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Case Report

Ureteral switch for bilateral ureteropelvic junction obstruction in a case of Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome

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KEYWORDS
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Abstract
Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome is characterized by congenital aplasia of the uterus and the upper part of the vagina. Secondary sexual characteristics and karyotype are normal. This syndrome affects at least 1 out of 4500 women.
MRKH may be isolated (type I) but it is more frequently associated with other malformations (MRKH type II).
We present a typical case of MRKH type II associated with bilateral pelvic kidneys ectopia, ureteropelvic junction (UPJ) obstruction and high inserting ureters. The ureteral switch was performed at the time of pyeloplasty to prevent postoperative obstruction secondary to the angulation.

Case presentation

A 34-year-old female with known Mayer-Rokitansky-Küster-Hauser syndrome (uterine and vaginal agenesis) was referred to the Urology Service of Cliniques Universitaires Saint Luc, Brussels for recurrent urinary tract infections and occasional hypogastric pain.

Habits of this patient include smoking of 10 cigarettes a day. The patient was socially and physically active and had a normal life.

No other malformations were recognized.

Renal function and urine analysis did not show any anomaly.

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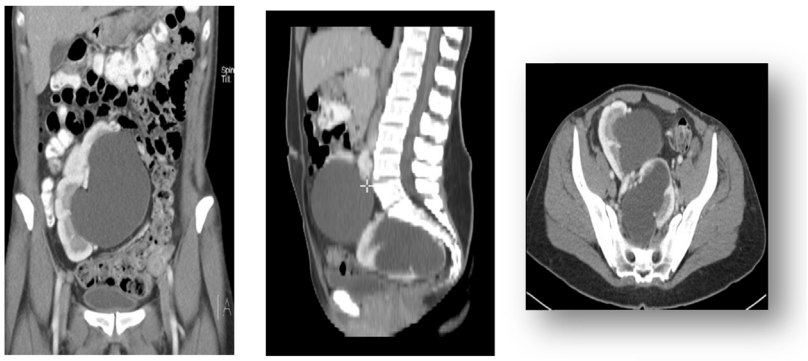


a b c

Figure 1 CT scan of abdomen showing bilateral pelvic kidneys (a: coronal, b: sagittal and c: transversal slices).

A contrast enhanced CT of the abdomen showed the two kidneys in pelvic position without fusion, slightly delayed excretion on both sides with major bilateral hydronephrosis.

Both renal arteries emerge from the common iliac arteries and describe an aciform path.

MRI of the pelvis showed two separated pelvic kidneys in close relationship to the iliac axis and no significant vascular compression of the UPJ (Fig. 1).

Retrograde uretero-pyelography showed displacement of each ureter to the opposite side due to the severity of the hydronephrosis.

A sub-umbilical laparotomy and exploration of the pelvis confirmed the diagnosis of MRKH type II syndrome with small amount of fibromatous tissue close to the right ovary, the absence of the uterus and a double pelvic kidney situation with high insertion of the ureters into largely distended renal pelves (Fig. 2).

A dismembered pyeloplasty was performed and kinking of ureters was prevented by crossing the ureters ("*Ureteral switch*").

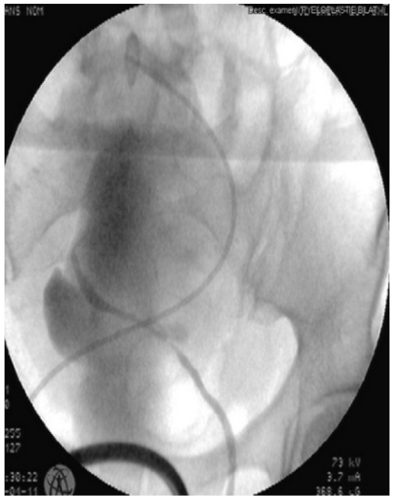


Figure 2 Preoperative retrograde uretero-pyelography showing shift of each ureter to the opposite side.

