Case Report

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Successful management of Herlyn–Werner–Wunderlich syndrome in a 17-year-old virgin girl in the challenging socio-cultural-logistic setting of Syria: a case report

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Introduction and Importance: Herlyn–Werner–Wunderlich syndrome (HWWS) is a rare developmental anomaly of the female reproductive system caused by a failure of fusion during Mullerian duct development. The triad of uterus didelphys, obstructed hemivagina, and ipsilateral renal agenesis characterizes HWWS. The most common presenting symptoms are dysmenorrhoea, pelvic pain, primary infertility in later years, and an abdominal mass due to hematometrocolpos.

Case Presentation: A 17-year-old girl presented to the authors' department with recurrent low back pain, which was neither responsive to analgesics nor associated with urinary complaints, vomiting, or fever. Imaging techniques confirmed she had the triad of uterus didelphys, obstructed hemivagina, and right renal agenesis.

Clinical Discussion: The genital system is the same for males and females before 6 weeks of pregnancy. HWWS is a rare congenital disorder as a result of the failure of fusion during Mullerian duct development. It consists of a didelphic uterus, hemivaginal septum, and unilateral renal agenesis.

Conclusion: Shame and social stigma associated with virginity continue to endanger the lives of many girls in Syria. To complicate matters further, the low resources in Syria produced by war pose a difficult challenge in managing many gynecological conditions, including HWWS, like this case, in which endoscopic technologies were not available, necessitating open surgery while keeping in mind preserving hymen intactness. So, the authors indicate that preserving virginity could be conducted even though the approach is open surgery by very careful intervention and experienced surgeons.

Keywords: case report, herlyn–werner–wunderlich syndrome, müllerian anomalies, obstructed hemivagina, uterus didelphys, virginity

HIGHLIGHTS

- Herlyn–Werner–Wunderlich syndrome (HWWS) is a rare case, characterized by the triad of uterus didelphys, obstructed hemivagina, and ipsilateral renal agenesis.
- The most common presenting symptoms are dysmenorrhoea, pelvic pain, and abdominal mass caused by hematometrocolpos. Pain could be the only presenting symptom.
- Delayed diagnosis may lead to significant complications, such as endometriosis and major infections.
- Open surgery becomes a necessity in a low-resource setting with preserving the fertility and virginity of the patients.

Introduction

HWWS is a rare developmental anomaly of the female reproductive system. It is characterized by the triad of uterus didelphys, obstructed hemivagina, and ipsilateral renal agenesis[1]. The most common presenting symptoms are dysmenorrhoea, pelvic pain, and abdominal mass caused by hematometrocolpos due to an anatomical obstruction precluding the evacuation of the menstrual blood[2]. Moreover, we should recall that HWWS could be asymptomatic during childhood and early puberty. The exact propagation of obstructive Müllerian malformations is still unknown, but it was reported to be about 0.1–3.8% in women[3,4]. The prevalence of HWWS is unknown and remains a subject of discussion. Greater awareness of HWWS will lead to early diagnosis and reduced complications caused by a delayed diagnosis, such as endometriosis, infertility, and spontaneous abortion. Ultrasound (US) and MRI are used to establish the diagnosis. Surgery is the primary treatment, which includes the removal of the vaginal septum and drainage of the effusion[5]. This case report has been reported in line with the SCARE Criteria[6].

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Case presentation

A 17-year-old Syrian virgin girl presented with spasmodic and periodic pelvic pain, which was neither responsive to analgesics nor associated with urinary complaints, vomiting, or fever. The past medical, surgical, drug, and family histories were unremarkable. She had a regular menstrual cycle and was not sexually active. Her parents were non-consanguineous. Physical examination was unremarkable, and laboratory findings, including complete blood count, kidney function, and urine analysis were normal.

Using the US, uterus bicornis was noted with a right endometrial cavity filled with fluid collection measuring 12 mm. The cervix was also extended by fluid collection measuring 48 mm (Fig. 1). The right kidney was not visualized in the right lumbar region or the pelvis. The left kidney was enlarged in size, measuring 14.2 cm.

