Case Series

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Anatomical Variations, Genitourinary Anomalies and Clinical Presentations in Obstructed Hemivagina and Ipsilateral Renal Anomaly Syndrome: Case Series

일측성 질 폐쇄 및 동측 신장기형 증후군에서의 해부학적 변이, 비뇨기계 이상 및 임상적 발현: 증례 시리즈

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Obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) syndrome is a rare Müllerian duct anomaly, commonly characterized by uterus didelphys, obstructed hemivagina, and ipsilateral renal agenesis. While these are the three most common genitourinary anomalies in OHVIRA syndrome, a spectrum of urogenital anomalies can be present. Knowledge of this spectrum is crucial for proper patient management and treatment planning. In this case series, we report on five patients with OHVIRA syndrome, each presenting with a urogenital anomaly other than the typical renal agenesis or uterus didelphys. We highlight the gynecological complications encountered, which clinicians and radiologists should be aware of.

Index terms OHVIRA Syndrome; Urogenital Anomaly; Ectopic Ureter; Classification; Vaginal Cancer

INTRODUCTION

Obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) syndrome, also known as Herlyn-Werner-Wunderlich syndrome, is a rare Müllerian duct anomaly

that was first reported by Purslow in 1922 (1). OHVIRA syndrome is usually characterized by urogenital anomalies such as uterine didelphys, blind hemivagina, and ipsilateral renal agenesis. Although these are the most common triad of urogenital anomalies of OHVIRA syndrome, there are several other important variants.

Zhu et al. (2) classified OHVIRA syndrome into two main categories based on whether the hemivagina was completely blocked or not. Each of these categories was further classified into two types. The completely obstructive hemivagina category was subdivided into one with a blinded hemivagina and another with concomitant cervicovaginal atresia. The incompletely obstructed hemivagina category was subdivided into one with partial reabsorption of the vaginal septum and another with a small communication between duplicated cervices, depending on the location of the communication (2, 3). OHVIRA syndrome with an incompletely obstructed hemivagina is not accompanied by hematocolpos or hematometra and is often diagnosed incidentally in adults because it is asymptomatic. Moreover, the literature highlights a diverse spectrum of Müllerian duct anomalies extending beyond the uterine didelphys, encompassing variations such as septate or bicornuate uterus (4). Additionally, urological anomalies including dysplastic or ectopic ureters have been documented in the reported cases (5).

