Case Report

ANNALS OF MEDICINE & SURGERY

OPEN

Acute urinary retention in an adolescent female with Herlyn–Werner–Wunderlich syndrome

Pradeep Shrestha, MBBS*[,a], Bibek Shrestha, MBBS[a], Amit K. Mishra, MBBS[a], Neeta Katuwal, MD[b]

Introduction and importance: Herlyn–Werner–Wunderlich syndrome (HWWS) is a rare congenital anomaly characterized by the triad of uterus didelphys, unilateral obstructed hemivagina, and ipsilateral renal agenesis. However, atypical presentations such as acute urinary retention are uncommon and pose diagnostic challenges. Early identification and treatment are crucial to prevent long-term complications like endometriosis and infertility.

Case presentation: A 12-year-old female presented with the unusual symptom of acute urinary retention, which was likely caused by compression of the bladder neck due to hematocolpos. Imaging via ultrasound and MRI confirmed the presence of uterus didelphys, obstructed hemivagina, and ipsilateral renal agenesis, consistent with HWWS. Surgical excision of the vaginal septum was performed to relieve the hematocolpos, with the patient experiencing a full recovery.

Clinical discussion: Congenital abnormalities of Mullerian Duct are rare, and atypical presentations such as acute urinary retention, vaginal discharge, and even pyocolpos or hematosalpinx have been reported. MRI played a crucial role in confirming the diagnosis and guiding treatment. The importance of early detection and long-term monitoring is highlighted, though limitations exist due to the inability to assess future reproductive outcomes.

Conclusion: This case emphasizes the importance of recognizing atypical presentations of HWWS, such as acute urinary retention. Timely diagnosis and appropriate surgical intervention are critical for preventing complications and restoring normal function. Early recognition of this rare syndrome, especially in patients with unusual symptoms such as urine retention, is essential for ensuring optimal clinical outcomes.

Keywords: acute urinary retention, case report, Herlyn–Werner–Wunderlich syndrome, OHVIRA syndrome

Introduction

Herlyn–Werner–Wunderlich syndrome (HWWS) is a rare congenital anomaly characterized by uterus didelphys, unilateral blind hemivagina, and ipsilateral renal agenesis[1]. The exact pathogenesis remains unclear, but it involves abnormal development of mesonephros and paramesonephros[2]. The condition typically presents after menarche with symptoms such as dysmenorrhea, pelvic pain, and vaginal bleeding[3]. The clinical features typically appear after menarche, including progressive pelvic pain and a palpable mass due to hematocolpos[4]. HWWS is a rare entity in surgical Gynecology incidence 0.1% and 3.8%[5]. Urinary retention is an uncommon symptom in these patients, potentially due to hematocolpos exerting pressure on the bladder neck, thereby obstructing urinary flow[6]. Management approaches differ based on age and symptom presentation, with asymptomatic pre-menarche patients recommended for regular follow-up until menarche onset, while post-menarche or symptomatic patients may require surgical intervention[7]. Treatment typically involves surgical resection of the vaginal septum to relieve obstruction and prevent complications such as endometriosis, adhesions, and infertility[8].

This report presents a case involving a 12-year-old girl with HWWS who presented with acute urinary retention which is an atypical and rare presentation. This case has been reported following the SCARE Guideline[9].

Case presentation

A 12-year-old female patient presented to the emergency department of Tertiary Hospital with a sudden onset of urinary retention, persisting for 1 day. Prior to this, she had experienced normal urinary frequency and flow. The urinary retention was accompanied by burning micturition and a constant, dull, aching pain in the lower abdomen, which was non-radiating. The patient had attained menarche at age 10, with regular menstrual cycles and normal flow over the past 2 years, without any clots or dysmenorrhea. Upon initial assessment, her general condition was stable, and her vital signs were within normal limits. Abdominal examination revealed a soft, non-tender, and non-distended abdomen with normal bowel sounds, and no palpable masses were detected. A vaginal inspection revealed a single vaginal opening with no visible bluish bulge. Examination of the vulva showed a normal hymenal opening.

