

Ectopic kidney: Associated diseases and treatment

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Abstract

Ectopic kidney (or “renal ectopia”) describes a kidney that isn’t located in its usual position.

The pelvic kidney is a rare congenital urinary malformation. However, it is the most common type of “ectopic” kidneys. It results from a lack of migration of the metanephros which remains in the pelvic position. It is often smaller than a kidney in the lumbar position, but functional. It can be associated with other malformations (urological, vascular, etc.), or present complications such as urolithiasis, hydronephrosis, urinary infection or even tumor.

In that case, Urolithiasis is the most common pathology of the pelvic kidney, it can be a source of serious complications. Its discovery requires an etiological investigation in search of a hereditary, metabolic or even infectious disease.

In practice, renal pelvic ectopia poses various diagnostic problems; the close relationships of ectopic kidneys with neighboring organs explain the borrowed semiology and diagnostic errors.

Alongside medical care, urological care is essential. The choice of treatment in pelvic kidney pathology depends on the pathology in question, its clinical and biological repercussions, as well as the patient's condition and its comorbidities. The use of open surgery and/or laparoscopy in cases of lithiasis pelvic kidney must remain exceptional after having eliminated all possibilities of LEC and endourology surgery.

Keywords: Ectopic; Kidney; Congenital; Malformation; Pelvic

1. Introduction

Ectopic kidneys are thought to occur in about 1 out of 900 births. But only about 1 out of 10 of these are ever diagnosed. They may be found while treating other conditions.

The most common type of “ectopic” kidneys is pelvic renal ectopia (60% of cases). The pelvic kidney is most often discovered incidentally, during an abdominal ultrasound examination. In this case, the kidney is often unharmed. However, the diagnosis can be made at the stage of complications.

In Morocco, the pathological pelvic kidney is a rare entity with no documented specific incidence rate.

Objective

This work aims to analyze the epidemiological aspects of pathological pelvic kidneys, and to evaluate their diagnostic and therapeutic management.

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2. Materials and methods

This is a retrospective study, covering a series of 30 cases presenting pathological pelvic kidneys, collected at the urology department of the Hassan II University Hospital in Fez over a period of 18 years from the year 2004 to the year 2022. In this study, we addressed the epidemiological aspect (frequency in relation to age, sex), but also the clinical and therapeutic aspects of pathological pelvic kidneys.

3. Results

Pathological pelvic kidney is found at a frequency of 0.8 cases/year, with a clear male predominance. The most common pathology of the pelvic kidney is urolithiasis. The average age is 35 years with a range of 17 years to 56 years, the peak is between 30 years and 50 years. The damage more frequently concerns the left side.

The clinical expression depends on the pathology of the pelvic kidney, but lacks specificity; the symptomatology is generally made up of abdominal pain like renal colic and hematuria, signs of urinary infection or the presence of an abdominal mass are possible.

The diagnosis is paraclinical, it is generally based on Plane urinary Tract film (X-ray kub). The CT scan is decisive in the event of a tumor on the pelvic kidney, it also allows a better study of kidney stones and their repercussions on the upper urinary tract.

The Plane urinary Tract film made it possible to highlight the presence of stones in the pelvic area in the case of lithiasic pelvic kidney, because in our study, all the stones were radiopaque, but was not conclusive in the case of hydronephrosis or tumor. IVU also made it possible to identify urolithiasis, but also the presence of hydronephrosis on a pelvic kidney, however it was not done in the case of tumor. Ultrasound, for its part, was unable to resolve the case of ureteral lithiasis or tumor. The abdominal-pelvic scan made it possible to make the diagnosis of pathological pelvic kidney and to highlight the pathology in question in 100% of cases. Finally, scintigraphy highlighted the presence of a silent kidney in 14.28% of cases.

Table 1 Sensitivity of imaging tests in the diagnosis of pelvic kidney pathology

Imaging tests	Sensitivity
X-ray KUB	78.57%
Ultrasound	85.71%
IVU	92.85%
TDM	100%

3.1. Pelvic kidney pathology

3.1.1. Urolithiasis

In our study, it is the most frequent pathology with a percentage of 78.57%. It was associated with hydronephrosis in 7.14%, with urinary infection in 28.57% and a nonfunctioning kidney in 14.28% of cases. The diagnosis was made by X-ray KUB , IVU , CT, scan and with less sensitivity by abdominopelvic ultrasound.