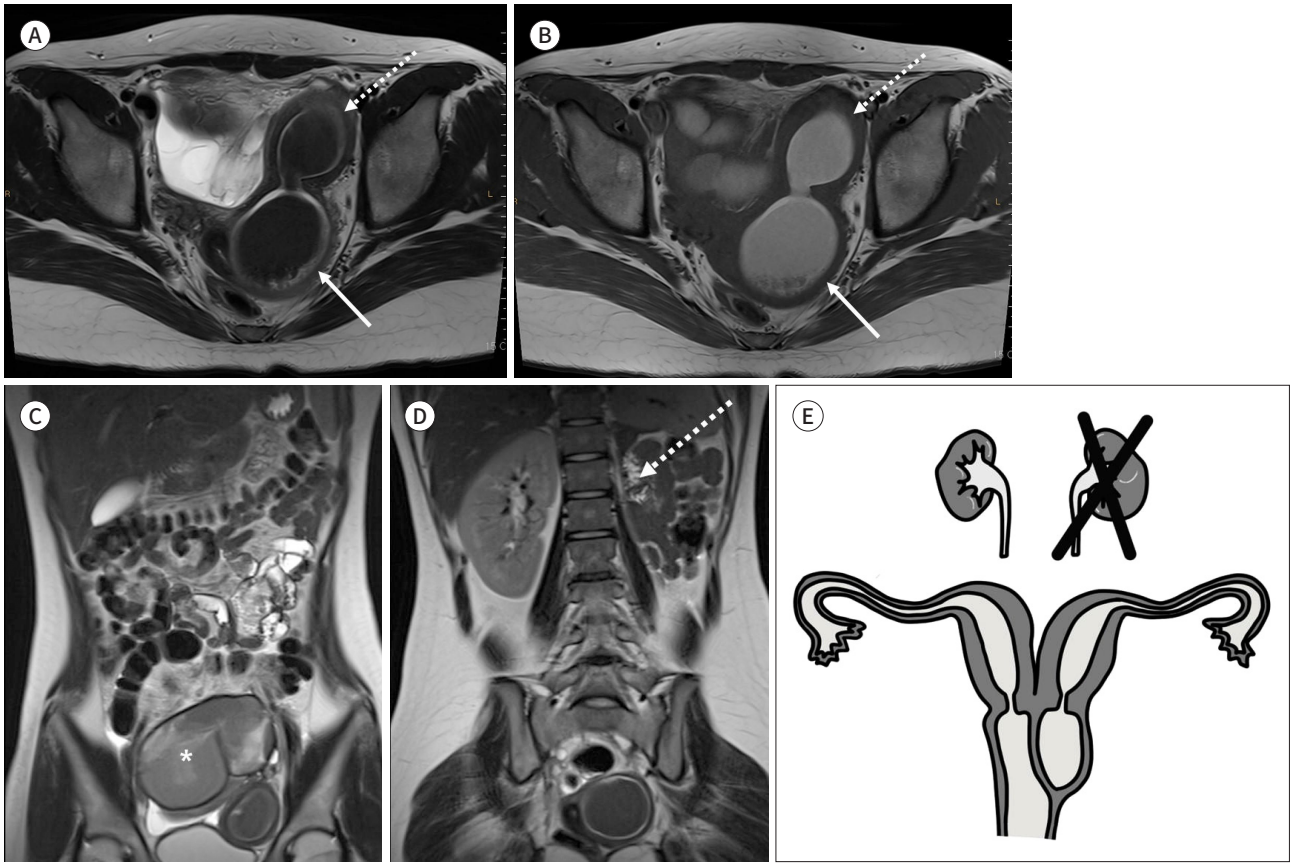
Therefore, awareness of these variants is crucial to facilitate optimal patient management and to effectively strategize treatment (6). In this report, we present our experiences with various OHVIRA syndrome anomalies and unusual but important urological anomalies associated with OHVIRA syndrome. We report cases of patients with OHVIRA syndrome, ranging from typical to rare manifestations, with gynecological complications, including malignancies, where preoperative diagnosis using imaging was difficult.

CASE SERIES

CASE 1: TYPICAL OHVIRA SYNDROME

A 12-year-old girl presented to the clinic with a medical history of menarche one year prior to her visit and persistent dysmenorrhea. The patient underwent pelvic MRI for further evaluation. Contrast-enhanced pelvic MRI revealed uterine didelphys with complete obstruction of the left hemivagina, accompanied by distension of the left upper vagina and endometrial cavity of the left uterus, which was filled with hemorrhagic fluid (Fig. 1A, B). Additionally, a dilated tubular-like structure in the left adnexa filled with hemorrhagic fluid, suggested a hematosalpinx (Fig. 1C). The left kidney could not be identified (Fig. 1D). Based on the imaging findings of the uterus didelphys, completely obstructed hemivagina resulting from hematometrocolpos and hematosalpinx, and unilateral renal agenesis, the patient was diagnosed with typical OHVIRA syndrome (Fig. 1E). Subsequently, laparoscopic left salpingectomy, removal of the left hematometra, and hysteroscopic resection of the left vaginal septum were performed. The patient was discharged with no immediate postoperative complications and remained symptom-free during outpatient follow-up visits.