[a]Maharajgunj Medical Campus, Tribhuvan University, Institute of Medicine, Maharajgunj, Nepal and [b]Department of Obstetrics and Gynaecology, Tribhuvan University Teaching Hospital, Maharajgunj, Nepal

Sponsorships or competing interests that may be relevant to content are disclosed at the end of this article.

*Corresponding author. Address: Maharajgunj Medical Campus, Tribhuvan University, Institute of Medicine, Kathmandu, Nepal.
Tel.: +977 9840968516.
E-mail: pradeepstha023@gmail.com (P. Shrestha).

Annals of Medicine & Surgery (2025) 87:331–334

Received 23 August 2024; Accepted 5 November 2024

Published online 9 January 2025

http://dx.doi.org/10.1097/MS9.0000000000002752

The patient was immediately catheterized, resulting in the drainage of 600 mL of urine, and the catheter was maintained for 5 days during her hospitalization. During her hospital stay, blood investigations, an abdominal ultrasound (USG), and a magnetic resonance imaging (MRI) scan of the abdomen and pelvis were performed. The blood tests, including hemoglobin (12.3 g/dL), routine urine analysis, and renal function tests, were all within normal ranges. The USG revealed a hemorrhagic ovarian cyst on the left ovary measuring 10.1 × 6.4 cm, approximately 256 mL of hematocolpos, a bicornuate bicollis uterus, and the non-visualization of the left kidney. MRI findings confirmed the presence of two distinct uterine horns consistent with a bicornuate uterus. (Fig. 1) Additionally, distended vagina was observed, suggesting hematocolpos (Fig. 2). The findings likely represented HWWS.

Upon review of her reports, her family was counseled regarding her condition, and informed consent was obtained for examination under anesthesia, followed by excision of the vaginal septum. For the management of Hematocolpos following sterile conditions painting and draping was done, and an incision was made at the site of the maximum bulge, resulting in the evacuation of approximately 300 mL of dark, chocolate-colored blood. During the operation, the longitudinal vaginal septum in the obstructed vagina was visualized (Fig. 3). The septum was excised transvaginally, and the resected septal wall was sutured to the anterior and posterior vaginal walls (marsupialization) was performed. Hemostasis was ensured, and the vaginal cavity was packed with a custom mold made of a sponge wrapped in a condom to prevent adhesions from forming. Ovarian cyst was asymptomatic for which regular follow-up with ultrasound was done to monitor the cyst.

Postoperatively, both the right and left cervices were visualized and palpated during a combined vaginal and speculum examination by the surgeon confirming the successful outcome of the intervention. After the surgery, the urinary catheter was removed, and the patient was able to void urine normally. Her postoperative recovery was uneventful, without any complications. During her follow-up visit after her next menstrual cycle, she reported no complaints, indicating a positive clinical outcome.

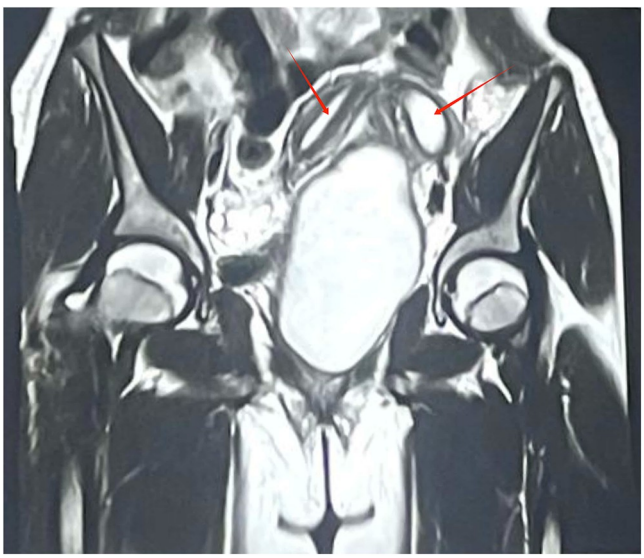

Figure 1. Axial MRI findings in a 12-year-old patient with OHVIRA syndrome arrow showing bilateral uterine horns.