Table 2 Sensitivity of imaging tests in the diagnosis of urolithiasis

Imaging tests	Sensitivity
X-ray	100%
Ultrasound	92.85%
UVU	100%
TDM	100%

The size of the stones varied between 4 and 30 mm with an average of 11 mm

The stones were multiple in 50% and single in 50% of cases.



Figure 1 Radiopaque stone over the left pelvic area (Hassan II University Hospital in Fez)



Figure 2 IVU showing an ectopic left pelvic kidney (Hassan II University Hospital in Fez)

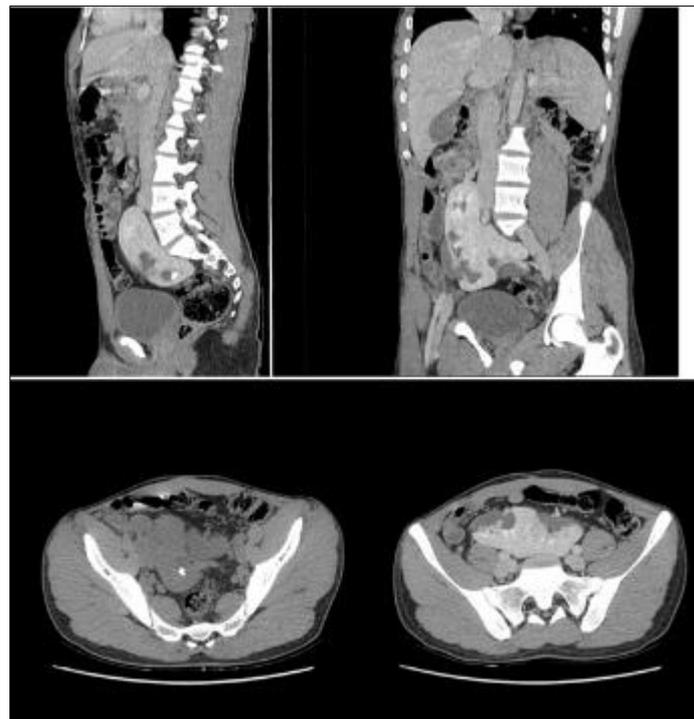


Figure 3 CT images showing a right horseshoe kidney in the pelvic position with the presence of multiple stones responsible for bilateral major hydronephrosis- images from the urology department of the Hassan II University Hospital in Fez

3.1.2. Hydronephrosis

Present in 14.28% of cases, associated with urolithiasis in half of the cases. The diagnosis is made mainly by IVU , ultrasound and CT scan.

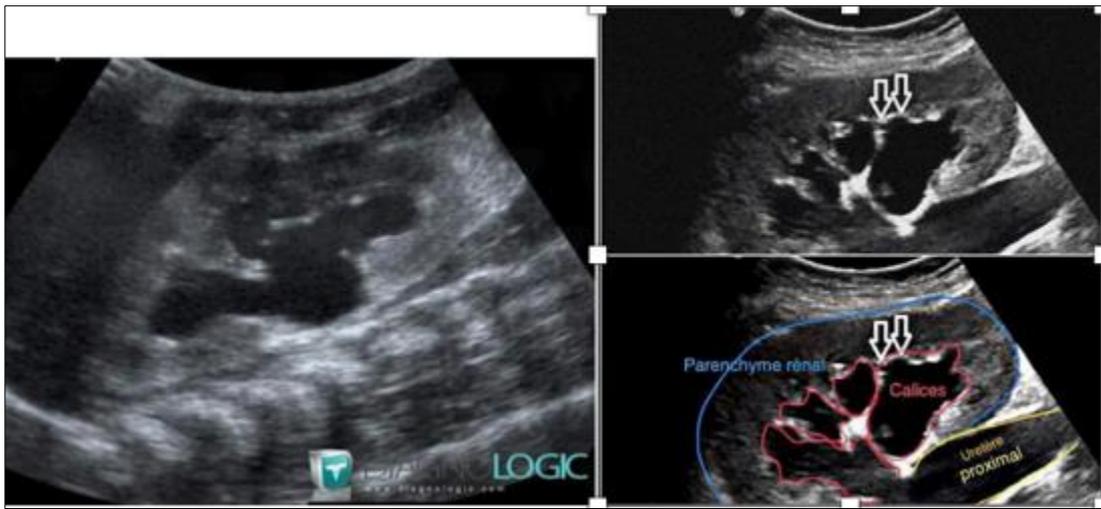


Figure 4 Hydronephrosis on renal ultrasound (pelvic kidney) (Hassan II University Hospital in Fez)

3.1.3. Kidney tumor

7.14% of cases with a diagnosis made on abdominopelvic CT SCAN .