Fig. 1. A 12-year-old female with dysmenorrhea (Case 1).
A, B. On pelvic MRI, axial T1 and T2 weighted images show uterine didelphys with a dilated left hemivagina (arrows), and endometrial cavity (dotted arrows) filled with T1 high and T2 low SI fluid, suggesting left hematocolpos and hematometra.
C. The dilated tubule-like structure in the left adnexa filled with heterogenous T2 low SI fluid also suggests a hematosalpinx (*).
D. The left kidney is not identified (dotted arrow).
E. The illustration shows a urogenital anomaly in Case 1, a typical finding of OHVIRA syndrome.
OHVIRA = obstructed hemivagina and ipsilateral renal anomaly, SI = signal intensity



CASE 2: A VARIANT TYPE OF OHVIRA SYNDROME WITH INCOMPLETELY OBSTRUCTIVE HEMIVAGINA

A 36-year-old female patient presented to the clinic with a complaint of intermittent abdominal pain over the past 10 years. A contrast-enhanced pelvic MRI revealed uterine didelphys, with no evidence of hematocolpos or hematometra (Fig. 2A, B). Communication between the two hemivaginas was observed (Fig. 2B, C), but the right kidney could not be identified (Fig. 2D). The patient was diagnosed with a variant of OHVIRA syndrome with communication between the duplicated hemivaginas (Fig. 2E). The patient was treated medically, resulting in resolution of symptoms.

CASE 3: A VARIANT TYPE OF OHVIRA SYNDROME WITH INCOMPLETELY OBSTRUCTIVE HEMIVAGINA WITH ECTOPIC URETER

A 20-year-old female patient presented with lower abdominal pain and turbid vaginal discharge, prompting clinical suspicion of pelvic inflammatory disease. Contrast-enhanced abdominal and pelvic CT revealed uterine didelphys with obstruction of the left hemivagina and

