ISSN: 2692-532X
DOI: 10.33552/AUN.2023.03.000575
Annals of Urology & Nephrology
Iris Publishers

Case Report

Long Segment Prolonged Right Ureteric Spasm Following Recent Passage of Stone Simulating Malignant Stricture in A Case of Hypoganglionosis

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Received Date: December 12, 2023

Published Date: December 18, 2023

Abstract

A very anecdotal, illustrative, and challenging case of a 51-year-old male with a background of chronic constipation since early childhood, dyslipidemia, vitamin D deficiency and musculoskeletal neck pain with appendicectomy at the age of 10 years presented in emergency with sudden severe right loin colicky pain radiating down to right groin associated with dysuria, nausea, vomiting, but no hematuria, fever or chills. Ultrasound and contrast computed tomogram (CT) showed long segment narrowing of the right distal ureter with hydroureter and hydronephrosis with concentric thickening of the wall and calcification, but no stone or signs of infection were evident, and possibility of malignant pathology led to further investigations of diagnostic cystourethroscopy at which the ureteroscope could not be passed into the right distal ureter but allowed JJ stent insertion. The retrograde ureterogram confirmed the findings. Radiological studies confirmed suspected associated hypoganglionosis and started on conservative management. At repeat laboratory, ultrasound, cystoscopy and ureteroscopy all were normal, and the JJ stent was removed. At 18 months follow up patient is doing well, remained asymptomatic, all investigations being normal and reversal of asymptomatic cholelithiasis was an additional bonus.

Keywords: Bowel motility disorders; hypoganglionosis; long segment; microbiome; micronutrient deficiency; ureteric spasm; stricture; stone; urolithiasis

Introduction

Urolithiasis is a common problem in India in general and its incidence is highest in the Saurashtra and Kutchh region of Gujarat state in particular [1]. Long segment ureteric stricture is very rare and mostly due to trauma, tropical diseases, and tumors and recently it has been reported following ureterorenoscopy for upper ureteric stone as an iatrogenic case and disastrous complication of endourology [2-6]. Recent observations that gut microbiome (GMB) has an impact on urolithiasis, and the conclusion that GMB disorder or dysbiosis may be involved in the occurrence and clinical characteristics of urolithiasis by influencing genetic and metabolic pathways [7,8]. We present a unique case of prolonged right ureteric long segment spasm following recent passage of a stone associated with hypoganglionosis with detailed investigations, interventions and treatment in a case simulating possible urothelial carcinoma.

Case Report

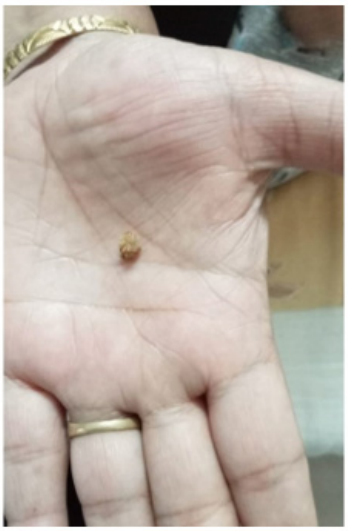

Figure 1: Recurrent spontaneous passage of small stones in the urine by patient's daughter after taking ayurvedic medications.

A 51-year-old male with a family history of urolithiasis and a background of chronic constipation since early childhood, dyslipidemia, vitamin D deficiency and musculoskeletal neck pain with appendicectomy for an acute appendicitis at the age of 10 years presented in emergency with sudden severe right loin colicky pain radiating down to right groin associated with dysuria, nausea, vomiting, but no hematuria, fever, or chills. There was a family history of urolithiasis with a patient's daughter who was regularly passing small stones in the urine (Figure 1). On examination, vital signs were normal. There was localized abdominal tenderness in the right lower quadrant but no right costovertebral angle tenderness, guarding, rebound or rigidity. Urinalysis showed microscopic hematuria but no signs of infection. Laboratory studies were all normal except white blood cells 14.80 (3.70 -11.00) k/uL and absolute neutrophil count of 12.05 (1.45 - 7.50) k/uL. Abdominal ultrasound showed cholelithiasis without any signs of obstruction or cholecystitis, trace of right perinephric fluid and borderline increased echogenicity of the right kidney.

