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Case Report

Case Report: A Giant Hydronephrotic Mass (Twelve Litres of Content) from a Neglected Left Congenital Ureteropelvic Junction Obstruction, in a Seventeen-Year-Old Patient Living in a Low-Resource Context

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Abstract: Introduction: Giant Hydronephrosis (GH) is characterised by an extensive collection of urine in the kidney due to ureteral obstruction, often exceeding one litre or a kidney weighing more than 1.6% of total body weight. It is commonly seen in under-resourced areas and is usually caused by congenital Ureteropelvic junction (UPJ) obstruction. Symptoms include abdominal distention, flank pain, dyspnoea, constipation, fever, and sepsis. Progressive renal damage can occur due to increased intrapelvic pressure. Case Report: A 17-year-old male from a rural area presented with a massive abdominal distention that had been growing since birth. The patient had limited access to medical care, leading to the condition being neglected for years. Upon examination, the abdomen was enlarged but not tender, and vital signs were normal. Laboratory tests and imaging revealed a massive fluid-filled mass, suspected to be GH. Surgery confirmed a hydronephrotic cavity due to a PUJ obstruction. The patient underwent a left nephroureterectomy and was discharged after an uneventful recovery. Discussion: Congenital anomalies of the genitourinary tract (CAKUT) are a significant part of paediatric surgical cases worldwide. Early diagnosis and treatment are crucial to prevent irreversible renal damage. In under-resourced areas, a lack of awareness and constrained access to medical care can delay diagnosis and treatment. Conclusions: The reported case is notable for the large volume of urine retained in GH and the minimal symptoms, which delayed medical attention. A high index of suspicion is required for CAKUT in young patients. Early surgical management is recommended to preserve renal function.

Keywords: Giant Hydronephrosis, Nephrectomy, Ureteropelvic Junction Obstruction.

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Introduction

Giant Hydronephrosis (GH) is a large pelvicalyceal collection of urine exceeding 1 litre or a kidney weighing more than 1.6% of the total body weight, secondary to ureteral obstruction [1].

Most of the reported cases of GH are from under-resourced contexts and due to a congenital Ureteropelvic junction (UPJ) obstruction, which is the most frequently observed obstructive nephropathy, especially in children and young patients. Retroperitoneal fibrosis and tumours, stones and trauma have also been reported among adult patients. Occasional association with renal dysplasia or neoplasm have been described in a few cases.

Half of the about 500 cases of GH in the literature went undiagnosed for a long time due to

nonspecific and often misleading presentations. Abdominal distention and flank pain, dyspnea, constipation, fever and sepsis are most frequently reported symptoms [2-5].

Progressive renal parenchymal damage related to increasing intrapelvic distension and high pressure leads to inflammation, proliferation/apoptosis, activation of the renin-angiotensin system, and ultimately, irreversible loss of nephrons [6].

Conservative treatment of GH by dismembered pyeloplasty, sometimes after a temporary percutaneous decompression, is strictly conditioned by the grade of parenchymal damage and consequently related to the precocity of referral and diagnosis [7].

We report the case of a seventeen-year-old male patient presenting with a progressively growing massive abdominal distention from a GH secondary to congenital PUJ obstruction that went undiagnosed since birth. Although the size of the anomaly was exceptional compared to other reported cases, the mildly symptomatic presentation, the lack of parental awareness, and the delay in seeking medical attention made a potentially correctable congenital anomaly run undetected for a long time, missing any organ-sparing treatment.

CASE REPORT

A 17-year-old male presented with a gradual and slow-growing painless massive abdominal distention since birth. He did not present or report vomit, constipation, difficulties in feeding or micturition. The patient came from a rural area, and the low intensity of associated symptoms, together with uneasy access to adequate medical attention, had made the condition neglected for years. On admission, vital signs were in the normal range: BP=136/76mmHg, PR=66bpm, RR=20 cycles per minute. On examination, the patient was alert, afebrile, not pale or dyspnoeic, with no oedema; the abdomen was massively and uniformly enlarged without tension or tenderness, dull at percussion, and with no palpable organ swelling. The urine output was normal for age and weight. Laboratory values were normal. WBC=6.49, Hb=13.7 g/dl, PLT=199. Renal function tests: creatinine = 63.4 micromol/L, urea = 4.84 mmol/L. Liver function tests were: ALT= 29.5 U/L, AST=36.4

U/L, AST/ALT= 1.2. Total protein 81.1g/Dl. Total cholesterol= 4.0 mmol/L.

Abdominal Ultrasound (US) revealed a massive fluid abdominal content, which was interpreted as ascites. A CT scan [Fig. 1, 2] with opacified gastrointestinal tract documented a fluid-filled uniloculated mass which extended from the left subdiaphragmatic region to occupy most of the abdominal cavity. The left kidney could not be identified, while a right functional kidney with a non-dilated ureter was well visible. The mass compressed the liver and displaced the stomach and the spleen upward, and the bowel laterally to the right. The lower end of the mass reached the pelvis, compressing the bladder and the rectosigmoid. A long-standing GH was suspected.

