Case Report

Rare complex malformations and iatrogenic postoperative stenoses of the urinary tract with late clinical effects and the steps towards restauration of normal functionality – clinical case

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Abstract

Congenital urinary obstruction is one of the most frequent conditions affecting the urinary tract in children. It may lead to significant long-term health consequences and the management of these pathologies represents a major challenge for urologists.[1] In cases where the patient presents with a complex pathology and an intricate medical and surgical history, retracing and understanding this history require a thorough anamnesis. Access to a state-of-the-art Imaging department is vital in order to have the highest degree of accuracy in determining the diagnosis. The doctor-patient relationship plays a very important role and all available treatment options must be discussed. The cases should be resolved through as few surgeries as possible. When the full reconstruction of the urinary tract is not feasible, as a last resort remains total nephrectomy or partial nephrectomy – when possible. The urologist's approach should always also bear in mind the patient's quality of life and it is preferable to avoid long-term indwelling catheters. Postoperative follow-up after reconstructive surgeries must be done through high-performance imaging studies of the urinary tract.

In this article we will present the case of a female patient with multiple congenital anomalies of the urinary tract and iatrogenic stenoses of the ureteropelvic junction and of the ipsilateral ureterovesical reimplantation site, following the reconstructive procedures she underwent during childhood.

Keywords:

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Introduction

Congenital urinary obstruction is one of the most frequent conditions affecting the urinary tract in children. It may lead to significant long-term health consequences and the management of these pathologies represents a major challenge for urologists.[1]

Vesicoureteral reflux (VUR) and ureteropelvic junction (UPJ) obstruction are two of the most common pathologies of pediatric urology and their coexistence is not unusual. VUR is associated in almost 40% of the cases of UPJ obstruction. [2,3] Whether their simultaneous presence is random or causally related is still uncertain. There have been discussions that an anomaly in the development of the ureteric bud could be the reason of the abnormal formation of both segments.[4]

In some cases of high-grade VUR, when the ureter is dilated and tortuous, kinking or even narrowing of the UPJ may exist. It is not always clear whether these can resolve spontaneously or if surgical correction of both pathologies is needed.[4,5] This association is specific to high-grade VUR, however a simultaneous surgical approach consisting in both uretero-vesical reimplantation and dismembered pyeloplasty is discouraged, due to concerns regarding the compromise of ureteral vascularization. Despite the lack of convincing evidence in this regard, either a conservative approach or a two-stage surgical approach, are still preferable.[1]

Infundibulopelvic stenosis refers to the hypoplasia of segments of the collecting system. It can affect the pyelocaliceal system at various sites and to various degrees. Focal forms involve only a very short segment of the pyelocaliceal system and may result in caliceal diverticula, cysts or UPJ stenosis. At the other end of the spectrum is the diffuse form, with the most severe cases resulting in a non-functional, multicystic kidney. [1,6,7]

In this article we will present the case of a female patient with multiple congenital anomalies of the urinary tract and iatrogenic stenoses of the ureteropelvic junction and of the ipsilateral ureterovesical reimplantation site, following the reconstructive procedures she underwent during childhood.

Case presentation

Patient O.S., age 27, having a complex history of surgical procedures of the urogenital tract performed in another hospital, currently with the suspicion of left ureteral duplication, hydrocalycosis of the upper unit and a percutaneous nephrostomy catheter inserted in the upper unit, is admitted into our service in February of 2019 for evaluation, requesting the removal of the nephrostomy catheter.

A thorough anamnesis reveals the patient's entire medical and surgical history:

At 11 years old she was diagnosed with left UPJ obstruction and bilateral VUR, for which both a left Hynes-Anderson dismembered pyeloplasty and a bilateral Cohen reimplantation were performed.

In October 2017 the patient was diagnosed with ovarian and pelvic endometriosis for which she underwent an exploratory laparoscopy, right ovarian cyst excision and aspiration of ascites.

One week postoperatively, she developed a left renal cholic and left grade III hydronephrosis was discovered. A right ureteroscopy was performed and a left ureteroscopy was attempted, upon retrograde injection of contrast discovering a normal right ureter and normal right collecting system, but a filiform left ureter which didn't allow the passing of the metal guidewire. She was also treated for a Klebsiella sp. urinary tract infection (UTI).

In July 2018, she undergoes a second laparoscopic surgery for the removal of left ovarian endometriomas.

A year after being diagnosed with left hydronephrosis, the patient is admitted to the local county hospital for severe urosepsis (urine cultures positive for E.Coli upon admission) and left grade II hydronephrosis, for which a left percutaneous nephrostomy was placed, draining turbid-looking urine.

