American Journal of Case Reports

ISSN 1507-6164
© Am J Case Rep, 2013; 14: 153-156
DOI: 10.12659/AJCR.889050

Received: 2013.02.14
Accepted: 2013.04.03
Published: 2013.05.15

Cystic adnexal mass in a 16-year-old female: Ovarian pathology or complication of a Müllerian anomaly?

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

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Patient: Female, 16
Final Diagnosis: Pelvic mass
Symptoms: None
Medication: None
Clinical Procedure: CT • MRI
Specialty: Diagnostic radiology • pediatrics

Objective: Unusual presentation of unknown etiology, Rare disease, Mistake in diagnosis

Background: Müllerian anomalies encompass a wide variety of malformations in the female genital tract, usually associated with renal and anorectal malformations. Of these anomalies, approximately 11% are uterus didelphys, which occurs when midline fusion of the müllerian ducts is arrested to a variable extent.

Case Report: We report the case of a 16-year-old female with uterine didelphys, jejunal malrotation, hematometra, hematosalpinx, and bilateral subcentimeter homogenous circular cystic-like renal lesions, who initially presented with left lower quadrant abdominal pain, non-bloody vomiting, and a history of irregular menstrual periods. Initial CT was confusing for an adnexal cystic mass, but further imaging disclosed the above müllerian anomalies.

Conclusions: Müllerian anomalies may mimic other, more common, adnexal lesions; thus, adequate evaluation of suspicious cystic adnexal masses with multiple and advanced imaging modalities such as MRI is essential for adequate diagnosis and management.

Key words: mullerian • uterus didelphys • hydrometrocolpos • intestinal malrotation • hematometra • hematosalpynx • renal cysts

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Background

Müllerian anomalies encompass a wide variety of malformations in the female genital tract, usually associated with renal and anorectal malformations [1]. Their occurrence is sporadic and there is no evidence of familial inheritance [1]. Of these anomalies, approximately 11% are uterus didelphys, which occurs when midline fusion of the müllerian ducts is arrested to a variable extent [2]. The full manifestation of this anomaly is defined by 2 hemiuteri, 2 endocervical canals, and fused cervices.

They are associated with ovarian malposition and obstruction due to the presence of a transverse vaginal septum [3]. Seventy-five percent of Didelphic anomalies also have a complete longitudinal vaginal septum [4]. There is a reported 20% incidence of renal anomalies in patients with uterus didelphys, with renal agenesis the most commonly reported. Although 1 case report showed renal cysts in a patient with complete septate (bicornuate) uterus, renal cysts have not been reported with uterus didelphys.

We report the case of a patient who was diagnosed with uterine didelphys, jejunal malrotation, and bilateral renal cysts. We describe the findings from computed tomography (CT) and magnetic resonance imaging (MRI). We also review the literature on associated renal anomalies and the management of uterine didelphys with obstructed vagina. To the best of our knowledge this is the first reported case to include renal cysts and uterine didelphys.

Case Report

A 16-year-old female without previous history of medical illnesses was admitted to our department due to non-bloody vomiting and left lower quadrant abdominal pain of 1-week duration. Review of systems was noncontributory except for a history of irregular menstrual periods. Physical examination did not disclose any abnormalities. Laboratory studies performed in-hospital were all negative. Previous imaging studies performed at another institution disclosed a complex cystic mass in the left adnexal region, suggesting a diagnosis of ovarian cystadenoma. Since the actual films where not available for review, an abdomino-pelvic CT was subsequently performed at our institution, which showed 2 distinct uterine cavities (Figure 1), hydrometrocolpos, intestinal malrotation with the jejunum in the right upper quadrant without evidence of obstruction, multiple bilateral renal cyst-like lesions (Figures 2 and 3), and a left retroaortic renal vein. Since the initial CT scan could not clearly differentiate the number of cervices, a pelvic MRI with contrast was ordered for clarification. MRI was performed and showed a duplicated uterus,

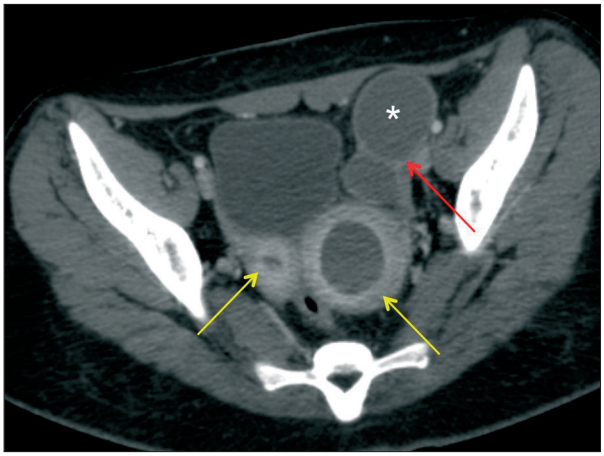

Figure 1. MDCT with IV contrast of the Abdomen and Pelvis shows two distinct uterine cavities with a dilated left cavity (yellow arrows). A tortuous and dilated left fallopian tube is also seen (red arrows). Both uterine cavities and the fallopian tube show hypodense intraluminal fluid (white asterisk).

which was consistent with uterus didelphys, and a rudimentary left cervix associated with distention of the left uterine cavity and left fallopian tube (hematometra and hematosalpinx) (Figures 4 and 5). An upper GI series was also performed for better assessment of the malrotation seen on the previous CT scan. The GI series confirmed jejunal mal-rotation (Figure 6). The patient was thus referred to OBGYN and Pediatric Surgery for further management. The patient has continued with follow-up at the OBGYN clinics and has not had surgery at this time. The patient will continue under medical management with eventual surgical intervention.

