Clinical Medicine and Health Research Journal (CMHRJ)
Volume 02, Issue 03, May - June, 2022
Page No. 125-128

Case Report

Bilateral Pyeloureteral Junction Syndrome on Horseshoe Kidney - A Case Report

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Received: 04 March, 2022 **Accepted: 30 April, 2022** **Published: 02 May 2022**

Abstract:

Horseshoe kidney is the most common type of congenital renal fusion defect in children. The horseshoe kidney occurs in approximately 0.25% of the general population. Isolated horseshoe kidneys, which account for one-third of horseshoe kidneys, may be asymptomatic and go undiagnosed until adulthood. However, the presence of a horseshoe kidney in the pediatric population warrants further evaluation because the horseshoe kidney is frequently associated with other abnormalities: 78.9% including pyelourethral junction syndrome. In addition, the horseshoe kidney is associated with an increased rate of renal complications in adults, such as stones, ureteropelvic junction (UPJ) obstruction, trauma, infection, and tumors. Ureteropelvic junction syndrome is a disease that causes a failure of urine flow from the pelvis to the ureter. It is the most common cause of hydronephrosis in children. The diagnosis was made on the basis of symptoms, mainly pain and urinary tract infection, and required surgical treatment.

Keywords: Pyeloureteral junction, Child, bilateral, horseshoe kidney, hydronephrosis

Introduction

Pyeloureteral junction syndrome is defined as a blockage or obstruction of urine flow from the kidney to the proximal ureter. Therefore, it is important to understand how to diagnose and treat this condition. Uroscanner is the radiological examination of choice for diagnosing hydronephrosis, prenatal diagnosis of hydronephrosis accounts for approximately 1-5% of all pregnancies [1-4].

Open surgical pyeloplasty (Anderson-Hynes technique) has long been the gold standard treatment for pyeloureteral junction anomaly. With the advent of minimally invasive surgery in the last decades, laparoscopic pyeloplasty (LP) with all its varieties has gained a great place as an alternative to open surgery. Other "minimally invasive" endoscopic therapeutic approaches have gained momentum in the last decade in the treatment of pyeloureteral junction obstruction(PUJ) [5-6].

Patient and Observation

Patient information: the age of our patient was 14 years, without any particular pathological history, the pyeloureteral junction syndrome was revealed by functional signs: abdominal pain associated with low back pain, no notion of urinary tract infection or fever or abdominal bloating.

Clinical findings: the clinical examination is usually poor in the context of pyeloureteral junction anomaly. Examination of the kidneys: tenderness of both lumbar fossae, absence of lumbar contact or renal ballooning.

Paraclinical assessment: the determination of urea, blood creatinine and glomerular filtration rate, all came back normal. The cytobacteriological examination of the urine was requested, it came back negative. The uroscanner showed that the two kidneys are joined by their lower pole of regular contour presenting a nephrography in excretory vascular time and sitting of an important pyelocalic dilatation predominating at the level of the lower calicium groups without dilatation of the sub-pyretic ureters. On the right side, there were 3 coralliform stones in the middle and lower calicium groups, measuring 5 mm in length, with an estimated density of 1200 HU.

Therapeutic intervention: the main operative indications were: the presence of painful symptomatology, the appearance on uroscanner of major dilatation of the cavities and the presence of associated renal lithiasis Figure 1,2.

The pyeloplasty performed in the Department was according to the technique of Anderson and Hynes. 1st time the patient benefited from a bilateral jj rise. The delay of management before the second surgical procedure was 3 days. The second procedure consisted of pyeloplasty and reduction of the pelvis and its suture by separate stitches with PDS 4/0.

Follow-up and outcomes: the average length of the

postoperative hospital stay was 3 days, the resumption of transit was the second day, the postoperative analgesia was assured for 48 hours. No postoperative complications were described.

Patient's perspective: during his hospitalization after the surgical treatment, the patient appreciated the functional results and the quality of care he received.

