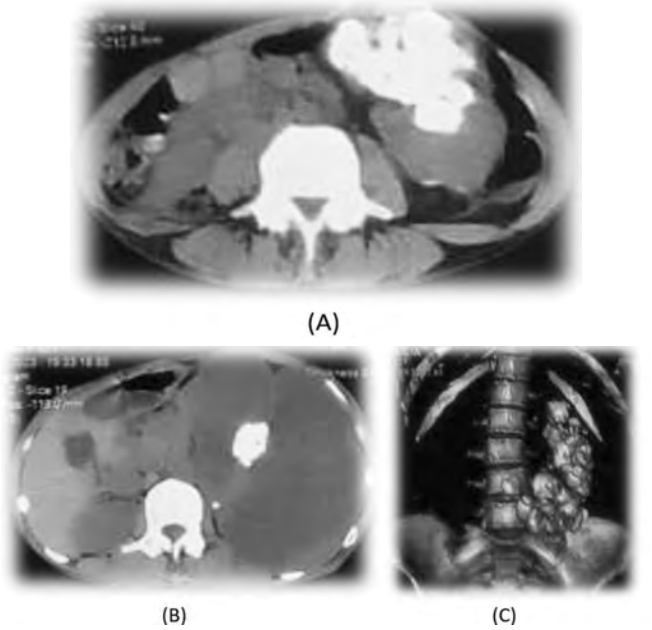

Figure 1. (A-C) Non-contrast enhanced CT scan of the KUB of the patient showing the presence of the staghorn calculus of the left kidney.

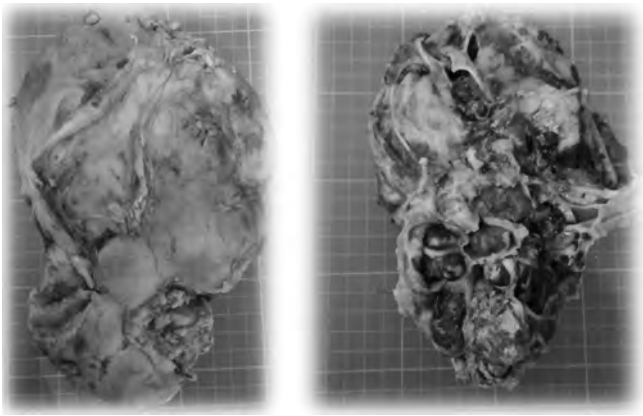

Figure 2. Gross and Opened-cut section of the kidney specimen of the patient status post open simple nephrectomy.

Patient was then transfused with 3 units of PRBC postoperatively, as the rest of the hospital stay was unremarkable. However, it turned out that

the surgical pathology report of the left kidney indicated that the patient had stage 1 (T1BNO) Squamous cell carcinoma. Patient was lost to follow up however three months post-surgery patient presented with low back pain and generalized body weakness.

Patient was then readmitted at this institution with the planned workup for the patient which included a whole abdominal CT scan with contrast. Unfortunately, the patient was discharged against the physician's advice.

Outcome and Follow-up

Upon further investigation, it was found out that the patient had developed bone metastasis presenting as low backpain and had expired at about 4 months post nephrectomy.

Discussion

A primary squamous cell carcinoma arising in the urinary tract is relatively uncommon, as SCCs usually arise from the urinary bladder and male urethra rather than the kidney.[5] Inversely, the patient reported in this case appears to have an incidentally acquired primary renal squamous cell carcinoma associated with calculi and severe hydronephrosis. Nevertheless, the connection between renal cancer and a kidney stone, particularly a staghorn calculus in this instance, appears to be an emerging trend of increasing significance in recent cases involving renal cell carcinoma diagnoses.

