Hematometrocolpos Secondary to Didelphic Uterus and Unilateral Imperforated Double Vagina as an Unusual Cause of Acute Abdomen

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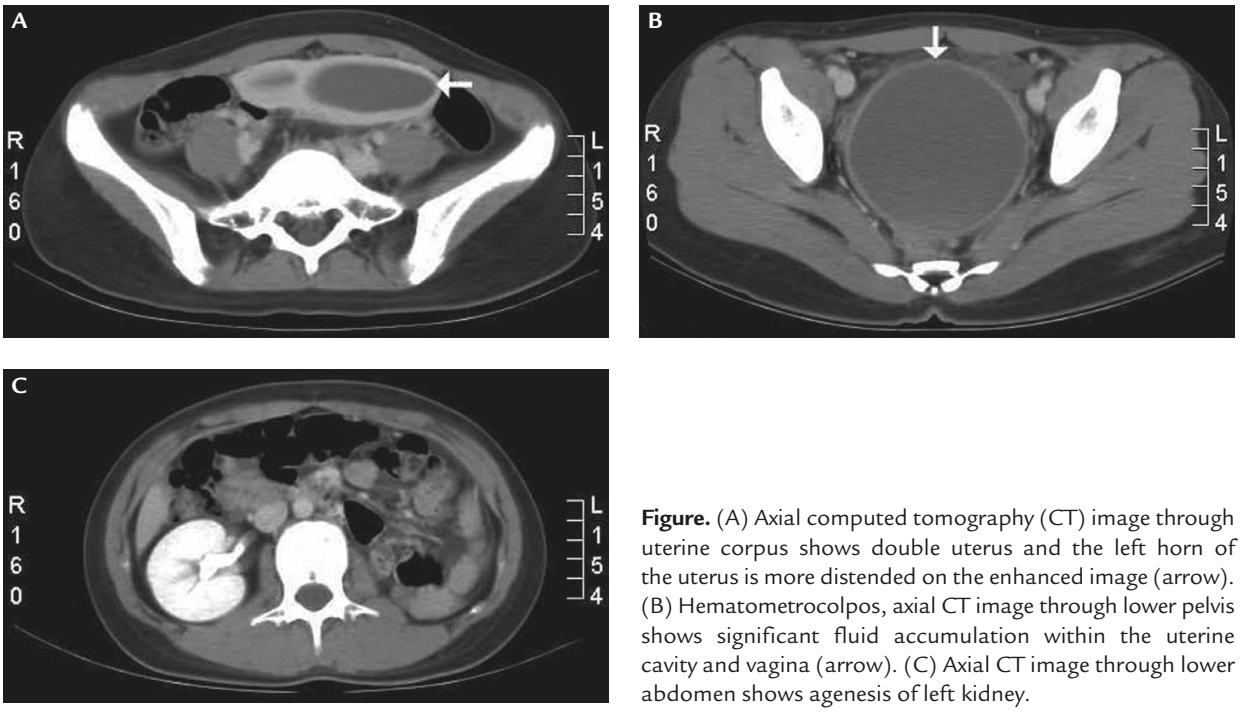

Figure. (A) Axial computed tomography (CT) image through uterine corpus shows double uterus and the left horn of the uterus is more distended on the enhanced image (arrow). (B) Hematometrocolpos, axial CT image through lower pelvis shows significant fluid accumulation within the uterine cavity and vagina (arrow). (C) Axial CT image through lower abdomen shows agenesis of left kidney.

The prevalence of müllerian duct abnormalities detected at ultrasound was thought to be about 3.87 per 1,000 women or approximately 1 in 250 women [1]. The true incidence of obstructive müllerian anomalies is unknown, but it is believed to be between 0.1% and 3.8% [2]. Didelphic uterus with longitudinal vaginal septum and unilateral imperforated hemivagina account for about 15–30% of these anomalies [3,4]. Acute abdomen caused by this anomaly is uncommon, and it is an embryologic malformation that affects the müllerian and metanephric ducts. These patients are usually asymptomatic and often have regular menstruation. These anomalies obstruct the outflow of menstruation and result in hematocolpos, hematometra and even hematosalpinx if an accurate diagnosis is not made in time. We describe herein an unusual case of acute abdominal pain.