Discussion

Horseshoe kidney is a malformation resulting from the symphysis of the two kidneys, usually by their lower pole. The incidence is estimated at 0.25% of the general population. There is a clear male predominance with a sex ratio of 2. All urological diseases can be seen but the incidence of some of them seems to be higher than in normal kidney. It is also important to note the frequent association with other malformative pathologies, notably the pyelourethral junction syndrome. Hydronephrosis due to pyelourethral junction anomaly is the most frequent complication, found in nearly one third of cases, most often unilateral and rarely bilateral.

Epidemiology [7] : the pyeloureteral junction anomaly is the most frequent malformative uropathy in children and represents more than 60% of neonatal urological anomalies; it is also the most frequent malformation found in adults. Thanks to the generalization of the practice of obstetrical ultrasound. The average age of discovery of the pyeloureteral junction anomaly has undergone a significant evolution.

Nowadays, anomalies of the pyeloureteral junction are diagnosed and treated before the age of one year [8]. VALLA and BELMONT [9] report an average age of no more than 13 months, it should be remembered that the cases in these series were diagnosed for the most part in the antenatal period and treated by laparoscopic approaches [10].

Postnatal diagnosis: the most common reason for consultation is abdominal or lumbar pain, of moderate intensity, dull, intermittent, and favored by heavy drinking that puts pressure on the pyelocalic cavity [11]. Recurrent urinary infectious episodes are quite frequent and require a search for a urinary malformation. Infectious episodes are often atypical, characterized by isolated febrile attacks, dehydration or febrile gastroenteritis [12].

Explorations:

Standard trans-abdominal ultrasound [13]: Ultrasound is used to detect persistent hydronephrosis, determine the level and severity of obstruction, and assist in diagnosis and appropriate treatment. The study of the contralateral kidney is essential to look for bilateral character, compensatory hypertrophy, or to screen for associated malformation, or renal lithiasis.

Uroscanner [14]: the scanner can detect the location and cause of the obstruction such as the polar vessels crossing the junction. Indirect signs that point to acute obstruction: ureteral dilatation, nephromegaly, density of the affected kidney compared to the contralateral kidney.

The arterial phase is very important; this time is well adapted to look for a polar vessel crossing the pyeloureteral junction. The Uroscanner has become a reference examination in the evaluation of the pyeloureteral junction syndrome in adults, because it allows to make the diagnosis and to search for the etiology and the associated renal anomalies.

The treatment options: pyeloplasty promoted by Küss, Anderson Hynes remains the reference treatment for pyeloureteral junction syndrome, whether it is performed open, laparoscopically or robotically assisted. In parallel, endoscopic techniques have been developed over the last ten years. They aim to reduce the morbidity of surgery and shorten hospitalization times.

Four most frequently discussed procedures in case of junction syndrome, including two endoscopic techniques.

Pyeloplasty according to küss anderson hynes: The procedure consists in resecting the stenotic portion of the pyeloureteral junction and the "chubby" part of the pelvis. The ureter is then spatulated with healthy tissue and sutured with fine absorbable suture to the lower part of the pelvis in a sloping position. This procedure is more and more often performed transperitoneally by laparoscopy or using the assistance of the Da Vinci robot. It is generally consolidated by the installation of a double J catheter, which allows to avoid or limit postoperative fistulas [16]. This surgical approach is associated, in case of associated compression by a polar vessel, with the uncoiling of this ureter with respect to the polar vessel, which limits the risk of secondary stenosis [16,17].

The calico-ureteral anastomosis [18]: this procedure is generally proposed in case of failure of a pyeloplasty if the lower pole of the kidney is atrophic. The procedure consists of resecting the atrophic renal parenchyma opposite the bottom of the lower calyx and anastomosing the "healthy" ureter directly to the calyx, the indications being mainly represented by failed pyeloplasty.

Endopyelotomy : the procedure consists of endoscopic incision of the pyeloureteral junction which has been previously prepared by a double J probe. The incision is longitudinal and concerns the whole wall to reach the peripyelic fat. Different techniques or energies can be used to section this junction endoscopically (endoscopic scissors, cold blade, cutting loop, laser).

