Scholars Journal of Medical Case Reports
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

Radiology

Giant Hydronephrosis Mimicking an Intraabdominal Mass: Case Report

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DOI: 10.36347/sjmcr.2023.v11i04.007 | **Received:** 07.02.2023 | **Accepted:** 24.03.2023 | **Published:** 09.04.2023

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Abstract **Case Report**

Giant hydronephrosis (GH) is a rare entity that may mimic progressive and benign abdominal cystic tumor, is defined as a collection of more than 1 litre of urine in the excretory cavities or as a dilation beyond the mid-abdominal line with a kidney length exceeding 5 vertebrae in children. It is rare, often due to a pyelo ureteral junction syndrome. The antenatal ultrasound must make the diagnosis before birth. We report the case of an 2-month-old infant, brought to the consultation for progressive abdominal distension since birth but aggravated since 1 month. The physical examination had revealed an abdominal mass occupying the whole abdomen, but more marked on the right, taut and matt to the percussion and whose limits and origin were difficult to determine. The abdominopelvic ultrasound had shown an echo-fluid structure mass extending from the right hypochondrium to the pelvis and well beyond the midline. The urotomodensitometry (uro-TDM) had shown at right giant hydronephrosis in the form of a voluminous liquid mass extending from the subhepatic region into the pelvis beyond the midline. This mass was surrounded by a thin parenchymal border with an elevation corresponding to the renal parenchyma.

Keywords: Hydronephrosis, infant, mass, pyelo-ureteral junction syndrome.

INTRODUCTION

The first case of GH was described in 1746. Since then, few cases have been described in the literature [1]. This condition is a rare urological entity, defined in the literature as a presence of more than 1 liter of urine in the collecting system [2]. Ureteropelvic junction (UPJ) obstruction is a common cause of hydronephrosis during childhood, and other causes include stone disease, trauma, and renal ectopia. Although giant hydronephrotic kidney may occasionally be found from intra-abdominal mass, intermittent abdominal pain, or hematuria, it is mostly asymptomatic because hydronephrosis progresses slowly to a GH without any prominent symptoms [3]. It is seen more often in males than in females (2.4:1). More than 500 cases of GH have been reported in the literature [4]. We report a case of GH in a 2-months-old children.

CASE REPORT

We report the case of an 2-month-old infant, brought to the consultation for progressive abdominal distension since birth but aggravated since 1 month. The physical examination had revealed an abdominal mass occupying the whole abdomen, but more marked on the right, taut and matt to the percussion and whose limits and origin were difficult to determine. The abdominopelvic ultrasound had shown an echo-fluid structure mass extending from the right hypochondrium to the pelvis and well beyond the midline.. According to the sonographic findings, it was thought to be of renal origin and perhaps represents a cystic neoplasm. Further investigation of the sonographic findings was undertaken with CT imaging. A contrast-enhancement CT scan of the abdomen obtained the next day (Fig 1a–b) showed the right kidney with enhancement to be replaced by giant cystic mass extending from the subhepatic region to the pelvis beyond the midline. This mass was surrounded by a thin parenchymal border with an elevation corresponding to the renal parenchyma. The extretory phase (Figure 2 a-b) demonstrating the excretion of contrast into the cystic mass. Furthermore, intra-abdominal extension of the cystic mass resulted in compression of the bladder and lateral deviation of all small bowels; however, there was no intestinal obstruction. The left kidney was normal, while the right one was not visualized. They presumably represented dilated pelvicaliceal system by obstruction, the diagnosis of giant hydronephrosis was suggested and a percutaneous nephrostomy had been placed and approximately 1.5 l of clear urine was drained. Then a Pyeloplasty was performed. On gross examination, a marked stenosis of the UPJ with a

dilated renal pelvis and calices. Besides, the hydronephrotic kidney had an extremely thinning cortex.

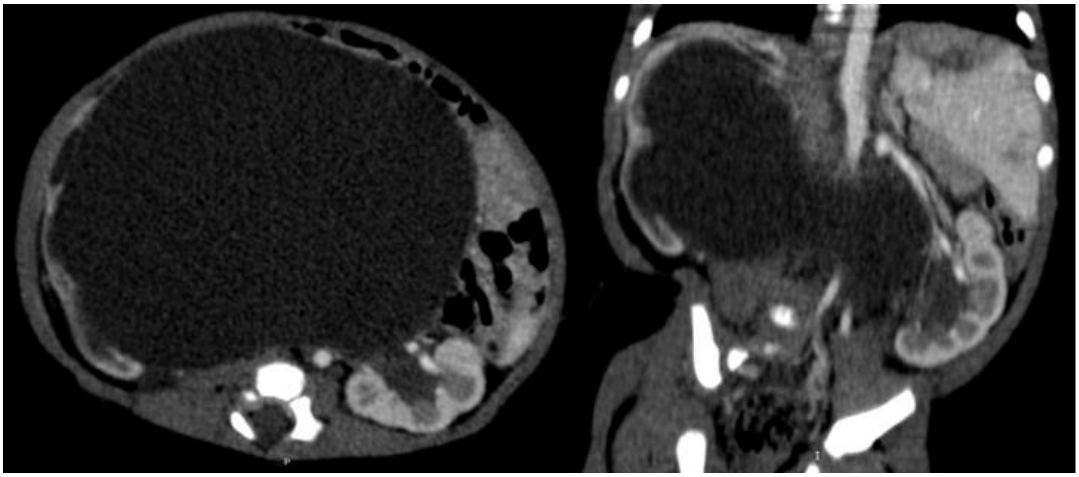

Figure 1: Axial (a) and coronal (b) Multiplanar CT portal venous phase contrast enhacement demonstrating a giant cystic mass occupying the right side of the abdomen and middleline