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Table. Reported cases of didelphic uterus with unilateral obstructed double vagina

Authors	Total cases	Age (yr)	Symptoms	Associated renal anomalies	Management
Skondras et al [5]	3	12–13	Abdominal pain, fever, dysmenorrhea, pelvic mass	Ipsilateral renal agenesis	Excision of vaginal septum and laparotomy
Brosera et al [6]	2	N/A	Acute urine retention, difficulty in micturition and abdominal pain	Ipsilateral renal agenesis	N/A
Stassart et al [7]	15	6–26	Abdominal pain, dysmenorrhea, pelvic mass, vaginal discharge, abnormal bleeding, acute urine retention and dysuria	Ipsilateral renal agenesis	Excision of vaginal septum (11/15), laparotomy with hemihysterectomy and/or salpingo-oophorectomy (8/15), laparoscopy (4), vaginal aspiration (1/15)
Shibata et al [8]	1	17	Macroscopic hematuria synchronous with menstruation, low grade fever, lower abdominal pain and purulent vaginal discharge, vaginal urinary discharge after incision vaginal septum	Ipsilateral renal agenesis, ipsilateral ectopic ureter communicating with the ipsilateral uterine cervix	Excision of vaginal wall, resection the ectopic ureter and communicating duct
Li et al [9]	24	13 (mean)	N/A	Ipsilateral renal agenesis (22/24) (2 ectopic ureters to Gartner's duct cysts), renal hypoplasia or dysplasia (2/24)	N/A
Hoeffel et al [10]	1	14	Abdominal pain, dysmenorrhea, pelvic mass	Ipsilateral renal agenesis	Excision of vaginal septum and laparoscopy
Candiani et al [3]	36	9–34	Dysmenorrhea, paravaginal mass, lower abdominal pain, vaginal discharge and pyocolpos	Ipsilateral renal agenesis	Excision of vaginal septum (30/36), laparoscopy (24/36), laparotomy with hemihysterectomy or hemicolectomy (6/36)
Tanaka et al [11]	7	11–30	Lower abdominal pain, paravaginal mass, vaginal discharge, dysmenorrhea, recurrent spontaneous abortion	Ipsilateral renal agenesis (6/7)	Excision of vaginal septum
Tsai et al [12]	1	12	Dysmenorrhea, pelvic mass	Ipsilateral renal agenesis	Resectoscopic resection of vaginal septum
Altintas [13]	8	N/A	Pelvic pain, dysmenorrhea, pelvic mass	Ipsilateral renal agenesis	Excision of vaginal septum
Haddad et al [14]	33	11–30	Dysmenorrhea, paravaginal mass, hematic vaginal discharge, abdominal pain, dyspareunia	Ipsilateral renal agenesis (32/33)	Excision of vaginal septum (27/33), hemihysterectomy and ipsilateral hemicolpectomy (5/33)