MRI of the pelvic organs was performed to confirm the US findings. The examination confirmed agenesis of the right kidney, the existence of two separate uterine bodies, and two separate cervixes, thus confirming the presence of uterus didelphys. There was evidence of pelvic fluid collection between the right cervix and the right body of the uterus which was suggested to be haematic (Fig. 2). According to these findings, HWWS was diagnosed.

Under general anesthesia, two separate uteri separated by a large septum were observed by an open abdominal approach (Fig. 3). A mildly dilated right uterus and a severely dilated right cervix were also noted. A probe was inserted through the hymen into the right vagina which was blind. Then we performed a longitudinal fissure between the two vaginas, removed a thin layer, and used simple interrupted sutures to establish a connec-

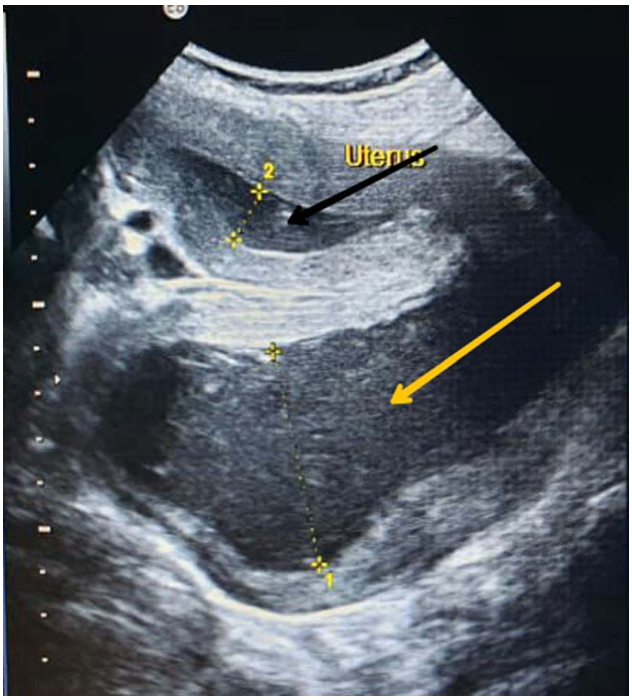

Figure 1. US of the pelvis showing right endometrial cavity filled with fluid collection with thickness 12 mm (the black arrow), and the right cervix with large, thick collection with thickness 48 mm (the yellow arrow). US, ultrasound

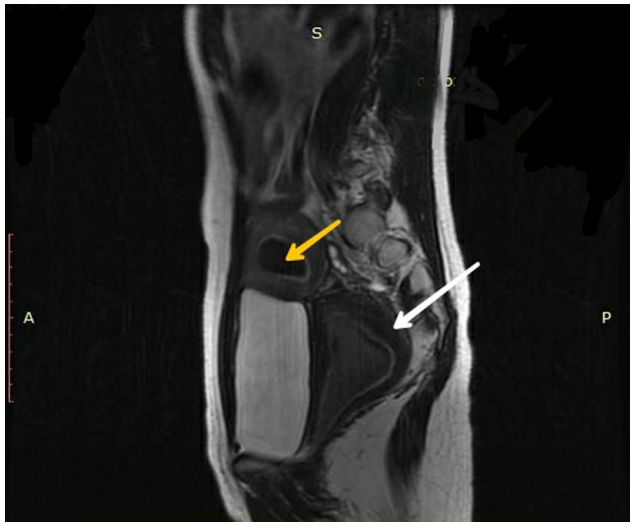

Figure 2. Sagittal T1-weighted image showing dilation of the cervical canal (the white arrow). The endometrial cavity was mildly dilated (the yellow arrow).

