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Single Case

A Case of Multicystic Dysplastic Kidney Presenting as a Single Midline Pelvic Cyst

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Multicystic dysplastic kidney · Congenital cysts · Pediatric nephrology · Case report

Abstract
We present an unusual case of a female neonate presenting with a single midline pelvic cyst. Prenatal imaging was suggestive of multicystic dysplastic kidney (MCDK), but postnatal imaging was atypical for this diagnosis given the location and singular cyst noted. The patient ultimately underwent surgical exploration and was diagnosed with an ectopic MCDK. Ectopic MCDK should be considered in the differential diagnosis of unilocular cystic pelvic lesions identified in the perinatal period.

Introduction

Fetal abdominal cystic lesions are relatively common, with an incidence of approximately 1 per 1,000 fetuses [1, 2]. However, the differential diagnosis is extensive and includes malignancies, anomalies of the GU tract, Müllerian duct anomalies, vascular and lymphatic malformations, and other cystic lesions of the bowel or spinal cord. We present the workup and management of an unusual case of an ectopic multicystic dysplastic kidney (MCDK) presenting as a unilocular pelvic cyst. The CARE Checklist has been completed by the authors for this case report, attached as online supplementary material (for all online suppl. material, see https://doi.org/10.1159/000530925).

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Case Presentation

A 4-week-old female was referred for evaluation of a possible left MCDK. This was first noted on prenatal ultrasound at 16 weeks of gestation; the fetus was noted to have no identifiable left kidney, as well as a large multicystic lesion in the abdomen. No other abnormalities with other organ systems were detected on prenatal imaging. Four prenatal ultrasounds were performed; by 36 weeks of gestation, the lesion was unilocular. Of note, the patient's mother has a history of left renal agenesis secondary to obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) syndrome.

The patient was born full-term via C-section. At time of presentation, she was asymptomatic. She did not have developmental or psychomotor delays. There was no palpable mass on examination.

She did have a renal ultrasound on the day of her visit, which showed no tissue in the left renal fossa and a 5.1 × 3.1 × 3.9-cm single cyst in the left lower quadrant with septations. A dedicated pelvic ultrasound re-demonstrated this pelvic cystic lesion crossing midline (shown in Fig. 1a). No ovarian tissue was seen. Radiology felt the most likely diagnosis was an ovarian lesion. The differential diagnosis also included lymphatic malformation, enteric duplication cyst, ectopic MCDK, cystic neuroblastoma, or obstructed Müllerian structure. Pediatric gynecology recommended a pelvic ultrasound in 4 weeks.

This repeat ultrasound was able to identify both ovaries, and the mass was felt to be non-ovarian (shown in Fig. 1b). The uterus was noted as bicornuate. Given these findings, magnetic resonance imaging (MRI) of the abdomen/pelvis was obtained. This confirmed the lesion was separate from the ovaries. Although this imaging was not consistent with ovarian pathology, it did expand the differential diagnosis to include paraovarian/paratubal lesion or neurenteric cyst (shown in Fig. 1c).

Ultrasound of the pelvis and kidneys was obtained 2 months later. These now identified a second multicystic structure in the left adnexa measuring 1.5 × 0.8 × 1.3 cm, concerning for a potential MCDK versus obstructed Müllerian structure (shown in Fig. 1d). Some nodular debris was also noted in the larger midline cyst, adding cystic ovarian neoplasm to the differential diagnosis.

Laboratory testing to evaluate for neuroblastoma (homovanillic acid and vanillylmandelic acid) and germ cell tumor (alpha-1 fetoprotein and beta human chorionic gonadotropin) was unremarkable. Estradiol and testosterone were checked to evaluate functional ovarian mass and were normal. No testing for genetic syndromes was performed.

Given that the diagnosis remained unclear and malignancy could not be excluded, we proceeded with surgical exploration. At age of 7 months, the patient underwent cystoscopy with retrograde pyelogram, vaginoscopy, and surgical exploration. Cystoscopy showed a normal bladder with an orthotopic right ureteral orifice and no visible left ureteral orifice. A right retrograde pyelogram did not show any abnormalities. We placed a right ureteral catheter to aid in identification of her solitary kidney's ureter during dissection of the pelvic mass. Vaginoscopy revealed a longitudinal vaginal septum with two normal cervices, one on either side of the septum.

