Double Uterus with Unilaterally Obstructed Hemivagina and Ipsilateral Renal Agenesis : A Variety Presentation and a 10-Year Review of the Literature

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Abstract

A double uterus with a unilaterally obstructed hemivagina is a rare condition, usually associated with ipsilateral renal agenesis. Herein, we report two cases, the first case presenting with abdominal pain and pelvic mass. Hemihysterectomy was performed leaving the contralateral uterus intact. The second case presented with chronic foul smelling vaginal discharge. The diagnosis was a double uterus and pyocolpos of the left vagina. Excision of the left vaginal septum and drainage were performed. The postoperative course of both cases was uneventful and the patients were well at the six-week follow-up. An accurate diagnosis, appropriate management and the prevention of future fertility problems are discussed.

Key word : Double Uterus, Hemivagina, Renal Agenesis, Pyocolpos

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The unique clinical syndrome consisting of a double uterus, obstruction of the vagina, and ipsilateral renal aplasia is a rare condition[1-6]. The clinical presentation varies greatly depending on whether or not the unilateral hemivagina is completely obstructed and on the location of its opening[2]. The most common clinical presentation is dysmenorrhea in association with a pelvic mass resulting from a hemiobstructed vagina. Other presentations are pelvic pain, hypermenorrhea, menometrorrhagia, intermittent vaginal spotting, malodorous vaginal discharge and urinary symptoms[2-6].

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Since the clinical presentations vary, arriving at a diagnosis is difficult[1]. Early and accurate diagnosis and management will prevent future fertility problems.

When a postmenarcheal young girl is found with a pelvic mass, an ovarian germ cell tumor should first be listed as the differential diagnosis. Despite their rare occurrence, Mullerian duct anomalies should be enlisted as well. One of the easiest clues is to demonstrate ipsilateral renal agenesis commonly associated with urinary and genital tract development.

In this study, we report two pubertal patients, one who presented with lower abdominal pain and pelvic mass with normal menstruation. The preoperative diagnosis was ovarian tumor. The second one presented with chronic foul smelling vaginal discharge. We discuss the pitfall of diagnosis and management in those cases.

CASE REPORT

Case 1

A 13-year-old girl was admitted to King Chulalongkorn Memorial Hospital because of right lower quadrant abdominal pain one month prior to admission. Her first menstruation had been 24 months previously and had been regular since then. Her last menstrual period was April, 17, 1999. She had never had complaints of dysmenorrhea. Her past medical record and family history was unremarkable.

Physical examination revealed a cystic mass, sized 8x10 cm, extending from the pubic symphysis to the right paraumbilicus with mild tenderness on pressure. Pelvic examination disclosed an intact hymen and normal external genitalia. On rectal examination, mild tenderness close to the small sized uterus and the cystic pelvic mass located on the right anterior wall of the uterus was palpated. Preoperative intravenous pyelography (IVP) and transabdominal ultrasonography demonstrated absence of the right kidney and a dumbell like mass with mixed echogenicity in the pelvic cavity. Preoperative diagnosis was right ovarian tumor.

At laparotomy, we found a double uterus with the left side normal in size and the right uterus enlarged to about 6x10x5 cm^3 (Fig. 1). Both uteri had only one adnexa each. There was a moderate degree of red spots of endometriosis-like appearance on the right fallopian tube and ovarian surface. Perituboovarian adhesion of the right adnexa was also observed. Aspiration of the fluid content from the right uterus was performed. A dark brown-coloured fluid, which looked like old menstrual blood, was thought to be hematometra. A vertical incision of the affected uterus was performed and a blind pouch vagina disclosed. A right hemihysterectomy was performed leaving the vaginal pouch open. The postoperative course was uneventful and the patient left the hospital on the 5th postoperative day. She had recovered well at the six-week follow-up.

Case 2

A 12-year-old girl was admitted because of chronic foul smelling vaginal discharge for two months prior to admission. Her first menstruation occurred three months previously and had been regular since then. Her last menstrual period was July 13, 1999 with the complaints of dysmenorrhea. Her past medical record and family history was also unremarkable.