Non contrast computed tomogram (CT) confirmed asymptomatic cholelithiasis and showed mild right-sided hydronephrosis and mild perinephric fat stranding but no calculi were visualized. Findings were interpreted to be due to a recently passed calculus and patient responded well to hydration, analgesics, anti-inflammatory and antispasmodic medications. After two days, the patient presented again with severe spasmodic right lower quadrant abdominal pain with localized tenderness and stable vitals. Urinalysis was not consistent with infection, but ciprofloxacin was started by primary care doctor for presumed urinary tract infection for leukocytosis, dysuria, and persistent flank pain. The patient required admission for rehydration and pain control. Repeat CT scan with contrast showed a long segment of somewhat concentric wall thickening in the distal right ureter, with questionable fullness in the lumen and tiny calcification. There was resultant mild hydronephrosis and hydroureter. The second focal area of mural enhancement of the posterior wall of the mid ureter was also observed.

Exact etiology was unclear and urine analysis did not suggest infection. The results and CT findings were discussed at the multidisciplinary meeting as the changes secondary to recent passage of stone defied in view of given failure of symptoms to improve despite antibiotics, analgesics, anti-inflammatory, and antispasmodic medications. An intraluminal or intramural lesion was suspected. Distal right ureter stenosis with hydroureter, hydronephrosis with mild acute kidney injury was thought due to focal structuring from possible stone induced stricture, pyelitis, retroperitoneal fibrosis unusual presentation of cancer etc. were in the differential diagnosis. Post appendicectomy ureteric injury was unlikely due to long period of over 4 decades. In view of persistent

pain, leukocytosis, absence of stone or sepsis, no improvement with conservative management and urine cytology being negative for high-grade urothelial cancer, never smoked, no exposure to carcinogen at work and no family history of any malignancy; diagnostic cystoscopy, right ureteroscopy with retrograde ureterogram and ureteral stent placement was planned.

Right ureteroscopy was attempted; but with distal ureteral obstruction present, ureteroscope was unable to be passed (Figure 2). The right retrograde ureterogram which confirmed CT scan findings followed by double 'J' ureteral stent placement was carried out (Figure 3). The radiological features of hypoganglionosis were seen on double contrast enema. Due to long segment of right distal ureteral partial obstruction and abnormal pyelogram, malignancy suspected, and further evaluation planned. Retrograde pyelogram which demonstrated distal right ureteral obstruction to the level of the pelvic brim with proximal hydronephrosis with findings concerning possible ureteral neoplasm versus a dense stricture. This was not amenable to ureteroscopic biopsy when the patient underwent stent placement. The management options which included repeat diagnostic ureteroscopy in the hopes of being able to access the distal ureter and potentially obtain biopsy and ureteroscopically evaluate the ureter proximal to the area of disease with possible laser end ureteric stricturotomy as our first logical preference to reassess the pathology before any further interventions or proceeding directly with distal ureterectomy and ureteral reimplantation for both diagnostic and therapeutic purposes via retroperitoneal robotic minimal invasive surgery.

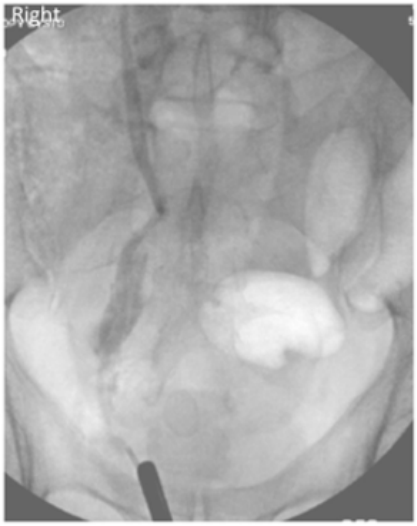

Figure 2: Right ureteroscopy and retrograde ureterogram. Note the long segment narrowing of the right distal ureter with inability of the scope to be negotiated just beyond the vesicoureteral junction, but open enough to allow contrast to pass through showing hydroureter above the narrow segment and mild right sided hydronephrosis.

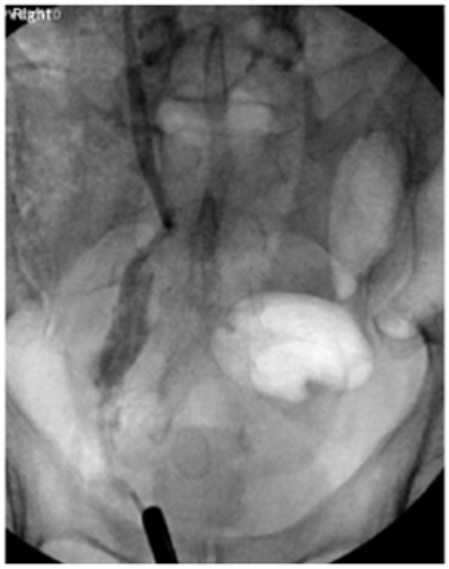

Figure 3: Attempted right ureteroscopy and JJ stent insertion. Note hugely dilated fecal loaded rectum with reversed rectosigmoid ratio and fecal loaded right colon secondary to hypoganglionosis in the background.