A similar study of a 54-year-old female complaining of left flank pain and an abdominal mass of six months presented with squamous cell carcinoma of the kidney, which was associated with a ureteral stone. Despite the patient being successfully treated after radical nephrectomy, as well as four cycles of tirelizumab as antibiotic therapy, the patient had developed adrenal, lymph, and uterine appendage metastases. The proponents mentioned that squamous cell carcinoma of the kidney has a poor prognosis and should be a consideration in patients with a renal mass, long-standing urinary calculi, or massive hydronephrosis.[2]

In another rare case, a 61-year-old male complains of right flank pain for five months. A CT scan then revealed multiple renal stones, as well as a necrotic mass in the right kidney lower pole, which was suspected as an ascending colon invasion. Radical nephrectomy, coupled with a right hemicolectomy and lymph node dissection was done. After which, an examination of the kidneys, along with a PET/CT led to the diagnosis of primary squamous cell carcinoma of the kidney. Adjuvant therapy was not administered to the patient, who remained alive throughout the 6-month follow-up period post-surgery. Notable features of this case were the presence of adjacent organ invasion, and the proponents remarked on how the mechanism of SCC pathology remains unclear, with its prognosis appearing to assimilate that of urothelial carcinoma when compared through differing stages.[5]

As to the presence of staghorn calculi in squamous cell carcinoma, a 59-year-old female presenting with a long-standing history of flank pain and hematuria was studied, in which the imaging modalities revealed the enlargement of the right kidney accompanied by the presence of multiple staghorn calculi. Subsequently, the patient underwent a radical nephrectomy right, and the histopathological analysis of the mass confirmed the presence of well-differentiated keratinized squamous cell carcinoma. Notably, as to the previous case, no primary source for this squamous cell carcinoma was identified.[3]

From the three cases previously discussed, common features arising from patients with SCC are: 1) the age range of the patients, which seem to be in the ranges of early 50s to early 60s; 2) a long-standing history of flank pain which may or may not include hematuria; and 3) the occurrence of staghorn calculus, which is present in one of the three case reports reviewed.[2,3,5,6]

In a broader case series consisting of 14 patients with SCC of the kidney[6], most were male (71.4%) and had a mean age of 56 years. Among the patients studied, flank pain emerged as the predominant presenting symptom, observed in 11 cases (78.6%), followed by fever, noted in 6 cases (42.9%). Interestingly, only 4 out of the 14 patients (28.5%) had received a preoperative diagnosis of squamous cell carcinoma (SCC), while the remaining 10 patients (71.4%) had an incidental finding of SCC upon examination of their histopathology specimens. The mean overall

survival duration stood at 5 months, with a standard deviation of 4.5 months. In a deeper look of the cases in the series, various modes of presentation were documented, encompassing flank pain, hematuria, fever, loss of appetite, and weight loss, with the addition of painful urination and wound discharge. These symptoms frequently overlap with those associated with the more prevalent forms of renal cancer, making primary renal SCC hard to diagnose. Six individuals, whose nonfunctional kidneys were surgically removed, were discovered to have remaining disease upon post-operative radiological assessment. These patients underwent excision of the residual disease.

Furthermore, among the ten patients who incidentally exhibited squamous cell carcinoma upon histopathological examination, two were identified to possess metastatic nodules within their lungs. Unfortunately, none of the patients provided consent for chemotherapy, while six of them did receive radiation therapy as a component of their pain management for secondary bony metastases. Unfortunately, all fourteen (14) died secondary to the extensive metastasis and disease progression.[6]

The cases reviewed emphasize the difficulties the recognition of squamous cell carcinoma in the affected cases, most especially, that a correct preoperative diagnosis cannot be made only based on the patient's symptomatology and radiological findings. Unfortunately, the confirmation of squamous cell carcinoma can only be done through differentiation of cells in pathological studies. It would be difficult to rely upon symptomatology and radiological studies, as they are not too dissimilar from those of other upper urinary tract neoplasms and chronic inflammatory diseases.