Physical examination showed well nourished and normally developed secondary sex characteristics. Pelvic examination under general anesthesia, revealed an intact hymen and normal external genitalia. Speculum examination revealed one cervix and one vagina. There was a cystic mass

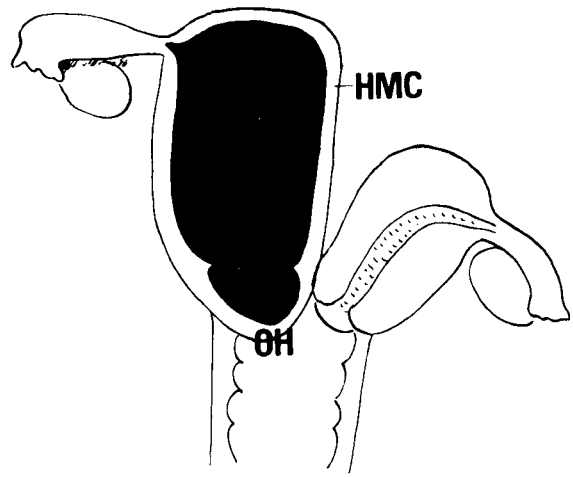

Fig. 1. Illustration of double uterus with right hematometrocolpos (HMC) and obstructed hemivagina (OH).

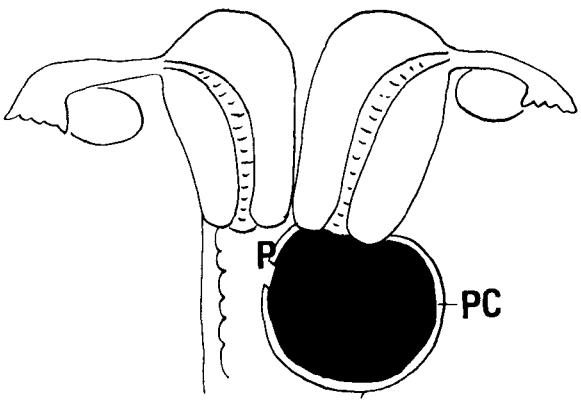

Fig. 2. Illustration of double uterus with left pyocolpos (PC) and pore at the right side (P).

of 6 cm in diameter at the left vaginal wall. A foul smelling purulent discharge was drained through a small hole at the right side of the cyst wall (Fig. 2). There were two separated small uterus with no palpable adnexal mass. Preoperative intravenous pyelography (IVP) demonstrated contrast media on the right but absence on the left kidney. Transabdominal ultrasonography confirmed a double uterus with a large hematocolpos. We decided to excise the vaginal septum of the left vagina wall and drain the discharge. After the drainage was completed, a normal left cervix could be visualized. The postoperative course was uneventful and the patient was discharged on the 7th postoperative day. She had recovered well at the six-week follow-up.

DISCUSSION

A double uterus with an obstructed hemivagina and ipsilateral renal agenesis is a relatively rare clinical presentation[1-6]. Renal agenesis on the side of the obstructed vagina associated with a double uterus and cervix is suggestive of an embryologic arrest occurring during the 8th week of gestation simultaneously affecting the Mullerian and metanephric ducts[1]. The exact cause of this developmental defect is unknown[1]. If one of the Wolffian ducts is absent, the kidney and ureter on the same side will fail to develop. The Mullerian ducts may also fail to fuse at midline, either completely or incompletely. If the failure to fuse is complete, a uterus didelphys is formed. The Mullerian duct on the side lacking the Wolffian duct, displaces itself laterally and cannot come into contact with the urogenital sinus in the center. Thus, the contralateral Mullerian duct gives way to the vagina, whereas, the already displaced component on the other side forms a blind sac, i.e, an imperforate or obstructed vagina. The distal part of the vagina, originating from the urogenital sinus, being distal from the hymenal ring, is not affected[4]. The more common defect was on the right side[3,5-8], but we found both types of defect with our two cases.