Throughout this nearly one month long hospital stay, the patient needed orotracheal intubation with several days of mechanical ventilation (also developing a respiratory tract infection with coagulase-negative white hemolytic staphylococcus), became anemic to the point of 6.7g/dL of Haemoglobin – requiring repetead transfusions of red blood cell units, underwent a third surgery – exploratory laparoscopy – with no pathological intraabdominal findings, was later on confirmed with a Clostridium Difficile infection which rescinded under dedicated treatment, and she developed a presacrate decubitus lesion which got infected with Candida Albicans, required multiple descalings and healed fully after 6 months.

At the beginning of November 2018 the patient was discharged from the county hospital and almost 3 weeks later she was again diagnosed with a UTI – this time with Serratia Marcescens, with sterile urine culture after Cotrimoxazol treatment.

In January 2019 she returned to the county hospital and the nephrostomy catheter was replaced. She was once again diagnosed with Serratia Marcescens UTI, was treated initially with Ceftazidim during her hospital

stay, after which she continued the treatment at home with Cotrimoxazol.

At the moment of presentation to our clinic at the end of February 2019, the patient was in good general condition, was afebrile, the left nephrostomy catheter was permeable and the urine both from the catheter and from the bladder was clear. A left anterograde pyelography through the nephrostomy catheter was performed, which revealed grade II hydronephrosis, a filiform left ureter with a winding trajectory, visible only until the lower lumbar segment. At that time, it was decided to keep the nephrostomy catheter and delay any further procedures in order to gain some distance from the septic episode previously described.

In May 2019 the patient returns to our clinic as it was scheduled. The urine culture taken on admission was positive for Klebsiella sp. ESBL (susceptible only to Amikacin and Imipenem) and Proteus sp. (susceptible to Amikacin, Amoxicillin/Clavulanic acid, Ceftazidime, Ceftriaxone, Ciprofloxacin and Meropenem). Specific antibiotic treatment is administered and the decision is made to continue the tests in order to establish the current status of the patient.

As the left nephrostomy catheter is once again replaced, the anterograde pyelography shows a bifid renal pelvis, with the upper unit containing the superior and middle calices and the lower unit consisting of just the inferior calix, an extremely narrow communication existing between the two units (either a focal form of infundibulopelvic stenosis or an iatrogenic stenosis), and the nephrostomy catheter having the loop in the pelvis of the upper unit and passing through the renal parenchyma posteriorly to the lower calix, without intercepting it [Fig. 1].

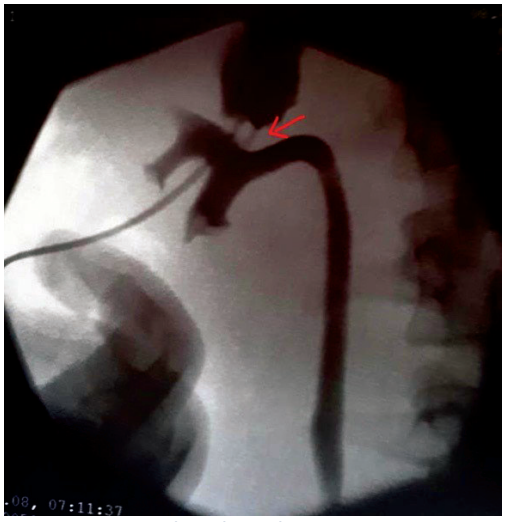

Figure 1. Anterograde pyelography aspect. Narrow communication (red arrow).

The patient desired the removal of the nephrostomy catheter, however at that stage that was not possible without resulting in another episode of renal cholic and possibly urosepsis. By mutual agreement with the patient, several options were considered:

Internal urinary drainage through a double-J ureteral stent – an option which would also require left ureterovesical reimplantation for ease of repeated access

Left re-pyeloplasty

Left partial upper pole nephrectomy

Left total nephrectomy – as a last resort.

Ureterocalicostomy was not considered, as it would not address the issue of the faulty communication between the upper and lower units. The option for a laser incision and balloon dilation of the stenosis was excluded, in the context of the post-pyeloplasty modified anatomy and the risk of intercepting vascular branches.

Furosemide and contrast enhanced computed tomography shows: Both kidneys have normal nephrogram and excretion times. Normal, nondilated right-side collecting system and ureter. Right kidney blood supply consisting of two arteries and one vein. Left kidney has nondilated lower pole calices which drain into the ureter, but dilated upper and middle calices (up to about 21 mm), with no indications of another ureter connected to them and without a clearly visible communication with the lower pole calices. Left nephrostomy catheter loop in the upper unit. Two left renal arteries – a permeable main artery and a small caliber supernumerary artery emerging from the abdominal aorta anteriorly to the main artery, visible only until it gets in the proximity of the anterior branch of the main renal artery – from this point on being no longer identifiable.

