ORIGINAL RESEARCH

Experience of a tertiary-level urology center in the clinical urological events of rare and very rare incidence. IV. Urological surprises: 2. Clinically visible giant hydronephrosis in adults: Is there a significant function?

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Abstract

Background: Giant hydronephrosis is rare with a controversy about the complete loss of renal functions. Our objective is to present our center's experience with the management of cases of clinically visible giant hydronephrosis considering the potential residual functions. Our study is a retrospective case series of clinically visible giant hydronephrosis which was managed during the period July 2001–June 2016. Demographic and clinical variables were studied with specific considerations to the potential residual functions.

Results: Of more than 82,000 urological interventions, only 47 cases (0.057%) were operated upon for clinically visible giant hydronephrosis. Group 1 included 21 patients (mean age $= 50.43 \pm 13.71$ years) who were treated initially by nephrostomy tube, and group 2 included 26 patients (mean age $= 42.96 \pm 15.16$ years) who were treated primarily by nephrectomy. The main clinical presentation was abdominal distention (61.7%), while 13 patients (27.7%) were unaware of the swellings. The commonest underlying causes of hydronephrosis were urolithiasis (68.1%) and bilharzial ureteral strictures (23.4%). The contralateral kidney was diseased in 22 cases (46.8%) including the bilateral clinically visible hydronephrosis in 7 cases (15%). Indications of placement of a nephrostomy tube included uremia, infections, and evaluation of renal functions, where 5 cases of group 1 regained significant split function ranged 14–33%.

Conclusions: Clinically visible giant hydronephrosis is an extreme form of renal dilatation with different etiologies such as urolithiasis and bilharziasis. Initial placement of a nephrostomy tube may save significant residual functions in these kidneys.

Keywords: Giant hydronephrosis, Hydronephrosis, Nephrectomy, Nephrostomy tube, Urolithiasis

1 Background

Hydronephrosis is the dilatation of the pelvicalyceal system of the kidney due to different etiologies [1–3]. The hydronephrotic kidney may expand gradually to reach a state of marked dilatation known as giant hydronephrosis which is almost a non-functioning kidney containing > 1000 ml of fluid [4, 5]. Giant hydronephrosis is rare with a few hundred case reports, and a few case series have been reported in the literature [1, 2, 6]. Moreover, clinically visible giant hydronephrosis is a rarer entity

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where cases of hydronephrosis containing > 2000 ml of fluid have been scarcely reported so far [4, 7]. This rarity warranted the conduction of this retrospective study in our locality in the context of prevalent underlying causes of urinary tract obstruction such as urolithiasis and bilharziasis [8–11]. The question is whether there are significant residual functions in these kidneys.

2 Methods

A retrospective search of the patients' records was done in our hospital for the cases of clinically visible hydronephrosis at their first clinical presentation in the period July 2001–June 2016. Each case was studied for the demographic characteristics including age, gender, job, residence, and education level. Clinical characteristics included a history of urolithiasis, first urological presentation, kidney size and parenchymal thickness, imaging features, underlying etiology, renal functions by serum creatinine and radio-isotope renal scanning, and initial and/or primary interventions. Patients were differentiated into two groups according to the approach of the initial line of treatment. Group 1 included the patients who were treated initially by the placement of a nephrostomy tube before definitive treatment, while group 2 included the patients who were treated initially by nephrectomy. Outcomes of the cases in group 1 were further differentiated and studied according to the amount, color, and concentration of produced urine per nephrostomy tube as < 100 ml, 100–400 ml, and > 400 ml per 24 h.

3 Results

Of more than 82,000 urological interventions, 47 cases (0.057%) presented with and were operated for clinically visible giant hydronephrosis. They included 39 males (83%) and 8 females (17%) with a mean age of 46.29 ± 14.86 years.

Demographic and clinical characteristics of the patients in both groups are summarized in Table 1. Group 1 patients were treated initially by the placement of a nephrostomy tube, while in group 2 patients were treated initially by nephrectomy. Clinical presentations were considered according to the main symptom at presentation (Figs. 1a, b, 2a, b). Thirteen cases (27.7%) were unaware of the renal swelling which was discovered by clinical examination, and they complained of manifestations of uremia or diffuse abdominal pain.

