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M.H. Kamran [1*], S.Elhan [1]., F.A.Iskakova[2]

[1] Assistant professor of the Kabul University of Medical science, Urology Department, Kabul, Afghanistan
[2]Associative Professor of Al-Farabi Kazakh National University, Almaty,Kazakhstan
*e-mail: m.hashimkamran@gmail.com

THE RARE CASE OF RETROCAVAL URETER IN UROLOGY SERVICE OF ALIABAD TEACHING HOSPITAL

Retrocaval ureter is an uncommon congenital abnormality in which the Inferior Vena Cava locates anterior to the ureter and causes its obstruction and hydronephrosis, as a result, and it is also considered one of the rare causes of hydronephrosis. The abnormality almost always occurs on the right because of the physiological position of the Inferior Vena Cava. It is a congenital abnormality, but the patients usually attend to the hospital due to hydronephrosis and right flank pain in the third and fourth decade of live. The abnormality needs surgical intervention and anastomosis of the ureter anterior to the Inferior Vena Cava.

A 40-year-old woman complaining from right flank pain and occasional nausea, vomiting and dysuria, was hospitalized in urology service of Aliabad Teaching Hospital on May 18, 2019. Renal ultrasonography and Intravenous Urography showed moderate hydronephrosis on the right, which was mistakenly diagnosed as uretero-pelvic junction obstruction. Consequently, after further exploration, retrocaval ureter was diagnosed and the retrocaval segment of the ureter was removed followed by end-to-end anastomosis of ureter on ureteral catheter.

The case implies that, clinically, retrocaval ureter seldom causes pain in the right flank and/or hydronephrosis, which can be mistaken for uretero-pelvic junction obstruction in IVU.

Key words: Inferior retrocaval ureter, Hydronephrosis, Pain in the flan.

Introduction

Retrocaval ureter is a rare congenital urologic anomaly. The incidence is reported about one in every 5000-10000 cases [1,2]. Since the Inferior Vena Cava is located on the right side; the abnormality usually occurs on the right. However, its occurrence on the left has also been reported in the literature in the presence of duplicated Inferior Vena Cava or, situs-inversus [3,4].

Inferior retrocaval ureter is also referred to as Circumcaval Ureter and/or Pre ureteral IVC, in which the ureter passes first medial and then posterior to the IVC. It, then, passes anteriorly, lateral to the IVC after orbiting around it. Eventually, it enters the bladder. The abnormality used to be considered a genetic defect which occurred during the development of the ureter. Nonetheless, now it is found out that the defect occurs in the development of IVC [3,4].

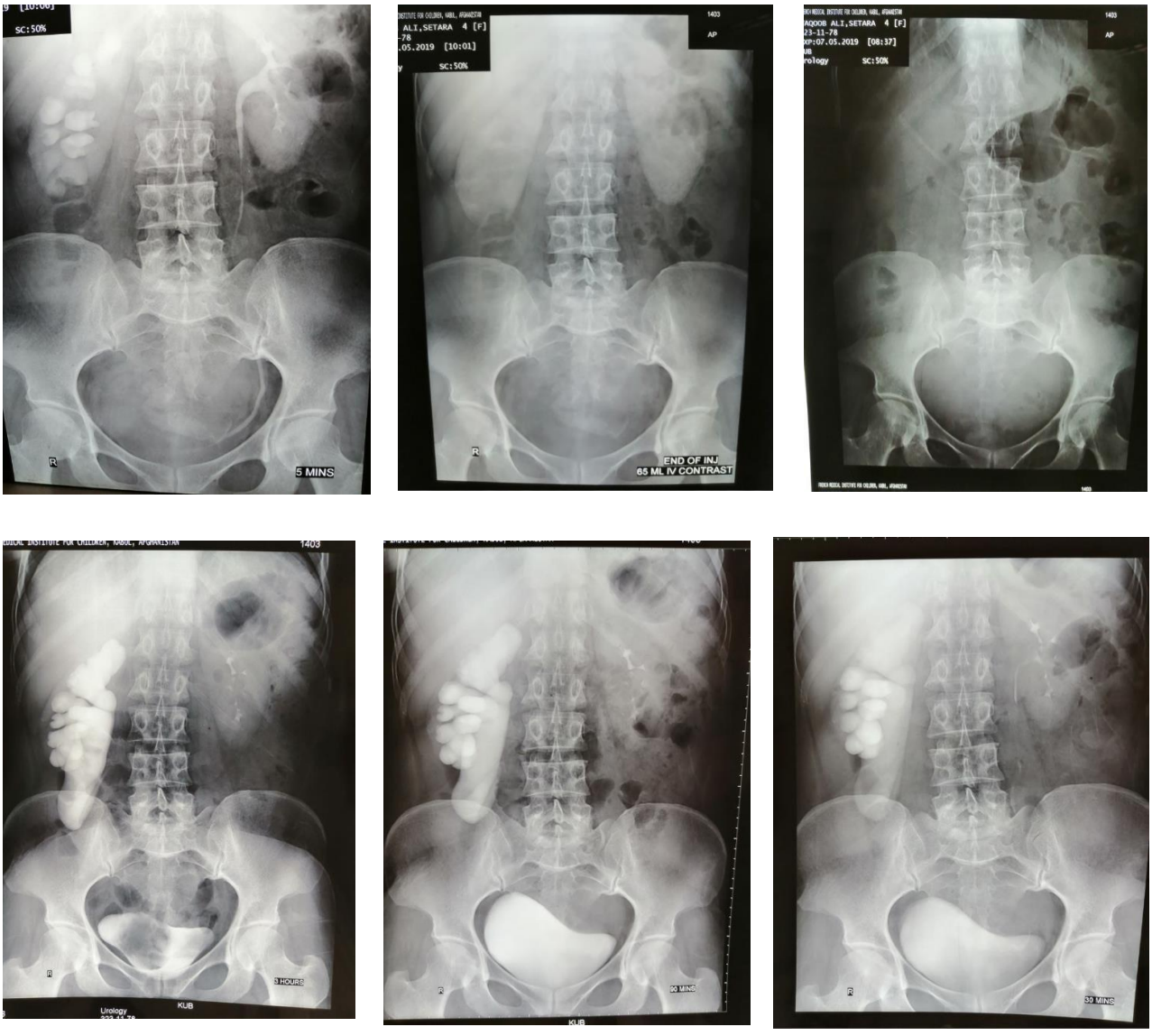
Despite the fact that it is a congenital abnormality, signs and symptoms of the disease appear in the third or fourth decade of life. The patients refer to hospital due to pain in the right flank and hydronephrosis, which is caused by immobilization of a segment of ureter and the pressure of IVC and psoas muscle on the ureter, and the ureter is trapped between IVC and psoas muscle. It is a congenital anomaly which causes hydronephrosis, so other causes of hydronephrosis specifically uretero-pelvic junction obstruction should be considered in differential diagnosis. This specific case indicates that not all hydronephrosis cases are caused by uretero-pelvic junction obstruction.