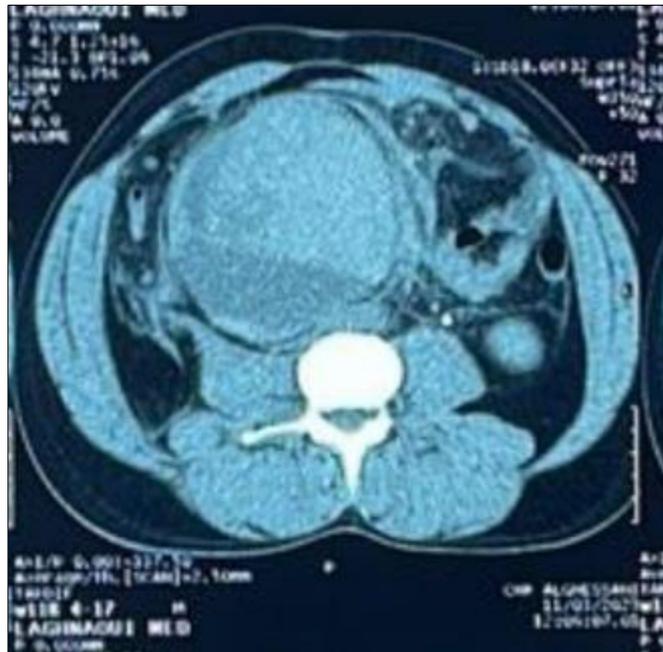


Figure 5 CT scan showing right pelvic kidney tumor - from the urology department of the Hassan II University Hospital in Fez

3.1.4. Nonfunctioning kidney

In 14.28% of cases, all associated with urolithiasis, demonstrated by renal scintigraphy.

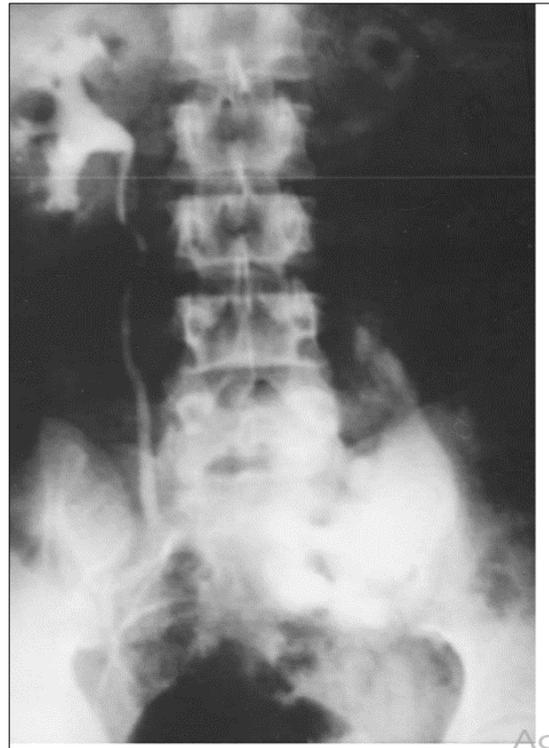


Figure 6 IVU showing an ectopic left presacral pelvic kidney with reduced cortical index, inferior calyx lithiasis, and stenosis of the pyeloureteral junction (PUJS) (Hassan II University Hospital in Fez)

3.2. Abnormalities associated with the pelvic kidney:

Imaging revealed congenital anomalies associated with the pelvic kidney, noting a horseshoe kidney in 21.42% of cases, and the syndrome of pyelo-ureteral junction in 21.42% of cases as well.

3.3. Impact on the urinary tract

An impact on the urinary tract is present in the majority of the cases; we find a dilationpyélo-calicielle in 78.57% , a reduction in the cortical index in 28.57% , a delay in secretion and/or excretion in 21.42% of cases, and finally a nonfunctioning kidney in 2 patients, or 14.28% of cases.