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Table. *(continued)*

Authors	Total cases	Age (yr)	Symptoms	Associated renal anomalies	Management
Cicinelli et al [15]	1	13	Dysmenorrhea	Ipsilateral renal agenesis	Resectoscopic resection of vaginal septum under ultrasound guidance
Frei et al [16]	1	26	Excessive chronic vaginal discharge	Ipsilateral renal agenesis	Excision of vaginal septum and laparoscopy
Lurie et al [17]	1	14	Dysmenorrhea, progressive right lower quadrant pain	Ipsilateral renal agenesis	Excision of vaginal septum
Heinonen et al [4]	9	21 (mean)	Dysmenorrhea, paravaginal mass, lower abdominal pain	Ipsilateral renal agenesis (8/9), renal malrotation (1/9)	Excision of vaginal septum, salpingectomy (1/9)
Phupong et al [18]	2	N/A	Abdominal pain and pelvic mass, chronic foul smelling vaginal discharge	Ipsilateral renal agenesis	Hemihysterectomy (1/2), excision of vaginal septum (1/2)
Pieroni et al [19]	1	13	Rectal pain, constipation	Ipsilateral renal agenesis	Incision of vaginal septum
Joki-Erkkila et al [20]	10	18.6 (mean)	Lower abdominal pain, dysfunction uterine bleeding, lower abdominal mass	Ipsilateral renal agenesis (6/10), renal hypoplasia (3/10), renal malrotation (1/10)	Excision of vaginal septum, laparotomy or laparoscopy (7/10)
Zurawin et al [21]	8	11–22	Cystic pelvic pain, pelvic mass	Ipsilateral renal agenesis (8/8), women also have contralateral renal anomaly (3/8)	Excision of vaginal septum, laparotomy (3/8), laparoscopy (2/8)
Gholoum et al [22]	12	11–15	Abdominal pain, menstrual irregularity, abdominal-pelvic mass, intra-abdominal abscess, dysmenorrhea	Ipsilateral renal agenesis	Excision of vaginal septum, laparotomy (1/12)
Patterson et al [23]	1	10	Chronic constipation, progressive rectal pain	Ipsilateral renal agenesis	Combine laparoscopy and vaginoscopy for neo-os creation
Smith et al [24]	27	10–29	Pain, irregular bleeding, fever	Ipsilateral renal agenesis (20/27), ipsilateral renal anomaly (3/27) (duplicated ureter, dysplastic or polycystic kidneys), contralateral duplex collecting system (1/27)	Excision of vaginal septum and marsupialization of vaginal cuff (26/27), laparoscopy (7/27), second vaginoplasty (7/27)
Present case	1	16	Acute abdominal pain, dysmenorrhea	Ipsilateral renal agenesis	Excision of vaginal septum

N/A = not available.

A 16-year-old girl presented to our emergency department with the complaint of severe lower abdominal pain. Abdominal pain was noted since 2 days previously and progressively worsened on the day of admission, on the third day of her recent menstrual cycle. She stated that her menstruation was regular before, and only mild dysmenorrhea had happened. Symptoms exacerbated in recent menstrual periods. She denied any sexual activity or underlying medical disease.

She went to a local clinic for help initially, and a huge adnexal cyst was found on ultrasound. Then, she was referred to our emergency department for further management. On physical examination, her abdomen was soft but tender. Transabdominal ultrasound demonstrated an echogenic mass arising from the left adnexal area and absence of the left kidney. Computed tomography revealed uterus didelphys and significant fluid accumulation within the left uterine cavity and vagina. The right horn of the uterus was less distended on the enhanced image (Figure). Agenesis of the left kidney was also noted. Uterus didelphys with left hematometrocolpos and ipsilateral renal agenesis was highly suspected. Intravenous urography showed congenital agenesis of the left kidney and normal nephrogram and pyelogram of the right kidney. This had allowed us to exclude the diagnosis of ectopic kidney.

Then, emergent operation was performed. Pelvic examination under anesthesia showed double vagina with left imperforated vagina. She underwent excision of the obstructed vaginal septum and drainage of 200 mL of old blood. The postoperative course was smooth, and she was discharged 2 days after the operation.

Acute abdomen caused by didelphic uterus and vagina with unilateral imperforated vagina and ipsilateral renal agenesis is uncommon. To date, at least 205 cases of didelphic uterus with unilateral obstructed double vagina have been reported since 1991 (Table) [3–24]. Most of the patients were diagnosed at a young age. The most common clinical findings were dysmenorrhea, abdominal pain, a pelvic or paravaginal mass, and vaginal discharge. Other reported symptoms included acute urinary retention, difficulty in micturition, hematuria, dyspareunia, irregular vaginal bleeding, rectal pain, and chronic constipation. All except five cases had ipsilateral renal agenesis or other urinary tract anomalies such as ipsilateral renal hypoplasia, malrotation, and contralateral renal anomalies. Excision of vagina septum was the most common surgical intervention. Laparotomy or laparoscopy was performed in some cases for the purpose of treatment or confirmatory diagnosis. Hysteroscopic resection of vaginal septum or combined laparoscopy and vaginoscopy for neo-os creation have also been reported. According to previous reports, an accurate diagnosis is not always easy because of various reasons [11,17,21,22]. First, the patients may have normal regular menstruation from a unilateral normal uterus via an unobstructed vagina. Second, they may take anti-inflammatory drugs and oral contraceptives for relieving their abdominal pain or cyclic dysmenorrhea. Third, it is not often thought of as a diagnostic possibility because of the uncommon condition. Finally, it is also difficult to distinguish hematometrocolpos from pelvic or adnexal mass in patients with acute abdomen, especially when the adnexal mass is associated with torsion, rupture or infection.