The site of the incision is determined by precise analysis of the CT scan in order to avoid haemorrhagic complications linked to accidental section of a polar vessel compressing the junction [19].

The electro-incision or Acucise® section of the junction: it consists of sectioning the pyeloureteral junction under radiological control using a low pressure angioplasty balloon with a conductive wire which, when applied posterolaterally to the ureter, delivers a sectioning current to the narrowed area. The procedure is performed under fluoroscopic control and the intraoperative effectiveness of the section is identified by the extravasation of the contrast medium around the excretory tract [20].

Conclusions:

Pyeloureteral junction syndrome is defined as an obstruction of urine flow from the kidney to the proximal ureter. This obstruction can lead to increased back pressure on the kidney, hydronephrosis and progressive deterioration of renal function. The most common reason for consultation is abdominal or lumbar pain in the form of renal colic. Ultrasound and scintigraphy are usually sufficient to make and confirm the diagnosis. Technical and scientific progress has contributed to the development of surgical techniques, more efficient and minimally invasive with attractive functional results.

Conflict Of Interest

The authors declare no conflicts of interest

Authors' Contributions

All authors contributed to the conduct of this work. All authors also declare that they have read and approved the final version of the manuscript.

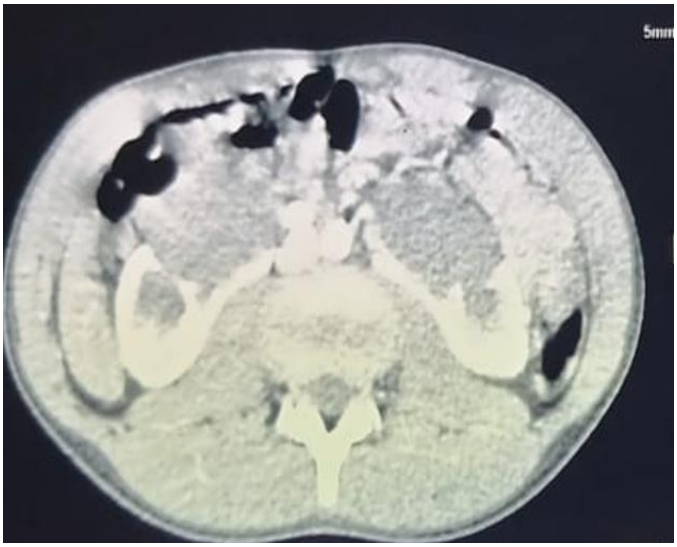

Figure 1: Cross section of a bilateral pyelo ureteral junction syndrome on a horseshoe kidney.

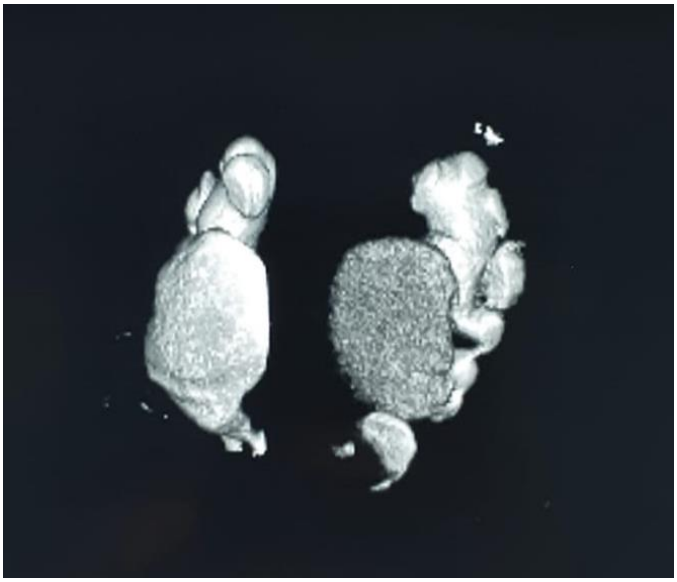

Figure 2: 3D reconstruction of a bilateral pyeloureteral junction syndrome on a horseshoe kidney.

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