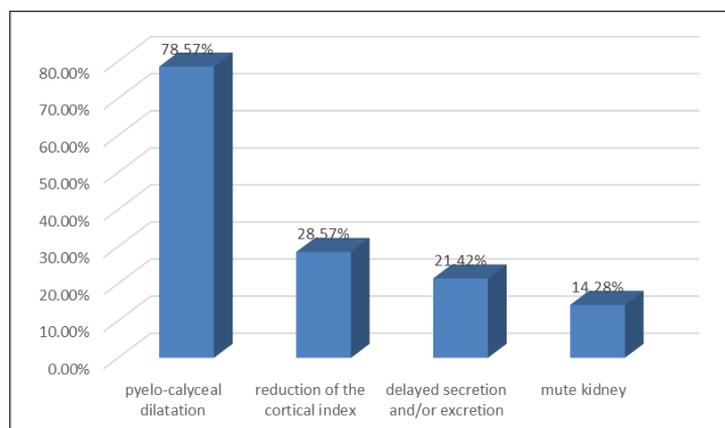


Figure 7 Distribution of patients according to the impact on the urinary system

3.4. Therapeutic modalities

The therapeutic modalities are the same as for a eutopic kidney with some difficulties related in particular to its pelvic position.

3.4.1. Urolithiasis

Flexible ureteroscopy-Laser

In our study, we report a series of 20 consecutive patients who underwent flexible ureteroscopy-Laser.

URS was performed under general anesthesia in 16 patients (80%) and under spinal anesthesia in 4 patients (20%). The postoperative length of stay varied between 1 and 8 days with an average duration of 2 days.

The average stone size was 11 mm (4-30).

The success of therapeutic ureteroscopy was 70%.

The main causes of failure of therapeutic ureteroscopy were bleeding, stone embedded in the ureteral wall, inability to remove through the Dormia catheter and inaccessibility to the ureter due to stenosis and the tortuous ureter often associated with the pelvic kidney which prevents the deviation of the flexible ureteroscope.

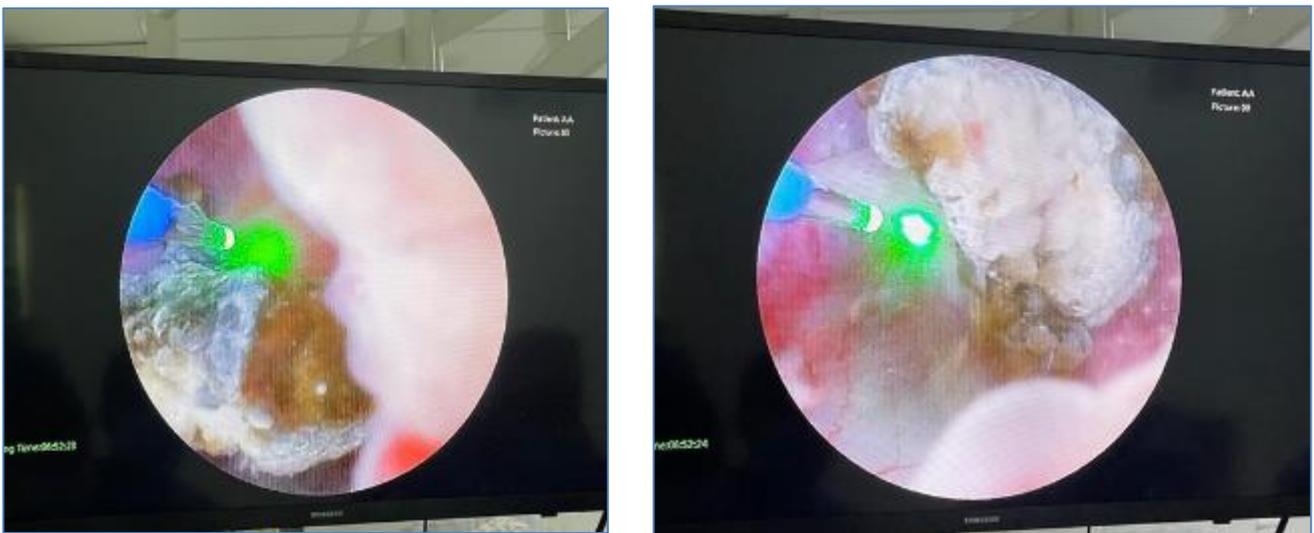


Figure 8 Images of Laser fragmentation (stones of pelvic kidney)–from the urology department of the Hassan II University Hospital in Fez