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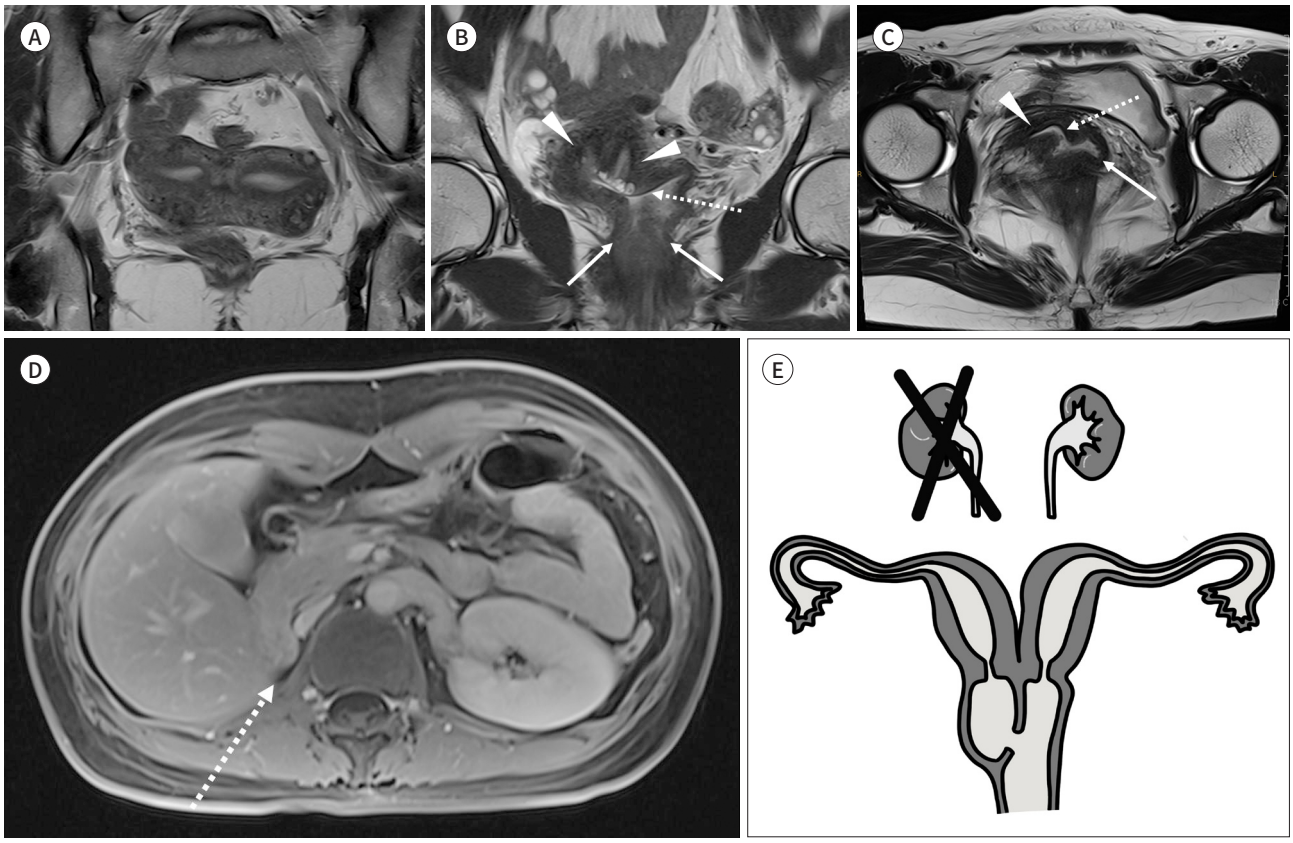
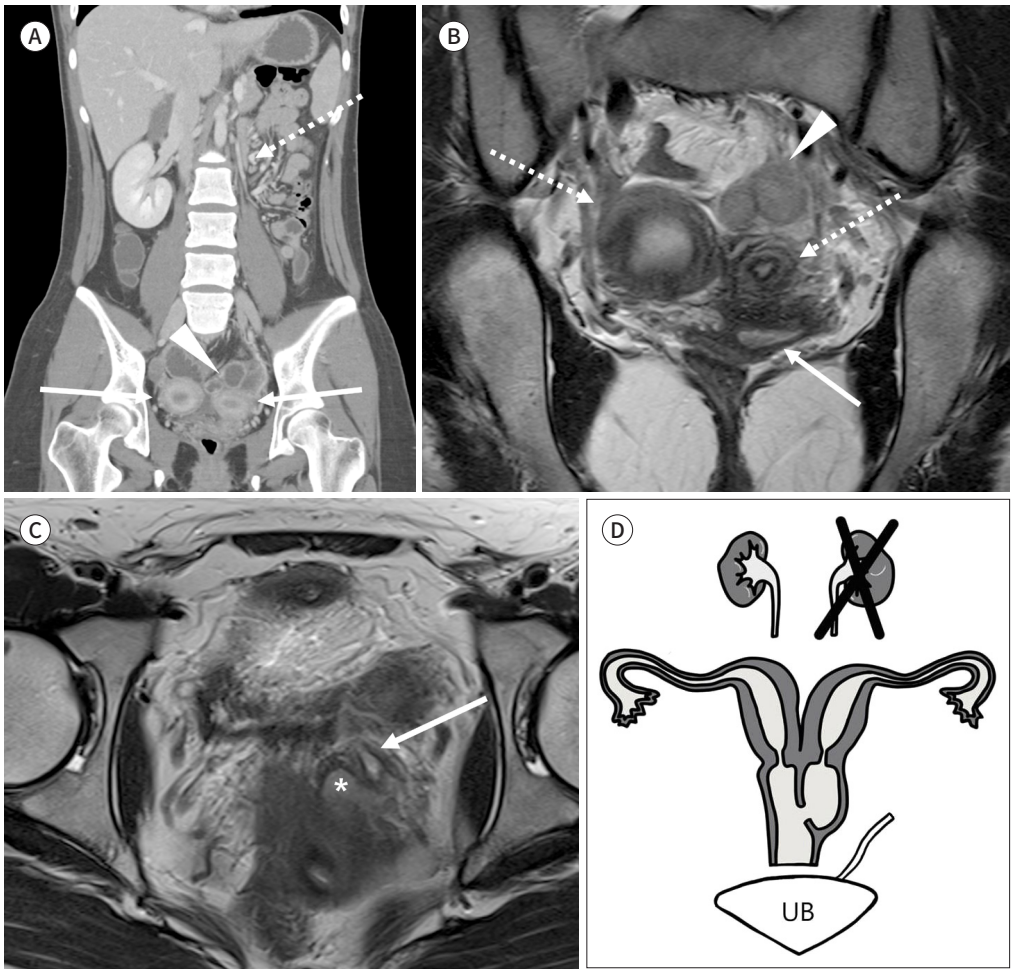
Fig. 2. A 36-year-old female with intermittent abdominal pain for 10 years (Case 2).
A, B. On pelvic MRI, the coronal T2 weighted image shows a duplicated uterine corpus (A) and cervix (arrowheads) with a duplicated vagina (arrows). A partial defect is observed in the vaginal septum of the right hemivagina (dotted arrow), suggesting communication between both hemivaginas.
C. Axial T2 weighted image also reveals a partial defect in the vaginal septum on the right hemivagina (dotted arrow), making communication possible between the hemivaginas (arrowhead, arrow).
D. The right kidney is not identified (dotted arrow).
E. The illustration shows a urogenital anomaly in Case 2.



