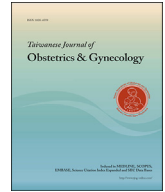




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Case Report

Herlyn–Werner–Wunderlich syndrome: An unusual case with presentation of menorrhagia

Hsin-I Liang ^{a, b}, Shao-Chi Fu ^{a, b}, Chih-Hsiang Yin ^b, Cheng-Chang Chang ^{b, *}^a Department of Obstetrics and Gynecology, Taichung Armed Forces General Hospital, Taichung, Taiwan, ROC^b Department of Obstetrics and Gynecology, Tri-Service General Hospital, National Defense Medical Center, Taipei, Taiwan, ROC

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ABSTRACT

Objective: Herlyn–Werner–Wunderlich (HWW) syndrome is a rare condition in which patients present with a palpable pelvic mass and pain caused by an obstructed hemivagina. Here we present a case of HWW syndrome characterized by prolonged menstrual bleeding.

Case report: A 19-year-old nonsexually active unmarried woman experienced irregular menstrual cycles and menorrhagia. The duration of menstrual bleeding was 10–14 days. She also suffered from mild dysmenorrhea since menarche at the age of 13. Transabdominal sonography revealed a double uterus and a heterogeneous myoma-mimicking mass over the left cervical region. The left kidney was absent. Magnetic resonance imaging revealed a double uterus, a double vagina with an unperforated left hemivagina, and ipsilateral renal agenesis. The patient underwent cervicovaginal orifice reconstruction surgery.

Conclusion: Left hematocolpos compression, a partially obstructed right vaginal channel, and an orifice with local venous drainage abnormalities resulted in prolonged menstrual bleeding. In HWW syndrome, the occurrence of a pelvic mass and pain is common; however, prolonged menstrual bleeding is rare.

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Introduction

Herlyn–Werner–Wunderlich (HWW) syndrome—or uterine didelphys associated with obstructed hemivagina and ipsilateral renal anomaly syndrome—results from a double uterus with a unilateral, imperforated double vagina and ipsilateral renal agenesis [1]. HWW syndrome was first reported in 1922 and was suspected in a young woman with regular menstruation who had experienced a gradual increase in pelvic pain and developed a palpable pelvic mass after menarche [2].

Case presentation

The patient was a 19-year-old unmarried woman with no history of sexual activity, systemic disease, or surgical procedures. The interval between her menstrual cycles was irregular, and the duration of menstrual bleeding was 10–14 days. She also suffered

from mild dysmenorrhea since menarche at the age of 13. She experienced progressively increasing pelvic pain in recent periods of menstruation. She was referred to our outpatient department because of a suspected cervical myoma diagnosis. Transabdominal sonography revealed a double uterus and one heterogeneous myoma-mimicking mass over the left cervical region (Fig. 1). The left kidney was absent. Magnetic resonance imaging (MRI) was used to further evaluate the patient's symptoms.

Fig. 2 (A) reveals that there were two uterine cavities with typical endometrial tissue separated by a myometrial septum. Two separate cervixes were found, which is compatible with uterine didelphys. There was a lobulated, mass-like lesion, measuring approximately $7.2 \times 6.9 \times 10.6$ cm in size, in the left hemivagina. The mass had a high signal intensity on T1-weighted MRI and an intermediate-to-low signal intensity on T2-weighted MRI. The mass was compatible with a dilated vagina containing collected blood, according to the image signal presentation, and an obstructive, low, transverse vaginal septum [Fig. 2 (B)]. The left kidney was absent, and left renal agenesis was suspected [Fig. 2 (C)]. Based on these findings, HWW syndrome was suspected.

Subsequently, surgical intervention was performed. Pelvic examination was performed with the patient in the dorsal lithotomy

* Corresponding author. Department of Obstetrics and Gynecology, Tri-Service General Hospital, National Defense Medical Center, No. 325, Sec. 2, Cheng-gong Rd., Neihs District, Taipei City, 114, Taiwan, ROC. Fax: +886 2 87927207.

E-mail address: obsynchang@gmail.com (C.-C. Chang).

position and under spinal anesthesia, and a protruding mass with a vaginal septum and an unperforated left hemivagina were identified. The patient underwent cervicovaginal orifice reconstruction. A wedged-shaped vertical incision was made in the left-side obstructed vaginal septum with a no. 15 blade, and approximately 250 ml of dark viscous fluid was evacuated. The lining of the obstructed vaginal septum was everted and approximated to the vaginal mucosa with interrupted sutures of 2-0 delayed absorbable material (Fig. 3).