To make the case more prominent, we want to present the report of the scarce case of inferior retrocaval ureter causing right flank pain and hydronephrosis, and mistakenly diagnosed as uretero-pelvic junction obstruction in IVU.

Material.Case Report

A 40-year-old woman was complaining from right flank pain and occasional nausea, vomiting and dysuria, was hospitalized in urology service of Aliabad Teaching Hospital on October 18, 2019. Based on her explanation, the patient had been suffering from it for three years. According to the patient's explanation, she had suffered from the condition for three years, and pain had always been present and was described as cricks in the right flank. The patient added that the pain sometimes had deteriorated to colic, accompanied with nausea and vomiting. During the three years, the patient had

also suffered from dysuria, which had disappeared after taking a lot of liquid or over-the-counter antibiotics. Medicines like alpha blocker have been taken for the treatment of hydronephrosis, which hadn't resulted in full recovery from the disease. Both right and left flanks were found normal in inspection. However, right flank was palpated tender in bimanual examination. Morphy's punch sign (tenderness at the costovertebral angle due to percussion) was positive on the right in percussion. Ultrasonography revealed moderate hydronephrosis on the right. KUB was normal, but IVU showed distended pelvis and calices in the right side. (picture 1)



Picture 1: scout film is normal no stone, nephrogram shows normal distribution of contrast bilaterally, but 5-minute, 30-minute, 90 minute and 3-hour films show right side calyceal and pelvis distention clearly.

Laboratory examination result of urine was as below: Glucose… nil
pH 5
Albumin trace
RBC… 7-8
WBC… 12-14
Epithelial cell many

Laboratory examination result of blood was as below: Monocyte… 2%
Lymphocyte… 31%
Neutrophil 65%
WBC… 7000
Hb 12.2
Urea… 30
Creatinine… 0.8

FBS… 108
HBS… negative
HCV negative

The patient, diagnosed with right moderate hydronephrosis due to right uretero- pelvic junction obstruction, was nominated for open pyeloplasty. The operation, then, was carried out on October 22.2019. Under general anesthesia, a lumbotomy incision was done on the right side. All the layers were incised one by one and the peritoneum was pushed medially. Consequently, the ureter was found posterior to the peritoneum. Unexpectedly, it was found out that the ureter, in the middle part, passed from medial to posterior and subsequently from lateral to anterior side of the Inferior Vena Cava. The distended superior ureter was dissected up to pelvis, which was also distended. The ureter was cut in the superior part and the retrocaval segment of the ureter, which was collapsed and stenotic, was removed. Then, ureteral stent was inserted into the ureter through pelvis and the ureter was given end-to- end anastomosis using suture vicryl 4.0. Drainage tube was inserted and the layers were repaired one by one. Finally, sterile dressing was applied and the patient was transferred to recovery room. Ureterostomy and drainage tube were removed on tenth and twelfth day of the operation respectively. The patient was discharged from the hospital on October 30.2019 after she got full recovery. Stent was removed by cystoscope 4 weeks after operation and intravenous urography of patient was normal with no hydronephrosis on 8 months of operation.