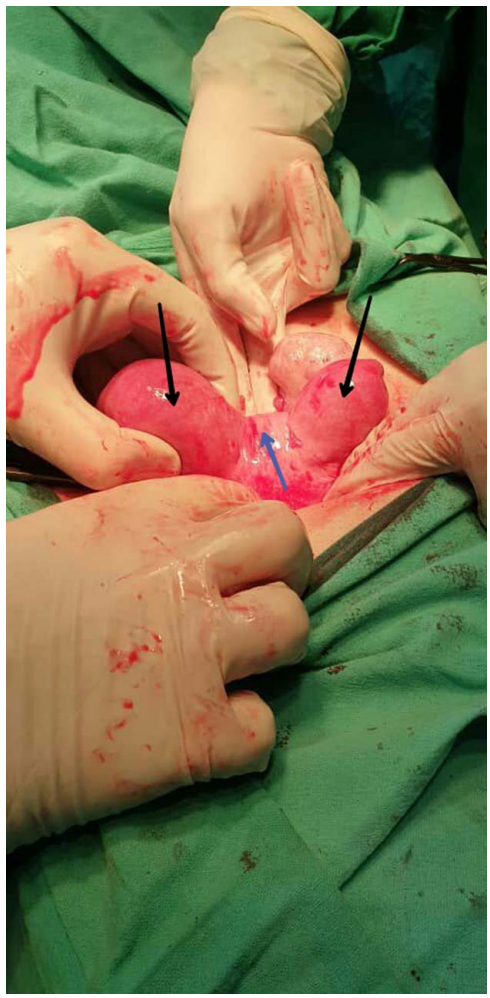

Figure 3. Open surgical approach shows the didelphic uterus, (the black arrows) and the separating septum (the blue arrow).

tion between the two cavities. The hymen was intact. Lastly, we put a drainage tube in the abdominal cavity and closed the abdomen layers. On follow-up after 3 months, the patient's symptoms were completely resolved.

Clinical discussion

The genital system is the same for males and females before 6 weeks of pregnancy. It consists of two pairs of symmetric genital ducts: the paramesonephric ducts (Mullerian) and the mesonephric ducts (Wolffian)[7]. Only the paramesonephric ducts develop in the absence of the Y chromosome, as testes produce Anti-Mullerian hormone, whereas mesonephric ducts do not. This distal segment of paramesonephric ducts gives later the uterine corpus, cervix, and the upper two-thirds of the vagina. The proximal segments remain unfused forming the fallopian tubes[7].

In the early stages, the female reproductive system is separated into two sections by a septum, and by the 12th week of pregnancy the septum is removed and the two sections merge to form a single uterine cavity. In the event of failure of fusion, the uterus will be bicornuate, and the cervix and endometrium will be separated as well[7]. HWWS is a rare congenital disorder as a result of a failure of fusion during Mullerian duct development. It consists of a didelphic uterus, hemivaginal septum, and unilateral renal agenesis[5]. As the reproductive system develops near the urinary tract and kidneys with some common embryonic structures, this genital malformation is always associated with homolateral renal agenesis. Obstructed hemivagina is a rare malformation that happens in 12% of women with congenital malformation of the Mullerian duct[8]. According to the literature, obstructed hemivagina was associated with a didelphic uterus in 18% of cases. Obstructed hemivagina and didelphic uterus are also reported with a 100% prevalence of renal agenesis on the same side of the obstruction, as in our case as well[9].

The most frequent symptoms of a didelphic uterus and a hemivaginal septum are periodical abdominal pain, dysmenorrhoea, and pelvic mass[9].

When a patient complains solely of abdominal pain such as in our case, many doctors usually prescribe anti-inflammatory medications, which leads to postponing the correct diagnosis of HWWS in patients with the condition. Dysmenorrhoea, due to the obstructed hemivagina was the most frequent symptom and has arisen in 30 patients (71%) in a study with a long-term follow-up of 49 cases with a didelphic uterus[9].

Nevertheless, other studies show that the most common symptom is pain after menarche with a vaginal or pelvic mass and normal periods. The diagnosis is usually made after the first menarche. Less frequently, the diagnosis is delayed such as in our case. Many factors may lead to a delayed diagnosis of HWWS such as having a regular normal menstrual period from unobstructed horn, and pain relieving, which might be the only warning, by an anti-inflammatory medication prescribed by doctors who did not pay enough attention to the possibility of this diagnosis due to its rarity and subtle presentation[5]. Early diagnosis is important to prevent endometriosis, infections, and other complications. Diagnosis is based on US and MRI. A pelvic mass and a vaginal septum can be shown by both ultrasonography and three-dimensional-ultrasonography. However, three-dimensional-ultrasonography is more effective in showing uterine malformations. Meanwhile, the vaginal septum is best shown on MRI with a very high sensitivity reaching 100%[8].