In terms of management, it was noted that nephrectomy became a necessary intervention in cases of metastatic squamous cell carcinoma, primarily for symptom management and, on occasion, to confirm the pathological diagnosis. In addition to the surgical management, the use of adjuvant cisplatin-based chemotherapy and palliative radiotherapy has been proposed to alleviate symptoms in cases of metastatic disease. However, their impact on overall survival remains uncertain, while the establishment of suitable treatment protocols for patients in this category warrants further extensive research through large-scale studies.[6]

Conclusion

Primary squamous cell carcinoma of the kidney is a rare malignancy involving the urinary tract system. While renal squamous cell carcinoma with sarcomatoid differentiation is extremely rare, and the prognosis is very poor. In dealing with patients that have long-standing staghorn calculus within the renal pelvis on top of the presence of massive hydronephrosis, the physician should always consider a differential diagnosis of a possible renal squamous cell carcinoma, even with its rare occurrence, as progression and metastasis have proven to be fatal. The clinical picture of the disease is also important as the gradual onset of vague symptoms, lack of pathognomonic signs, and inconclusive radiological features make the squamous cell carcinoma of the kidney unsuspected in most cases, therefore delaying diagnosis and treatment.

Conventional imaging methods may not reliably detect this type of malignancy, and in most instances, the use of non-contrast and contrast enhanced CT scan in conjunction with MRI imaging plays a vital role in nailing down the diagnosis. It is also prudent for specialized physicians to be tactical in offering prompt surgical interventions combined with immunotherapy if suspicion is high likely, as it may improve survival rates of these patients.

Squamous cell carcinoma of the kidney usually presents at an advanced stage, with often poor prognosis. Therefore, not only should a high index of suspicion be warranted in patients with chronic kidney stone disease, but more studies should be conducted to discern the common features of patients with squamous cell carcinoma of the kidney associated with renal calculi. This would enable the identification and perspective evaluation of risk factors which would greatly help in the treatment and management of this condition in the susceptible patient population.

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CASE REPORT

Renal Calculus in an Ectopic Pelvic Kidney – A Case Report and Review of Literature

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Pelvic kidneys are anatomical abnormalities that occur when the kidney does not rise from the pelvis during embryogenesis. The majority of cases are asymptomatic, though they are associated with higher risks for traumatic injury, infections, renal calculi, and other urological issues.

Because of its advantages of flexion and deflection, retrograde intrarenal surgery (RIRS) employing flexible ureterorenoscopy (fURS) is an alternative treatment method for small- to medium-sized calculi in anatomically aberrant kidneys. Presented here is a case of a 43-year-old male with a renal stone in a pelvic left kidney with the ureter crossing the midlineand is located at the prevertebral region at the level of L4 to S1.

Key words: pelvic kidney, ectopic kidney, retrograde intrarenal surgery, flexible ureteroscopy

Introduction

Congenital renal anomalies are among the most common birth deformities, exceeded only by cardiac and skeletal defects.[1] An ectopic kidney's vascular supply is not consistent, and may receive vascular access from a range of vessels as the fetal blood supply can be retained. Multiple vascular sources may supply the ectopic kidney; the iliac arteries, direct branches from the aorta, mid sacral vessels, or the hypogastric arteries have all been found supplying ectopic kidneys. Surgery on an ectopic kidney requires thorough knowledge of this anatomy.[2]

Ectopic kidneys are also associated with several other congenital abnormalities. In females, this may be in the pelvis, such as Mullerian agenesis or unicornuate uterus. Ectopic kidneys can be a feature of multisystem congenital syndromes such as CHARGE syndrome (coloboma, heart disease, atresia choanae, retarded growth, genital hypoplasia, and ear abnormalities) or VACTERL malformations (vertebral, anal, cardiac, tracheal, esophageal, renal, and limb anomalies).[3] With an ectopic kidney, the kidney may become malrotated or the ureter may cross the midline as it approaches the kidneys. This may prove a challenge on performing retrograde ureteroscopic procedures such as diagnostic ureteroscopy and retrograde intrarenal surgery. The aim of this report is to present a case of a pelvolithiasis on a pelvic kidney with ureter crossing the midline that underwent retrograde intrarenal surgery as well as the challenges the surgeons encountered as well as tips on how to successfully clear the stone.

The Case

A 43-year-old male initially presented with periumbilical pain radiating to the epigastric area, described as colicky, dull, associated with vomiting episodes. Consult with private physician was done, wherein he was managed as a case of Acid Peptic Disease (APD) and was prescribed Omeprazole 40mg/tab 1 tab once daily. However, progression of symptoms and intermittent low-grade fever were noted. Work-up included urinalysis which showed microscopic hematuria. Serum Creatinine was within normal limits, and WBC count was slightly elevated (12.3).

