Herlyn-Werner-Wunderlich syndrome: An unusual presentation with pyocolpos

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Introduction

Herlyn-Werner-Wunderlich syndrome (HWWS) is a rare congenital anomaly of the urogenital tract, which is characterized by the triad of uterus didelphys, obstructed hemivagina, and ipsilateral renal agenesis. It usually presents at puberty with pelvic pain, dysmenorrhea, and a vaginal or pelvic mass. Although rare, it may present with purulent vaginal discharge due to secondary infection of the obstructed hemivagina, making diagnosis difficult. A careful pelvic examination to identify the cervix and vagina is the key to the diagnosis of Müllerian duct anomalies and magnetic resonance imaging can provide additional useful information. The optimal treatment is full excision and marsupialization of the obstructing vaginal septum so that both uteri can drain through the patent vagina. The authors report a case of a 22-year-old female with an unusual presentation of Herlyn-Werner-Wunderlich syndrome complicated by pyocolpos, which was successfully managed by vaginal septum resection and drainage of pus.

Keywords: Herlyn-Werner-Wunderlich syndrome; Renal agenesis; Urogenital anomalies; Uterus didelphys

Case report

A 22-year-old woman (gravida 0 and para 0) visited our gynecology department with the chief complaint of a foul-smelling vaginal discharge with intermenstrual bleeding. The patient denied fever, chills, nausea, vomiting, or diarrhea. Past medical and surgical history was unremarkable. Menarche had occurred at the age of 13 years. She still had some brownish discharge although her last menstrual period was 2 weeks ago. She had no complaints of dysmenorrhea, dyspareunia, or dysuria. Physical examination revealed no tender...
ness or palpable masses in the abdomen. Pelvic examination demonstrated a patent cervix and vagina with purulent discharge and bulging of the right lateral vaginal wall (Fig. 1A). The discharge was cultured for *Neisseria gonorrhoeae, Chlamydia trachomatis, and Trichomonas vaginalis*, but the results were negative. Laboratory tests revealed leukocytosis (18,220/mm³), C-reactive protein level of 1.18 mg/dL, and normal renal function. Transvaginal ultrasound images revealed uterus didelphys with a hypoechoic heterogeneous cystic mass measuring 4.8×5 cm behind the bladder. The right kidney was not visualized on subsequent renal ultrasound. Abdominopelvic MRI was performed to better identify the pelvic anatomy and verify the absence of the right kidney. This study confirmed the ultrasound findings of uterus didelphys with a distended right hemivagina measuring 3.1×3.5×4.8 cm (Fig. 2A), suggesting a turbid fluid collection (Fig. 2B), and right renal agenesis (Fig. 2C). The patient was diagnosed with HWWS complicated with pyocolpos. Resection of the vaginal septum was planned and prophylactic antibiotics were administered before surgery. Initially, a puncture was made into the right vaginal wall to identify the obstructed vaginal septum before the resection began. After confirmation of the septum, it was incised with electrocautery, and 200 mL of foul-smelling purulent material was drained (Fig. 1B). A specimen was sent for culture and sensitivity testing. The vaginal septum was widely excised until the cervix was reached (Fig. 1C). Laparoscopic exploration revealed uterine didelphys, powder-burn appearing endometriosis on the serosal surface of the uterus (Fig. 1D), and pelvic adhesions (Fig. 1E). The patient underwent laparoscopic ablation of the endometrial implants. Cultures from the specimen grew group B beta-hemolytic streptococcus, for which the patient was given a 7-day course.
of amoxicillin-clavulanate. She was discharged 2 days after surgery, having had an uneventful recovery.

Discussion

Uterine anomalies can result from failure of the Müllerian ducts to fuse in the midline, to connect with the urogenital sinus, or to create the appropriate lumen in the upper vagina and uterus by resorption of the central vaginal cells and the septum between the fused Müllerian ducts [2,4]. Lack of fusion of the two Müllerian ducts results in duplication of corpus and cervix [5]. Occasionally, uterus didelphys is associated with an obstructed hemivagina and often with ipsilateral renal agenesis [6]. Renal agenesis on the side of the obstructed vagina can be explained by embryologic arrest at 8 weeks of pregnancy, simultaneously affecting the Müllerian (paramesonephric) and Wolffian (metanephric) ducts [3,7]. This unique clinical syndrome was first described as HWWS in 1971. Estimates of its incidence range from 0.1% to 3.8% [8,9].

HWWS is usually discovered during puberty. Symptoms vary depending on the uterovaginal relationship in individual cases. A 10-year review of this anomaly showed that most patients presented with dysmenorrhea (73%), a pelvic or paravaginal mass (71%), and an affected right uterus and vagina (63.5%) [10]. In the present case, the patient presented unusually with excessive mucopurulent discharge. Pyocolpos can occur in patients with HWWS due to secondary infection of retained menstrual blood in the obstructed hemivagina. The two vaginal cavities can communicate through a partially fenestrated septum, or the two cervices can communicate through a fistula [11].

A careful pelvic examination to identify the cervix and vagina is the key to the diagnosis of Müllerian duct anomalies. Sonography can detect uterine malformations, but MRI is the study of choice for HWWS. MRI provides more detailed information regarding the uterine contour, the shape of the intrauterine cavity, the continuity with each vaginal lumen, the character of the septum, and the nature of the fluid content, as well as associated aspects such as endometriosis, pelvic adhesions, or renal anomalies [12].

The optimal treatment is full excision and marsupialization of the obstructing vaginal septum so both uteri can drain through the patent vagina [2]. In cases of pyocolpos or hematocolpos, two-step surgery may be recommended. Otherwise, distention and stretching of the septal tissue may increase the risk of inadequate resection and possible postoperative stenosis [13]. Laparoscopy is not mandatory but can be helpful in confirming the diagnosis when radiologic imaging is inconclusive. In addition, as in the reported case, laparoscopy is useful in the diagnosis and treatment of endometriotic lesions and pelvic adhesions, caused by retrograde menstruation from the obstructed hemivagina.

Uterine anomalies have long been associated with pregnancy loss and obstetric complications, but affected women generally are not infertile. A retrospective study of 36 patients with HWWS over a 30-year period revealed that 87% of the patients wanting children had a successful pregnancy, 23% had abortions, 15% had preterm births, and 62% had full-term pregnancies, for an overall live birth rate of 77% [14].
To our knowledge, HWWS is rare and only 10 cases have been reported in Korea [15]. Moreover, there have been no reports discussing the rare complication of pyocolpos in HWWS. Pyocolpos can occur in HWWS patients due to secondary infection of the obstructed hemivagina and thus may be difficult to diagnosis. Although it is not very common, it is important for clinicians to be aware of this entity, since timely diagnosis requires a high index of clinical suspicion.

Conflict of interest

No potential conflict of interest relevant to this article was reported.

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