Ultrasound is a simple method for primary diagnosis. It can help to identify the anatomy of uterus and the associated urogenital anomalies. Magnetic resonance imaging can help to define the anatomy and classification of the müllerian duct anomalies [25–27]. These patients may have pelvic endometriosis due to retrograde menstruation [28]. Endometriosis has been found in 16–35% of women with a didelphic uterus, especially in obstructive forms [4,29,30]. If the diagnosis is delayed, infection and subsequent obstetric complications may occur.

In conclusion, müllerian duct anomalies, such as didelphic uterus with unilateral imperforated vagina, should be included in the differential diagnosis in female adolescents with acute abdominal pain. Early diagnosis and appropriate therapy can relieve symptoms, preserve fertility, and decrease complications in such patients.

References

1. Byrne J, Nussbaum-Blask A, Taylor WS, Rubin A, Hill M, O'Donnell R, Shulman S. Prevalence of müllerian duct anomalies detected at ultrasound. *Am J Med Genet* 2000; 94:9–12.
2. Burgis J. Obstructive müllerian anomalies: case report, diagnosis, and management. *Am J Obstet Gynecol* 2001;185: 338–44.
3. Candiani GB, Fedele L, Candiani M. Double uterus, blind hemivagina, and ipsilateral renal agenesis: 36 cases and long-term follow-up. *Obstet Gynecol* 1997;90:26–32.
4. Heinonen PK. Clinical implications of the didelphic uterus: long-term follow-up of 49 cases. *Eur J Obstet Gynecol Reprod Biol* 2000;91:183–90.
5. Skondras KG, Moutsouris CC, Vaos GC, Barouchas GC, Demetriou LD. Uterus didelphys with an obstructed hemivagina and ipsilateral renal agenesis: a rare cause of acute abdomen in pubertal girls. *J Pediatr Surg* 1991;26:1200–1.
6. Brosera E, Boronat F, Ruiz JL, Alonso M, Osca JM, Jiménez-Cruz JF. Urological complications associated to uterus didelphys with unilateral hematocolpos. A case report and review of the literature. *Eur Urol* 1991;20:85–8.

