CLINICAL CASE

HYDRONEPHROSIS IN A HORSESHOE KIDNEY

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Abstract

Horseshoe kidney is the most common fusion anomaly of the kidney with a prevalence of 1/400, being more frequent in males. This anomaly is characterized by malrotation, abnormal blood supply and other anatomic anomalies and has a propensity to form UPJ obstruction in up to one-third of the cases, being more common on the left side (89% of the cases). UPJ obstruction on the right side, being so rare, we report the case of a 5 years and 6 months old male patient suffering of horseshoe kidney and right side UPJ obstruction.

Keywords*: horseshoe kidney, hydronephrosis, pyeloplasty, ureteropelvic junction obstruction*

Introduction

Horseshoe kidney is the most common fusion anomaly of the kidney. The incidence of horseshoe kidneys is between 1/400 to 1/1800 autopsies and is more frequent in male patients.[1]. Horseshoe kidney is characterized by malrotation, abnormal blood supply and other anatomic variations.[2] The most common complications of this anomaly are hydronephrosis, reflux, lithiasis, recurrent UTIs.

Case report

We present the case of a male patient, aged 5 years and 6 months, with the prenatal diagnosis of right pyelocaliceal system dilation and suspicion of horseshoe kidney.

At the age of 3 months the patient underwent IVU showing malrotation, a slightly dilation of the renal pelvis and normal calyces on the left side but dilated pyelocaliceal system with ballooned calyces on the right side. The right ureter couldn't be visualized. The deviation to vertical of the long axis of the two pyelocaliceal systems highly suggested the horseshoe kidney diagnosis.

Another IVU at 9 months showed the same as the one at 3 months. The patient underwent also a VCUG indicating no VUR and no post-micturition residual urine. At the same age (May 5th 2010) the patient underwent surgery: right Hynes-Anderson pyeloplasty. During the operation the horseshoe kidney diagnosis was confirmed. Three months after the operation the IVU indicated normal left kidney, fusion anomaly - horseshoe kidney, secretion and excretion present on the right side, low degree dilation of the right pyelocaliceal system.

One year after the operation IVU showed a minimal degree dilation of the right pyelocaliceal system. Two years after the operation IVU indicated residual dilation of the right pyelocaliceal system, but in a lower degree than in the past. At that time the ureter could be visualized at 3 hours sequence. On September

30th 2013 a new IVU showed stationary dilation of the pyelocaliceal system and no ureter.

On Oct 1st 2013 the patient underwent exploratory cystoscopy and right ureter double-J stent placement. Three months after the placement, the stent was removed. In February 2014 the patient presented an UTI episode. On May 16th 2014 the IVU indicated bilateral secretion and excretion, stationary aspect on right side with persistent pyelocaliceal dilation.

On September 16th 2014 the patient was admitted on our ward for recurrent UTIs and caliceal dilations for further investigation and continuing the treatment. The physical examination was normal. Abdominal ultrasound examination showed horseshoe kidney, 8 mm isthmus thickness, anteroposterior right pelvis diameter of 30 mm, renal parenchymal thickness of 6-13 mm; on the left side: anteroposterior left pelvis diameter of 20 mm, renal parenchymal thickness of 13-14 mm, unseen ureters, thin urinary bladder walls. The abdominal CT scan showed horseshoe kidney having the isthmus placed anteriorly of L4 body, 8-12 mm thickness. On the right side: 8-12 mm parenchymal thickness, dilated pelvis: 30/40 mm; on the left side: 14 mm parenchymal thickness, 20/17 mm pelvis; normal ureters.

In spite of the first surgery, pyelocaliceal dilations were persistent and several UTI episodes were repeated. Taking into consideration this aspect and also the test results from above, the patient underwent revision pyeloplasty with classic approach. Before the operation the patient was rehydrated and 50-100 mg/kg/day of Ceftriaxone was administered. The surgical intervention was performed under general anesthesia with orotracheal intubation. An incision beginning at the right costo-vertebral angle and continuing lateral and caudal, ending at the vertical line passing through the anterior superior iliac spine was practiced. After the dissection of the abdominal wall, the Zuckerkandl fascia, covering the kidney, was discovered. The adipose capsule of the kidney was very adherent to the renal capsule due to postoperative fibrosis. The renal pelvis was dilated and anteriorly rotated. The renal pedicle was formed by the main renal artery and aberrant polar arteries (anteriorly and posteriorly of the renal pelvis). The renal parenchyma was about 8 mm thickness and the proximal ureter was surrounded by postoperative adhesions. The ureter was high inserted and stenotic from the UPJ to the isthmus (2mm diameter). Then, right pyeloplasty was performed, placing a transanastomotic double J stent of 4.7 Fr.

Postoperative evolution of the patient was good and he was discharged 5 days after the operation.

At one year follow-up after the intervention we found signifiant improvement of the pyelocaliceal dilation with satisfactory urinary flow.

Discussions

Horseshoe kidney, a developmental defect occurring during embryogenesis involves the fusion of the lower poles of the two kidneys. The two kidneys are joined by a bridge of normal renal tissue or a dysplastic, fibrous tissue called isthmus [1]. This common fusion defect is a rare condition having an incidence of 1/400 [1]. In this anomaly there are a lot of anatomical malformations of blood vessels, ureters and renal pelvises.

Some of the authors report an impaired drainage and hydronephrosis in 15% of the horseshoe kidney patients, due to a distortion of the ureter along the isthmus, high insertion and abnormal take off of the ureter or due to compression by abnormal blood vessels [3]. Other authors report that the propensity of a patient suffering of horseshoe kidney to form UPJ obstruction is up to one-third of cases[2] (15-33% [4]).

The peculiarity of the case presented is the rare association between horseshoe kidney and UPJ obstruction. Furthermore the obstruction of the UPJ on the right side, as in our patient, is even more rare. Stephen Faddegon et al. reported a series of patients having horseshoe kidney and UPJ obstruction in which 89% of the UPJ obstructions were on the left side [5].

Normally, the surgical treatment of the horseshoe kidney should be done only if complications like UPJ obstruction or urolithiasis occur. In case of UPJ obstruction, the most common indication for surgical intervention in a patient with horseshoe kidney, there are several approaches that have to be

considered. There could be robotic, laparoscopic or open approach and the management of UPJ obstruction could be open dismembered pyeloplasty with isthmectomy and nephropexy of the ipsilateral kidney[6] or simple Anderson–Hynes pyeloplasty without division of the isthmus and lateropexy of the kidney, which is a highly effective and safe procedure [7]. In our case we preferred an open correction of the obstruction of the UPJ due to postoperative adhesions and fibrosis which could have made the laparoscopic procedure very difficult.

Conclusion

We presented a rare case of right side hydronephrosis in a horseshoe kidney due to UPJ obstruction, a challenging case who needed 2 surgical interventions. The surgical interventions should be performed only by experienced surgeons because of the anatomical anomalies of the renal pelvises, ureters, blood vessels, which can be sometimes challenging.

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