We proceeded with removal of the cyst trans-abdominally via a Pfannenstiel incision. The cystic structure was identified in the retroperitoneum via a transmesenteric approach (shown in Fig. 2a). It was carefully dissected circumferentially, during which we did identify a small, possibly vascular stalk holding it posteriorly. This was transected with cautery. The cyst was removed intact. Grossly, it appeared thin walled without solid components (shown in Fig. 2b). We inspected the retroperitoneum and found no communications from the cyst to other structures. The didelphic uterus was observed, each with a corresponding normal-appearing fallopian tube and ovary. There was no dilation of the gynecologic structures. The patient did well postoperatively and was discharged on postoperative day 1.

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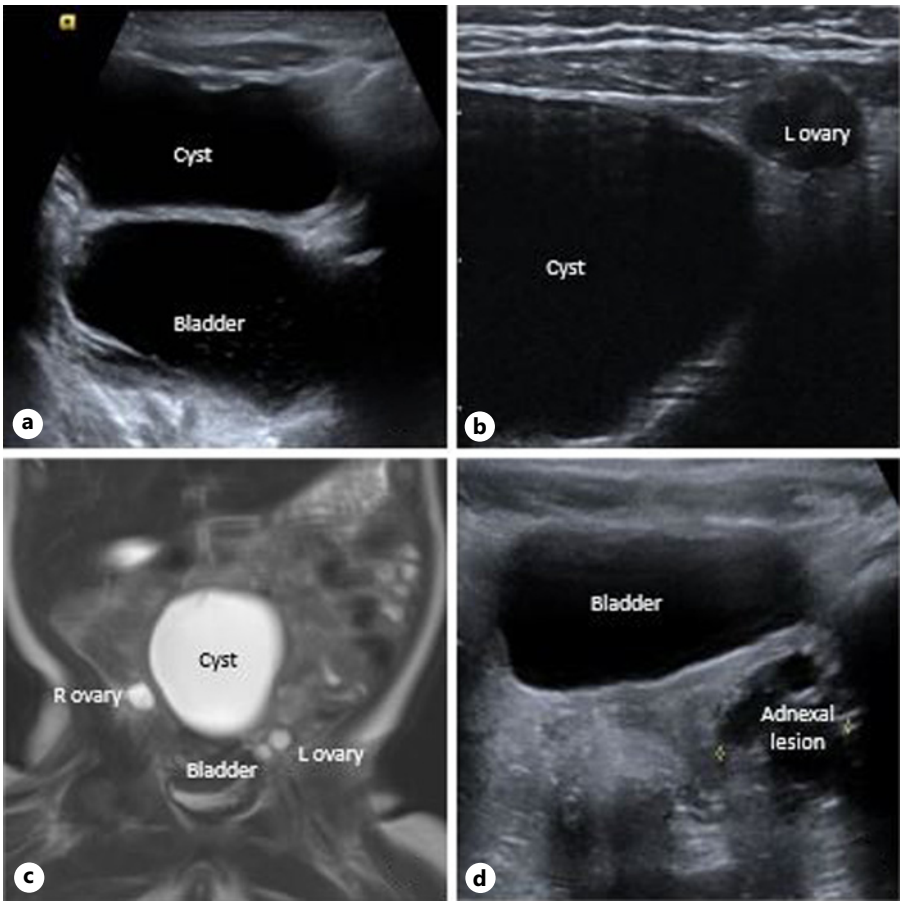

Fig. 1. a Midline pelvic cystic lesion superior to the bladder on pelvic ultrasound. **b** Cyst noted to be separate from left ovary on pelvic ultrasound (right ovary not pictured). **c** Cyst identified in the retroperitoneum and identified as separate from both ovaries and the bladder on T2-weighted MRI. **d** Multicystic lesion in left adnexa on pelvic ultrasound.

Surgical pathology returned as benign cystic tissue containing abortive glomeruli and renal tubules and mesenchyme-like tissue (shown in Fig. 2c). These findings were most consistent with a cystic dysplastic kidney.

The patient was seen 2 months postoperatively and was doing well. The family was counseled on the implications of a solitary kidney. She is planned to be seen yearly with a renal ultrasound.

Discussion

We report on a case of an MCDK presenting as a large, midline, unilocular pelvic cyst in a female infant; this unusual morphology presented a diagnostic challenge. In females, ovarian cysts are one of the most common fetal cystic lesions [3]; these form due to stimulation of the fetal ovaries by maternal and placental hormones [4]. However, the differential diagnosis of a fetal abdominal cyst is extensive. In many cases, ultrasound alone is able to make the diagnosis. MRI is a useful adjunct, as it may more clearly define any association between the cyst and a specific organ.