Abdominal ultrasonography and radiography were the basic imaging, which were done for all cases. Lost parenchymal thickness in imaging was the sign of a non-functioning state and on which decisions of treatment were scheduled (Figs. 1c, 2c). In group 2, accordingly, 21 cases were considered non-functioning kidneys. Five cases only had renal radio-isotope scanning studies to establish

the non-functioning state in 3 relatively young patients and 2 patients asked for accurate documentation before nephrectomy. Intravenous urography and computed tomography were used alternatively in 24 patients for anatomical studying (Table 1).

Twenty-two cases (46.8%) had bilateral renal pathologies. They included bilateral clinically visible hydronephrotic kidneys in 7 patients (31.8%), contralateral non-visible hydronephrosis in 8 patients (36.4%), chronic pyelonephritic kidney in 3 patients (13.6%), absent kidney in 1 patient (4.6%), and non-obstructing nephrolithiasis in 3 patients (13.6%).

A percutaneous nephrostomy tube was placed bilaterally in 8 cases. Indications were infected hydronephrosis (19%), uremia (38.1%), hematonephrosis (4.8%), and evaluation of residual function (38.1%). Major complications of nephrostomy tube were septic peritonitis and death in 1 patient and hematonephrosis that indicated wide-bore drainage in 2 patients. Crude evaluation of function was done according to urine output per nephrostomy tube (Table 2). Further evaluation of function and definitive treatment were planned accordingly. A significant number of patients regained residual renal functions enough to correct the cause of obstruction and postpone regular dialysis (Table 2).

Besides the unilateral visible giant hydronephrosis with lost parenchymal thickness, normal contralateral kidney (Fig. 2c), and serum creatinine, indications of initial nephrectomy in group 2 included old patient's age (> 55 years) and patient's refusal to the placement of nephrostomy tube. Apart from the generous classic flank incision, no major complications were reported among the patients of group 2.

Follow-up periods ranged as 4–20 months. In group 1, significant improvement in function was detected in 5 cases. In group 2, only 3 patients experienced deterioration in the renal function through their follow-up due to causes other than nephrectomy (hypertension and nephrotoxic medications).

4 Discussion

The term "hydronephrosis" could be confused with non-synonymous terms [12]. Here, however, we adhered to this term to express the state of massive renal dilatation. Hydronephrosis is commonly reported from the developing countries where its common causes such as urolithiasis and bilharziasis are endemic [1, 6, 8, 10]. Congenital pelvi-ureteral junction obstruction has been reported as the commonest cause of giant hydronephrosis with more occurrences among pediatrics [2, 7, 13]. However, the matter seems to be different among adults, especially in developing countries. The commonest causes of hydronephrosis are ureteral stones [8, 14] and bilharziasis, especially in tropical countries [9–11]. Variable degrees

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Table 1 Demographic and clinical characteristics of the patients differentiated according to the initial line of treatment into two groups

Characteristics	Group 1 (*n* = 21, treated initially by PCN)	Group 2 (*n* = 26, treated initially by nephrectomy)
Age range (Mean ± SD)/years	22–72 (50.43 ± 13.71)	21–83 (42.96 ± 15.16)
Gender		
Male	18 (85.7%)	21 (80.8%)
Female	3 (14.3%)	5 (19.2%)
Job		
Farmer	15 (71.4%)	16 (61.5%)
Others	6 (28.6%)	10 (38.5%)
Residence		
Local governorate	14 (66.7%)	18 (69.2%)
Distant governorate	7 (33.3%)	8 (30.8%)
Education level[a]		
High	1 (4.8%)	None
Middle	3 (14.3%)	4 (15.4%)
Low	17 (80.9%)	22 (84.6%)
Anatomical side		
Right	9 (42.9%)	14 (53.8%)
Left	8 (38.1%)	12 (46.2%)
Bilateral	4 (19%)	None
Clinical presentation		
Abdominal distention	8 (38.1%)	13 (50%)
Abdominal pain	2 (9.5%)	13 (50%)
Fever	5 (23.8%)	None
Uremic symptoms	6 (28.6%)	None
History of known previous urolithiasis	10 (47.6%)	11 (42.3%)
Underlying etiology		
Urolithiasis	13 (61.9%)	19 (73.1%)
Bilharzial stricture	6 (28.6%)	5 (19.2%)
Congenital PUJO	2 (9.5%)	2 (7.7%)
Serum creatinine range (Mean ± SD)	0.8–12.3 (3.13 ± 3.13)	0.56–1.4 (0.94 ± 0.21)
Imaging		
MSCT	8 (38.1%)	16 (61.5%)
IVU	2 (9.5%)	10 (38.5%)
RI	None	5 (19.2%)
Only basic imaging[b]	11 (52.4%)	None
Parenchymal thickness (mm)	0–4	0