The advantages and disadvantages of each of these approaches were reviewed and discussed with the patient. Based on the findings at ureteroscopy and retrograde pyelography, the long segment of affected ureter would not be amenable to endourological management if this was a benign stricture. Furthermore, repeat attempt at diagnostic ureteroscopy may not be successful given the difficulty encountered at the time of the initial ureteroscopy two weeks ago. Furthermore, the limitations of ureteroscopic biopsy and this may be nondiagnostic and that regardless of potential biopsy results, surgical reconstruction would likely be necessary to definitively manage the obstruction. The option of distal ureterectomy with ureteral reimplantation would favor robotic surgical approach. Given the length of the segment of ureter that would be respected, the need for a psoas hitch and possibly Boari flap to fully reconstruct were anticipated. We gave time to the patient and family to think over these options and in the meantime, conservative treatment of associated hypoganglionosis was instituted. The patient opted for the first option and after couple of weeks the repeat ultrasound was normal and at cystoscopy and right ureteroscopy, there was no stricture and the ureteroscope passed easily.

Whole of the ureter was normal and the ureterogram showed normal ureter and pelvis without any obstruction. It was concluded that the recent passage of the ureteric stone might have caused mucosal ulcerations and irritation of the muscles leading to severe diffuse long segment ureteric spasm and due to associated hypoganglionosis multiple micronutrient deficiencies would have led to prolonged effect mimicking the stricture. The conservative management of hypoganglionosis with holobiotics, dietary changes, laxatives, lifestyle changes, dietary supplements and adequate hydration has led to replacement of multiple micronutrient deficiencies and long-term relief of partial functional obstruction due to spasms and symptoms. Following up at 18 months, the patient is progressing well, asymptomatic, and the urinalysis, complete blood count, bio metabolic profile and lipid profile is normal without any statins. The abdominal ultrasound was normal with dissolution of the cholelithiasis, and the renal ultrasound is normal without any renal calcification or stones and both ureters and pelvis are normal.

Discussion

Our case is a reminder of the fact that we should consider uncommon presentations of common disorders and diseases rather than thinking in terms of common presentations of an uncommon disease. Colorectal motility disorders are very common and may present with variety of extraintestinal symptoms and defies their detection as most patients may have some gastrointestinal symptoms but being long term and chronic makes one to ignore them and focus on the current presenting problem with a tunnel vision. In urology, the dictum is once a stone, always a stone as recurrence is the rule rather than exception and is rightly so as the cause of stone formation in most cases is originated somewhere else and unless that is diagnosed and treated; it's going to come back. Our team and institution have deep interests in urolithiasis and secretomotility disorders of the bowel and want to add more observations based on our extensive experimental, investigative, translational, clinical, and interventional diagnostic and therapeutic studies.

Our initial work in urolithiasis started in the 1980s during which gut nutrition in general and macro and micronutrient deficiency in particular in relation to pediatric vesical stones was established while working with the WHO project but it took some time to find out the role of gut secretomotility disorders and subsequent effects on the gut microbiome (GMB) leading to macronutrient excess and micronutrient deficiency for renal stones in apparently healthy patients. In the 1990s, it was expanded to special interest in experimental fetal surgery studies in gastroschisis in general and functional gastrointestinal secretomotility disorders and gut microbiome in particular [9]. The further establishment of specialized nephrological and urological hospitals and research centers at the millennium allowed us to consolidate intensive clinical integrative combined traditional and alternative medicine holistic functional gastroenterological (gut) and genitourinary tract (GUT) clinical and research work simultaneously for decades helped us to explore the relationship of the gut - GUT axis [10]. A recent systemic review and meta-analysis has confirmed that there is a characteristic gut microbiota dysbiosis in kidney stone patients.