One month later, the patient visited our outpatient clinic, and hysteroscopy was performed. We identified the external orifice of the right cervix and retained the absorbable sutures in the left vaginal orifice. Hysteroscopy revealed a right proximal tubal ostium, left cervical external orifice and left proximal tubal ostium were also identified (Fig. 4).

Discussion

Pelvic pain and a palpable pelvic mass are common symptoms of an obstructed vagina and ipsilateral renal agenesis. The diagnosis age ranges from 10 to 29 years with a median age of 14 years [3]. A complex malformation of the female genital tract has been found in rare cases [4]. These malformations are frequently incorrectly identified and inappropriately treated due to asymptomatic or non-specific symptoms that can interfere with diagnosis. In development, the paramesonephric ducts of the genetically female embryo midline fusion are found in the uterus, cervix, and the upper four-fifths of the vagina. The lower 20% of the vagina is formed from the sinovaginal bulbs, which are protrusions of the urogenital sinus [5]. The genital and urinary systems arise from a common ridge of the mesoderm along the dorsal body wall. The genital and urinary systems rely on normal mesonephric system development. Thus, mesonephric and paramesonephric duct malformations may be associated with renal abnormalities [5]. Female genital tract malformations are the consequences of disturbances or interruptions of embryologic development of the mesonephric and paramesonephric ducts caused by multiple factors such as unknown environmental, teratogenic, and genetic factors [6]. In the general population, the prevalence of paramesonephric duct abnormalities ranges approximately from 0.17% to 6.7%. A double uterus with a unilateral, unperforated double vagina accounts for 15%–30% of these anomalies [7]. HWW syndrome results from damage to the caudal portion of the mesonephric duct. Obstruction of the right side accounts for approximately 66% of cases and is more frequently accompanied by ipsilateral renal agenesis. However, our case with left-sided vaginal obstruction and left renal agenesis is even rarer.

The most common clinical presentation of HWW syndrome is pelvic pain, dysmenorrhea, and a palpable mass due to

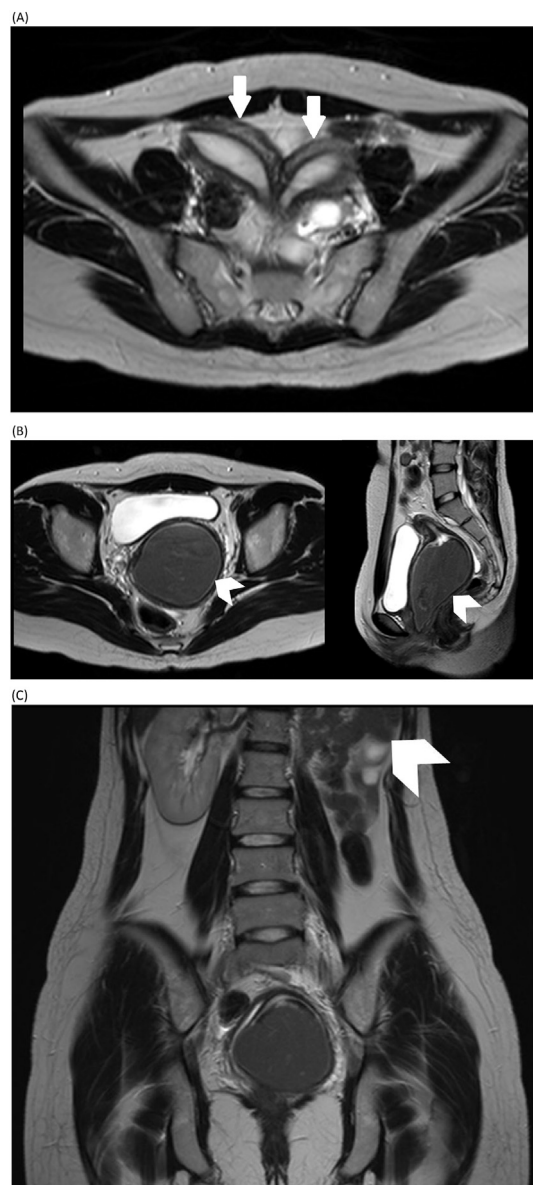


Fig. 2. (A). Axial T2-weighted magnetic resonance (MR) image demonstrating two uterine cavities with a normal endometrium (arrow). (B). Axial and sagittal T2-weighted MR images revealing a dilated hemivagina, with suspected collection of pooled blood according to the image signal presentation (arrowhead). (C). Absence of the left kidney (arrowhead).