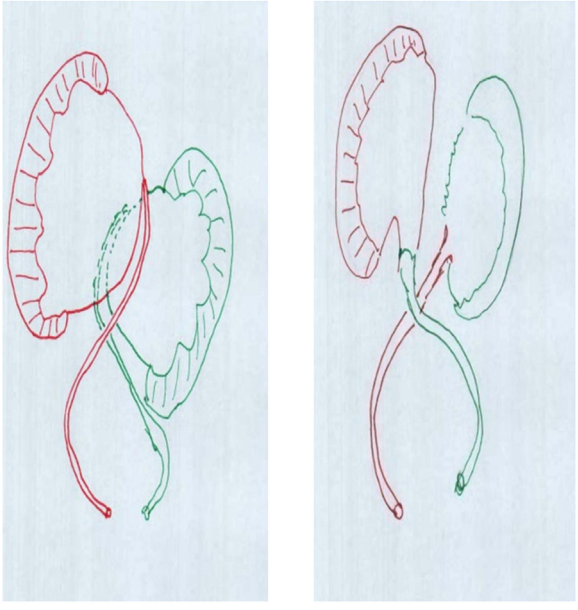
Postoperative evolution was unremarkable. Renal function remained normal. The patient was discharged on p.o. day five (Fig. 3).

Intravenous urography performed 5 months postoperatively showed good opacification of the switched ureters.

At 24 months p.o. patient was asymptomatic and ultrasound control was normal (Fig. 4).

Discussion

Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome affects at least 1 out of 4500 women; it is due to a variable developmental arrest in the distal two thirds of the Müllerian tubes. This syndrome is characterized by a congenital aplasia of the uterus and the upper



a b

Figure 3 Drawing of pathology and operative results. Preoperative: bilateral hydronephrosis with high insertion of the ureters. Postoperative results after dismembered pyeloplasty and ureteral switch.

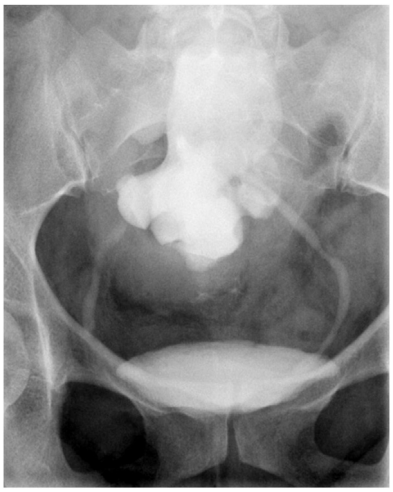


Figure 4 Intravenous urography performed 5 months postoperatively showed good opacification of the switched ureters.

part of the vagina. Secondary sexual characteristics and karyotype are normal [1–7].

The uterovaginal aplasia is either isolated (type I) or more frequently associated with other malformations (type II or Müllerian Renal Cervico-thoracic Somite (MURCS) association) [1–7].

Congenital anomalies of kidneys and upper urinary tract are found in about 40% of cases with MRKH syndrome. They mainly include unilateral renal agenesis (23–28%), ectopia of one or both kidneys (17%), renal hypoplasia (4%), horseshoe kidney and hydronephrosis [1].

Cyclic abdominal pain is often due to renal pelvis and UPJ anomalies.

For the UPJ stenosis due to high insertion of the ureter, several techniques have been proposed including bypass pyeloplasty, transposition pyelo-pyelostomy, renal pelvis cuff pyeloplasty and Foley Y-V pyeloplasty [8–12].

In our case, association of pelvic kidneys and UPJ stenosis represented a high risk of angulation of the UPJ after pyeloplasty. An original surgical method ("*ureteral switch*") was designed for the anatomic situation in this particular case. The right ureter was connected to the left renal pelvis and the left ureter to the right renal pelvis. This bilateral switch provided a progressive and gradual path to the UPJ.

Conclusion

We confirm the exceptional and unusual situation of our patient presenting a bilateral non-fused pelvic kidney with bilateral intrinsic UPJ obstruction. To the best of our knowledge, we believe that our case presents an original technique of ureteral switch at the time of pyeloplasty to prevent postoperative obstruction.

Conflict of interest

All the authors confirm that there is not any conflict of interest.

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