When a cystoscopy was performed, the ureteral orifices could not be identified; during this procedure, methylene blue was injected through the nephrostomy catheter, however no expression of it could be seen in the bladder.

Subsequent selective arteriography of the left renal vessels revealed 3 left renal arteries emerging individually from the abdominal aorta:

A main artery which supplies blood to the upper 2/3 of the left kidney;

A second narrow artery emerging from the abdominal aorta anteriorly to the main artery, which provides for a portion of the anterior half of the lower third of the kidney;

A third artery emerging on the left lateral wall of the aorta, inferiorly to the inferior mesenteric artery, having a diameter of 2.8mm and supplying the posterior half and the convexity of the lower third of the kidney. [Fig. 2, 3]

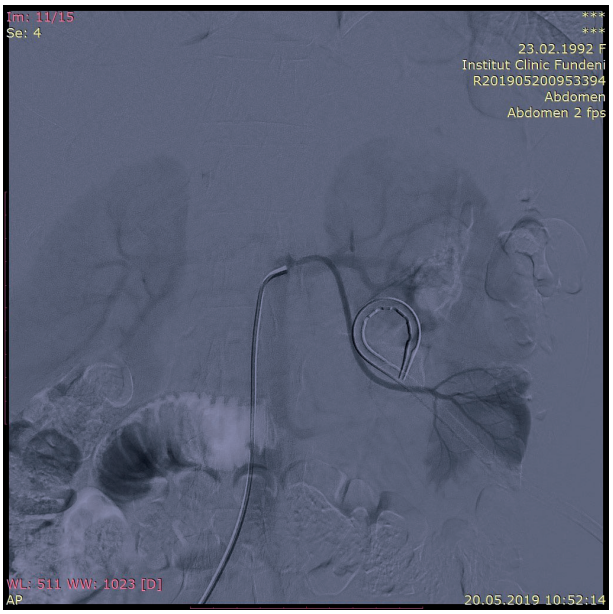

Figure 2. The main artery and the second artery.

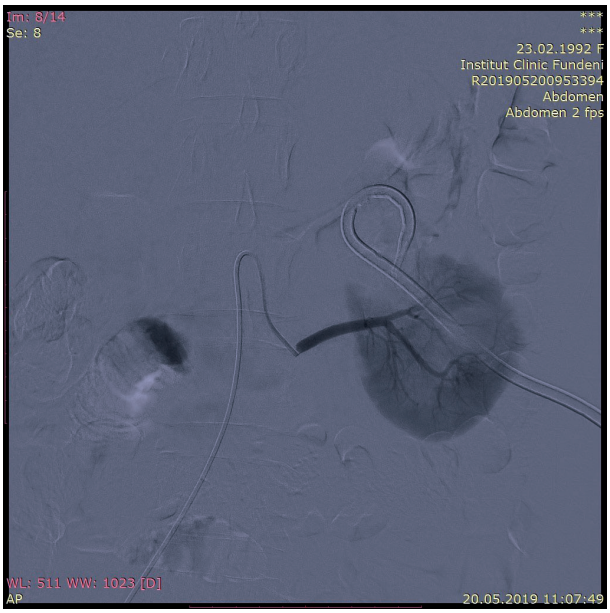

Figure 3. Third (lower polar) artery

The complex vascularization of the left kidney eliminates the option of a left partial upper pole nephrectomy. Also, considering the current anatomy of the collecting system, as well as the left pyeloplasty the patient underwent during childhood – which unavoidably created some degree of perirenal fibrosis, the option of a redo pyeloplasty is not considered feasible.

Taking into account the lack of identifiable ureteral orifices during cystoscopy, the lack of expression of the methylene blue inside the bladder when injected through the nephrostomy catheter as well as the impossibility for the passage of a guidewire into the left ureter during the attempted ureteroscopy in October 2017, together with the patient we decided for a left ureterovesical reimplantation, as a first step towards the reconstruction of the anatomy of the left upper urinary tract.

We carried out a Lich-Gregoire direct left ureterovesical reimplantation combined with a psoas hitch, over a 7CH 24cm double-J stent which was placed intraoperatively. The postoperative period was uneventful.

After three weeks the 7CH double-J stent was replaced with a 6CH stent under fluoroscopy guidance, managing to place it with the cranial loop inside the upper unit, by passing through the filiform communication between the two renal units. By utilizing an angle tip ureteral catheter twisted so that the tip pointed cranially, after a few attempts the surgeon managed to pass the guidewire through the small opening between the two renal units. The only size of stent that would pass through this area was 6CH.