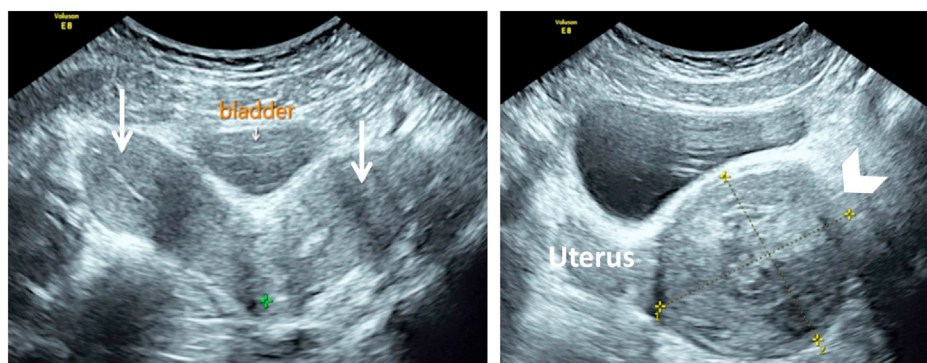


Fig. 1. Double uterus (thin arrow) and one heterogeneous, myoma-mimicking mass over the left cervical region (arrowhead).

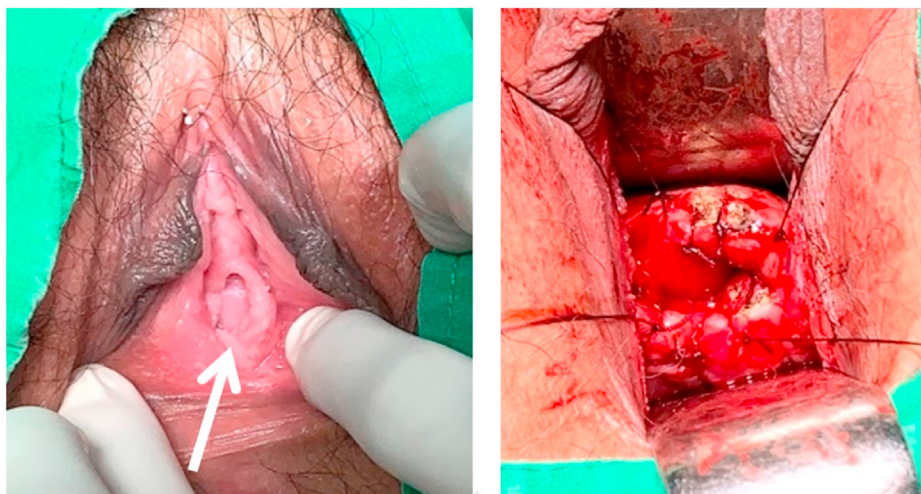


Fig. 3. A protruding mass over the vagina of an imperforated left hemivagina is shown (thin arrow). Image showing the reconstruction of the left cervicovaginal orifice with 2-0 delayed absorbable material.