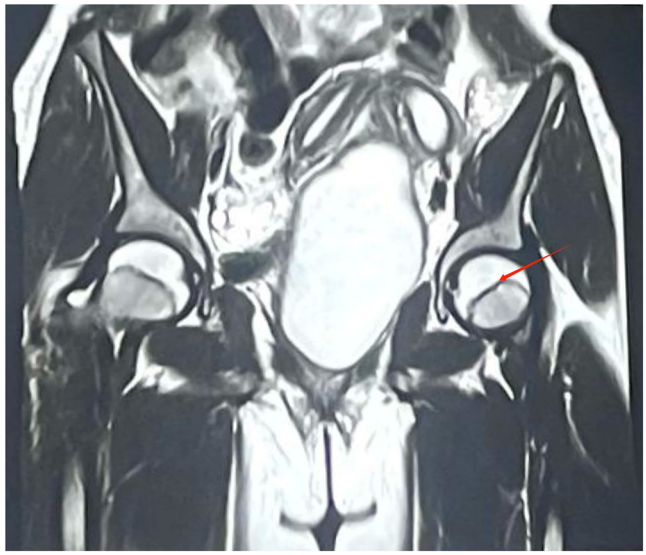

Figure 2. Axial MRI findings in a 12-year-old patient with OHVIRA syndrome arrow showing distended ovary: left hematocolpos.

Discussion

Congenital abnormalities of the Müllerian tract are estimated to occur in 2% to 3% of women overall[10]. HWWS also known as OHVIRA is characterized by uterus didelphys, unilateral blind hemivagina, and ipsilateral renal agenesis typically asymptomatic until menarche, patients often present with abdominal pain, dysmenorrhea, and pelvic mass due to hematocolpos or hematometra[11,12]. It is often associated with delayed diagnosis, misdiagnosis, and errors in management[12]. However, atypical presentations such as acute urinary retention, vaginal discharge, and even pyocolpos or hematosalpinx have been reported[10]. In this case, a 12-year-old female patient presented with the sudden onset of urinary retention with dull aching constant lower abdominal pain and burning micturition. This unusual presentation of urine retention might be due to obstructed hemivagina. Nandan *et al* also reported the rare presentation as urine retention due to obstructed hemivagina[10].

Diagnosis can be achieved through various imaging techniques, such as ultrasound, CT, and MRI[13]. Ultrasound is usually the initial diagnostic tool, but MRI is superior for detailed characterization of the uterine and vaginal anatomy, as well as associated renal anomalies[14]. MRI findings include two uterine horns, two separate vaginal canals, and distention of the obstructed hemivagina which is a characteristic feature. In this case, USG revealed hematocolpos, a bicornuate bicollis uterus, and the absence of visualization of the left kidney. To further confirm these findings, MRI was performed, which demonstrated the presence of two distinct uterine horns and two cervical canals, consistent with the diagnosis of a bicornuate bicollis uterus. Early diagnosis and appropriate surgical management are crucial for preventing complications and ensuring normal menstrual cycles post-surgery. Delayed diagnosis can lead to complications such as endometriosis, which is present in approximately 14% of cases[15].Treatment