left renal agenesis, indicative of OHVIRA syndrome (Fig. 3A). Within the left pelvic cavity, a fluid-filled dilatation with enhanced wall thickening of the left salpinx was observed, suggesting pyosalpinx (Fig. 3A). Contrast-enhanced pelvic MRI further revealed a small amount of hematocolpos in the obstructed left hemivagina and left hematosalpinx (Fig. 3B). Additionally, a small tubular cystic structure adjacent to the lateral wall of the left hemivagina and extending toward the left posterior wall of the urinary bladder with blinded proximal and distal ends suggested a remnant of the ectopic left ureter (Fig. 3C). Although MRI did not demonstrate communication between the duplicated cervix or hemivagina, a small amount of hemorrhagic fluid draining from the left hemivaginal septum was noted on vaginal speculum examination, confirming the presence of a small defect in the left hemivaginal septum (Fig. 3D). Subsequently, hysteroscopic removal of the left hemivaginal septum was performed, and the patient was discharged without any immediate complications. She has been asymptomatic during follow-up.

Fig. 3. A 20-year-old female with lower abdominal pain and vaginal discharge (Case 3).
A. The contrast-enhanced coronal CT image shows left renal agenesis (dotted arrow), uterine didelphys (arrows), and left pyosalpinx (arrowhead).
B. On pelvic MRI, a coronal T2 weighted image shows uterine didelphys with a duplicated uterus (dotted arrows) and obstructive left hemivagina (arrow) with a small amount of hematocolpos. The left hematosalpinx is also visible (arrowhead).
C. The axial T2 weighted image shows a thin-walled, fluid-filled tubular structure with a blinded proximal end (arrow) abutting the lateral wall of the obstructive left hemivagina (*).
D. This structure extends to the posterior wall of the urinary bladder with a blinded distal end, suggestive of a remnant ectopic distal ureter. The illustration shows a urogenital anomaly in Case 3.



CASE 4: VAGINAL CANCER IN A PATIENT WITH A VARIANT TYPE OF OHVIRA SYNDROME WITH INCOMPLETELY OBSTRUCTIVE HEMIVAGINA AND ECTOPIC URETER

A 48-year-old female patient presented at our hospital with lower abdominal pain and vaginal bleeding. Contrast-enhanced abdominal and pelvic CT showed right renal agenesis and uterine didelphys. A heterogeneously enhanced mass was identified in the right upper anterior vaginal wall, extending into a tubular cystic structure in the right pelvic cavity (Fig. 4A). Contrast-enhanced pelvic MRI revealed uterine didelphys, duplication of the uterine cervix and vagina with a right obstructive hemivagina, and a communicating tract between the two duplicated uterine cervices, with no evidence of hematocolpos or hematometra (Fig. 4B). No-