[a] Education level is classified as high for education to college level or higher, middle for secondary school level, and low for education below the secondary school level
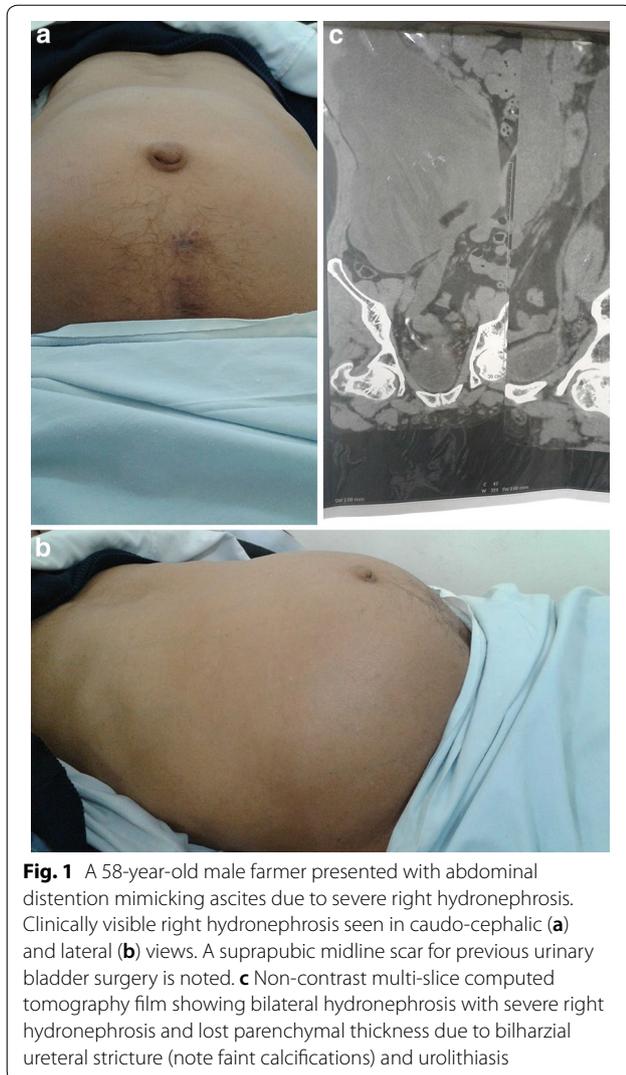
[b] Basic imaging included abdominal ultrasonography and radiography

IVU intravenous urography, *MSCT* multi-slice computed tomography, *PCN* percutaneous nephrostomy, *PUJO* pelvi-ureteral junction obstruction, *RI* radio-isotope scanning, *SD* standard deviation

of hydronephrosis may result from renal obstruction at different levels along the urinary tract [12, 15]. Giant hydronephrosis in adults may have widely variable causes and mechanisms than pediatrics [3, 4].

The natural course of giant hydronephrosis has always no definite start, where most of the patients deny any previous urinary troubles. However, unnoticed initial events such as stone migration or acute bilharziasis may pass unnoticed [9–11]. During the process of chronic renal dilatation and atrophy, vague abdominal discomfort is the predominant symptom [7]. However, the neglected acute onset symptoms and the chronic renal pain could be accompanied by the advancement of severe degrees of hydronephrosis [16].