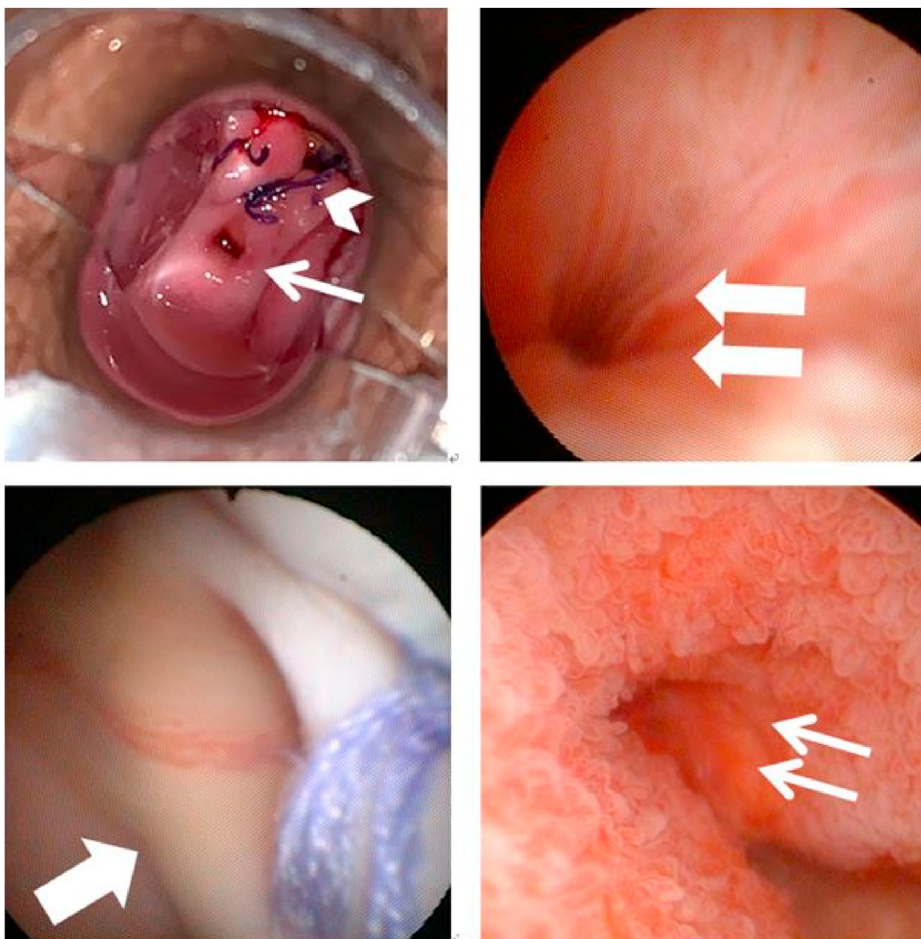


Fig. 4. The right cervical external orifice (thin arrow) and left vaginal orifice with the retained suture (arrowhead). Upon performing hysteroscopy, a right proximal tubal ostium (paired arrow), left cervical external orifice (arrow), and left proximal tubal ostium (paired thin arrow) were identified.

hematocolpos [8]. However, our case initially presented with menorrhagia and occasional dysmenorrhea. Because she was an unmarried woman with no sexual exposure, based on her clinical symptoms, a cervical myoma was suspected upon assessment by

transabdominal sonography at a local medical clinic. Transvaginal or transrectal sonography is an excellent tool for the differential diagnosis of hematocolpos and cervical myoma. Due to the left hematocolpos compression, a partially obstructed right vaginal

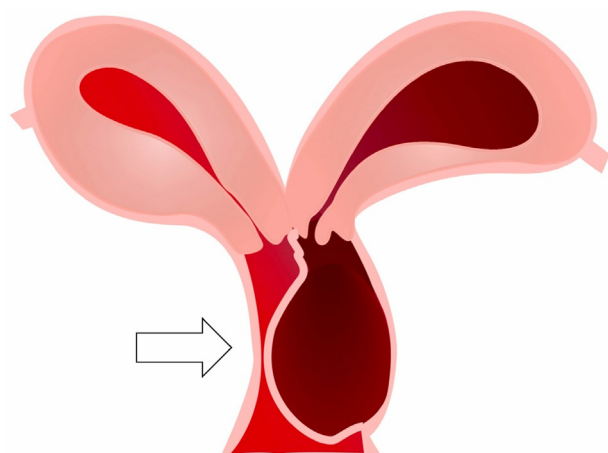


Fig. 5. A partially obstructed right vagina due to left hemocolpos was identified (arrow).

channel and an orifice with local venous abnormalities resulted in prolonged menstrual bleeding (Fig. 5). As demonstrated in this study, MRI is the gold standard technique for evaluating the exact condition of the female reproductive anatomy [9].

Surgical intervention with reconstruction of the vaginal orifice is indicated for vaginal obstruction with secondary hemocolpos. The primary purpose of this treatment is to relieve symptoms and ensure that a patient's fertility is uncompromised [10]. Current reports have confirmed that HWW syndrome is associated with an elevated risk of endometriosis [11]. The potential mechanism for endometriosis may be a result of vaginal outflow obstruction and retrograde blood flow through the endometrial tissue and fallopian tubes [12]. The rate of successful pregnancies in patients with HWW syndrome is approximately 87% [13].

In conclusion, although the occurrence of a pelvic mass and pain is common, prolonged menstrual bleeding is a rare symptom in young women with HWW syndrome. A correct diagnosis of HWW syndrome and timely treatment can relieve these symptoms, ensure the fecundity of a patient's fertility, and decrease complications.

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Declaration of competing interest

The authors have no conflicts of interest relevant to this article.

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