

Ectopic Pelvic Kidney with Giant Hydronephrosis

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Abstract: Pelvic ectopic kidney associated with giant hydronephrosis is rare. It may be responsible for an extrinsic compression of adjacent structures. The authors report a case of giant hydronephrosis on a left pelvic ectopic kidney caused by obstructive lithiasis in a patient of 25 years. First ultrasound guided percutaneous nephrostomy allowed urinary drainage. Definitive treatment consisted in nephrolithotomy with removal of kidney stone. The outcome was favorable.

Keywords: Giant hydronephrosis ; Ectopic pelvic kidney ; Uro-CT scan ; Percutaneous nephrostomy ; Prenatal diagnosis.

1. Introduction

The association of pelvic ectopic kidney and a giant hydronephrosis is exceptional. Only five cases have been reported in the literature [1]. We report a case of hydronephrosis on a pelvic ectopic left kidney.

2. Observation

A 25 years old young patient consult emergency surgical in Ibn Sina University Hospital in Rabat of pelvic pain accentuated on the left side in a context of deterioration of general condition and fever of 38.5 °C. Palpation of the left side showed a large pelvic mass sensitive beyond the umbilicus. In laboratory tests, there was a significant inflammatory syndrome (a *C-reactive protein* of 180 mg/L) and a renal failure (Creatinine: 16.5 mg/l, Urea: 0.88 g/l). The urinary catheterization brought 200 cc of cloudy urine. Radiologically, the abdominal-pelvic ultrasound revealed a paravesical left compartmentalized cystic mass with impure content, an empty left renal fossa and normal right kidney. The URO-CT scan showed a large compartmentalized cystic mass involving pelvic left kidney and measuring about 36X42 cm with the presence of obstructive calculus of 3 cm above the ureteropelvic junction (Figure 1). The diagnosis was a giant hydronephrosis on a left pelvic ectopic kidney caused by obstructive lithiasis.



Figure 1: URO-CT scan: large compartmentalized cystic mass involving pelvic left kidney and measuring 36X42 cm

Three liters of purulent urine were drained by percutaneous nephrostomy. The cyto-bacteriological examination of urine objectified *Escherichia coli* sensitive to fluoroquinolones and third generation cephalosporins. Renal DMSA (Dimercaptosuccinic acid labeled with technetium 99) has objectified a functional value of the left kidney to 13%. Definitive treatment consisted in nephrolithotomy with removal of kidney stone after sterilization of urine and establishment of a double J stent. The outcome was favorable with improving renal function. Monitoring was regular and based on echography and assessment of renal function.

3. Discussion

The giant hydronephrosis is rare. Described by Glass in 1749 during an autopsy, it is defined for the first time in 1939 by Sterling as the accumulation of more than one liter of urine in the renal excretory cavities [2]. Its causes are represented by ureteropelvic junction syndrome (UPJ) in 80 % of cases, obstructive lithiasis, congenital malformations, ureteral stenosis, retroperitoneal fibrosis and trauma [3]. It is diagnosed most often during an abdominal mass syndrome associated with pain and signs of digestive, urinary, pulmonary or venous compression. Rupture of the giant hydronephrosis is a serious complication [4]. Although the association of renal pelvis and calyces dilatation and ectopic kidney is relatively frequent, the presence of giant hydronephrosis is exceptional. Hsieh and al, in a recent review of the literature, report only five cases and always an UPJ syndrome was causing giant hydronephrosis [1].

Ultrasound is usually the first-line examination. It shows the importance of dilatation and cortical atrophy. Its main interest lies in prenatal diagnosis [5]. Abdominal and pelvic CT seems, in our consideration, the gold standard. It makes the diagnosis of giant hydronephrosis and ectopic pelvic kidney, assesses renal functional impairment (secretion, excretion, cortical thickness) and search etiology [6]. In our case, CT allowed the diagnosis and the etiology of giant hydronephrosis on left pelvic ectopic kidney and the appreciation of its importance and impact. The magnetic resonance imaging (Magnetic Resonance Urography MRU) is an excellent diagnostic examination, which allows

morphological and functional accurate assessment [7]. It may be suggested as first line. In our context, because of its limited availability, we prescribe it only in case of contraindication for CT. Renal DMSA can be used to assess renal relative function. Its other interest is the location of an ectopic kidney, especially if it is atrophic and poorly functional up to a relative functional value of 5% [8].

The therapeutic management of ectopic pelvic kidney disease depends on the renal disease and its impact. Chiang recommends draining the giant hydronephrosis by ultrasound-guided percutaneous nephrostomy [5]. This helps relieve secondary symptoms such as intestinal obstruction, but also reduce the risk of cardio-respiratory failure caused by a sudden intra-abdominal decompression. In our case, the percutaneous nephrostomy draining has permitted urinary diversion and clinical relief of the patient.

A conservative therapeutic approach is increasingly advocated. Nowadays, it is considered that the kidney may be preserved if its functional relative value is greater than 10 % or less according to Aziz et al ; because there is no correlation between the degree of expansion and the degree of obstruction [8,9] .

4. Conclusion

Pelvic ectopic kidney associated with giant hydronephrosis is an extremely rare entity with difficult diagnostic. Prenatal diagnosis and monitoring are important. CT remains the diagnosis investigation of choice. A conservative approach should be considered as first-line when the functional value of kidney permits.

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