In addition to the genetic and metabolic pathway, we believe that congenital colorectal secretomotility disorders or allied variants of Hirschsprung's disease including that associated with prematurity, birth method (vaginal>cesarean for neonatal GMB), beneficial effects of breastfeeding [11,12], acquired lifestyle-related factors such as infant nutrition, diet, exercise, environmental exposures of toxins/pathogens and stress manifesting through psycho-neuro-endocrine-immune axis all affect the GMB initially leading to colonic dysbiosis followed by intermittent or continuous ileocecal reflux with consequent small intestinal bacterial overgrowth (SIBO), leaky gut and multiple micronutrient deficiencies. Hippocrates believed all diseases begin in the gut. Implications of the associated hypoganglionosis while shedding light on the intricacies of its manifestation in our specific patient case. Our case was unique in which the initial obstructing stone has passed away leaving some indirect evidence, but the patient has not seen or felt coming out and the CT scan did show some calcification.

However, persistent severe long segment right distal ureteric spasm not being relieved by even general anesthesia and muscle relaxants responded very well to the appropriate treatment of underlying colorectal motility disorder is striking and we could not find similar case in the extensive search for the literature. Hypoganglionosis is seen mainly in children and young adults [13-15]. It has been reported in adults, but it is rare to see it in elderly patients [16]. Our hypothesis is that in our case perhaps the association of the urolithiasis with the congenital hypoganglionosis has been a risk factor in predisposition of the dyslipidemia causing macronutrient excess and multiple micronutrient deficiencies and prolonged spasm of smooth muscles leading to spams and colic secondary to the deficiencies of calcium, magnesium, and other key micronutrients. The hypoganglionosis leads to chronic fecal retention and alteration of the microbiome of the hind gut causing dysbiosis initially in the hindgut with intact ileocecal valve predisposing to appendicitis during teenage years of adolescent

growth spurt which happened in our patient at the tender age of ten years.

This was later followed by midgut colonization of dysbiosis via ileocecal reflux and small bowel bacterial growth secondarily making midgut in general and distal ileum functionally absent which led to hyperlipidemia and cholelithiasis in our patient both of which got reversed after conservative treatment of underlying hypoganglionosis and tertiarily via fecal-oral route altering oral and foregut microbiome which has been implicated in the development of gut and gut derived recurrent infections in children especially for those children who are not breast fed as per recommendations recently. Our multicenter multinational prospective integrated holistic combined prospective cohort study on the congenital and acquired functional disorders of the embryonic gut and their derivatives in general and that of the GUT, in particular, is still in progress for firmly establishing the gut-GUT axis, and Wang et al's study had been limited to hypocitraturia urolithiasis (HCU) and GBM. Therefore, more prospective randomized control studies would enable us to establish relationships and research more on this intriguing subject of urolithiasis and associated gut and GMB-related factors that would greatly interest the patients, parents, public, press, politicians, policymakers, and professionals.

Conclusion

A simple recent passage of ureteric stone led to such a prolonged severe right ureteric spasm and must escalate to oncology as the stone has passed but left some indirect evidence by exclusion of serious illness like cancer and was very scary for the patient and their families. Occult congenital anomalies in the form of hind gut secretomotility disorder can predispose the patient to not only gastrointestinal consequences like appendicitis and cholelithiasis with or without cholecystitis and their consequences but a wide range of gastrointestinal and extraintestinal associated predisposition to urolithiasis. The relationship of colorectal motility disorder and urolithiasis needs to be explored. Traditional approach of treating urolithiasis and recurrent irritative and infective complications needs to be reviewed in view of increasing evidence of changing the microbiome with the help of diet, laxatives, supplements and holobiotics which are safe, effective, non-invasive, and cheaper patient friendly option.

Funding

The authors are very grateful to Dr Dhaval R Govani MB BCh, MBA, Medical Director, PGICHR and the associated University Teaching Hospitals, Rajkot for his personal interest and supervision for the ongoing gut secretomotility disorders and microbiome project and providing much needed financial support for the project through the Dhaval Govani Educational and Research Foundation Trust, Rajkot, Gujarat, India.

Acknowledgements

The authors are grateful to Dr. Bhaumik Upadhyay MD, Ekal Vidyalaya Foundation of India; Dharmesh Mistry, Ekal Vidyalaya Foundation of UK; and Ramesh Shah CEO and former President-Director of Ekal-USA for implementing our gut-GUT axis findings for the promotion, prevention and protection of diseases in general and urolithiasis in particular via Ekal active and trusted international network of volunteers with integrated holistic targeted remote rural development with digital literacy, women empowering, healthcare provision, telemedicine and in-person care, healthy lifestyle, hygiene practices, nutrition education and planting nutritious family gardens to reboot the GMB to healthy one with prebiotic, probiotic and postbiotic food grown at home.

Conflict of interest

The authors declare that they have no conflict of interest.

Ethical approval

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards.

Informed consent

Informed consent was obtained from the patient.

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