Previously unseen 80L giant hydronephrosis

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ABSTRACT

Here we describe the case of a patient who was referred to our institution with an immense abdominal volume of unknown origin. The patient was unable to stand up and therefore was bedridden. A giant hydronephrotic kidney was diagnosed and total volume of urine removed was 80L. Nephrectomy was uneventful and, despite his acquired thoracic and abdominal deformities, he was able to recover completely. This is the largest reported hydronephrosis in the literature.

Introduction

Hydronephrosis is defined by ischemic atrophy of renal parenchima due to dilation of the renal collecting system and is secondary to obstruction or urinary reflux [1]. Stirling [2] defined giant hydronephrosis when total volume is over 1000ml.

Giant hydronephrosis is more common in children due to congenital causes, such as ureteropelvic junction obstruction and duplicated collecting system or ureteral ectopia [3]. In adults, acquired conditions are more frequent, namely ureteral calculus, trauma, carcinoma or retroperitoneal fibrosis [4,5].

Giant hydronephrosis is usually painless and patients present with progressive increasing abdominal volume. Rarely, it can cause respiratory distress by diaphragmatic pressure and cardiovascular changes [5]. When untreated, long term complications include renal failure, hypertension, malignant transformation and renal rupture [4,5].

This case highlights challenges in the management of a patient with an 80L hydronephrosis.

Case presentation

A 62-year-old man presented to the emergency room of a tertiary hospital with increasing abdominal volume, superficial abdominal wall bleeding and infection after a minor blunt trauma. The patient declared that his abdominal volume had been increasing slowly and progressively for the last 12 years, affecting his daily activities. For the last two years, he had been restricted to household activities. Due to worsening of the skin condition, he was taken to a local hospital and later transferred to our institution for specialized care.

At first evaluation, the patient was hemodynamically stable, with no fever. Physical examination showed giant abdominal volume with phlogistic signals and ischemic-looking wounds on the abdominal wall (Fig. 1a). Body weight was 153kg.

With the hypothesis of an abdominal wall infection in a patient with ascitis, a CT scan was ordered to confirm the etiology of the abdominal volume. The patient, however, did not fit the CT scanner due to his abdominal circumference. Paracentesis was performed and 24L of a brown and thick liquid were removed. Liquid creatinine was 8,24mg/dL and potassium 14,5mEq/L, and cultures were negative.

The CT findings after first initial paracentesis (Fig. 1b and c) showed an extensive liquid formation, from the diaphragm to the pelvis, repelling the liver and colons laterally. Dimension measures were 67 × 26 × 38 cm (CC x LL x AP), after the first partial drainage. The right kidney wasn’t visualized and the report suggested a giant hydronephrosis as etiology.

After determining the abdominal volume origin, a percutaneous nephrostomy was performed. We decided to drain 3 L on a daily basis, considering physiological adaptations regarding venous return, respiratory mechanics and liquid distribution the patient would have to undergo. During this gradual drainage, he was unable to stand up on his own, experiencing dyspnea and tachycardia even after sitting up. He received motor and respiratory physical therapy twice a day as well as
Fig. 1. Pre-operative patient status. 1a – Patient’s abdominal volume and infectious skin lesions. 1b – Sagittal CT scan image after removal of 24L of urine. Note extensive intra-abdominal organ compression. 1c – Axial CT scan image demonstrating patient’s kidney occupying his pelvis.
nutritional support. He was also given Ceftriaxone and oxacillin, and the abdominal wall wounds were treated with daily dressings.

By the 18th day after admission, a total of 80 L of urine were removed (Fig. 2a and b) and CT scan showed a severe abdominal deformity (Fig. 2c). From the 18th day until surgery (30th day), nephrostomy was maintained opened and had no urine output.

At this time, the patient was able to walk with assistance, improved his nutritional status (albumin increased from 2.5 g/dl to 3.4 g/dl) and
We planned a total right nephrectomy on the 30th day after admission along with abdominal reconstruction and dermolipectomy. The surgery was performed under general anesthesia, on dorsal horizontal decubitus, through a median longitudinal incision. The findings included a redundant and strong aponeurosis and a giant hydronephrotic kidney, occupying most of the abdominal cavity and retracting the viscera laterally to the left. There were no firm adherences and nephrectomy was uneventful. Fig. 3a shows the aspect of the abdominal cavity after kidney removal.

The aponeurosis closure was made using an overlap technique, with good strength and no need for mesh reinforcement, followed by a standard abdominoplasty. The total surgery time was 5 hours, estimated blood loss was 300ml and the patient was admitted to ICU immediately after surgery. The removed kidney weighed 2970g (Fig. 3b) and excess skin weight was 3835g. Pathologic findings showed no malignancies and a severe ureteropelvic junction obstruction.

After 48h of ICU care, the patient had an uneventful evolution throughout ward care. At follow-up (Fig. 3c), he appeared well groomed and cheerful. After 12 years of seldom leaving his home, he now walks 4Km a day without any symptoms.

Discussion

We report a case of severe hydronephrosis in a 62-year-old man who had numerous limitations due to a 12 years-long progressive increase in abdominal volume. The first reported case of a giant hydronephrosis was in 1746, albeit exact volume is questioned. Despite more than 600 cases reported worldwide, this is the largest reported hydronephrosis in the literature.

Roughly half of the cases are adequately diagnosed due to nonspecific symptoms and increasing abdominal volume [5]. However, better imaging may allow early diagnosis and treatment. Differential diagnosis that should be considered at first evaluation includes massive benign or malignant ascites, as well as hepatobiliary cysts, mesenteric cysts, pseudomyxoma, cystic, retroperitoneal haematoma, and splenomegaly [4,5].

Although the definitive treatment for hydronephrosis with no functioning kidney is total nephrectomy, we initially considered a percutaneous drainage, intensive nutrition, physiotherapy support and treatment of abdominal wall infection. This temporary drainage effectively relieved compressive symptoms, allowed adjacent organs to return to normal position, lowered body weight before definitive treatment and, most importantly, allowed vascular and hydro-electrolytic balance to be achieved. In addition, this was consistently performed by others centers [4]. After 30 days of hospitalization, with significant motor and respiratory rehabilitation, complete drainage of urine and better nutritional status, we performed an extensive surgical procedure with total right nephrectomy with wall reconstruction and abdominoplasty with no major complications.

Conclusion

Giant hydronephrosis requires individualized patient management. Timing for nephrectomy involves nutritional and physical evaluation. Renal drainage should be used to allow improvement of patient conditions and reduce postoperative complications. This case shows that hydronephrosis leading to decreased global activity and abdominal wall deformities, was safely managed with drainage and delayed surgery.

Declarations of interest

None.
Fig. 3. —Post-operative status. 3a Abdominal cavity after kidney removal. 3b– Giant hydronephrotic kidney (weight 2970g). 3c – Late (40 days) patient’s status.
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