Through a flank access, the thick wall of the cystic mass was exposed and incised [Fig. 3A]. Twelve litres of clear, straw-coloured fluid were evacuated under tension. The empty collapsed mass confirmed to be a huge hydronephrotic cavity associated with a thigh pelvic ureteric junction stenosis [Fig. 3B]. A compressed and thinned (0.5 cms) residual renal parenchymal area was still identifiable in the mass wall. The dilated pelvis was thickened and adherent to surrounding anatomical structures. The renal artery and vein were ligated separately, and the ureter was interrupted with a transfixed stitch. After the left nephroureterectomy, the abdominal wall was closed in layers, and a drain was left in the empty subdiaphragmatic left region. The patient was discharged after an uneventful outcome.



Figure 1: Multilayers coronal views of Left Giant Hydronephrosis H (Deep: A, Intermediate B, Superficial C)

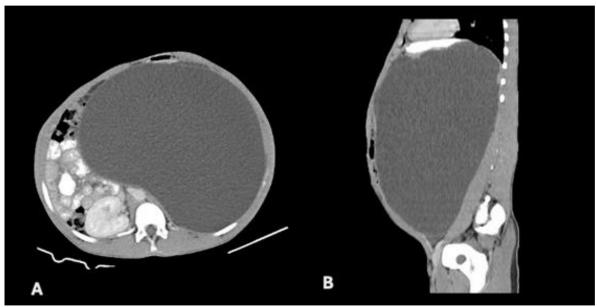


Figure 2: Axial (A) and sagittal (B) views of Left Giant Hydronephrosis

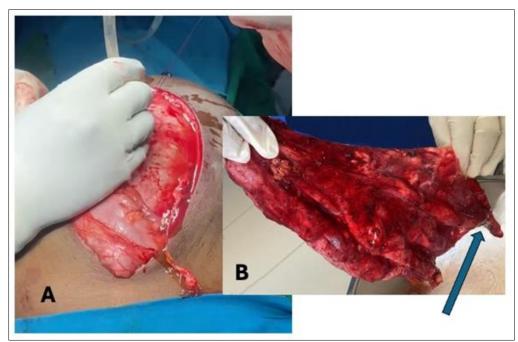


Figure 3: Intraoperative view. A: The left Kidney has been exposed, and the urine content (12 litres) has been aspirated through a small incision on the distended pelvis. B: The collapsed hydronephrotic left kidney has been removed, showing a PUJ obstruction (arrow)

DISCUSSION

Congenital anomalies of the genitourinary tract (CAKUT) account today for a significant portion of the worldwide paediatric surgical workload. Hydronephrosis resulting from UPJ obstruction is the most frequent anomaly discovered and is amenable to successful treatment [8, 9]. Widespread use of prenatal or perinatal Ultrasound (US) screening alerts on a still asymptomatic CAKUT case since foetal life or early infancy. Precocious management reduces the risk of irreversible renal damage from obstructive or reflux anomalies. Children living in areas underserved by adequate

medical services cannot benefit from the advantages of these simple, reliable diagnostic tools for early diagnosis, which are accessible only through a few specialist facilities in major urban areas.

Many CAKUT cases could so far remain undetected throughout childhood until adult age, before more severe symptoms like acute sepsis or abdominal pain led the patient to medical attention.

Poor parental awareness, even of the most evident clinical signs, associated with financial and transportation limitations affecting low-income

individuals residing in rural areas, delays seeking medical advice and restricts access to specialist care. In these contexts, the burden of urinary anomalies may be underestimated, and their epidemiology may be significantly understated. The prevalence of CAKUT in Sub-Saharan Africa is quoted at 1.6 per 1000 births, and Hospital admissions are reported at a mean age of 4.7 years, mainly for sepsis, abdominal pain or voiding symptoms. Forty-five per cent of these children have an already elevated rate of irreversible renal damage. In better-resourced contexts, the higher prevalence of CAKUT, estimated at 3-7 cases per 1,000 births, and a median age at referral of 48 months for postnatally diagnosed cases, reflect a higher and more prompt detection rate [10-12].

The case reported has a special interest due to the exceptional volume of fluid entrapped by the GH (12 litres). To our knowledge, only one case with a higher amount (15 litres) had been reported in a sixty-five-year-old Indian woman. The second aspect of interest lies in the few associated symptoms that contrast with the volume silently reached by the mass, which, in a certain way, justifies a lack of parental awareness leading to delayed referral to medical care, thereby precluding a possible organ-sparing surgery.

CONCLUSIONS

Congenital PUJ obstruction, with its prevalence estimated at 1 in 500 to 1 in 1,500 live births, is the most common cause of hydronephrosis, affecting boys twice as often as girls and the left kidney twice as often as the right [13]. Early surgical management is recommended to preserve renal function. Diagnosis relies on a simple US investigation, which is an excellent screening examination for CAKUT in early life. US effectiveness is enhanced by a high index of suspicion for a specific condition within a particular age group. Unfortunately, in under-resourced contexts, the high occurrence rate of CAKUT in young patients is still underestimated, and the risk of missed or delayed diagnosis remains elevated [11].

Ethical Considerations

This case report does not contain any patient identifiers, and neither does it show picture(s) of any identifiable patient. Confidentiality has been strictly adhered to. Only de-identified information is presented for the benefit of the medical fraternity and community at large.

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