Uterus didelphys with blind hemivagina and ipsilateral renal agenesis complicated by pyocolpos and presenting as acute abdomen 11 years after menarche: presentation of a rare case with review of the literature

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Summary

Background: Congenital anomaly of the Müllerian duct system can result in various urogenital anomalies including uterus didelphys with blind hemivagina and ipsilateral renal agenesis. The diagnosis of this condition is usually made after menarche, but its rarity and variable clinical features may contribute to a diagnostic delay for years after menarche. *Case:* A 24-year-old woman presented to the emergency room of the Department of Obstetrics and Gynecology complaining of severe worsening lower abdominal pain, vomiting and pus-like vaginal discharge. Physical examination revealed acute abdomen with diffuse lower abdominal tenderness, rebound and muscular resistance. Cervical and vaginal observation was impossible because of the patient's discomfort. Bimanual gynecological examination showed high tenderness cervical motion. Transabdominal ultrasound scan was performed and the radiologist interpreted the ultrasonographic findings as existence of a pelvic mass with mixed echogenicity. The preoperative diagnosis was ruptured tubo-ovarian abscess and emergency laparotomy was performed. Free pus in the pelvis was found. Also, a double uterus with an elongated and inflammatory right fallopian tube with pus passing out of its fimbrial end was observed. Vaginal examination under general anesthesia revealed an obstructed right hemivagina with vaginal pus-like discharge from a small fistula hole on the septate vagina. The final diagnosis was uterus didelphys with unilateral imperforate right hemivagina and pyocolpos. Transvaginal resection of the vaginal septum was performed and a large amount of pus and blood was spilled out. Postoperatively, intravenous pyelography demonstrated agenesis of the right kidney. *Conclusion:* We demonstrated the difficulty in making a correct diagnosis of this rare congenital anomaly of the female genital tract, especially after many years since menarche. This condition should be considered to prevent misdiagnosis or suboptimal treatment and decrease morbidity and unnecessary surgical procedures.

Key words: Pyocolpos; Uterus didelphys; Genital malformation; Urogenital abnormalities; Renal agenesis; Obstructed hemivagina

Introduction

Uterine didelphys with blind hemivagina and ipsilateral renal agenesis is an extremely rare congenital malformation of the female genitourinary system, occurring between the 12th and 16th week of gestation [1, 2]. In neonates, the obstruction of the blind hemivagina blocks the outflow of the cervical mucinous discharge caused by maternal estrogens, resulting in hydrocolpos with or without hydrometra [3]. After menarche, the blockage of the menstrual cycle results in hematocolpos with or without hematometra and hemosalpinx. Reflux of the blood during menstruation into the peritoneal cavity might cause endometriosis [4]. The interval from menarche to the onset of symptom ranges from a few months to several years and the manifesting symptoms consist of dysmenorrhea, acute or chronic pelvic pain, fever, peritonitis and purulent vaginal discharge [4-6]. The most suitable management of this condition is excision of the obstructing vaginal septum as complete as possible [7].

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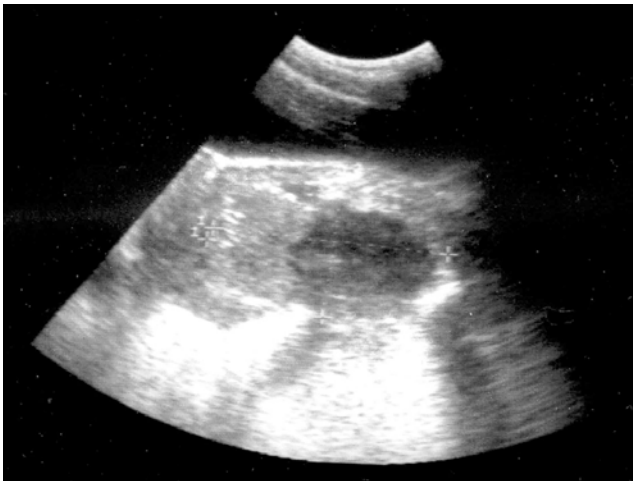
We present the case of a blind hemivagina associated with uterus didelphys and ipsilateral renal agenesis in a patient who presented with acute abdomen because of pyocolpos and reflux of pus into the peritoneal cavity 11 years after menarche.