7. Stassart JP, Nagel TC, Prem KA, Phipps WR. Uterus didelphys, obstructed hemivagina, and ipsilateral renal agenesis: the University of Minnesota experience. *Fertil Steril* 1992; 57:756–61.
8. Shibata T, Nonomura K, Kakizaki H, Murayama M, Seki T, Koyanagi T. A case of unique communication between blind-ending ectopic ureter and ipsilateral hemi-hematocolpometra in uterus didelphys. *J Urol* 1995;153:1208–10.
9. Li YW, Sheih CP, Chen WJ. Unilateral occlusion of duplicated uterus with ipsilateral renal anomaly in young girls: a study with MRI. *Pediatr Radiol* 1995;25(Suppl 1):S54–9.
10. Hoeffel C, Olivier M, Scheffler C, Chelle C, Hoeffel JC. Uterus didelphys, obstructed hemivagina and ipsilateral renal agenesis. *Eur J Radiol* 1997;25:246–8.
11. Tanaka YO, Kurosaki Y, Kobayashi T, et al. Uterus didelphys associated with obstructed hemivagina and ipsilateral renal agenesis: MR findings in seven cases. *Abdom Imaging* 1998; 23:437–41.
12. Tsai EM, Chiang PH, Hsu SC, Su JH, Lee JN. Hysteroscopic resection of vaginal septum in an adolescent virgin with obstructed hemivagina. *Hum Reprod* 1998;13:1500–1.
13. Altintas A. Uterus didelphys with unilateral imperforate hemivagina and ipsilateral renal agenesis. *J Pediatr Adolesc Gynecol* 1998;11:25–7.
14. Haddad B, Barranger E, Paniel BJ. Blind hemivagina: long-term follow-up and reproductive performance in 42 cases. *Hum Reprod* 1999;14:1962–64.
15. Cicinelli E, Romano F, Didonna T, Schonauer LM, Galantino P, Di Naro E. Resectoscopic treatment of uterus didelphys with unilateral imperforate vagina complicated by hematocolpos and hematometra: case report. *Fertil Steril* 1999;72:553–5.
16. Frei KA, Bonel HM, Haberlin FC. Uterus didelphys, obstructed hemivagina and ipsilateral renal agenesis with excessive chronic vaginal discharge. *Acta Obstet Gynecol Scand* 1999;78:460–1.
17. Lurie S, Feinstein M, Mamet Y. Unusual presentation of acute abdomen in a syndrome of double uterus, unilaterally imperforated double vagina, and ipsilateral renal agenesis. *Acta Obstet Gynecol Scand* 2000;79:152–3.
18. Phupong V, Pruksananonda K, Taneepanichskul S, Tresukosol D, Virutamasen P. Double uterus with unilaterally obstructed hemivagina and ipsilateral renal agenesis: a variety presentation and a 10-year review of the literature. *J Med Assoc Thai* 2000;83:569–74.
19. Pieroni C, Rosenfeld DL, Mokrzycki ML. Uterus didelphys with obstructed hemivagina and ipsilateral renal agenesis. A case report. *J Reprod Med* 2001;46:133–6.
20. Joki-Erkkila MM, Heinonen PK. Presenting and long-term clinical implications and fecundity in females with obstructing vaginal malformations. *J Pediatr Adolesc Gynecol* 2003;16: 307–12.
21. Zurawin RK, Dietrich JE, Heard MJ, Edwards CL. Didelphic uterus and obstructed hemivagina with renal agenesis: case report and review of the literature. *J Pediatr Adolesc Gynecol* 2004;17:137–41.
22. Gholoum S, Puligandla PS, Hui T, Su W, Quiros E, Laberge JM. Management and outcome of patients with combined vaginal septum, bifid uterus, and ipsilateral renal agenesis. *J Pediatr Surg* 2006;41:987–92.
23. Patterson D, Mueller C, Strubel N, Rivera R, Ginsburg HB, Nadler EP. Laparoscopic neo-os creation in an adolescent with uterus didelphys and obstructed hemivagina. *J Pediatr Surg* 2006;41:E19–E22.
24. Smith NA, Laufer MR. Obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) syndrome: management and follow-up. *Fertil Steril* 2007;87:918–22.
25. Ballesio L, Andreoli C, De Cicco ML, Angeli ML, Manganaro L. Hematocolpos in double vagina associated with uterus didelphus: US and MR findings. *Eur J Radiol* 2003;45:150–3.
26. Fischetti SG, Politi G, Lomeo E, Garozzo G. Magnetic resonance imaging of müllerian duct anomalies. *Radiol Med (Torino)* 1995;89:105–11. [In Italian]
27. Troiano RN, McCarthy SM. Müllerian duct anomalies: imaging and clinical issues. *Radiology* 2004;233:19–34.
28. Pfeifer SM, Gosman GG. Evaluation of adnexal masses in adolescents. *Pediatr Clin North Am* 1999;46:573–92.
29. Fedele L, Bianchi S, Di Nola G, Franchi D, Candiani GB. Endometriosis and nonobstructive Müllierian anomalies. *Obstet Gynecol* 1992;79:515–7.
30. Ugur M, Turan C, Mungan T, Kuscu E, Senoz S, Agis HT, Gokmen O. Endometriosis in association with Müllerian anomalies. *Gynecol Obstet Invest* 1995;40:261–4.