Discussion

The clinical presentation of uterus didelphys depends on the coexistence of hemivaginal obstruction. Non-obstructive uterus didelphys is usually asymptomatic until menarche and can be associated with second-trimester spontaneous abortions. Those with hemivaginal obstruction may present with dysmenorrhea, progressive pelvic pain, unilateral pelvic mass, marked rectal pain, and constipation secondary to hematocolpos infringement.

Correcting obstructed anomalies is important because hematometra and hematosalpinx may develop from retrograde menstruation [5]. If corrected, risk of poor reproductive outcomes and simultaneous pregnancies may be reduced [6]. Uterine didelphys should be worked-up with hysterosalpingogram, MRI, and intravenous pyelogram to diagnose the typically identified 2 separate uterine horns and cervices and to rule out urinary tract anomalies [7]. Transvaginal ultrasonography and sonography can also be used [8].

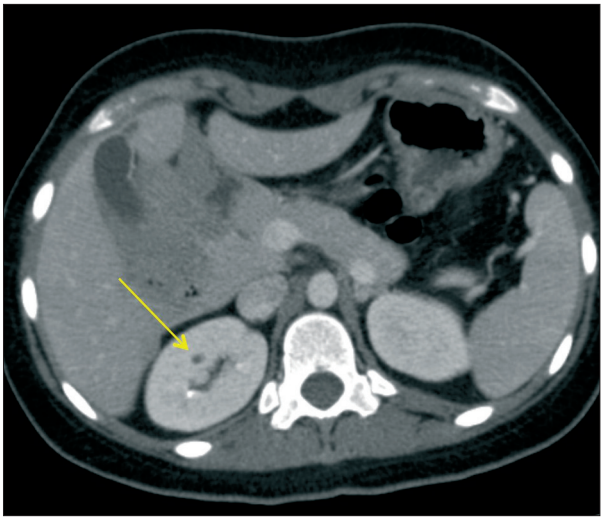

Figure 2. MDCT with IV contrast of the abdomen and pelvis shows a hypodense well circumscribed lesion in the right kidney favoring a simple renal cyst (yellow arrow). Other similar lesions were seen in the same kidney, not shown.

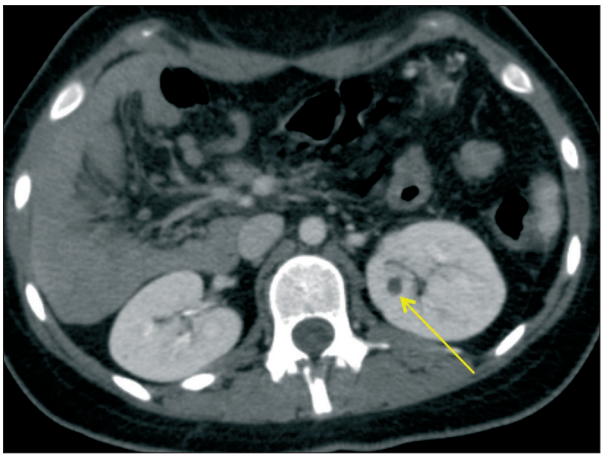

Figure 3. MDCT with IV contrast of the abdomen and pelvis shows a hypodense well circumscribed lesion in the left kidney favoring a simple renal cyst (yellow arrow). Other similar lesions were seen in the same kidney, not shown.

The reported incidence of renal anomalies in patients with uterus didelphys is 20% [9]. This is thought to be due to erroneous induction of nephrogenesis and malpositioning of ipsilateral müllerian ducts from Wolffian duct developmental arrest. The 2 hemiuteri resulted from unsuccessful fusion of the malpositioned müllerian duct. Other described related anomalies include ureteral agenesis, bladder exstrophy, and congenital vesicovaginal fistula [10–12]. Renal agenesis is the most commonly reported associated renal anomaly. Renal cysts have been reported once with complete septate (bicornuate) uterus, but to the best of our knowledge never with uterus didelphys [13]. Familial cases have been reported, and recently mutations in the hepatocyte nuclear factor-1β (HNF1B) gene have been commonly found in combined uterine and renal malformations.

