RETOCAVAL URETER IN CHILDREN. CASE REPORT AND BIBLIOGRAPHIC REVIEW

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Summary.- OBJECTIVE: Retrocaval ureter is a rare disease which is due to a abnormal development of the inferior cava vein. It usually presents with ureteral obstruction, and surgery is needed for symptomatic cases.

METHOD: We present a case of retrocaval ureter and a revision of the literature.

INTRODUCTION

The ureter retrocavo or circumcavo is a congenital, not very frequent anomaly, due to the anomalous development of this vein, where the ureter surrounds the vein at the level of the third or fourth lumbar vertebra. It almost invariably affects to the right ureter and the embryological event that precipitates this anomaly is the persistence of the subcardinal vein in the right side (1). The exceptional cases described in the left side asso- ciated to situs inversus (2,3). It was described initially by Hochstetter in 1893 (4). Its prevalence is of 0’06% (3-4 times more frequent in males than in females), its incidence is of 1 of each 1000 births, with a morbidity of 1 each 1500 (5,6).

Batenson and Atkinson performed a radiological classification, in two types: type I (90% of the cases) with

RESULTS: A 9 year-old boy who had consulted for macroscopic hematuria. The intravenous urography (IVU) suggested the diagnosis of retrocaval right ureter, which is confirmed by magnetic resonance imaging (MRI). Make us resection of the retrocaval segment and relocation of the ureter anterior to the inferior vena cava. Follow-up IVU showed good resolution of hydronephrosis.

CONCLUSIONS: Retrocaval ureter is a rare disease, with easy diagnostic and effective treatment. Other associated anomalies could be associated.

Keywords: Ureter. Urogenital abnormalities. Inferior vena cava. Ureteroureterostomy.


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FIGURE 1. Image of Urogramy IV that it shows a dilation pielocalicial and of the superior third of the ureter, with the typical image of overturned “J” and medialized ureter.

hidronefrosis and type II, without obstructive uropathy (7). The obstruction is caused by the compression of the ureter between the inferior cava vein and the vertebrae.

The usual symptoms are abdominal pain, hematuria, infection or urolithiasis, and it usually starts at the third or fourth decades (8).

The diagnosis is based on imaging tests. The ultrasonography usually shows ureterohidronefrosis according to the obstruction grade. The intravenous urography (IVU) can show an ureteropyelocalicial dilation of the upper ureter, with a typical image of inverted “J” and a medialized ureter at the level of L3-L4. A retrograde pyelography shows the characteristic form of “S” in the anteroposterior view and the sign of Randall-Campbell (9) (ureter close to the column vertebral) in the lateral view. CT and MRI are the best diagnosis modalities and they have displaced the oldest tests (cavographies) (8). They confirm the diagnosis when revealing the itinerary of the ureter behind the ICV. The isotopic tests contributes to the diagnosis of the obstructive pattern, offering essential data in the therapeutic decision (10).

The differential diagnosis includes retroperitoneal masses and retroperitoneal fibrosis. The treatment is mandatory when a significant obstruction, infection, urolithiasis or progressive hidronefrosis has been diagnosed. In these cases the best option is the division of the ureter and a termino-terminal anastomosis (11,12).

METHOD

We present a case of retrocaval ureter and a revision of the literature.

RESULTS

CASE REPORT

A 9 years-old boy came to the hospital because a sudden macroscopic haematuria. He was operated on of hypospadias (Snodgrass technique) four years ago. The physical exploration didn’t reveal any abnormality. The urine and blood tests only revealed hematuria and proteinuria (albumin: 150 mg/gl). A renal ultrasonography was carried out, showing a right hydronefrosis (diameter of the pelvis of 34mm), suggestive of ureteropelvic junction stenosis. The intravenous pyelography informed of a delay of the right kidney, with a pyelocalicial ectasia (grade III) from L4, and a medialized ureter, with a kinking suggestive of retrocaval ureter (Figure 1). A MRI confirmed the diagnosis.

With diagnosis of retrocaval ureter causing obstructive uropathy, a surgical treatment was proposed. A right ureteroplasty and decrossing of the ureter was performed. (Figure 2) The patient was discharged from the hospital at the 4th day. The JJ was retired at the 15th day of post-op. A posterior IVU showed the appropriate resolution of the obstructive uropathy.

DISCUSSION

From the original description of the retrocaval ureter by Hochtetter (4), about 400 cases have been published in the literature, mainly in adult patients (10). The retrocaval ureter can be associated, up to 20% of the cases, to congenital anomalies, mainly cardiovascular, renal, genitals and in the urethra. Our patient suffered for a hypospadias, whose association has already been described in the bibliography (13).

The pain in the right flank is the main symptom, although most of the cases are asymptomatic. Other frequent symptoms are the hematuria, as the case that we present; infection or urolithiasis (8). It is frequent, as in our case, that the diagnosis of suspicion settles down starting from the discoveries of the UIV. This one demonstrates, a characteristic pattern of a pyelocaliceal dilation and of the superior segment of the right ureter,
with “J shape” and medial deviation, at level of the vertebrae L3-L4 (9) (Figure 1).

The definitive diagnosis was carried out through a Nuclear Magnetic Resonance. Although it is also useful the CT, some authors prefer the RMN due to the good quality of images of the retroperitoneum and to its inoquity (absence of ionic radiation), in theses young patients. The isotopic tests, introduced recently in the diagnosis of this pathology, can offer essential information in the therapeutic decision. This way asymptomatic patients with a pathological diuretic renograma (elimination of the inferior plotter to 50%) should be operated on (10). The ultrasonography is very useful in the follow-up of theses patients.

If surgery is necessary, an individualized boarding, depending on the alteration grade and associate malformations, should be done. The classical way is the section of the ureteral and a termino-terminal anastomosis (11,12).

In patients with only a kidney a treatment of ureter and cava vein (division a transposition) could be advocated.

The transperitoneal laparoscopic access is used in this patients from 1994 (15). The laparoscopic retroperitoneal access is used since 1999 (16). From then on, many other authors have used this road with good results, communicating some surgical times in descent according to the improvement of the technique, with progressive decrease in operation time, from 9h20 in the first case to 2h10 o’clock’ communicated by Tobias-Machado (17-22).

Different studies have compared open versus laparoscopic approaches in adults, with less invasiveness, less hospital stay, best stetic appearance and reduction of pain in the laparoscopic arm (16,23). Since laparoscopic approach is the gold standard in the treatment of pyeloureteral syndrome, some authors some authors think that this approach is also the gold standard in theses patients (24). Laparoscopic robotical approach offer more ergonomy and better suturing over standard laparoscopy (25).

Nephrectomy could be necessary when pyelonephritic kidney.

CONCLUSION

Retrocaval ureter is a rare disease, with easy diagnost and effective treatment. Other associated urinary tract abnormalities could be associated.

REFERENCES AND RECOMMENDED READINGS

(*of special interest, **of outstanding interest)

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