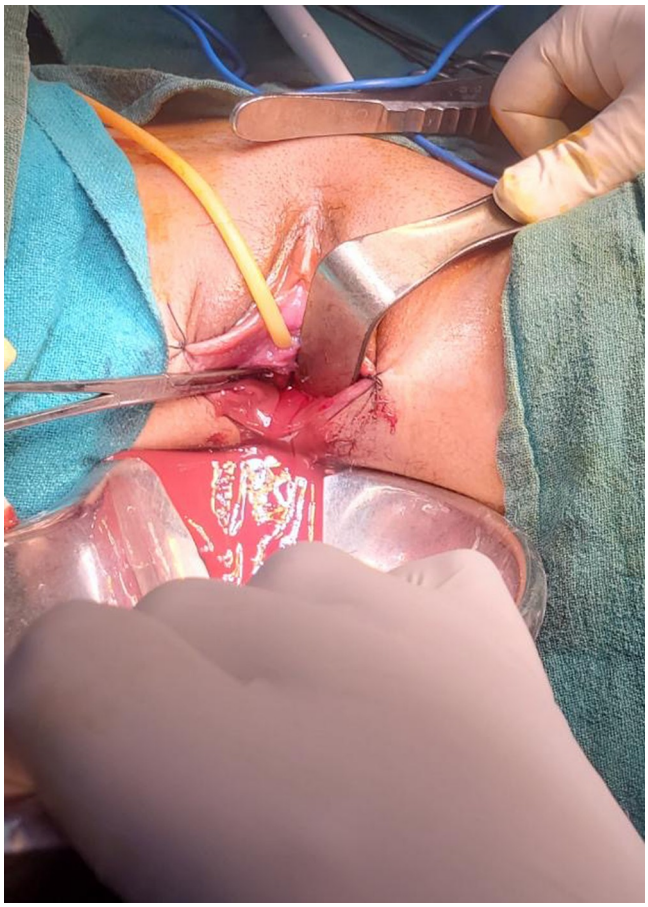

Figure 3. Intraoperative view showing the longitudinal vaginal septum in the obstructed vagina, with evidence of blood evacuation from the hematocolpos.

primarily involves surgical drainage of hematocolpos and resection of the vaginal septum[16]. A combined laparoscopic and transvaginal approach has been reported as useful for treating OHVIRA with oviductal hematoma[17]. Innovative instruments like specialized vaginal retractors can facilitate the procedure in young patient[18]. Kudela *et al* suggested vaginal septectomy as the most common and sufficient surgical treatment for hematocolpos in OHVIRA syndrome[19]. In this case, the evacuation of approximately 300 mL of dark, chocolate-colored blood and septal resection by transvaginal approach and sutured by Marsupialization technique was done and custom mold made of a sponge wrapped in a condom to prevent adhesions from forming. Long-term follow-up is essential to monitor potential complications such as endometriosis and renal issues[7]. Surgical correction of obstructed hemivagina in OHVIRA patients can lead to a variety of reproductive risks, though successful term and near-term pregnancies are possible[20]. The limitation of the study was that since the patient was just 12 years old, the long-term impact of the surgery on preventing reproductive issues could not be assessed.

Conclusion

This case of HWWS presenting with acute urinary retention highlights an unusual manifestation of this rare congenital anomaly. Prompt diagnosis through ultrasound and MRI are essential for identifying the condition and guiding appropriate management. Surgical excision of the vaginal septum and drainage of hematocolpos successfully relieved the obstruction, allowing for normal urinary function and an uneventful recovery. This case emphasizes the importance of considering HWWS in adolescent females with atypical symptoms, as early diagnosis and timely surgical intervention are critical for preventing complications such as endomsetriosis and preserving future fertility.

Ethical approval

None.

Consent

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

Sources of funding

None.

Author's contribution

P.S., B.S. and A.K.M. wrote the original manuscript, reviewed, and edited the original manuscript and N.K. reviewed and edited the original manuscript.

Conflicts of interest disclosure

None.

Research registration unique identifying number (UIN)

Not applicable.

Guarantor

Dr. Neeta Katuwal.

Provenance and peer review

Not commissioned, externally peer reviewed.

Data availability statement

Research data associated with case series are available.

Acknowlgedgements

Not applicable.

References

[1] Mittal R. Herlyn–Werner–Wunderlich syndrome. J Obstet Gynecol India 2015;66:128–30.

[2] Panaitescu AM, Peltecu G, Gică N. Herlyn-Werner-Wunderlich Syndrome: case report and review of literature. Diagnostics 2022;12:2466.

