THE ASEAN JOURNAL OF RADIOLOGY
ISSN 2672-9393

Classic Case

Imaging findings of OHVIRA syndrome with surgical correlation in an 11-year-old girl: A classic case

Fon Suratsukkasem, M.D.
From Department of Radiology, Mahidol Bumrungrak Nakhonsawan Medical Center, Nakhonsawan, Thailand.
Address correspondence to F.S. (email: fonfon212224@gmail.com)

Received 5 March 2026; revised 25 March 2026; accepted 28 March 2026
doi:10.46475/asean-jr.v27i2.1021

Abstract

OHVIRA syndrome (Obstructed Hemivagina and Ipsilateral Renal Agenesis) is a rare congenital Müllerian duct anomaly characterized by uterine duplication, obstructed hemivagina, and ipsilateral renal agenesis. Early recognition is essential to prevent long-term complications such as endometriosis and infertility. We report the case of an 11-year-old girl who presented with acute lower abdominal pain for one day without fever. Physical examination revealed a 10-cm vaginal mass. Transabdominal ultrasonography demonstrated a cystic mass in the mid-pelvic cavity. Contrast-enhanced computed tomography confirmed uterine didelphys with marked distension of the right hemivagina and absence of the right kidney. Surgical exploration revealed a bulging right hemivagina with approximately 400 mL of retained brownish fluid, and vaginal septum excision was performed. The postoperative course was uneventful. This case highlights the importance of multimodality imaging in the early diagnosis and management of OHVIRA syndrome.

Keywords: Computed tomography, Obstructed hemivagina, OHVIRA syndrome, Renal agenesis, Ultrasound.

Introduction

OHVIRA syndrome, also known as Herlyn–Werner–Wunderlich syndrome, is a rare congenital Müllerian duct anomaly characterized by the triad of uterine didelphys, obstructed hemivagina, and ipsilateral renal agenesis [1]. The estimated incidence of Müllerian duct anomalies ranges from 0.1% to 3.8% in the general population [2].

The Müllerian (paramesonephric) ducts normally develop into the fallopian tubes, uterus, cervix, and upper vagina through a process of elongation, fusion, canalization, and septal resorption. At 9 weeks of gestation, the ducts elongate and reach the urogenital sinus. The uterovaginal canal is formed and inserted into the urogenital sinus at the Müller tubercle [2].

The mesonephric (Wolffian) ducts play an important role in the development of the urinary system and are closely related to the Müllerian ducts during embryogenesis. Disruption of this coordinated development may result in combined genital and urinary tract anomalies. In OHVIRA syndrome, failure of Müllerian duct fusion leads to uterine didelphys, while ipsilateral renal agenesis is attributed to abnormalities of the mesonephric duct.

Patients typically present shortly after menarche with cyclic lower abdominal pain, dysmenorrhea, and a tender vaginal mass. Other reported symptoms include recurrent urinary tract infections, urinary retention, and vaginal discharge [3]. Delayed diagnosis may lead to complications such as endometriosis, pelvic adhesions, and an increased risk of infertility and other adverse reproductive outcomes [4].

Ultrasonography is usually the first-line imaging modality because it is readily available, cost-effective, and free of ionizing radiation. Magnetic resonance imaging is considered the reference standard because of its multiplanar capability and superior soft-tissue characterization [5]. Early recognition is essential for timely surgical management and prevention of long-term reproductive complications.

Herein, we report a classic case of OHVIRA syndrome in an 11-year-old girl presenting with acute lower abdominal pain, emphasizing the role of multimodality imaging in diagnosis and surgical management.

THE ASEAN JOURNAL OF RADIOLOGY
ISSN 2672-9393

Case summary

An 11-year-old girl presented with acute lower abdominal pain for one day. She denied fever, abnormal uterine bleeding, or urinary symptoms. Physical examination revealed a palpable lower abdominal mass and a 10-cm bulging vaginal mass. Transabdominal ultrasonography demonstrated a well-defined hypoechoic cystic mass measuring approximately 9.4 × 5.9 cm, located in the mid-pelvic cavity, with a suspected connection to the right uterine horn. The left uterine horn was separately identified in the left pelvic cavity (Figure 1).