Case Report

A 24-year-old, sexually active, nulligravid Caucasian female presented to the emergency room of the Department of Obstetrics and Gynecology, "Tzaneio" General State Hospital, Pireaus, complaining of severe worsening lower abdominal pain. In addition, the patient was suffering from nausea and vomiting. There were no associated bowel or bladder symptoms. Her past medical history was uneventful. She reported menarche at the age of 11 years. Menstruation had been regular and she was on day 16 of a 24-day menstrual cycle. She denied ever having had a cervical smear test, gynecological examination or abdominal surgery. She reported having desmenorrhea, dyspareunia and excessive chronic vaginal discharge. Physical examination revealed an acute abdomen with diffuse lower abdominal tenderness and rebound. Her abdomen showed muscular resistance. The patient was hemodynamically stable. Her

Fig. 1



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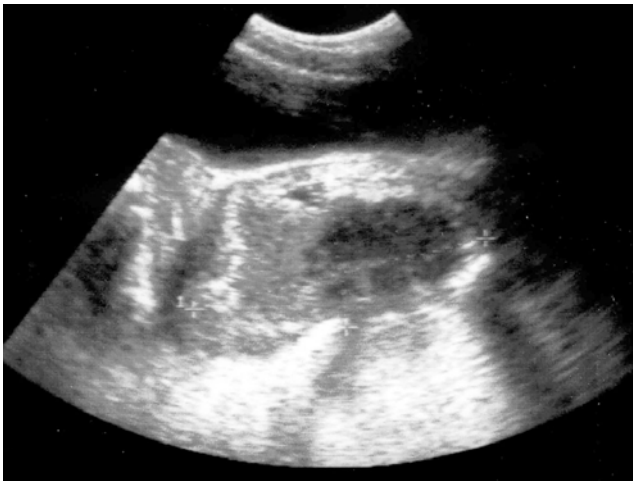
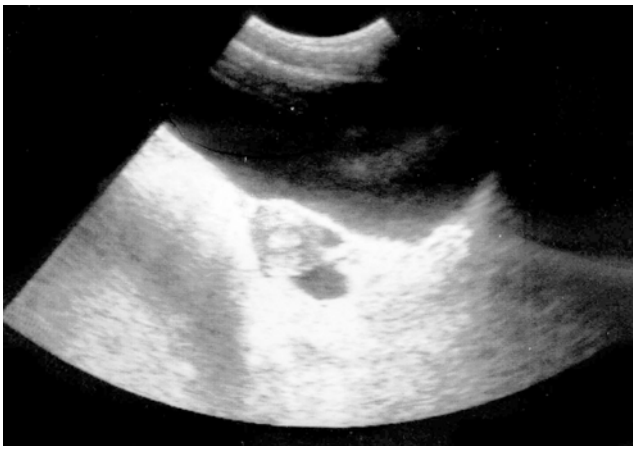


Fig. 3



Figures 1, 2, 3. — Transabdominal ultrasonography showing the uterus didelphys with the right pyocolpos. These ultrasonographic findings were incorrectly interpreted by the radiologist as the existence of a pelvic mass with mixed echogenicity measuring 11 x 5.4 cm.

temperature was 38.4°C. Inspection of the external genitalia revealed no unusual findings. A sufficient amount of pus-like vaginal discharge was noted. Observation of the cervix and vagina was impossible because of the discomfort of the patient. Bimanual gynecological examination showed high tenderness cervical motion. Estimation of the uterus and adnexa was difficult because of the muscular resistance of her abdominal wall. Hemoglobin concentration was 11.7 g/dl, hematocrit 34.6%, and white blood count 21,000 cells/ml with 94.2% polymorphonuclear leukocytes. Serum beta-hCG was negative. Renal and liver function tests were normal. Transabdominal ultrasound scan was performed and the ultrasonographic findings were interpreted by the radiologist as the existence of a pelvic mass measuring 11 x 5.40 cm with mixed echogenicity. Free pelvic fluid was also seen (Figures 1, 2, 3).