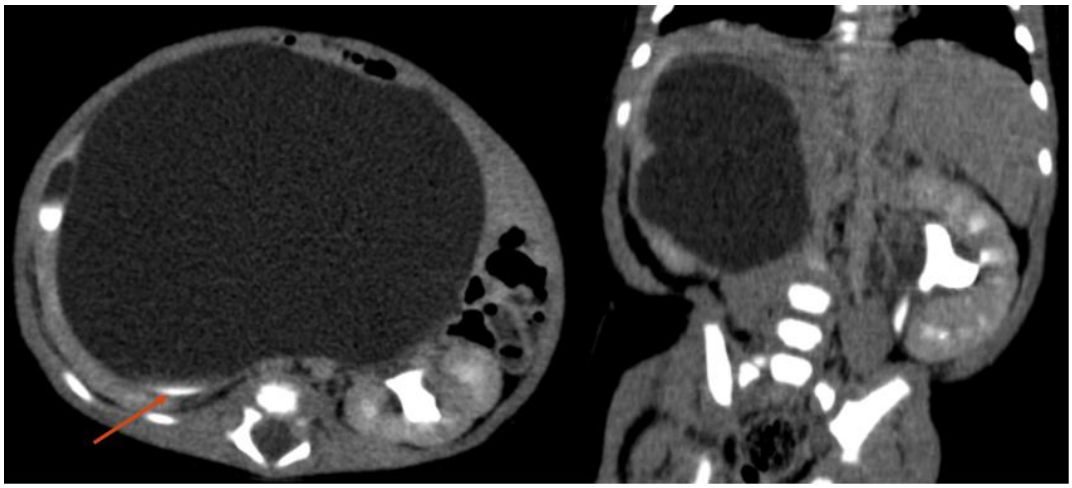

Figure 2: Axial (a) and coronal (b) Multiplanar CT extretory phase demonstrating a giant cystic mass occupying the right side of the abdomen and middleline and showing the contrast is excreted into the calices and into the renal collecting systems

DISCUSSION

Hydronephrosis is the most common mass in pediatric patients. It may occur due to the obstructive or non obstructive causes [5]. Obstructive causes include UPJ obstruction, ureterovesical junction obstruction, obstructed upper moiety of a duplex kidney with ectopic ureterocele, and posterior urethral valves. Congenital UPJ obstruction or congenital hydronephrosis is a common disorder that can be seen in patients of all ages. The majority of congenital obstructions at the UPJ are most likely due to abnormal musculature preventing relaxation at the junction that would allow filling of the ureteral cone and emptying of the renal pelvis.

Whatever the cause, the effects of the obstruction are the same and result in a dilatation of the pelvis and calices from mild to severe. The pelvis and calices fail to empty normally and become variously dilated with a corresponding degree of atrophy of the renal parenchyma [5]. In extreme cases, like this one, a shell of renal cortex remains present. In these cases, UPJ obstruction may cause substantial renal damage. They mostly have interstitial fibrosis, glomerulosclerosis with inflammation, medullary dysplasia, and glomerular changes [7]. Giant hydronephrosis is defined as a hydronephrotic kidney containing greater than 1 l of urine [5]. Al Saleh *et al.*, [8] reported two adult cases with giant pelviureteric hydronephrosis in which the hydronephrotic kidneys contained 12 and 11.5 l of urine.

Newborns and young infants with UPJ obstruction commonly present with a diagnosis already established on prenatal sonography [9]. Most infants appear clinically normal except for a palpable flank mass in some cases.

Older children with UPJ obstruction may present with intermittent abdominal pain, hematuria, or urinary tract infection [9, 10]. Fewer delayed cases were presented, with progressive abdominal distension. Dahniya *et al*., [11] reported a 7-year-old female with giant pelviureteric hydronephrosis. She presented with progressive abdominal distension and evaluated by only plain radiographs and antegrade pyelography [11]. The diagnostic approach to suspected hydronephrosis in children and neonates consists of sonography as the firstline investigation to confirm the diagnosis. Although diagnosis is readily accomplished by sonography in severe hydronephrosis, as ours, it may be confused with other cystic masses of the kidney [6, 10]. CT and MR images were helpful in the differentiation of a giant hydronephrosis from a cystic neoplasm or a large simple cyst by showing absence of the compressed renal parenchyma adjacent to the cyst margins and absence of an enhancement solid component within the cystic mass. In additional, multiplanar MR images demonstrated the location and extent of the hydronephrotic kidney in better detail. The most widely accepted surgical procedure is a type of pyeloplasty in moderate UPJ obstruction. Historically, severe hydronephrotic cases with longstanding obstruction were treated by total nephrectomy. The use of a staged surgical approach with slow decompression has been advised because of the potential cardiopulmonary collapse that might be caused from a sudden decrease in intraabdominal pressure [12]. The management of GH should be treated on a case-by-case basis depending on salvageability of the affected kidney and accessibility of kidney-sparing therapy.

Early diagnosis with accurate preoperative delineation of the anatomy of the affected kidney is important for treatment strategy [5]. Therefore, additions to sonographic findings CT and MR appearances were also useful in to rule out the other cystic masses of the kidney and to confirm the final diagnosis.

CONCLUSION

Giant hydronephrosis is exceptional in children and new born. The pyelo-ureteral junction syndrome is the most frequent cause. The urotomodensitometry (uro-TDM) and MRI are the key examination of the diagnosis. The treatment is surgical, consisting the most often in a nephrectomy. Waiting nephrostomy allows the evaluation of the functional state of the kidney and facilitates the extra-peritoneal approach to the kidney.

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