[3] Nhu QV, Trong BL, Thanh TN. Herlyn-Werner-Wunderlich syndrome: a report of three cases in adolescents and adult woman. Radiol Case Rep 2021;16:2792–98.

[4] Fachin CG, Rocha JLAS, Maltoni AA, *et al*. Herlyn-Werner-Wunderlich syndrome: diagnosis and treatment of an atypical case and review of literature. Int J Surg Case Rep 2019;63:129–34.

[5] Burgis J. Obstructive Müllerian anomalies: case report, diagnosis, and management. Am J Obstet Gynecol 2001;185:338–44.

[6] Gajendran I, Uzamere O, McSurdy K, *et al*. An unusual cause of recurrent urinary retention in an adolescent female. Cureus 2019;11:e5136.

[7] Zarfati A, Lucchetti MC. OHVIRA (Obstructed Hemivagina and Ipsilateral Renal Anomaly or Herlyn-Werner-Wunderlich syndrome): is it time for age-specific management? J Pediatr Surg 2022b;57:696–701.

[8] Siu Uribe A, Vargas Cruz V, Murcia Pascual FJ, *et al*. Síndrome de OHVIRA: características clínicas y complicaciones, nuestra experiencia [Clinical characteristics and complications in patients with OHVIRA (obstructed hemivagina and ipsilateral renal anomaly) syndrome. Our experience]. Cir Pediatr 2019;32:11–16.

[9] Sohrabi C, Mathew G, Maria N, *et al*. The SCARE 2023 guideline: updating consensus Surgical CAse REport (SCARE) guidelines. Int J Surg Lond Engl 2023;109:1136.

[10] Nandan N, Tanneru A, Rai P, *et al*. OHVIRA syndrome with a rare presentation. J South Asian Federation Obstet Gynaecol 2023;15:354–56.

[11] Dias JL, Jogo R. Herlyn–Werner–Wunderlich syndrome: pre- and post-surgical MRI and US findings. Abdominal Radiol 2015;40:2667–82.

[12] Zaborowska Ł, Stefania T, Inga L, *et al*. Overview of patients with OHVIRA syndrome: diagnosis delay, misdiagnosis, and errors in management. Eur J Obstet Gynecol Reprod Biol 2024;293:54–55.

[13] Bajaj S, Misra R, Thukral B, *et al*. OHVIRA: uterus didelphys, blind hemivagina and ipsilateral renal agenesis: advantage MRI. J Hum Reprod Sci 2012;5:67.

[14] Hayat AM, Yousaf KR, Chaudhary S, *et al*. The Herlyn-Werner-Wunderlich (HWW) syndrome – a case report with radiological review. Radiol Case Rep 2022;17:1435–39.

[15] Kudela G, Wiernik A, Drosdzol-Cop A, *et al*. Multiple variants of obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) syndrome – one clinical center case series and the systematic review of 734 cases. J Pediatr Urol 2021b;17:653.e1–653.e9.

[16] M'hamdi O, Boujida S, Bensghir I, *et al*. Hematocolpos in women with uterus didelphys, imperforate hemivagina and ipsilateral renal agenesis: a case report and review of the literature. Int J Surg Case Rep 2022;95:107277.

[17] Arakaki R, Yoshida K, Imaizumi J, *et al*. Obstructed hemivagina and ipsilateral renal agenesis (OHVIRA) syndrome: a case report. Int J Surg Case Rep 2023;107:108368.

[18] Satirapod C. Optional method with innovative instrument for OHVIRA syndrome treatment: case reports and review of literature. J Gynecol Res Rev Rep 2023;5:1–6.

[19] Kudela G, Wiernik A, Drosdzol-Cop A, *et al*. Multiple variants of obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) syndrome – one clinical center case series and the systematic review of 734 cases. J Pediatr Urol 2021;17:653.e1–653.e9.

[20] Bunnell M, Laufer M, Cipres DT. Case series of reproductive outcomes after surgical correction of obstructed hemi-vagina in OHVIRA. Am J Perinatol Rep 2023;14:e26–e30.