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Giant hydronephrosis secondary to ureteropelvic junction obstruction, a rare occurrence, case report

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ABSTRACT

Giant hydronephrosis (GH) is very rare in adults. We report a case of a 46-year-old man who presented with progressively increasing abdominal swelling. A contrast-enhanced CT scan of the abdomen and pelvis was used for the diagnosis. Left side open nephrectomy was performed and 22 L fluid was evacuated. UPJ obstruction was noted intraoperatively. No recurrence of abdominal swelling was noted on follow up examination.

1. Introduction

Giant hydronephrosis is a very rare urological disease. In the published literature the majority of giant hydronephrosis caused by ureteropelvic junction (UPJ) obstruction, occur in the pediatric population. It is not common in adults and diagnosis is often elusive. The objective of this paper is to present one case of GH in an adult patient as well as to review the current literature.

2. Case presentation

A 46-year-old man presented to the urology clinic of a tertiary hospital with abdominal swelling that has been gradually and progressively increasing in size over 10 years. Associated with this he had early satiety, unquantified weight loss but no fever or night sweats. Three months before his presentation, he had profound limitations on his activities of daily living but was never confined to bed. He claims that he was treated for pulmonary tuberculosis and improved 20 years ago.

On physical examination, the patient had stable vital signs. His body weight was 78 kg. The abdomen was hugely distended. Thinned out skin, rectus diastasis, and visible veins were noted on the abdomen but no lesions (Fig. 1).

The mass filled the whole abdomen from Xiphisternum extending to the suprapubic area with a bulge in the flanks and dull to percussion. There was a positive sign of fluid collection with no tenderness and ballotable mass.

Laboratory investigations were performed and a complete blood count showed mild anemia with hemoglobin of 8.3 g/dl(14–18g/dl). Other parameters of complete blood count were in the normal range. The serum creatinine (Cr.) and blood urea nitrogen (BUN) were 0.64mg/dl (0.6–1.2mg/dl) and 20 mg/dl (10–49mg/dl), respectively. Liver enzymes and serum electrolytes were in the normal range. Serologic tests for hepatitis B and C were negative. The fluid was tapped and microscopic analysis showed necrotic background with degenerated inflammatory cells, mainly lymphocytes but no malignant cells. A contrast-enhanced CT (CECT) scan of the abdomen was obtained and showed a huge multiseptated intra-abdominal mass arising from the left abdomen with septal and wall calcifications and dimensions of (CC 42cm LL32 and AP 25 cm) with an index of massive left side hydronephrosis. The left kidney was not visualized and there was no lymph node enlargement. The right kidney was normal in size, shape, contrast enhancement, and excretion with mild pelvicalyceal dilatation but no stone or mass (Fig. 2).

The left kidney was approached using a left subcostal flank incision. In total, 22 L of coffee-ground color fluid was evacuated. The anatomy of the left retroperitoneum was distorted and obstruction at the left UPJ area was noted with a massively dilated pelvis (Fig. 3). Left side nephrectomy was performed in the standard fashion. The nephrectomy specimen was sent for histopathologic examination. A retroperitoneal non-suction drain was placed in the left renal fossa because it was a large cavity. The drain output was 600ml over the next 24 hours. The drain output subsequently decreased and was removed on the 7th postoperative day when it became null.

Histopathologic examination of the nephrectomy specimen revealed

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Fig. 1. Preoperative picture of the patient A: frontal view B: left lateral view.

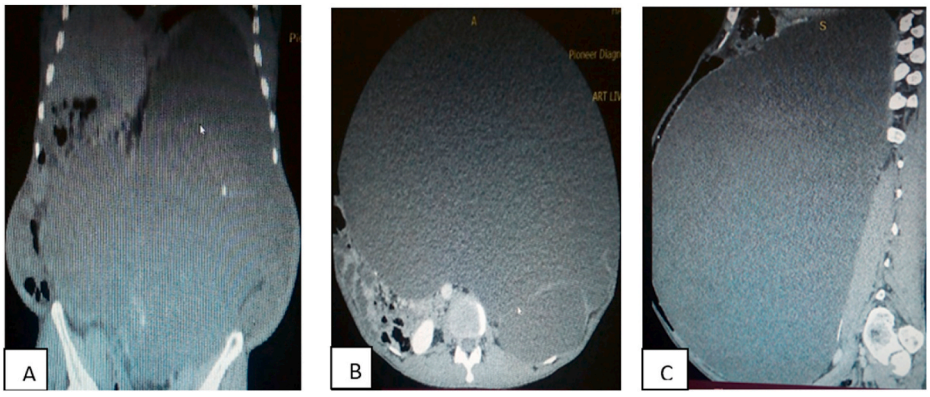

Fig. 2. Preoperative CECT of the abdomen and pelvis. A: Coronal reformatted image showing left giant hydronephrosis filling the whole abdomen B: Axial image C: Sagittal reconstruction.