Discussion

Retrocaval ureter is an uncommon congenital abnormality the occurrence of which is 1 in every 1500 cases. Moreover, it is 3-4 times more common in men then in women. The first retrocaval ureter case was reported by Hoschstter in

1893 This is the first retrocaval ureter case being reported in a female patient in Urology Service of Aliabad Teaching Hospital.

First it was thought the abnormality is related to the ureter but embryologic studies revealed that it is related to Inferior Vena Cava [2,4]. Therefore, the term Pre Ureteral IVC is a more appropriated one for the abnormality.

It almost always occurs on the right side, as in the mentioned patient. However, if in some cases it occurs on the left, it will be accompanied by partial or complete situs inversus or duplicated IVC [3,4].

The ureter crosses the IVC medial to posterior. Then, it rotates around IVC following its way posterior and lateral to it and eventually turns medially and locates anterior to IVC. It, then, follows its normal route and leads to the urinary bladder, as it is seen in this specific case. Pelvis and the superior segment of ureter, prior to moving posteriorly, are dilated and tortuous forming a J or Fish hook shape [3,4]. Although it is a congenital abnormality, clinical manifestations appear in the third or fourth decade of life, and most of the patients refer with flank or abdominal pain [1,2,4,5]. It is mostly a vague pain or aching which is due to hydronephrosis and occurs intermittently.

Hydronephrosis is caused by immobility of a segment of the ureter and compression of ureter between IVC and Psoas muscle [5,6]. Nonetheless, some patients refer to hospital due to repeated urinary infection, hematuria, kidney stone and pyonephrosis, and the retrocaval ureter is sometimes diagnosed accidentally via radiographic examination carried out to find out other conditions. In this specific case, a 40-year-old woman is reported to have complained about right flank pain, whereas hematuria, repeated urinary infections and pyonephrosis were not found. Retrocaval ureter is divided to two clinical types.

Type 1, which is more common and forms 50% the cases, results in severe ureter obstruction and moderate to severe hydronephrosis. Type 1 includes the middle third or ureter, in which severe deviation of ureter is seen posterior to IVC. Type 2, which occurs in 10% of the cases, includes mild deflection of the ureter posterior to IVC. Furthermore, it either causes mild hydronephrosis or doesn't cause it at all [3,4,7,8,9]. Surgical operation (robotic, laparoscopic or open payeloplasty) is indicated to reform Type 1 because it is symptomatic. In the mentioned patient existed Type 1 which involved middle third of the right ureter, and resulted in moderate hydronephrosis. Thus, surgical open operation was carried out for the purpose of treatment.

Kidney ultrasonography revealed hydronephrosis and proximal hydroureter, in addition pelvis and proximal ureter distension are seen in IVU, whereas middle and inferior ureter is not usually seen. Hence, retrograde ureteropyelography is necessary for diagnosis, which was not carried out in this specific case. Retrograde ureteropyelography is important to carry out even though a CT-Scan has been done. MRI indicates the pathway of ureter and provides more details than CT-Scan about the abnormality. In spite of not being invasive, it exposes smaller amount of radiation to the patient than CT-IVU [2,4,10,12,13]. In this case, ultrasonography and IVU were carried out, while retrograde pyelography and CT-Scan were not performed because patient already had

IVU and due to economic issue we didn't advise CT-IVU, resulting in misdiagnosis because the middle and inferior parts of ureter are not visible in IVU.

The best treatment for Type 1 retrocaval ureter is surgery, which can be either robotically laparoscopically or open. The retrocaval segment of the ureter, which is immobile, is either removed or saved. Then, an end-to-end anastomosis is performed on ureteral JJ Stent between the two ends of ureter or between pelvis and ureter [1,2,3,4,14]. The retrocaval segment of ureter was removed, but anastomosis was performed on ureteral JJ Stent.

The abnormality should be differentiated from retroperitoneal fibrosis and retroperitoneal tumors both of which push the ureter and displace it. Retrograde pyelography, CT-Scan and MRI can help differentiate the two conditions. The abnormality was mistakenly diagnosed as uretero-pelvic junction obstruction in this patient [15]. Moreover, retrograde pyelography, CT-Scan and MRI were not carried out for diagnosis.

Conclusion

Retrocaval ureter is an uncommon congenital abnormality which is mostly ignored due to its uncommonness. The patients usually refer to hospital complaining about right flank pain in third or fourth decade of life. In IVC, the abnormality can easily be misdiagnosed as all the other conditions causing external pressure on the ureter such as uretero-pelvic junction obstruction, retroperitoneal fibrosis and tumors, so not all hydronephroses are caused by uretero-pelvic junction obstruction, which should be seriously considered. Some additional radiographic examinations such as retrograde pyelography, CT-IVU and MRI are required in order to make sure that the condition is accurately diagnosed. If the abnormality is symptomatic, robotic, laparoscopic or open surgery is indicated for treatment.

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