Pelvic pain and worsening dysmenorrhea since menarche associated with the finding of a pelvic mass is the common presentation[2,4-6,9,10]. Most of the cases have a normal and regular menstruation period[2,4,11]. Occasionally, patients may present with symptoms of urinary tract infection[7]. Acute urinary retention[12] or chronic vaginal discharge[11] as the initial symptom has also been reported. Rarely, necrosis and rupture of the hematosalpinx resulting in typical peritonitis has been reported[10]. As for our cases, in contrast with previous reports[2,4-6,9,10], in which the first presented with right lower quadrant abdominal pain and the subsequent finding of a right cystic pelvic mass. The first case had normal menstruation and no dysmenorrhea since menarche. The second case presented with chronic foul smelling vaginal discharge and dysmenorrhea which is similar to the case report by Frei et al[11].

The diagnostic tool most valuable to confirm the diagnosis of this syndrome is ultrasonography[2,6,13]. It is both noninvasive and useful in the evaluation of renal agenesis[2]. There have been reports about magnetic resonance imaging (MRI) also being useful for diagnosing this syndrome[14]. Another investigative method is IVP which not only confirms the absence of a normal kidney on the affected side but also detects abnormalities of both the contralateral kidney and the ureter[5]. In our first case, both transabdominal ultrasonography and IVP were not helpful but rather indicated ovarian neoplasm. But both methods proved helpful to diagnose the second case.

We analyzed the literature using MEDLINE between the years 1989 to 1999 and found 113 cases reported. We analyzed and compared them with the 2 cases in the present study. They are summarized in Table 1. Their age range was between 9 and 37 years. The most common symptom at presentation was dysmenorrhea (84/115, 73%), and

Table 1. Review 11 literature and analysis of 115 reported cases.

Authors (year)	N	Age range (Years)	Most common presentation (N)	Side of vaginal obstruction		Management			
				Right	Left	Vaginal septum resection	Vaginal aspiration	Hemihysterectomy	Total hysterectomy
1. Haddad (1999)(9)	42	11-30	Dysmenorrhea (35) Paravaginal mass (35)	22	20	37	-	5	-
2. Frei (1999)(11)	1	26	Chronic vaginal discharge (1)	1	-	1	-	-	-
4. Tanaka (1998)(14)	7	11-30	Abdominal pain(4) Paravaginal mass(2)	3	4	7	-	-	-
5. Candiani (1997)(6)	36	9-36	Dysmenorrhea (29) Paravaginal mass(27)	26	10	30	-	4	2
6. Shibata (1995)(15)	1	17	Urinary incontinence(1)	1	-	1	-	-	-
7. Stassart (1992)(5)	15	6-26	Pelvic pain , dysmenorrhea and vaginal or pelvic mass (10)	11	4	9	1	5	-
8. Erdogan (1992)(4)	1	15	Pelvic mass , abdominal pain and dysmenorrhea(1)	1	-	1	-	-	-
9. Skondras (1991)(10)	3	12-13	Acute abdominal pain and dysmenorrhea (3)	2	1	3*	-	-	-
10. Lin (1991)(2)	5	18-37	Dysmenorrhea and pelvic mass(5)	3	2	5	-	-	-
11. Altintas (1991)(16)	1	16	Abdominal pain, dysmenorrhea and pelvic mass (1)	1	-	1	-	-	-
12. Burgos (1989)(13)	1	14	Abdominal pain and pelvic mass(1)	1	-	1	-	-	-
13. Present study (1999)	2	12-13	Abdominal pain and pelvic mass(1) Chronic vaginal discharge(1)	1	1	1	-	1	-
Total	115	9-37	Dysmenorrhea(84,73%) Pelvic / paravaginal mass(82,71%)	7 3 (63.5%)	42 (36.5%)	97	1	15	2

* and unilateral salpingo-oophorectomy

pelvic/paravaginal mass (82/115, 71%). The right uterus and vagina were commonly affected (73/115, 63.5%). Almost all cases (97/115, 84.3%) were managed by resection of the vaginal septum.