Figure 9 Images of extraction of stones using Dormia Basket(stones of pelvic kidney)–from the urology department of the Hassan II University Hospital in Fez

Open surgery

In our study, we report a series of 5 consecutive patients who underwent open surgery, the approach to the pelvic kidney was via a midline subumbilical incision in 40% of cases, an iliac incision in 40% of cases and a lombotomy in 20% of cases.

3.4.2. *Pyelo-ureteral junction syndrome on pelvic kidney*

Pyelo-ureteral plasty

Mostly carried out under laparoscopy, pyeloplasty consists of the removal of the junction between the pelvis and the ureter and therefore the narrowed area, plasty of the pelvis and the suture between the pelvis and the ureter.

The procedure requires the installation of a so-called double J stent for several weeks as well as a bladder catheter.

The results are above all functional, allowing good drainage from the kidney to the ureter in 90% of cases. On the other hand, the pelvis remains dilated for a long time, even permanently.

In our study, one patient benefited from pyelo-ureteral plasty, in 7.14% of cases.

Nephrectomy

A PUSJ prevents the drainage of urine which stagnates upstream of the stricture, which can lead to hydronephrosis or recurrent pyelonephritis. Thus, prolonged pyelo-calyceal dilation can cause destruction of the kidney with total loss of its function, causing a nonfunctioning kidney. In this case, there is an indication for nephrectomy for pyelo-ureteral junction syndrome.

The procedure involves removing the entire kidney along with the fat surrounding it.

In our study, one patient benefited from a nephrectomy on PUSJ (7.14%).

Ectopic kidney tumor

The radical treatment of the tumor is nephrectomy, subsequently associated with adjuvant anti-cancer treatment.

In our study, we have 2 patients that benefited from a nephrectomy (7.14% of cases of nephrectomy on pelvic kidney tumor)

3.5. Evolution

3.5.1. *Post-operative consequences*

The postoperative course was simple in all cases.

We encountered immediate complications such as hematuria in 21.42% of cases, and late complications such as chronic renal failure in 14.28% of cases.

3.5.2. *Length of hospitalization*

The duration of postoperative hospitalization varied between 4 and 12 days with an average duration of 6 days.

3.5.3. *Follow-up*

The success of the intervention is defined by the absence of residual fragment of lithiasis on the postoperative X-ray KUB in the case of lithiasis and the absence of tumor residue in the case of tumor on the CT scan.

In our series, the absence of residual fragment was observed in all our patients with urolithiasis.

Removal of the double J lead was generally done 6 weeks postoperatively.

In 71.42% of patients, the evolution was marked by an absence of stone recurrence and the progressive improvement of renal function in patients who presented renal failure; the evolution towards chronic end-stage renal failure occurred in 2 patients.

Stone recurrence was mentioned in 3 patients

In the two patients with the renal tumor, the postoperative CT scan result was satisfying.

4. Discussion

4.1. Pelvic kidney and associated anomalies:

In our study, the most common pathology is urolithiasis, with 78.57% of cases. A urinary infection was found in 42.85% of patients, most often associated with lithiasis. 7.14% of cases presented a tumor on the pelvic kidney, 14.28% hydronephrosis and the same for the nonfunctioning kidney. Benchekroun et Al (1), in their study, spoke of 45.45% of cases of renal lithiasis, the same for hydronephrosis on SJPU, 18.18% of cases of nonfunctioning kidney and 9% of cases of pyonephrosis. A urinary infection was found in 36.36% of cases. In the study by LAGOU D.A. (2) they found renal lithiasis in 8.33% of cases, urinary infection in 8.33% of cases as well, and 9.30% of cases presented a urological anomaly associated with the pelvic kidney (PUJ) in 25% of cases, hydronephrosis in 25% of cases, and malrotated excretory tract in 50% of these cases), anomalies of the contralateral kidney and the genital tract have been highlighted; the rest of the patients did not present any pathology, the diagnosis of pelvic kidney was made only during the exploration of chronic pelvic pain. DRETLER, OLSSON AND PFISTER (3) identified 29.16% of cases of infection, and 10.46% of cases presented gynecological anomalies associated with the pelvic kidney (all female)

