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Case Report

Massive Congenital Giant Megaureter Associated with a Complete Duplex Collecting System Mimicking a Mesenteric Cyst in a Neonate: A Diagnostic Challenge

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Abstract

Introduction: Congenital giant megaureter (CGMU) is a rare urinary tract anomaly that may present as a large abdominal cystic mass in neonates, creating significant diagnostic challenges.

Case Presentation: A 15-day-old male neonate presented with progressive abdominal distension and decreased urine output. Ultrasound and contrast-enhanced computed tomography (CECT) demonstrated a large multiloculated abdominopelvic cystic lesion, initially suspected to be a mesenteric cyst. Exploratory laparotomy revealed a massive urine-filled CGMU associated with a right complete duplex collecting system and a blindly ending distal ureter. Surgical management included ureterectomy with proximal ureteroureterostomy. The postoperative course was uneventful.

Clinical Discussion: This case highlights the diagnostic difficulty of CGMU when standard imaging fails to demonstrate communication with the urinary tract. Marked ureteral dilatation and distorted anatomy may obscure the lesion's origin.

Conclusion: CGMU should be considered in the differential diagnosis of large neonatal abdominopelvic cystic masses. Delayed excretory-phase imaging or MR urography may improve preoperative diagnosis in suspected complex genitourinary anomalies.

Introduction

Congenital Giant Megaureter (CGMU) is an uncommon congenital abnormality in the pediatric population, defined as dilatation of the ureter by more than 10 times the normal size, with a normal urinary bladder [1]. CGMU has been reported in association with several congenital genitourinary anomalies, including duplex collecting systems, megacalicosis, contralateral hydronephrosis, and cryptorchidism [2]. This case report highlights the limitations of standard ultrasound and portal venous phase CT in establishing the genitourinary origin of a poorly communicating CGMU.

Case Presentation

A fifteen-day-old full-term boy weighing 2.8 kg presented with progressive abdominal distension and decreased urination for one week. The antenatal period was unsupervised. The first passage of meconium was normal. The abdominal examination revealed an ill-defined, soft mass with a smooth surface occupying the majority of the abdominopelvic cavity and bilaterally empty scrotum. Laboratory investigations, including complete blood count, liver function tests, and urinalysis, were unremarkable except for a mildly elevated serum creatinine level of 1.5 mg/dL, which normalized following resuscitation. An ultrasound of the abdomen showed a large anechoic cystic mass occupying the majority of the abdominopelvic cavity, with thin incomplete septation, but did not accurately identify the origin of the mass. The mass insinuated into the pelvis, and there was mild hydronephrosis on the right side. Both kidneys had normal shape and size. The bladder was mildly dilated with a smooth wall of normal thickness. A Contrast-Enhanced Computed Tomography (CECT) scan of the abdomen and pelvis [Figure 1 A and C] showed a large, non-enhancing, irregularly shaped cystic lesion involving the entire abdomen and pelvis, insinuating into and causing compression of adjacent structures. There was associated dilatation of the proximal ureter and pelvicalyceal system on the right side (Figure 1 B). The CT scan also revealed normal-shaped bilateral kidneys. No contrast opacification of the cystic lesion was observed during the acquired imaging phase.

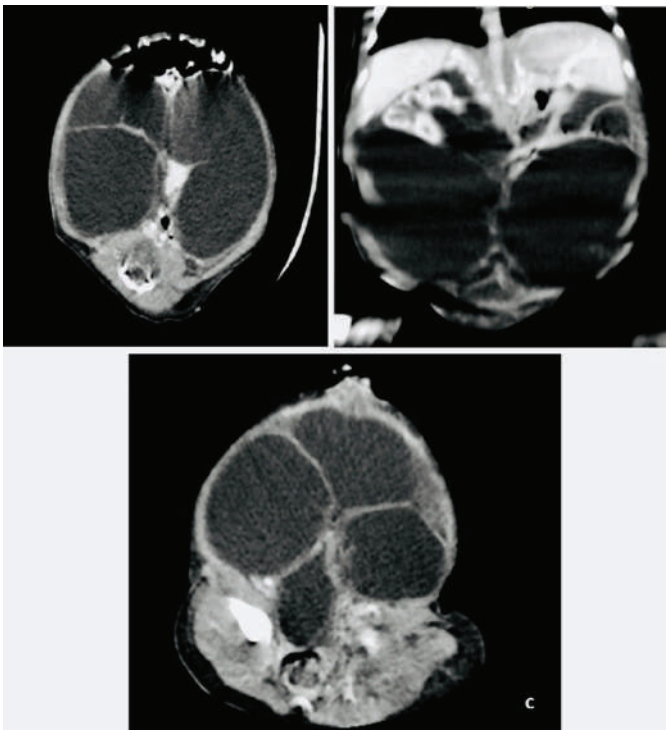

Figure 1: Contrast enhanced computed tomography findings (a) Large Cystic mass occupying the upper abdomen (b) large cystic lesion involving the entire abdomen and pelvis and causing compression of the right kidney (c) large cystic mass extending into the pelvis.