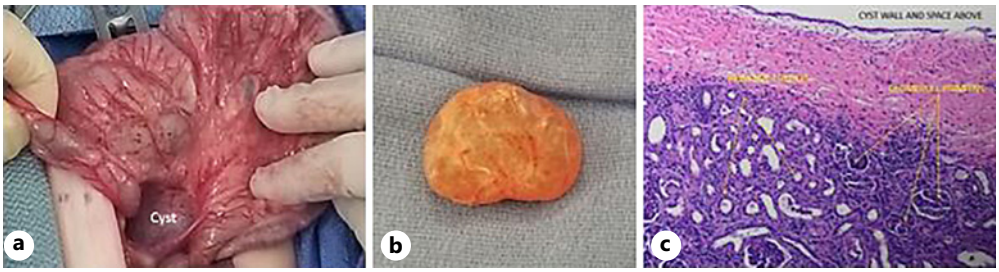

Fig. 2. **a** Intraoperative view of cyst identified in the retroperitoneum, posterior to small bowel mesentery. **b** Gross appearance of excised cyst. **c** Primitive glomeruli and renal tubules identified on pathology (hematoxylin and eosin stain).

Given that this patient had no renal tissue in the left renal fossa and a family history of OHVIRA, an ectopic renal anomaly was included in the differential diagnosis for this cystic mass. OHVIRA is theorized to be caused by mesonephric duct-induced Müllerian anomalies [5] and consists of uterine didelphys, unilateral obstruction of one hemivagina, and ipsilateral renal anomalies, including MCDK or renal agenesis. Its incidence is unknown; the syndrome has been primarily reported in small case series from tertiary care centers [6]. It can present in infancy because the obstructed hemivagina can collect secretions made secondary to maternal hormones. Our patient did not have evidence of hemivaginal distention on her imaging; however, patients often do not develop symptoms from obstructed hemivagina until after menarche [7]. Given findings of her ectopic MCDK, didelphic uterus, and vaginal septum, this patient very likely has OHVIRA like her mother. Congenital anomalies of the kidney and urinary tract including OHVIRA may have a monogenic cause in an estimated 14–20% of patients [8]; genetic testing could have been considered for this patient given the positive family history but was not done.

MCDK, a congenital renal anomaly where the kidney is replaced with multiple variable-sized non-communicating cysts, is an anomaly often seen in association with OHVIRA. The incidence of MCDK is between 1 and 2,400–4,300 live births [9, 10]. Its etiology may be related to congenital renal obstruction, as it is often associated with ureteral atresia [11, 12]. MCDK has been described as a single predominant cyst [11, 13, 14], but this is an uncommon presentation that is difficult to distinguish from severe hydronephrosis with parenchymal thinning. Magnetic resonance urography can help differentiate between MCDK and severe hydronephrosis [15]. In this case, MRI was unable to make a definitive diagnosis because of the unique midline pelvic location of the single cyst.

When imaging cannot make a clear diagnosis, the decision must be made to either continue with observation or proceed with surgical exploration. We recognize many physicians may want to take a conservative route in a pediatric patient; however, we elected to proceed with exploration given that malignancy could not be ruled out. Although the absence of a left ureteral orifice suggested an MCDK, we were unable to intraoperatively identify a ureter or any communication from the cyst to other structures. Pathologic examination ultimately confirmed the diagnosis of MCDK. Ultimately, the patient's family was satisfied that the patient underwent surgery, as it was able to confirm the diagnosis.

Conclusion

Ectopic MCDK should be considered in the differential diagnosis of unilocular cystic pelvic lesions identified in the perinatal period. Ultrasound and MRI/magnetic resonance urography are likely to identify the correct diagnosis in most cases, but surgical exploration may be warranted in the case of equivocal imaging, particularly when malignancy cannot be excluded.

Statement of Ethics

Ethics approval was not required for this study per guidelines of the Institutional Review Board at Nationwide Children's Hospital (IRB16-00249). Written informed consent was obtained from the patient's parents for publication of the details of their medical case and any accompanying images.

Conflict of Interest Statement

The authors of this study have no conflicts of interest to disclose.

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Author Contributions

Christina Ching encouraged Kristin Ebert to investigate and publish this case. Kristin Ebert performed chart review and wrote the manuscript under the supervision of and with critical feedback from Christina Ching.

Data Availability Statement

All data generated or analyzed during this study are included in this article. Further inquiries can be directed to the corresponding author.

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