Non-contrast Enhanced Helical CT Scan showed a left pelvic kidney located at the prevertebral region, at the level of L4 to S1, with a lithiasis in the interpolar/pelvic region measuring 0.7cm x 1.3cm x 1.3cm (AP x T x CC), 550HU, Moderate dilatation of the upper and mid calyces. The patient was then scheduled for elective definitive surgical intervention, and underwent Retrograde Intrarenal Surgery (RIRS). Intra-operatively, retrograde pyelography was performed and an S shaped ureter was noted from the left ureterovesical junction insertion crossing the midline then back to the ipsilateral renal pelvis at the pelvic kidneys (Figure 3). A 0.35 Sensor wire was placed and an F11 ureteral access sheath placement was attempted. With the configuration of the ureter, the F7.5 flexible ureteroscope was inserted through the guidewire under C-arm guidance. Since the stone migrated to the inferior pole, there was diffulty in the visualization and access of stone. The flexible scope was fully deflected and lithotripsy was performed. The procedure took around 2.5 hours for both fragmentation and dusting of the stone. Some less than 5 mm stone fragments were left for medical dissolution and expulsion. The patient tolerated the procedure well, and postoperative course was uneventful.

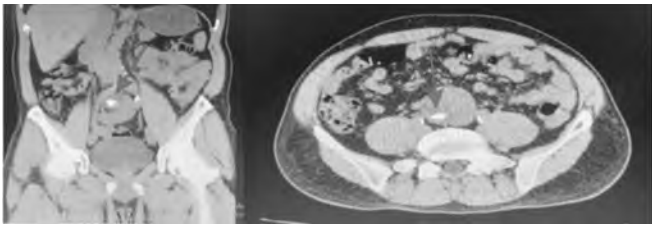

Figure 1. Non-contrast Enhanced Contrast Helical CT scan
Left: Coronal view. Right: Axial view. Arrows indicate the lithiasis.

Figure 2. Intraoperative findings – Flexible Ureteroscopy
Left: Guidewire inserted into the left ureteral orifice.
Middle: Fragmentation of the nephrolithiasis using LASER lithotripsy. Right: Post-fragmentation.

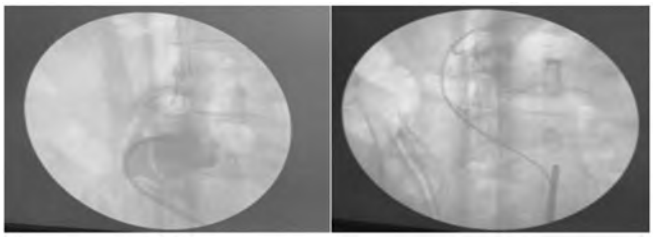

Figure 3. Intraoperative findings - Fluoroscopy
Left: Retrograde pyelography with tip of fURS within the renal pelvis
Right: Ureteral stent placement with coiling of the distal tip noted

Discussion

The kidney develops between weeks 6 and 8 after conception, and the embryologic kidney rises from the pelvis into the lumbar region in the 9th week. If the kidney fails to pass above the fork of the umbilical arteries, the blood supply degenerates, or there are other factors inhibiting renal migration, then the kidney fails to rise to its normal anatomical location and instead becomes ectopic. The exact location can be varied, with most cases being in the contralateral pelvis, but in the cases of crossed renal ectopia, both kidneys can be on the same side of the spine or, more rarely, the kidney can be outside the pelvis or retroperitoneal space entirely and even become located within the thorax. Ectopic renal units are usually a unilateral condition, but there are documented cases of bilateral ectopic kidneys.[4]

The incidence quoted is variable worldwide but is often approximately 1 in 1000 births.[4] A retrospective study of 13,701 antenatal scans in Turkey found an incidence of pelvic kidneys of 1 in

571, although this study only included scans with a normal amniotic fluid volume.[4] A Taiwanese study screening 132,000 school children found a lower incidence of only 1 in 5000, but it is thought this may have underestimated the actual incidence due to the screening method used.[5] An ectopic, pelvic kidney is the most likely finding in a fetus where the prenatal ultrasound finds an absent or missing renal fossa, but the amniotic fluid is normal.[6]