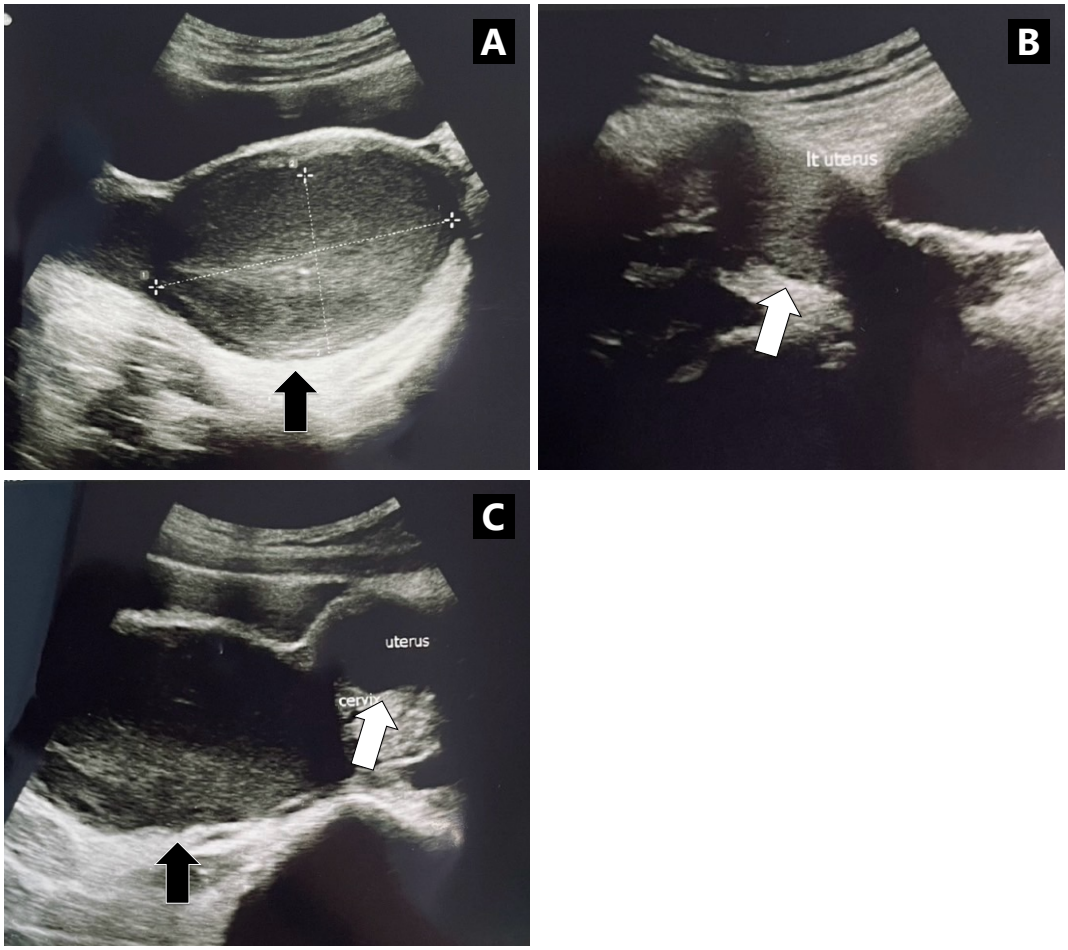

Figure 1. *Transabdominal ultrasonography shows a well-defined hypoechoic cystic mass in the mid-pelvic cavity (A and C, black arrow) and the left uterine horn in the left pelvic cavity (B and C, white arrow).*

Contrast-enhanced computed tomography of the lower abdomen confirmed uterine didelphys. Hydrocolpos of the right hemivagina was seen, measuring 6.9 × 7.7 × 13.9 cm (AP × transverse × craniocaudal dimensions). It communicated with the right uterine horn, which demonstrated hydrometra. The right kidney was not visualized, while the left kidney appeared normal. The left hemivagina, left uterine horn, and bilateral ovaries were unremarkable (Figure 2).