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Fig. 1 A 58-year-old male farmer presented with abdominal distention mimicking ascites due to severe right hydronephrosis. Clinically visible right hydronephrosis seen in caudo-cephalic (**a**) and lateral (**b**) views. A suprapubic midline scar for previous urinary bladder surgery is noted. **c** Non-contrast multi-slice computed tomography film showing bilateral hydronephrosis with severe right hydronephrosis and lost parenchymal thickness due to bilharzial ureteral stricture (note faint calcifications) and urolithiasis

Incomplete renal obstruction and absent infection may allow the obstructed kidney to dilate up to large sizes under the brunt of prolonged building-up back pressure on the renal parenchyma [15]. Complete loss of function and irreversibility are the results of unresolved complete obstruction and/or infection of the kidneys. Compensation of the contralateral kidney is a part of the natural history of severe unilateral hydronephrosis [17]. Abdominal distention is the common presenting symptom of giant hydronephrosis [1, 5]. It seldom fills the whole abdomen to be confused with tense ascites [7, 16]. This finding was more obvious in our patients who presented for the first time with diffuse abdominal distention due to giant hydronephrosis (Fig. 1). Besides the endemic underlying pathologies in the current study, low socio-demographic characteristics were predominant and could be proposed as risk factors for the development of clinically visible giant hydronephrosis.

Most of the cases of visible giant hydronephrosis in the current series occurred in surgically fresh kidneys. This finding could be attributed to the expansible characters of the surrounding inter-fascial retroperitoneal planes which may allow the kidney to expand complying with the long-standing internal forces of renal obstruction [18]. Such characters could be lost in cases of chronic pyelonephritis and previous renal surgeries which may limit its expansion due to the acquired adhesions and altered anatomical planes [19].

Although the renal mass of giant hydronephrosis is occasionally visible, it is usually soft and could be missed on abdominal palpation in these cases [20–22]. Imaging investigations for hydronephrosis are directed to the determination of anatomical issues such as the cause of obstruction and functional issues such as the total and split renal functions [17]. In the old eras, abdominal radiographs and intravenous urography were the main studying tools [22]. However, introduction of ultrasonography has positively changed the quality of diagnosis of the abdominal disorders [7]. Also, the new advances in computed tomography may describe more anatomical details. Renal isotope scanning represents the most accurate method to describe the split renal functions, especially in the situations of bilateral renal affection [17]. In the current series, the need for renal isotope scanning was reduced by the huge size of the kidneys that was the basic state to decide nephrectomy. Also, the emergency and acute presentations in group 1 were obligations to postpone or ameliorate this need.

These acute presentations made the initial placement of nephrostomy tube a mandatory intervention and helped in the differentiation of those patients into two groups. From this differentiation, a significant proposal for the preservation of residual functions in those compromised kidneys could be drawn. This principle issue may evolve on the management of giant hydronephrosis in relatively young patients and bilateral cases [23]. However, the initial placement of a temporary nephrostomy tube, before deciding elective nephrectomy or correction of the primary cause, is still controversial [1, 6, 7]. In the current study, however, although imaging tools evaluated the kidneys as non-functioning, the initial placement of nephrostomy tube improved the kidney function significantly in 5 patients. This observation could be attributed to the distribution of the residual functioning nephrons over a wide surface area of the giant kidney. We proposed that the recoil of a large surface area after placement of nephrostomy tube provided a considerable cortical thickness and, therefore, a reasonable function preservation. This interpretation may be parallel to the recent promising results of using the renal parenchymal-to-hydronephrosis area ratio as an early predictor for surgery in