The management of choice by most authors is excision of the vaginal septum[2,4-6, 9-11,13-16]. However, hemihysterectomy is recommended in patients with a high, thick-walled obstruction, massive ovarian involvement, endometriosis and adenomyosis[10]. Pyocolpos due to closure of the septal incision and dysmenorrhea due to cervical stenosis have been reported[10]. Hence, careful follow-up of conservatively managed cases is essential.

We managed our first case in contrast with other recommendations[2,4-6,9-11,13-16] with hemihysterectomy and left the blind pouch of the vagina open. The reasons for this management were the thick-walled obstruction, as well as the educational and economic background of this patient in which she might be lost to follow-up when managed conservatively. The advantages of this management are: a decrease in longterm complications after conservative management such as pyocolpos due to closure of the septal incision, dysmenorrhea due to cervical stenosis and recurrent hematometrocolpos. But the second case, we managed similarly to other recommendations[2,4-6, 9-11,13-16] by excising the vaginal septum and draining the discharge. There have been few reports about endometrial carcinoma occurring in cases of a double uterus[17,18]. In these situations correct diagnosis may be difficult due to anatomic difficulties or failure to recognize a second uterine cavity. If we leave both uteri intact without correction, we must be concerned about the risk of endometrial carcinoma in the future.

In conclusion, when ipsilateral renal agenesis is found in an adolescent girl presenting with abdominal pain and a pelvic mass, or with chronic vaginal discharge, the syndrome of double uterus, obstructed hemivagina and ipsilateral renal agenesis should be considered. These clinical findings should allow clinicians to make an early diagnosis, appropriate management and reduced morbidity.

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REFERENCES

1. Rock JA. Congenital out flow tract obstruction. In: Adashi EY, Rock JA, Rosenwaks Z, editors. Reproductive endocrinology, surgery, and technology. New York : Lippincott-Raven, 1996, 1445-74.
2. Lin CC, Chen AC, Chen TY. Double uterus with an obstructed hemivagina and ipsilateral renal agenesis : report of 5 cases and a review of the literature. J Formos Med Assoc 1991 ; 90 : 195-201.
3. Rock JA, Jones HW. The double uterus associated with an obstructed hemivagina and ipsilateral renal agenesis. Am J Obstet Gynecol 1980 ; 138 : 339-42.
4. Erdogan E, Okan G, Daragenli O. Uterus didelphys with unilateral obstructed hemivagina and renal agenesis on the same side. Acta Obstet Gynecol Scand 1992 ; 71 : 76-7.
5. Stassart JP, Nagel TG, Prem KA, Phipps WR. Uterus didelphys, obstructed hemivagina, and ipsilateral renal agenesis : the Universtiy of Minnesota experience. Fertil Steril 1992 ; 57 : 756-61.
6. 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.
7. Allan N, Cowan LE. Uterus didelphys with unilateral imperforate vagina. Obstet Gynecol 1963 ; 22 : 422-6.
8. Intaraprasert S, Benchakan V. Mullerian duct anomalies and unilateral renal agenesis : report of 21 cases from Ramathibodi Hospital. J Med Assoc Thai 1985 ; 68 : 183-9.
9. Haddad B, Barranger E, Paniel BJ. Blind hemivagina : long term follow-up and reproductive performance in 42 cases. Hum Reprod 1999 ; 14 : 1962-4.
10. Skondras KG, Moutsouris CC, Vos GC, Barouchas GC, Demetriou LD. Uterus didelphys with obstructed hemivagina and ipsilateral renal agenesis : a rare cause of acute abdomen in pubertal girls. J Pediatr Surg 1991 ; 26 : 1200-1.
11. 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.
12. Adams WG, Wilson EC, Holloway HJ. Uterus