The preoperative diagnosis was ruptured tuboovarian abscess and an emergency laparotomy through a Phannestiel incision was performed. The main operative findings are summarized as follows: (i) free pus in the pelvis; (ii) grossly normal left uterus, left fallopian tube and left ovary; (iii) grossly normal right uterus, elongated and inflammatory right fallopian tube with pus passing out of its fimbrial end and normal right ovary; (iv) no obvious endometriosis; (v) no evidence of abscess formation; (vi) absence of adhesions (Figure 4). The abdominal cavity was copiously irrigated with sterilized normal fluid. The abdominal wound was closed in layers. A uterus didelphys with obstructed hemivagina was suspected. Vaginal examination under general anesthesia revealed a pus-like discharge from a fistula hole on the distended and obstructed right hemivagina. Also, using a needle and syringe, pus was aspirated from the right hemivagina (Figure 5). Drainage of the pyocolpos with transvaginal right lateral colpotomy was performed using conventional scalpel diathermy causing a large amount of pus and blood to spill out (Figures 6, 7). Almost complete resection of the vaginal septum was achieved; the edges of the septum were sutured with interrupted Vicryl sutures (Figure 8). The right cervix was clearly evidenced. Postoperatively, the patient did well and was afebrile 24 hours after surgery and treatment with antibiotics. Histopathologic examination of the removed septum demonstrated vaginal tissue and cervical epithelium on the open vaginal side. An ultrasound scan showed absence of the right kidney, while the left kidney was increased in size (longitudinal diameter 14.5 cm). Intravenous pyelography showed agenesis of the right kidney and hypertrophy of the left kidney (Figure 9). A diagnosis of double uterus with unilaterally unperforated right hemivagina (Figure 10) and ipsilateral renal agenesis was made. At postoperative examination six months after surgery, the patient was free of symptoms and had regular menses. An ultrasound scan showed no reaccumulation of fluid and a normal vagina, uterine bodies, cervices and adnexa. Additionally, the Papanicolaou test from each cervix was negative for cytologic abnormalities.

Discussion

The male and female genital systems are identical at six weeks of intrauterine development, formed by two pairs of symmetric genital ducts: the paramesonephric