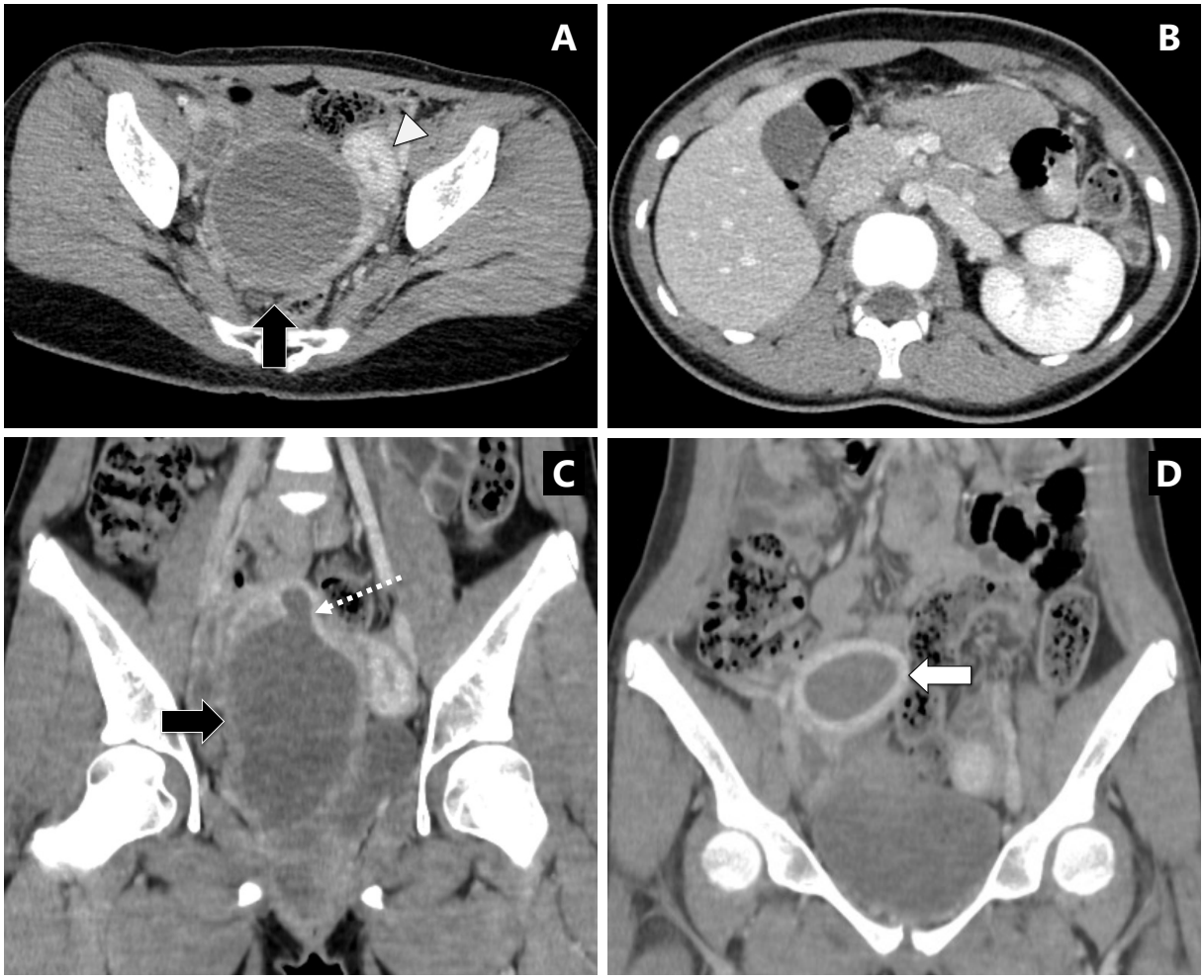

Figure 2. *Axial (A, B) and coronal (C, D) contrast-enhanced CT images of the lower abdomen demonstrate hydrocolpos of the right hemivagina (black arrow), communicating with the endocervix (white dotted arrow) and extending into the right uterine horn, which shows hydrometra (white arrow). The left uterine horn is unremarkable (arrowhead). The right kidney is not visualized (B).*

THE ASEAN JOURNAL OF RADIOLOGY
ISSN 2672-9393

The patient underwent vaginoplasty with resection of the vaginal septum under general anesthesia. Intraoperative findings revealed a bulging right vaginal canal, from which approximately 400 mL of thick brownish fluid was drained. The vaginal septum was incised to establish continuity of the vaginal canal. A partial vaginal septum, located approximately 5 cm above the hymen, was subsequently excised to reconstruct a single vaginal canal. The procedure was completed without complications, and the postoperative course was uneventful.

Discussion

OHVIRA syndrome represents a rare spectrum of Müllerian duct anomalies resulting from abnormal development of the Müllerian and mesonephric ducts. The condition is classically characterized by uterine didelphys, obstructed hemivagina, and ipsilateral renal agenesis [1,6]. The close embryologic relationship between the genital and urinary systems explains why renal anomalies frequently coexist in affected patients.

Several classifications have been proposed based on the degree of vaginal obstruction and uterine communication. Our case corresponds to Type 1.1 [7], defined as a completely obstructed hemivagina with a blind hemivaginal pouch. This subtype typically presents at an earlier age, often shortly after menarche due to the progressive accumulation of menstrual blood, leading to hematocolpos and hematometra. Early recognition is particularly important in Type 1.1 cases, as prolonged obstruction may predispose patients to endometriosis, pyosalpinx, and even pyocolpos [7,8].