didelphys with unilateral imperforate vagina : a rare cause of acute urinary retention. J Urol 1979 ; 121 : 131-2.
13. Burgos FJ, Matorras R, Rivera M, Sanchez E, Mayayo T. Double uterus associated with unilateral vaginal obstruction and ipsilateral renal agenesis : ultrasonographic diagnosis. J Clin Ultrasound 1989 ; 17 : 296-8.
14. 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.
15. Shibata T, Nanomura K, Kakizaki H, Murayama M, Seki T, Koyanaki T. A case of unique communication between blind-ending ectopic ureter and ipsilateral hemi-hematocolpometra in uterus didelphys. J Urol 1995 ; 153 : 1208-10.
16. Altintas A, Aridogan N, Atay Y, Koker I. Uterus didelphys with unilateral imperforate vagina and ipsilateral renal agenesis. Aust N Z J Obstet Gynecol 1991 ; 31 : 288-9.
17. Holub Z, Shomani A. Uterine reduplication, unilateral ureteral and renal aplasia syndrome associated with endometrial cancer : a case report. Eur J Gynecol Oncol 1998 ; 19 : 575-6.
18. Kosinski A, Dini M. Endometrial cancer in a double uterus : a report of two cases. J Reprod Med 1994 ; 39 : 926-7.

ภาวะมดลูกสองอันร่วมกับช่องคลอดปิดกั้นข้างเดียวและภาวะไม่มีไตข้างเดียวกัน: ความหลากหลายของอาการนำ และการรวบรวมรายงาน 10 ปี

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ภาวะมดลูกสองอันร่วมกับช่องคลอดปิดกั้นข้างเดียวกันที่พบร่วมกับภาวะไม่มีไตข้างเดียวกัน เป็นภาวะที่พบได้น้อย คณะผู้รายงานได้รายงานผู้ป่วย 2 ราย ซึ่งรายแรกมาด้วยอาการปวดท้องน้อยและตรวจพบก้อนในอุ้งเชิงกรานตรวจภายในและคลื่นเสียงความถี่สูงพบมีก้อนที่รังไข่ข้างขวา ตรวจเอ็กซเรย์ไตและหลอดไตด้วยสารทึบแสง ไม่พบไตและหลอดไตข้างขวา เมื่อทำการผ่าตัดพบว่าก้อนที่โตเป็นมดลูกที่มีเลือดระดูอยู่ภายในมดลูกด้านขวาโดยแยกกับมดลูกด้านซ้ายอย่างชัดเจน ปีกมดลูกด้านขวามีพังผืดอยู่รอบรังไข่และหลอดมดลูก ได้ตรวจพบว่ามีผนังช่องคลอดกั้นทางติดต่อของปากมดลูก ได้ทำผ่าตัดมดลูกข้างขวาร่วมกับช่องคลอดออกเหลือบางส่วนของช่องคลอดเปิดไว้ ส่วนรายที่สองมาด้วยอาการมีตกขาวกลิ่นเหม็นออกทางช่องคลอดมา 2 เดือน ตรวจภายในและคลื่นเสียงความถี่สูงพบมีมดลูก 2 อัน ปากมดลูก 2 ข้าง ช่องคลอดข้างขวาปกติ ช่องคลอดด้านซ้ายอุดตันเป็นถุงหนอง ตรวจเอ็กซเรย์ไตและหลอดไตด้วยสารทึบแสง ไม่พบไตและหลอดไตข้างซ้าย ได้ทำผ่าตัดผนังช่องคลอดที่อุดตันและระบายหนองออก และให้ยาปฏิชีวนะทางหลอดเลือดดำต่อหลังผ่าตัด ภาวะหลังผ่าตัดและการตรวจติดตามเมื่อ 6 สัปดาห์พบว่าผู้ป่วยทั้งคู่ปกติดี คณะผู้รายงานได้วิจารณ์การวินิจฉัยและการรักษาในผู้ป่วยสองรายนี้ การวินิจฉัยที่ถูกต้องและการรักษาที่เหมาะสมจะช่วยลดอาการและป้องกันผลกระทบต่อปัญหาการเจริญพันธุ์ได้

คำสำคัญ : มดลูกสองอัน, ช่องคลอดปิดกั้น, ภาวะไม่มีไต, ถุงหนองของช่องคลอด

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