After stabilization of the neonate, the parents were ounselled thoroughly and the neonate was taken to the operating room after informed consent was obtained. The surgery was undertaken under general anesthesia and the abdominal cavity was entered using a supraumbilical transverse incision. During surgery, a cystic fluid-filled mass occupying almost the whole of the pelvis and abdomen was discovered (Figure 2A). Further exploration demonstrated that the lesion was identified as a urine-filled CGMU associated with a right complete duplex kidney. The distal duplicated ureter terminated blindly near the bladder without communication to the urinary bladder. Gross renal parenchymal architecture appeared preserved intraoperatively in both upper and lower moieties (Figure 2B). Both testes were found intra-abdominally, with the

right testis attached to the duplicated ureter and twisted with ischemic changes (Figure 2C). The right testis appeared ischemic and non-viable secondary to torsion and dense attachment to the duplicated ureter; therefore, right orchiectomy was performed. Left orchidopexy and right ureterectomy with proximal ureteroureterostomy were performed (Figure 2D). The postoperative course was uneventful. Serum creatinine normalized to 0.4 mg/dL, and follow-up ultrasonography demonstrated decompression of the collecting system.

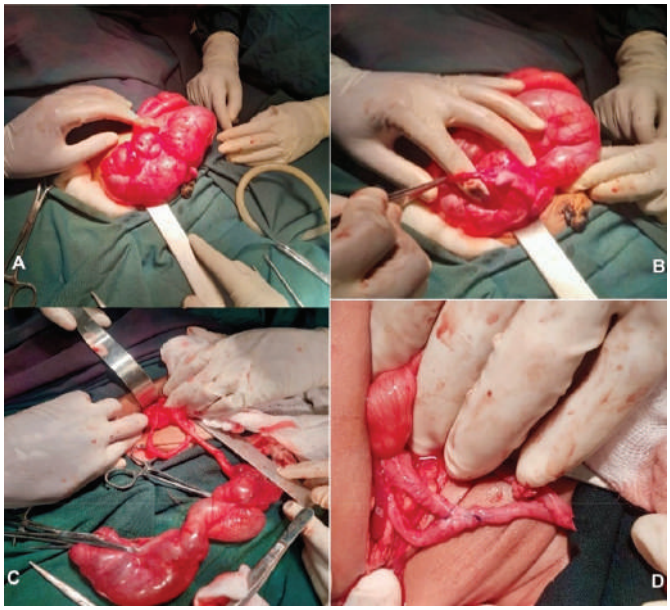

Figure 2: Intraoperative findings. (A) Giant cystic abdominal mass occupying the abdominopelvic cavity. (B) Surgical dissection of the cystic lesion demonstrating the giant megaureter. (C) Complete mobilization of the giant megaureter following dissection. (D) Bilateral intra-abdominal testes identified during surgery, with the right testis demonstrating torsion and ischemic changes.

Histopathology

Histopathology of the resected ureteral specimen confirmed markedly dilated ureteral tissue with muscular hypoplasia and chronic inflammatory changes. The right testicular specimen demonstrated ischemic necrosis consistent with prolonged torsion.