Fig. 4

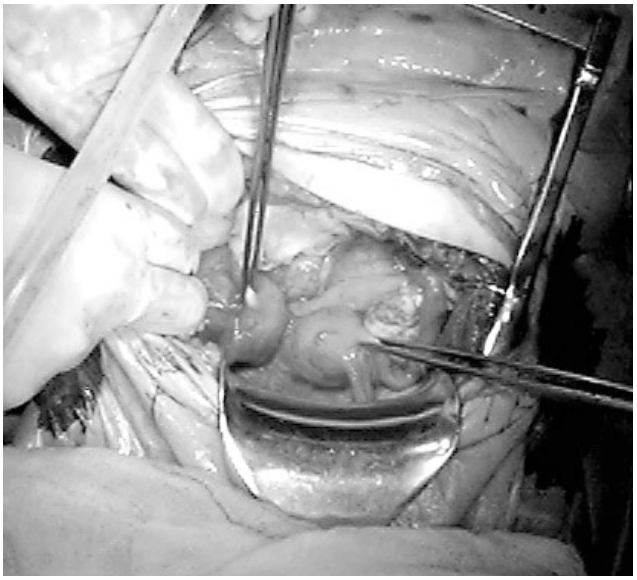


Fig. 5

Fig. 6

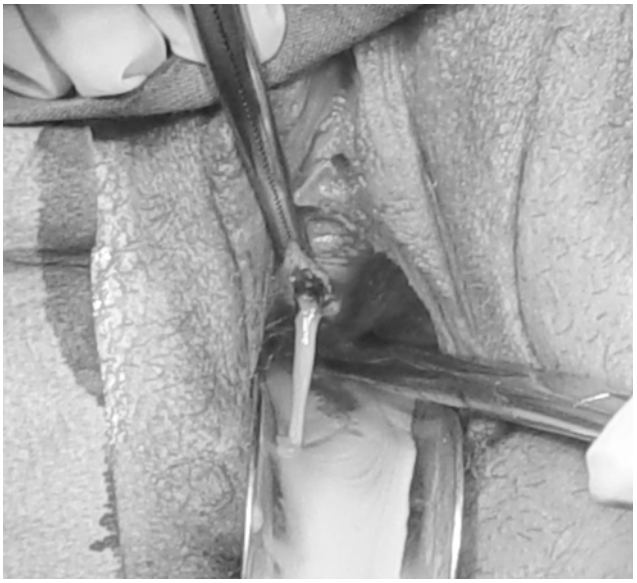


Fig. 7

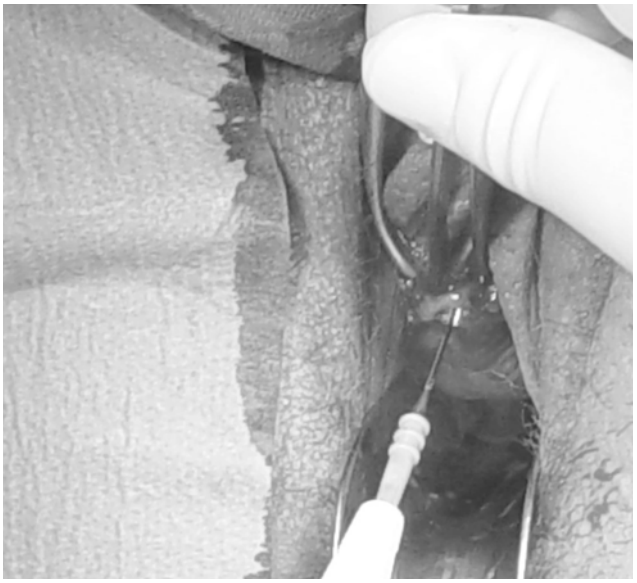


Fig. 8

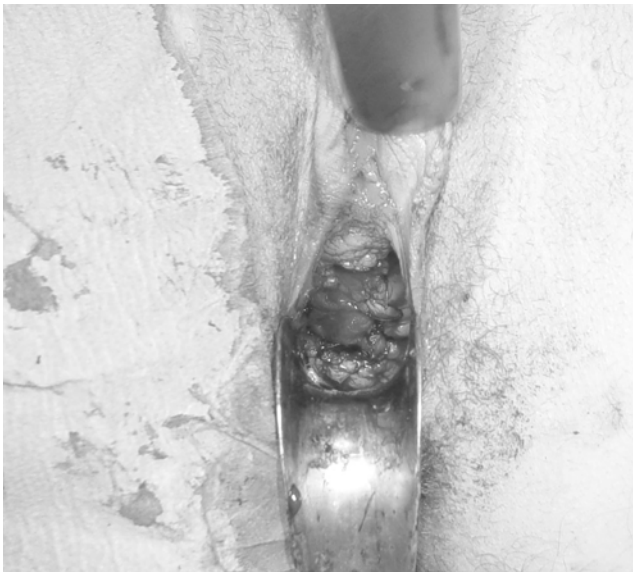


Figure 4. — Uterus didelphys was observed during laparotomy.

Figure 5. — Pus was aspirated from the right hemivagina using a needle and syringe.

Figures 6, 7. — A large amount of pus and blood was spilled out during the drainage of pyocolpos with conventional scalpel diathermy.