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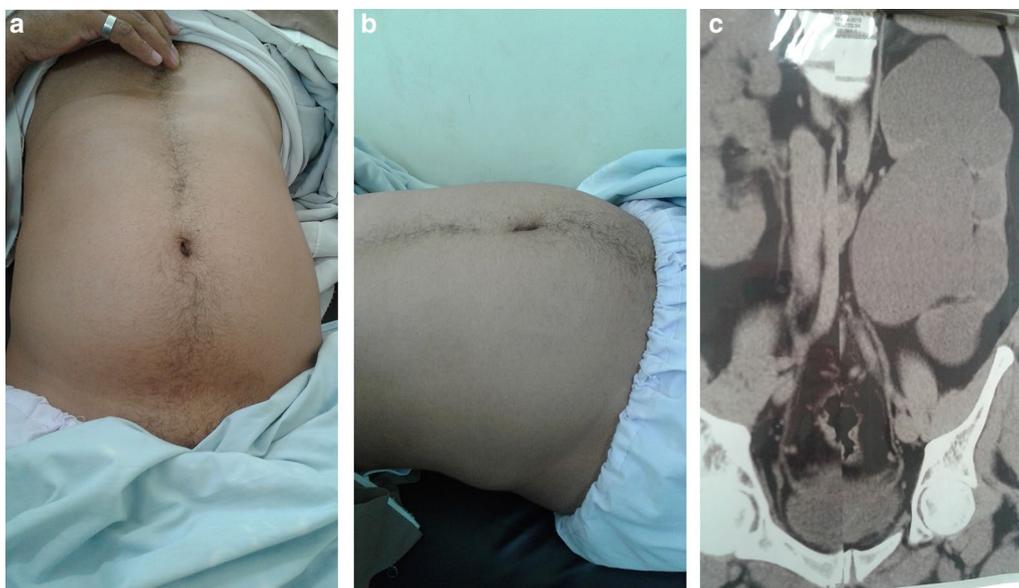
Fig. 2 A 45-year-old male patient presented with diffuse abdominal pain and distension due to left giant hydronephrosis. Clinically visible left hydronephrosis seen in caudo-cephalic (**a**) and lateral (**b**) views. **c** Non-contrast multi-slice computed tomography film showing severe left hydronephrosis with lost parenchymal thickness due to pelvi-ureteral junction obstruction and normal right kidney

Table 2 Functional outcomes of the patients in group 1 after placement of nephrostomy tubes

Fluid output characters	Volume/24 h	<100 ml (n = 11)[a]			100–400 ml (n = 5)		>400 ml (n = 4)
	Nature	Pus	Urine	Blood	Pus	Urine	Urine
	Patients	5	6	1	2	3	4
Anatomical side	Unilateral	4	5	1	2	2	3
	Bilateral	1	1	0	0	1	1
RI scanning split function: split GFR and percentage (done after the placement of PCN)		None	None	None	0–8 ml/min (0–5%)	15–18 ml/min (7–14%)	18–25 ml/min (20–33%)
Definitive treatment	None	2	1	0	0	0	0
	Nephrectomy	2	5	1	2	2	0
	Correction of the cause	0	0	0	0	1	4

RI radio-isotope scanning, *GFR* glomerular filtration rate, *PCN* percutaneous nephrostomy

[a] One patient in group 1 died due to septicemia that followed immediately the placement of nephrostomy tube. Nephrectomy was done for 7 of the 11 patients. The other 4 patients were unfit for surgery and followed up with exchange upon being slipped, besides regular dialysis

pediatric hydronephrosis [13]. It is a striking indicator to not consider any hugely dilated and clinically visible hydronephrotic kidney with apparent lost parenchymal thickness as a non-functioning one.

Nephrectomy for a hugely dilated kidney may indicate a generous incision [20]. More surgical morbidities may supervene without providing more benefits than nephrectomy of a decompressed dilated kidney like in the current series. Laparoscopic resection of giant hydronephrosis has been reported recently [24]. However, it may have more challenges with the enormous hydronephrosis.

In spite of the descriptive retrospective methodology that was employed for this study, it is mostly the only way

for studying these rare events. The current study is the first and largest case series in the literature that targeted this extreme form of giant hydronephrosis looking for the potential residual functions in these kidneys which are primarily assumed to be non-functioning in most of the instances. Although the matter of initial placement of nephrostomy tube has been studied before, these studies included giant hydronephrosis in different ages and etiologies [1]. However, the current study targeted only the visible kidneys in adults. Placement of initial nephrostomy tube for the clinically visible giant hydronephrosis is recommended. Also, the role of the underlying etiology

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such as urolithiasis and bilharziasis relative to potential residual functions may warrant larger comparative studies.

5   Conclusions

In spite of the clear appearances of lost parenchymal thickness in imaging, a hugely dilated and clinically visible hydronephrotic kidney may be still a functioning one after decompression. So, placement of a percutaneous nephrostomy tube should be the initial intervention in these cases. It may unmask significant residual renal functions which may be preciously needed in cases such as bilateral giant hydronephrosis and relatively young patients.

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