The pelvic kidney is often accompanied by urological pathologies, the most frequent found in different studies being urolithiasis and hydronephrosis (often the result of an obstruction of the pyelo-ureteral junction), urinary infection is also frequently found, but renal tumors are rarer. The association of the pelvic kidney with malformations, particularly urological, is common. The frequency varied from 20.23 to 66% according to the different studies carried out on this subject (ARENA et al., 2007 (4)), (VAN DEN BOSCH et al., 2010 (5)), (UBETAGOYENA et al., 2011 (6)). The pyelo-ureteral junction syndrome was the most frequently found urological anomaly; cases of vesicoureteral reflux, mega ureter and retrocaval ureter have also been reported. In our study, the pelvic kidney was associated with urological malformations in 35.71% of cases (3 cases of horseshoe kidney and 3 cases presenting pyelo-ureteral junction syndrome).

Certain authors have highlighted genitourinary, cardiovascular, digestive or even bone malformations (Malek RS (7)) (LAGOU D.A. (2)) (EARL F. NATION, M.D (8)) (DRETLER, OLSSON AND PFISTER (3))

4.2. Imaging tests

4.2.1. X-ray KUB

In our series, the X-ray KUB made it possible to objectify the presence of stones in the pelvic area, 100% of the stones were radiopaque and therefore visible on the X-ray KUB.

But, it was not conclusive in the case of hydronephrosis or tumor.

Benchekroun et Al (1) report the same percentage; while Rohit Bhoil (9), in his study, the X-ray KUB was not conclusive and the diagnosis was made on the data from the IVU coupled with the scanner.

The X-ray KUB only provides information on the presence of a stone but does not allow us to identify its possible complications or to diagnose other pathologies of the pelvic kidney.

In our study, ultrasound was not able to decide in the case of ureteral lithiasis, and in the case of renal tumor. On the other hand, it made it possible to make the diagnosis of lithiasis in cases where the lithiasis was renal, as well as that of hydronephrosis; its sensitivity was 78.57% compared to 100% in the study of Benchekroun and AL (1) and that of Rohit Bhoil (9) and 90% for LAGOU D.A (2).

4.2.2. Ultrasound

Ultrasound, coupled with X-ray KUB, is the first-line examination to look for renal lithiasis; it is used for radiolucent stones not visible with X-ray alone; it makes it possible to objectify the impact on the upper urinary tract and sometimes to specify the location and size of the stones (visible on ultrasound generally from 4mm). (10)

4.2.3. IVU

In our study, IVU made it possible to identify urolithiasis in 100% of cases, and also the presence of hydronephrosis on a pelvic kidney, however it was not done in the case of tumor; either the IVU made it possible to make the diagnosis of pelvic kidney in 100% of cases and of the associated pathology in 92.85% of cases.

IVU also made it possible to highlight a nonfunctioning kidney in 14.28% of cases, confirmed by renal scintigraphy, thus, indicating nephrectomy.

In the study by LAGOU D.A. (2), the UIV made it possible to make the diagnosis of renal pelvic ectopia, and to specify the state of the kidneys and the excretory pathways, both homo and contralateral.

Benchekroun et Al (1) reported that IVU made it possible to make the diagnosis of pathological pelvic kidney in 100% of cases by showing the situation of the kidney and any associated lithiasis or malformations. EARL F. NATION M.D (8), as well as CINMAN ET AL. (11), spoke in their respective studies of the usefulness of UIV in the diagnosis of the pelvic kidney and the evaluation of its function allowing in several cases to confirm the presence of a silent kidney posing the indication for a nephrectomy.

In the study by Dretler, Olsson and Pfister (3), the pelvic kidney was not opacified with IVU in 16.27% of cases, visualization of the kidney was therefore possible by retrograde ureteropyelography.

4.2.4. CT scan

In our study, the abdominal-pelvic scan made it possible to make the diagnosis of pathological pelvic kidney and to highlight the pathology in question in 100% of cases. It is the same in the series of Benchekroun et Al. (1), David M. Hoenig (12), Soltani et Al (13) and LADOU D.A. (14) where the abdominopelvic CT was conclusive in 100% of cases.