Discussion

Congenital Giant Megaureter (CGMU) is a rare congenital anomaly characterized by marked dilatation of the ureter, conventionally defined as enlargement exceeding 10 times the normal ureteral diameter in the presence of a normal urinary bladder [3, 4]. The underlying mechanism is thought to involve abnormal development of the distal ureteral muscular layer, resulting in impaired peristalsis and progressive dilatation [5, 6]. CGMU may occur as a primary obstructive, refluxing, or non-refluxing non-obstructive megaureter and has been reported in association with other genitourinary anomalies, including duplex collecting systems, megacalicosis, hydronephrosis, and cryptorchidism [2, 7-9]. Most reported cases of CGMU are diagnosed during the prenatal period or are recognized postnatally because imaging clearly demonstrates continuity between the cystic lesion and the urinary tract [1, 2, 7]. In contrast, the present case posed a significant diagnostic challenge. Despite ultrasonography and contrast-enhanced CT, the lesion was interpreted as a large mesenteric cyst because neither imaging modality established its urinary tract origin. The coexistence of a complete duplex collecting system with a blindly ending distal duplicated ureter created a poorly communicating, non-draining collecting system that masked the diagnosis until surgical exploration. To our knowledge, this combination of a massive CGMU, complete duplex collecting system, blind-ending duplicated ureter, and imaging findings mimicking a mesenteric cyst has rarely been reported. Previous reports describing CGMU associated with duplex collecting systems generally established the diagnosis preoperatively using ultrasonography or complementary imaging modalities. In contrast, our patient presented with a giant non-communicating megaureter whose markedly distorted anatomy and absent functional communication with the urinary tract prevented identification of its origin on both ultrasonography and contrast-enhanced CT, ultimately necessitating exploratory laparotomy to establish the definitive diagnosis.

This case also illustrates important limitations of conventional imaging in evaluating large neonatal cystic abdominal masses. Ultrasonography remains the first-line investigation for neonatal abdominal and genitourinary abnormalities; however, when a cystic lesion becomes extremely large, distortion of the normal anatomy may obscure its site of origin. Similarly, the CT examination in our patient was performed during the portal venous phase and failed to demonstrate contrast opacification of the lesion. The absence of communication between the giant megaureter and the functioning collecting system, together with poor drainage, likely prevented contrast filling of the dilated ureter, resulting in an imaging appearance more consistent with a mesenteric cyst than a urinary tract abnormality. This emphasizes that a normal-appearing kidney or the absence of contrast within a cystic lesion does not exclude a congenital urinary tract anomaly. Surgical exploration established the definitive diagnosis of a urine-

filled congenital giant megaureter arising from a complete duplex collecting system with a blind-ending distal ureter. Ureterectomy with proximal ureteroureterostomy successfully relieved the obstruction while preserving the functioning renal moiety. An additional noteworthy finding was ipsilateral intra-abdominal testicular torsion with irreversible ischemia, necessitating orchiectomy. Although cryptorchidism has been described in association with CGMU, the coexistence of a giant megaureter, duplex collecting system, and torsion of an undescended testis is exceptionally uncommon and further highlights the complexity of this congenital anomaly.

Our report has several limitations. Functional imaging with renal scintigraphy and MR urography was unavailable, limiting preoperative assessment of differential renal function and detailed delineation of the collecting system. Furthermore, delayed excretory-phase CT imaging was not performed and may have demonstrated urinary tract communication if residual drainage had been present. This case provides an important clinical lesson. Congenital giant megaureter should remain an important differential diagnosis in neonates presenting with a large cystic abdominopelvic mass, even when initial ultrasonography and conventional CT findings appear to favor a gastrointestinal or mesenteric lesion. When imaging findings are inconclusive and a genitourinary origin remains possible, delayed excretory-phase CT urography or MR urography should be considered to improve preoperative diagnostic accuracy and facilitate appropriate surgical planning.

Conclusion

Congenital giant megaureter is a rare but important differential diagnosis in neonates presenting with large cystic abdominopelvic masses, particularly when conventional imaging findings are inconclusive or suggest a non-genitourinary lesion. This case demonstrates that markedly dilated, poorly communicating megaureters associated with complex congenital anomalies may obscure their urinary tract origin on ultrasonography and routine contrast-enhanced CT. When a genitourinary abnormality remains a diagnostic possibility, delayed excretory-phase CT urography or MR urography should be considered to improve preoperative diagnosis, guide surgical planning, and avoid unexpected intraoperative findings.

Research registration number: NA.

Ethical Approval: The report was conducted as per the Declaration of Helsinki. At Haramaya University's Health Science Campus, the Institutional Review Board granted ethical clearance, including publishing this patient's case information. The patient's privacy and confidentiality were protected.

Data Availability: The data used to support the findings of this study are available from the corresponding author upon reasonable request.

Consent: The patient's parents gave written informed consent for this case report and any related images to be published.

Conflicts of Interest: The authors declare that they have no conflicts of interest.

Authors' Contributions: All authors contributed to data analysis, drafting, or revising of the article, have agreed on the journal to which the article will be submitted, gave final approval of the version to be published, and agree to be accountable for all aspects of the work.

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