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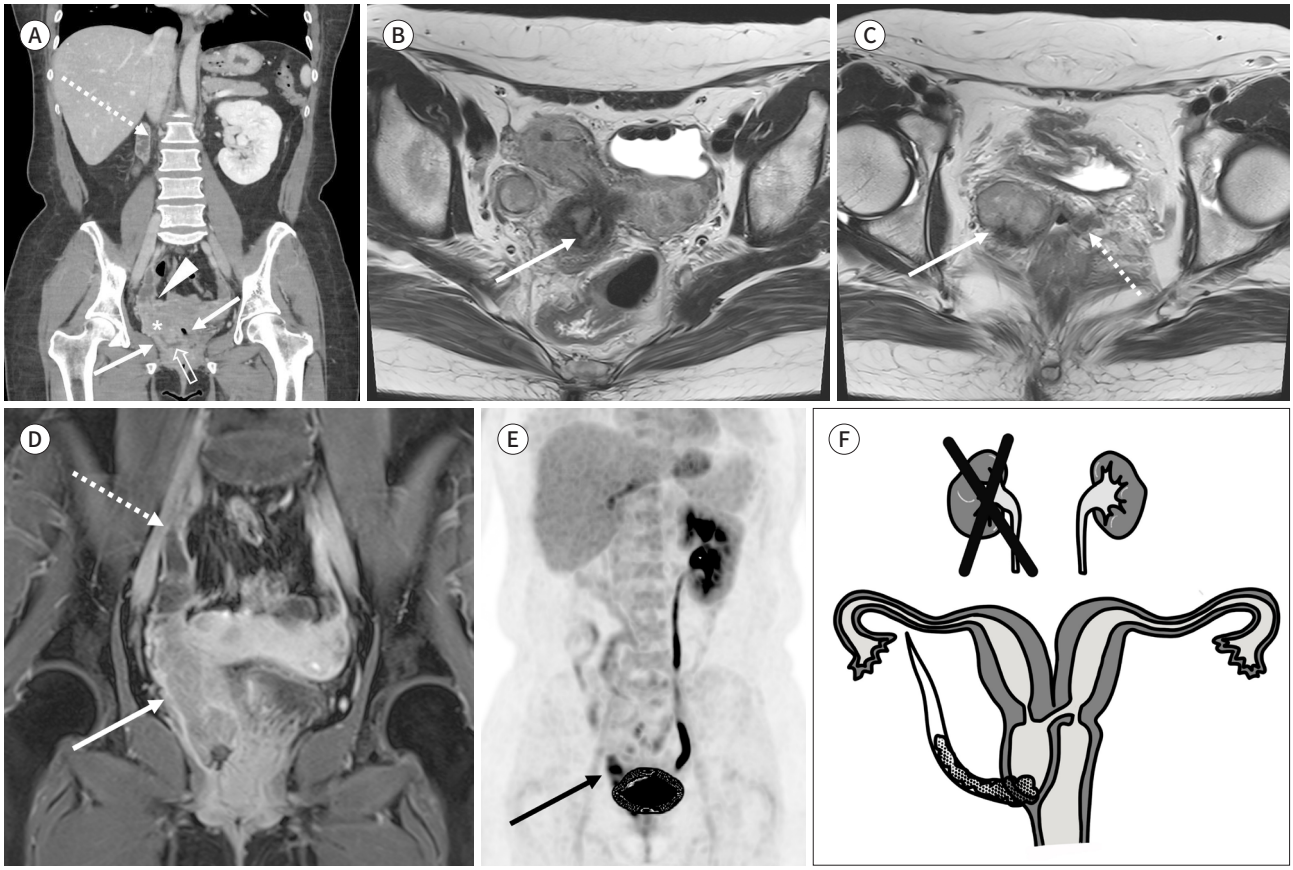
tably, there was a T2 heterogeneous signal intensity-enhancing solid lesion measuring >7 cm originating from the right obstructed hemivagina, extending into a tubular cystic structure with a blinded proximal end on the right side of the pelvic cavity (Fig. 4C, D). The lesion showed hypermetabolism on PET-CT (Fig. 4E). Speculum examination showed a 3 cm necrotic tissue at the vaginal entrance, and the lesion was diagnosed as clear cell carcinoma of the vagina on biopsy. Based on the imaging and pathological results, the patient was diagnosed with an underlying variant of OHVIRA syndrome with communicating cervices, right ectopic distal ureter, and vaginal cancer arising from the right obstructive hemivagina with tumor extension within the right ectopic ureter (Fig. 4F). The patient was subsequently transferred to another hospital for concurrent chemoradiotherapy.