Five days later the nephrostomy cat que regard the difficulty of retrograde catheterization of the ureteral orifice positioned superolateral, as an important disadvantage in cases where retrograde imaging studies, insertion of ureteral stents or urolithiasis management are needed.[25,26]

One of the long-term complications of this technique is obstruction, which in some cases might require reintervention. In cases like these, reimplantation is technically more difficult. The dissection and extensive mobilization of the ureter are crucial to the creation of an adequate submucous tunnel. It is preferable to create a new hiatus with its own submucous tunnel. If the ureter is too short, the psoas hitch method can ease the construction of the anti-reflux mechanism.[1]

Both the pyeloplasty and the ureterovesical reimplantation, regardless of access, are complex surgical procedures that in the hands of experienced urologists can lead to the recovery of the function of the affected kidney and can significantly improve patients' quality of life. Their success depends in equal measure both on the postoperative healing ability of the patient's body and on the training and experience of the surgeon.

Infundibulopelvic stenosis is is a very rare congenital malformation of the collecting system, is usually bilateral and it is commonly associated with vesicoureteral reflux, suggesting an abnormality of the entire UB, as well as with other congenital anomalies.[7,27] Renal biopsies of patients with various degrees of infundibulopelvic stenosis and end-stage renal disease revealed renal dysplasia proximal to the stenotic infundibuli and

varying degrees of glomerulosclerosis of the glomeruli which were not part of the regions with dysplasia. [27] Current therapeutic approach recommendations are regular imaging studies to evaluate the degree of dilation and the concurrent presence of other urinary tract anomalies such as VUR, as well as monitoring of renal function to include a baseline and yearly serum creatinine level, estimation of glomerular filtration rate, and urinalysis, in order to timely detect a potential decrease in the total mass of functional renal tissue which would lead to hyperfiltration injury in the remaining glomeruli, subsequent glomerulosclerosis and progressive renal disease. If vesicoureteral reflux is identified, either pharmacological or surgical management should be used to preserve renal function. Regular follow-up should be tailored to the patient needs and maintained throughout the patient's life. Progressive hydronephrosis warrants surgical treatment. Surgical management is based on the size and location of the stenosis.[1,27,28]

In cases like the one we described, where the patient presents with a complex pathology and an intricate medical and surgical history, retracing and understanding this history are always difficult and require a thorough anamnesis.

Access to a state-of-the-art Imaging department (with the necessary equipment and personnel for high-performance investigations such as URO-CT, MRI and interventional radiology procedures) is vital in order to have the highest degree of accuracy in determining the diagnosis and the current local anatomy, before establishing the course of treatment.

The doctor-patient relationship plays a very important role. All available treatment options must be explained to the patient in such a way that he or she understands the risks and the benefits, as well as the steps of the treatment plan, so that the doctor and patient choose together the best available option.

Seeing as the reconstructive surgeries must be performed on previously operated-on anatomical segments, they are expected to have a higher degree of difficulty. It is for this reason that we must strive to resolve the case through as few surgeries as possible, as each new surgery creates some degree of postoperative fibrosis, which can result in its own set of complications.

The full reconstruction of the urinary tract is not always possible in cases like the one here presented and as a last resort remains total nephrectomy – when the contralateral kidney is normal, or partial nephrectomy – when possible – in an attempt to preserve a number of nephrons that is as high as possible.

In the above presented case, due to the complex vascularization of the left kidney which eliminated the option of a partial nephrectomy, the peculiar anatomy of the collecting system and the expected post-pyeloplasty perirenal fibrosis – both of which indicated against a redo pyeloplasty – and the desire of both patient and surgeons to keep the kidney, which took nephrectomy off the table, the only viable option remained the internal urinary drainage via a double-J stent, which in turn imposed the reimplantation of the left ureter in a manner that would allow long-term periodic ureteral retrograde access.

The urologist's approach should always also bear in mind the patient's quality of life. When possible, it is preferable to avoid long-term indwelling catheters, which predispose to chronic UTIs (with microorganisms which may, in time, become multidrug-resistant) that can induce a state of permanent discomfort for the patient and might lead to recurrent hospital admissions, multiple invasive procedures, in some cases even requiring long-term antibiotic treatment.

Postoperative follow-up after reconstructive surgeries must be done through high-performance imaging studies of the urinary tract – which allow comparison with the preoperative description and the adequate evaluation of local postoperative recovery, as well as the early detection of possible complications.

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