In studies made by CINMAN ET AL. (11), DRETLER, OLSSON AND PFISTER (3), and EARL F. NATION, M.D. (8), the scanner made it possible to provide a lot of precision concerning the exact position of the pelvic kidney, its orientation but also its relationships with the neighborhood bodies.

Also, CT scanning is indicated when a tumor pathology is grafted onto the pelvic kidney (15); it makes it possible to study loco-regional and distant extension as well as the relationship of the tumor with neighboring organs. (1)

It may prove interesting in cases of bilateral pelvic kidneys looking for a possible renal symphysis. (16)

4.3. Therapeutic modalities

4.3.1. Urolithiasis

Flexible ureteroscopy

Weizer (17) report a success rate of flexible ureteroscopy estimated at 75% in the case of the pelvic kidney.

A study by Omer Faruk Boskurt et al (18), and associates, showed that flexible ureteroscopy is a minimally invasive, safe and effective treatment for small to medium sized stones on a pelvic kidney, with a rate of 84.6% success.

the choice of approach in surgery must be judicious and thoughtful, taking into account the anatomical relationships of the pelvic kidney and its vascularization, which are different from the kidney in normal position.

Open surgery

Boujnah (19) propose the extraperitoneal iliopelvic route for simple lithiasis and PUJS, and the transperitoneal route in cases of conditions requiring control of the renal pedicle. Sarvjeet Meravi (20), in their series, opted for a midline abdominal incision to easily access the retroperitoneal space because the vascular pedicle of the kidney was posterior.

4.3.2. *Pyelo-ureteral junction syndrome on pelvic kidney:*

Pyelo-ureteral plasty

In our study, one patient benefited from JPU plasty, i.e. 7.14% of cases.

In the study by Benchekroun et Al. (1), 36.36% of patients benefited from JPU plasty.

Nephrectomy

In our study, one patient benefited from a nephrectomy on SJPU or 7.14%.

In the study by Benchekroun et Al (1), 18.18% of patients underwent nephrectomy on SJPU.

4.3.3. *Ectopic kidney tumor:*

Cases of tumor pathologies of the pelvic kidney are not widely described in the literature.

C. Terrone (21), in his study reports that renal cancers in ectopic kidneys are exceptional, and that total nephrectomy is appropriate in this situation.

El Moussaoui and Aboutaib (22) also used nephrectomy in their study of a case of ectopic kidney cancer.

4.4. Evolution

4.4.1. *Post-operative complications*

The postoperative course was simple in all cases.

We encountered immediate complications such as hematuria in 21.42% of cases, and late complications such as chronic renal failure in 14.28% of cases.

The complications described in the literature are dominated by infection of the operating wound. (16)

4.4.2. *Length of hospitalization*

The duration of postoperative hospitalization varied between 4 and 12 days with an average duration of 6 days.

Esgbi (23) report an average similar to ours, while Meravi et al (24) report a duration of 8 days.

4.4.3. *Monitoring*

Prolonged follow-up of patients is essential to avoid recurrences, this is done by regular clinical examination with search for a possible urinary infection, evaluation of renal function, and X-ray and/or ultrasound if necessary.

In the series of Desai and Jasani (25), no stone recurrence was reported, and renal function improved in all cases who had impaired renal function with a rate of 100%, these results are similar to those of Aboutaib R. (22) Benchekroun et Al (1) also report a 100% surgical success rate with good progress whether for nephrectomies or conservative treatments.

C. Terrone (21), in his study about a case of pelvic kidney cancer, reports a good progress of the surgery without complications with a good long-term evolution after nephrectomy without recurrence on control examinations.

5. Conclusion

The pathological pelvic kidney is a rare entity, which can evolve quietly for a long time. Urolithiasis is the most common pathology of the pelvic kidney, it can be a source of serious complications. Its discovery requires an etiological investigation in search of a hereditary, metabolic or even infectious disease.

Alongside medical care, urological care is essential. The choice of treatment in pelvic kidney pathology depends on the pathology in question, its clinical and biological repercussions, as well as the patient's condition and its comorbidities.

The use of open surgery and/or laparoscopy in cases of lithiasis pelvic kidney must remain exceptional after having eliminated all possibilities of LEC and endourology surgery.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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