Figure 8. — Demonstration of the vagina after an almost complete resection of the right hemivaginal septum; the edges of the vaginal septum were sutured with interrupted Vicryl sutures.

Fig. 9

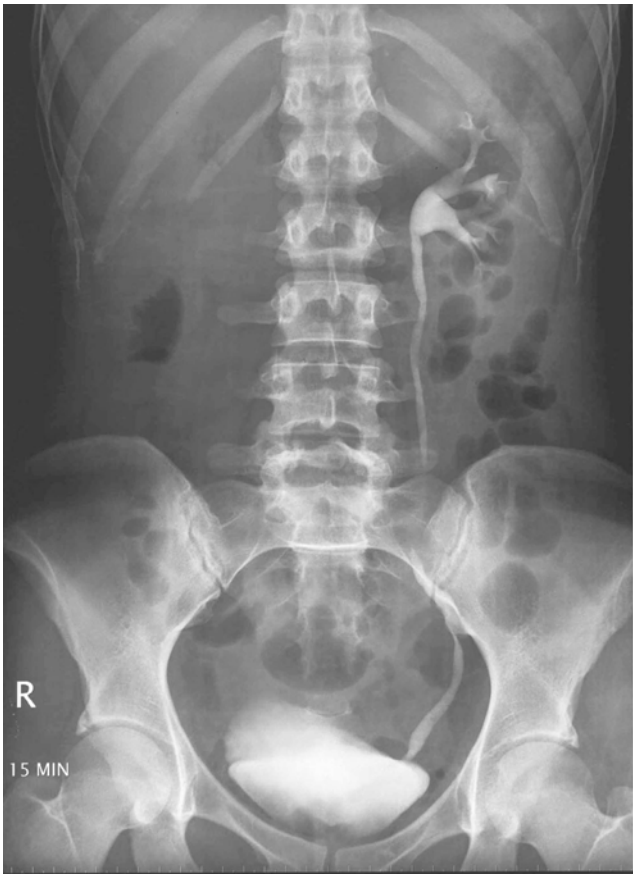


Fig. 10

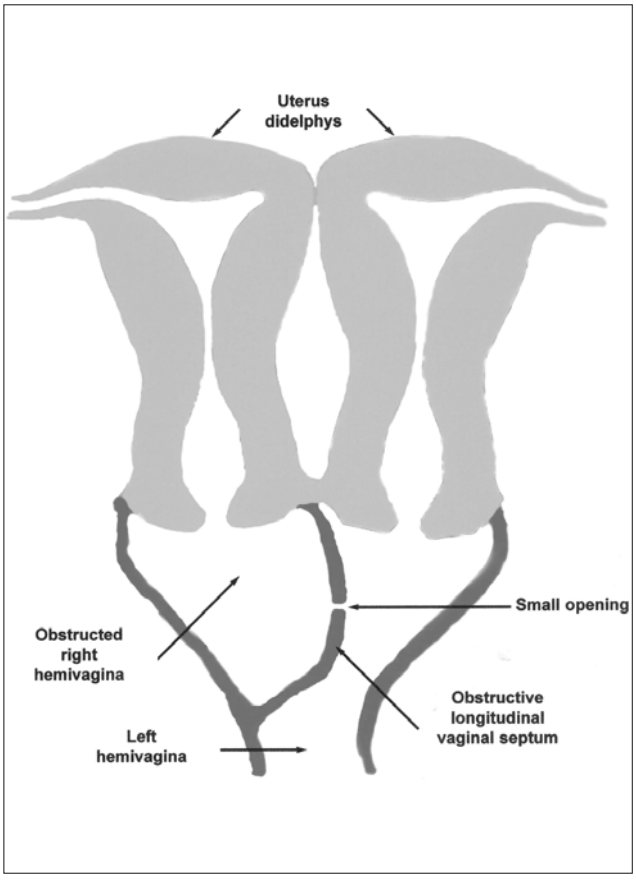


Figure 9. — Intravenous pyelography 15 min after injection showed agenesis of the right kidney and hypertrophy of the left kidney.
Figure 10. — Illustration of the surgical findings: uterus didelphys with obstructed right hemivagina and a small fistula hole on the right vaginal septum.