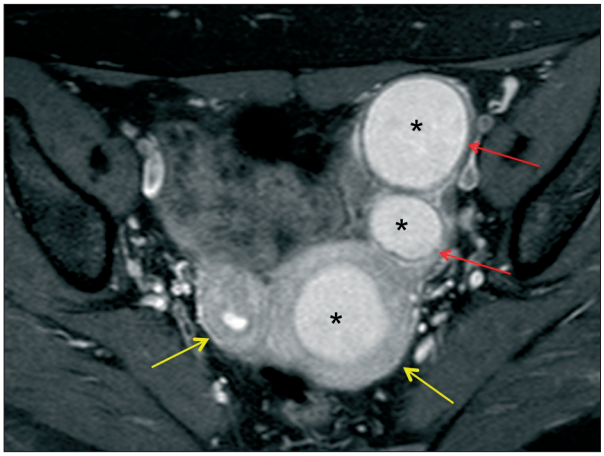

Figure 4. Fluid sensitive Pelvic MRI sequence shows two distinct uterine cavities with a dilated left uterine cavity (yellow arrows). A tortuous and dilated fallopian tube is also seen (red arrows). Hyperintense material is visualized within the uterine cavities and fallopian tube (asterisks)

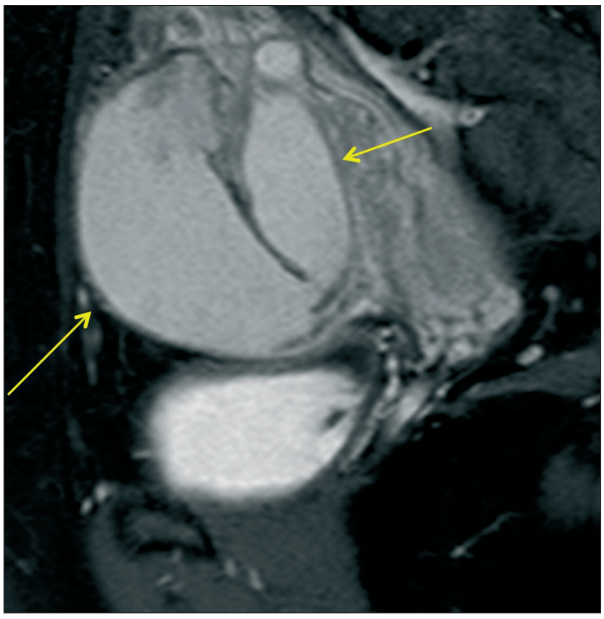

Figure 5. Fluid sensitive MRI sequence shows a sausage shaped mass in the pelvis representing a folded and tortuous fallopian tube (yellow arrows).

Surgical management of uterine didelphys with obstructed hemivagina includes full excision and marsupialization of the vaginal septum, leaving enough pedicle to decrease possible bleeding in case the vaginal mucosa retracts [14]. Laparoscopy can then be used to treat coexisting endometriosis and/or adhesions [15]. Non-obstructed manifestations have few indications for septal resection because it may be difficult and may result in cervical incompetence or stenosis [16]. The decision to perform metroplasty should be individualized and

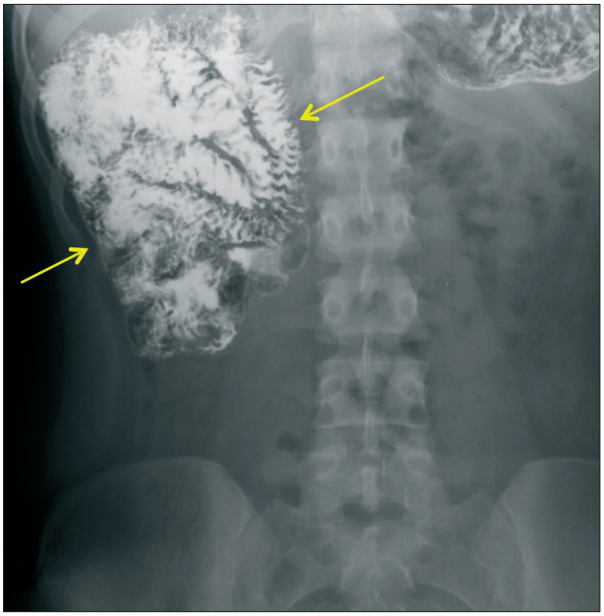

Figure 6. UGIS shows small bowel malrotation (yellow arrows).

may take into consideration dyspareunia, recurrent abortions, or preterm births.

Our patient initially presented with left lower quadrant abdominal pain, non-bloody vomiting, and a history of irregular menstrual periods. Initial CT was confusing for a cystic mass, but further imaging disclosed the above müllerian anomalies. The patient continues under close medical management at our institution.

Conclusions

Müllerian anomalies encompass a wide variety of genital and renal abnormalities. Diagnosis can be difficult and challenging, and the presenting symptoms are usually vague and non-specific, as in our case. Initial imaging studies can also be confusing and lead to an initially incorrect diagnosis, hence delaying management and treatment. We recommend careful assessment of young females presenting with abdominal complaints and abnormal uterine bleeding, utilizing accurate imaging modalities such as CT and MRI for accurate identification and diagnosis.

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