Fig. 4. A 48-year-old woman with lower abdominal pain and vaginal bleeding (Case 4).
A. Enhanced coronal reformatted CT image shows right renal agenesis (dotted arrow) and uterine duplication (arrows) with an obstructive right hemivagina (empty arrow). A solid enhancing mass (*) arises from the right hemivagina and extends to the tubular structure of the right pelvic cavity (arrowhead).
B. On pelvic MRI, an axial T2-weighted image reveals a uterine duplication with an obstructive right hemivagina, and there was communication (arrow) between the duplicated uterine cervices with no evidence of hematocolpos or hematometra.
C. Masses originating from the right obstructive hemivagina (arrow) and left hemivagina (dotted arrow) are noted separately.
D. The mass shows heterogeneous enhancement (arrow) and extension into a tubular cystic structure with a blinded proximal end in the right pelvic cavity (dotted arrow), which is presumed to be a right ectopic distal ureter.
E. Maximum intensity projection image of the ^{18}F PET-CT scan showing right renal agenesis and hypermetabolism of the mass (arrow).
F. The illustration shows a urogenital anomaly in Case 4.

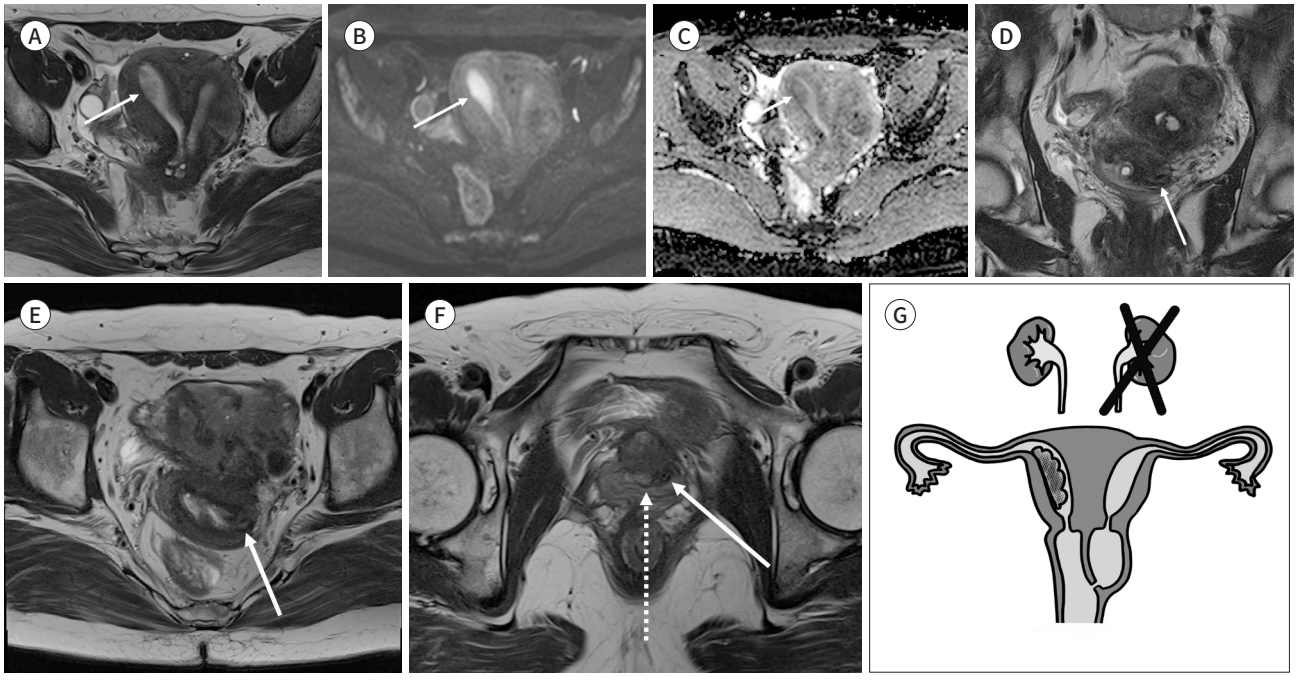


CASE 5: ENDOMETRIAL CANCER IN A PATIENT WITH A VARIANT TYPE OF OHVIRA SYNDROME WITH A COMPLETE SEPTATED UTERUS AND INCOMPLETELY OBSTRUCTED HEMIVAGINA