(Müllerian) ducts and the mesonephric (Wolffian) ducts. In the male embryo, a factor associated with the SRY region or sex-determining region of Y chromosome is responsible for the differentiation of the undifferentiated gonads to testes. If testes differentiate normally, Sertoli cells produce anti-Müllerian hormones, which results in the loss of the Müllerian duct and Leydig cells produce testosterone, which promotes Wolffian duct development. In 46, XX individuals, in the absence of testosterone, the Wolffian ducts begin to degenerate. Also, the lack of anti-Müllerian hormones results in the synchronous caudal elongation of Müllerian ducts along the lateral aspect of the gonads reaching the urogenital sinus. The adjacent mesonephric ducts are responsible for the fusion of the paramesonephric ducts. Fusion of the paramesonephric ducts results in the formation of the uterine body, cervix and upper two-thirds of the vagina. The lower one-third of the vagina is of urogenital sinus origin. Failure of Müllerian duct fusion leads to uterus didelphys (2 uterine bodies, 2 cervices, and vaginal septum). Because the genital system arise from a common embryonic mesoderm and the development of the uterovagianl complex is derived from the mesonephric ducts, maldevelopment of the paramesonephric ducts also may disturb embryogenesis of the kidneys and ureters [8-11]. Therefore, the association of uterus didelphys with blind hemivagina and ipsilateral renal agenesis simultaneously affects the mesonephric and paramesonephric ducts [2, 7, 12].

Most patients with uterus didelphys and blind hemivagina present with abdominal pain that starts right after menarche. It is presumed that hematocolpos is responsible for abdominal pain [13]. Menses in patients with this syndrome are often regular [12]. The main diagnostic problem of this syndrome is its low incidence, which makes the preoperative diagnosis difficult, although the clinical presentation is quite typical: dysmenorrhea, severe abdominal pain and pelvic mass after menarche [5]. Very rarely pyocolpos caused by infected fluid collection within the obstructed hemivagina may occur, as in the current case. Reflux of the pus into the peritoneal cavity causes salpingitis and acute abdomen and this is a life-threatening condition requiring emergency surgery. In young girls, after menarche the differential diagnosis of acute abdomen should include hematometra, hematocolpos/pyocolpos, pelvic inflammatory disease, pregnancy complications or torsion of an adnexal mass [14-19].

The treatment of patients with uterus didelphys associated with blind hemivagina is resection of the obstructing vaginal septum as complete as possible using scissors or scalpel or conventional scalpel diathermy or resectoscope

[4, 7]. If the patient complains of cyclic abdominal pain after surgical treatment, a suspicious endometriosis should be studied and treated [5, 7]. This syndrome has the best prognosis in terms of alleviation of symptoms. Successful pregnancy rates range from 37% to 40% of cases [20]. Annual Papanicolaou tests are recommended for each cervix. In the current study, the preoperative diagnosis was not correctly made because of the rarity of this condition and its late clinical presentation 11 years after menarche. The most likely preoperative diagnosis was a ruptured tuboovarian abscess because of (a) the acute abdomen of the patient; (b) the pus-like vaginal discharge; (c) the patient's fever; (d) the increased white blood cells; and, (e) the weakness of the radiologist to interpret the ultrasonographic findings. During surgery, the presence of a didelphys uterus and tuboovarian abscess or pyosalpinx made us consider the possibility of an obstructed vagina and avoid incomplete treatment.

In conclusion, obstetrician-gynecologists and the radiologists should keep in mind the very rare case of uterus didelphys with blind hemivagina and ipsilateral renal agenesis in order to avoid complications, diagnostic delay and incomplete management.

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