Ultrasonography is usually the first-line imaging modality because it is readily available and free of ionizing radiation [5]. In this patient, transabdominal ultrasonography demonstrated a large midline cystic pelvic mass with a separately identified uterine horn, raising suspicion for a Müllerian duct anomaly. Although magnetic resonance imaging is considered the reference standard for evaluating complex Müllerian anomalies due to its multiplanar capability and superior soft-tissue characterization [5], contrast-enhanced CT was performed due to the patient's acute presentation and institutional availability. CT adequately delineated uterine duplication, marked distension of the right hemivagina containing slightly hyperattenuating fluid suggestive of retained old blood products, and absence of the right kidney. These findings were essential for confirming the diagnosis and guiding surgical planning.

Definitive management consists of surgical resection of the obstructing vaginal septum to restore normal outflow and prevent long-term complications [9]. In our case, surgical findings correlated well with imaging, revealing a markedly distended right vaginal canal containing retained old blood. Septum excision successfully established a single vaginal canal, and the postoperative course was uneventful.

This case underscores the pivotal role of multimodality imaging in the diagnosis of OHVIRA syndrome and highlights the importance of early detection, particularly in completely obstructed subtypes, to prevent reproductive sequelae.

This report provides additional educational value by demonstrating the diagnostic role of CT in emergency settings and highlighting precise imaging-surgical correlation in a classic OHVIRA subtype.

Conclusion

This case demonstrates the characteristic imaging findings of OHVIRA syndrome and emphasizes the importance of considering this diagnosis in adolescent females presenting with pelvic pain and a cystic pelvic mass, particularly in the presence of ipsilateral renal agenesis. Multimodality imaging plays a pivotal role in accurate diagnosis. Although magnetic resonance imaging remains the reference standard, contrast-enhanced CT can provide sufficient anatomic detail in acute settings to confirm the diagnosis and guide surgical management. Early recognition is essential to prevent long-term reproductive complications.

References

1. Taliento C, Rondoni A, Scutiero G, Pavone M, Mordenti M, Vizzieli G, et al. Uterus didelphys with hematocervix in the unilateral obstructed hemivagina and ipsilateral renal agenesis: a case report of Herlyn-Werner-Wunderlich syndrome. Reprod Med [Internet]. 2024 [cited 2026 Mar 28];5:197-203. Available from: https://doi.org/10.3390/reprodmed5040018

2. Burgis J. Obstructive Müllerian anomalies: case report, diagnosis, and management. Am J Obstet Gynecol 2001;185:338–44. doi: 10.1067/mob.2001.116738.

3. Devanabanda B, Chatterton B, Hassan IN, Patel J. Obstructed hemivagina with ipsilateral renal agenesis (OHVIRA) syndrome: typical presentation of a rare syndrome. Radiol Case Rep 2023;18:2437-40. doi: 10.1016/j.radcr.2023.04.016.

4. Del Vescovo RD, Battisti S, Di Paola VD, Piccolo CL, Cazzato RL, Sansoni I, et al. Herlyn-Werner-Wunderlich syndrome: MRI findings, radiological guide (two cases and literature review), and differential diagnosis. BMC Med Imaging 2012;12:4. doi: 10.1186/1471-2342-12-4.

5. Behr SC, Courtier JL, Qayyum A. Imaging ofMüllerian duct anomalies. Radiographics 2012;32:E233-50. doi: 10.1148/rg.326125515.

6. Kim HJ, Lee EJ, Chang YW, Hong SS, Hwang J, Nam B, et al. Anatomical variations, genitourinary anomalies and clinical presentations in obstructed hemivagina and ipsilateral renal anomaly syndrome: case series. J Korean Soc Radiol 2025;86:129-40. doi: 10.3348/jksr.2024.0018.

7. Zhu L, Chen N, Tong JL, Wang W, Zhang L, Lang JH. New classification of Herlyn-Werner-Wunderlich syndrome. Chin Med J (Engl) 2015;128:222-5. doi: 10.4103/0366-6999.

8. Tong J, Zhu L, Chen N, Lang J. Endometriosis in association with Herlyn-Werner-Wunderlich syndrome. Fertil Steril 2014;102:790-4. doi: 10.1016/j.fertnstert.2014.05.025.

9. Smith NA, Laufer MR. Obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) syndrome: management and follow-up. Fertil Steril 2007;87:918-22. doi: 10.1016/j.fertnstert.2006.11.015.