Patients with definite symptoms should receive proper management. The best approach is to undergo resection of the vaginal septum and drainage of the hematocolpos, which is believed to restore the functionality of the didelphys uterus, evading Hemihysterectomy. Like any other procedure, HWWS correction surgery has its complications such as dyspareunia, recurrence of obstruction, and vaginal stenosis. The extent of the surgical operation may have a big part in the emergence of these complications[8]. Other treatments are still under discussion. Moreover, having a second or an ipsilateral uterus reflects positively on fertility, therefore it is not recommended to have an ipsilateral hysterectomy[10]. Other complications related to this procedure are bladder and rectum injuries[9]. However, for such a rare congenital oddity, like a vaginal septum, a resectoscope allows exploring the vaginal pathology of the girls with an intact hymen[11].

For discussing the overall future prognosis of such patients, the fertility aspects must be included. A study has shown that 94% of women who wanted to conceive had at least one pregnancy and 89% of them had at least one living infant despite the possibility of preterm delivery, cesarean section, and spontaneous abortion[12].

The probability of hymen damage should be discussed with the patient and her family. As with any other malformation in the uterus, there is a risk of reproductive loss, premature delivery, and adverse obstetric outcomes[12].

In particular, hymen intactness is considered proof of virginity, an important sociocultural issue in Syria. The hymen is commonly referred to as "the tissue of virginity" in Arabic[13,14]. However, there are many misconceptions about the hymen in such societies, like the fact that the hymen is naturally perforated to allow menses egression. Hymen varies greatly in its natural shape or may have many congenital abnormalities. In addition, the hymen's flexibility increases after puberty allowing intercourse without bleeding. These facts are misunderstood by many people in conservative cultures which may lead to social disasters ([15]).

Honor-based violence is a pronounced problem in middle-eastern societies, such as Syria, where hymen intactness and proof of virginity are socially linked to the local concept of masculine honor. The male-decided presumptuous violation of such a concept causes many women and girls to face all kinds of sexual violence, including murder [13,14].

Such sociocultural pressure further burdened the attending healthcare providers of this case, adding to the logistic pressure of the low-setting Syrian healthcare environment caused by prolonged periods of conflict and national crisis.

Because of all of the above, knowledge of the hymen's anatomy and its abnormalities are essential to eliminate misconceptions and protect females from violent assault ([15]).

Conclusion

Shame and social stigma associated with virginity, leading to acts of sexual violence in the name of honor, including murder, continue to endanger the lives of many women and girls in Syria. To complicate matters further, the low-resource setting of the Syrian

healthcare environment produced by a decade of war poses a difficult challenge in managing many gynecological conditions, including the rare HWWS, like our case, in which endoscopic technologies were not available, necessitating open surgery while keeping in mind the difficult but essential task of preserving hymen intactness of the 17-year-old virgin patient. So, we would indicate that preserving virginity could be successfully conducted even though the approach is open surgery by very careful intervention and experienced surgeons.

Also, we would highlight that HWWS is a tricky rare condition that poses a great challenge to healthcare providers. Therefore, solid knowledge coupled with high suspicion of this syndrome is imperative.

Ethical approval

Not applicable.

Consent for publication

Written parental informed consent was obtained from the patient's parents for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

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Author contributions

N.M. contributed to drafting, editing, reviewing, and bibliography. E.A. contributed to drafting, reviewing, and editing. B. A.-G. contributed to drafting, reviewing, and editing. I.A. contributed to data collecting, reviewing, and editing. N.T. is the mentor and contributed to reviewing and supervising. All authors read and approved the final manuscript.

Conflicts of interest disclosure

The authors declare that they have no competing interests.

Registration of research studies

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