Even in asymptomatic patients, the ectopic kidney often has reduced function relative to the contralateral kidney. Most patients with ectopic kidneys are asymptomatic, and if recognized at all, the diagnosis tends to be an incidental finding while investigating other pathology or on routine antenatal ultrasonography. However, urinary tract complications can develop, and patients may present with a range of pathologies, including an increased incidence of urinary tract infections, ureteropelvic junction obstruction in the ectopic kidney, or increased risk of renal calculi. The most common associated abnormality is vesicoureteral reflux, which occurs in 30% of patients with simple renal ectopia.[6]

Patients with ectopic pelvic kidneys are more likely to develop urolithiasis compared with the normal population. This is linked to impaired drainage of the kidney due to altered anatomy, as well as metabolic reasons. Therefore, treatment of pelvic kidney stones remains challenging for the urologist because of the structural and architectural anomalies of such kidneys. The exact incidence of renal calculi in pelvic kidneys is unknown, but is thought to be higher than the general population due to altered anatomy and impaired urinary flow rates. The altered anatomy does have significant implications in the operative management of renal calculi, and the risk of vascular injury is increased relative to the general population.[7]

Extracorporeal Shockwave Lithotripsy (ESWL) in the prone position was first recognized as a non-invasive method for treatment of patients with pelvic kidney stones in 1988. Several studies have indicated that ESWL can be recommended as a first-line treatment option for anomalous kidney stones because of the high rates of stone clearance. However, Demirkesen et al. reported that ESWL for normal kidneys had a higher stone-free rate than that for aberrant kidneys (78% vs. 56%, respectively) and that aberrant kidneys had a higher rate of clinically insignificant residual fragments than did normal kidneys (37% vs. 18.5%, respectively).[8] Consequently, ESWL was been viewed as the first-choice non-invasive treatment modality with a relatively poor success rate for pelvic kidney stones.[9]

Flexible ureteroscopy has emerged as an alternative treatment method for small- to medium-sized renal calculi because of its advantages of flexion and deflection. Several studies have shown that the success rates of flexible ureteroscopy in patients with pelvic kidneys range from 75.0% to 84.7%.[10] Nonetheless, anatomic alterations including a tortuous ureter and malrotated kidney have been suggested as significant factors that increase the difficulty of the procedure and influence the stone clearance rate in treatment by flexible ureteroscopy. The use of a ureteral access sheath has been noted to assist in performing the procedure efficiently and safely.[11]

Other treatment options for stone disease in ectopic kidneys include Pyelolithotomy [open vs laparoscopic (transperitoneal vs retroperitoneal vs trans-mesenteric) vs robot-assisted], as well as Percutaneous Nephrolithotomy (PCNL). Although some authors have suggested the use of ultrasound or computed tomography guidance to achieve percutaneous access in patients with pelvic kidney stones, there is still a high risk of injuring the surrounding viscera and major vessels.[12]

A 11/13Fr ureteral access sheath was used for this patient, and flexible renoscopy was done under C-arm guidance. Stone fragmentation was done using Boston Scientific® Auriga™XL LASER Lithotriptor using Dusting method (high frequency, low energy, low pulse duration). A 6Fr x 24cm ureteral stent was placed post-procedure. Indwelling urethral catheter was removed first post-operative day, and patient was discharged on the second post-operative day. Imaging on follow-up one month post-procedure showed no lithiasis, and DJ stent was subsequently removed.

Conclusion

Although fURS in patients with anomalous kidneys can be technically challenging, advancements in endourological techniques have

made it a safe and effective procedure. In these patients, the stone-free rates are good with a low risk of major complications.

Patients with stone disease in anomalous kidneys need individualized management and probably should involve an interdisciplinary treatment with interventional radiology colleagues with interventions carried out in high-volume endourology centers. Although randomized trials between treatment modalities would be difficult given the rarity of this condition, perhaps large prospective multicentric studies with long-term follow-up and standardized references would be able to provide with high-quality insightful data.

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