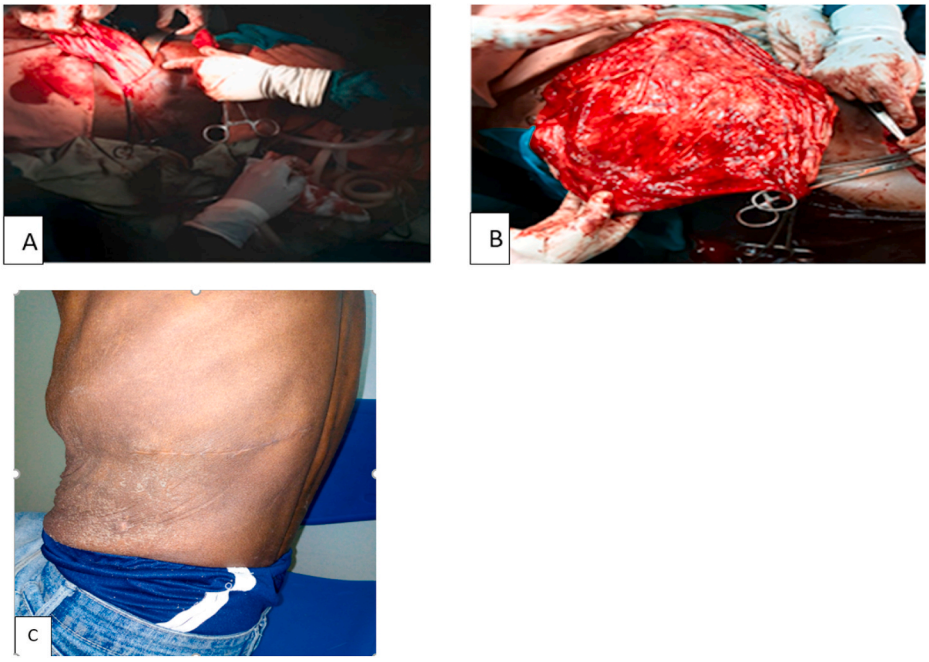

Fig. 3. Intraoperative pictures and post operative pictures. A: Partially decompressed hydronephrotic sac of the left kidney B: Fully decompressed and excised left kidney C: Post operative picture at second follow up visit.

a thinned-out cortex with thyroidisation of tubules, extensive inflammatory cell infiltrate, fibrosis, arterial wall calcification, and arteriosclerosis but no features of malignancy whatsoever.

The patient was discharged with improvement with normal serum creatinine on the 8th postoperative day.

On the first and second follow-up visits, the patient reported

unquantified weight gain, significant improvement in abdominal pain and swelling.

3. Discussion

Giant hydronephrosis is a rare urological condition defined as the presence of more than 1 L of fluid contained in the collecting system.[1]

It has also been radiologically defined as the kidney occupying hemi-abdomen with midline crossing and/or that equals the height of five vertebral bodies.[2]

The first case of more than 600 cases reported so far in the literature was published in 1746. Differential diagnoses for giant hydronephrosis include mesenteric cyst, pancreatic cyst, adrenal cyst, extra-renal tumor, echinococcal cyst of the liver, retroperitoneal (non-renal) cyst, tuberculous peritonitis, and ascites. Even though about 600 cases of giant hydronephrosis have been reported in the literature, accurate diagnosis of giant hydronephrosis remains a challenge to date.[3] Our patient posed such a diagnostic dilemma before cross sectional imaging was obtained which subsequently showed giant hydronephrosis. He was being investigated in the medical outpatient in the line of ascites but after the imaging, he was sent to urology unit.

Giant hydronephrosis, although rare should be considered when evaluating patients with massive abdominal swellings.[1]

UPJ obstruction is the commonest cause of giant hydronephrosis, other causes include urinary stones, trauma, renal ectopia, ureterovesical junction obstruction, and rarely malignancies.[4]

Since the majority of cases with giant hydronephrosis present with severely damaged kidneys with no expectation for improvement, nephrectomy is usually performed, particularly when the contralateral kidney is fully functional.

The diagnostic accuracy, even of modern imaging modalities, is limited since functioning renal parenchyma with adequate contrast enhancement might be absent.[1] Hence, the differential diagnoses of any abdominal/retroperitoneal cystic mass should include giant hydronephrosis.

4. Conclusion

This case elucidates the diagnostic challenge and the rare presentation of UPJ obstruction in adults. As is the case in the majority of patients with giant hydronephrosis, our patient had well-functioning right kidney with severely damaged and ballooned out left kidney where we opted for simple open nephrectomy along with evacuation of 22L of coffee ground color fluid. This type of presentation makes our patient one of the few cases of adult giant hydronephrosis caused by UPJ obstruction as it has been reported in the literature.

The histopathology report only showed evidence of long-standing obstructive damage and inflammatory changes in the left kidney. However, based on the long duration of the patient's clinical presentation and the size of hydronephrosis, congenital cause of UPJ obstruction was highly likely in our patient.

Consent for publication

An informed written consent was obtained from the patient for publication of this article. A copy of the written consent is available for review by the Editor-in-Chief of this journal up on reasonable request.

Availability of data and material

Patient data are available from the corresponding author on reasonable request.

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Authors' contributions

SMH, FOM and FHI performed the operation. FHI and IKH wrote the draft. All authors contributed to, read and approved the final manuscript.

Declaration of competing interest

The authors declare that they have no competing interests.

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