A 49-year-old female presented with a chief complaint of vaginal bleeding. Contrast-enhanced pelvic MRI showed a completely septate uterus. An irregular T2 intermediate signal intensity lesion was noted in the right cornu of the endometrial cavity (Fig. 5A), showing diffusion restriction (Fig. 5B, C). The left kidney was not identified on the scan of upper abdomen. Axial and coronal T2-weighted images showed a small tubular structure arising from the left separated cervical external os with a blinded distal end, suggesting an obstructed left hemivagina (Fig. 5D-F). Small communication was observed between the right hemivagina and the obstructed left hemivagina (Fig. 5F). The patient was diagnosed with OHVIRA syndrome showing a complete septate uterus with communication between the two hemivaginas concomitant with endometrial cancer (Fig. 5G). The patient underwent vaginal hysterectomy and bilateral salpingo-oophorectomy, which revealed endometrioid carcinoma and received adjuvant chemotherapy.

This study, consisting of five case presentations, was approved by our Institutional Review Board (IRB No. 2023-12-007-001). Given the retrospective nature of the study, the need for in-

Fig. 5. A 49-year-old woman with vaginal bleeding (Case 5).
A-C. On pelvic MRI, the axial T2 weighted image shows a completely septate uterus and an irregular T2 intermediate signal intensity lesion (arrow, A) in the right endometrial cavity, which shows diffusion restriction on the axial high-b-value diffusion-weighted image (arrow, B) and apparent diffusion coefficient map (arrow, C).
D-F. The coronal and axial T2 weighted image shows a small tubular structure arising from the left separated cervical external os (arrows, D and E). Axial T2 weighted image shows a small tubular structure arising from the left duplicated uterine cervix blinded to the distal end (arrow, F), suggesting an obstructed left hemivagina. There is a small communication between the right hemivagina (dotted arrow, F) and obstructive left hemivagina.
G. The illustration shows a urogenital anomaly in Case 5.



formed consent was waived.

DISCUSSION

OHVIRA syndrome is a Müllerian duct anomaly typically characterized by uterine didelphys, obstructed hemivagina, and ipsilateral renal anomalies. Although most patients with OHVIRA syndrome have uterine didelphys with obstructive hemivagina and ipsilateral renal agenesis, OHVIRA syndrome is accompanied by a variable spectrum of Müllerian duct and urological anomalies. Thus, it is important for clinicians and radiologists to understand the various anomalies of OHVIRA syndrome for appropriate diagnosis and management.

Zhu et al. (2) proposed a classification system for the OHVIRA syndrome, categorizing it into two main categories depending on whether the hemivagina was completely obstructed. In cases of typical OHVIRA syndrome, characterized by the absence of communication between the hemivagina and cervix, as observed in Case 1, the diagnosis typically occurs around the onset of menarche. It is often accompanied by hematocolpos, hematometra, and hematosalpinx due to the accumulation of menstrual blood in the obstructed side. Symptoms typically manifest a few months after the onset of menarche and present with progressively worsening abdominal pain or dysmenorrhea. The reported symptom onset and diagnosis occur at mean ages of 12.86 and 13.00 years, respectively (2). In Case 1, the onset of symptoms occurred at the age of 12 years. When communication between the hemivagina or cervix is present, the mean age at onset of symptoms and diagnosis tends to be later (21.68 and 25.74 years old, respectively) (2), as observed in our patients aged 20 to 49 years. A retrospective study of 70 patients by Tong et al. (7) also showed that the average age at diagnosis for patients with complete obstruction was 13 years, whereas, for patients with an obstructed hemivagina with communication, the mean age at presentation was notably later at 24.7 years.

The fusion of the Müllerian ducts occurs along the midline, extending in a cranio-caudal direction, and the ducts fuse and form the uterus and upper one-third of the vagina. The distal portions of these ducts merge with the urogenital sinus at a structure known as the Müllerian tubercle (6). The presumed embryonic mechanism of the Müllerian anomaly in OHVIRA syndrome is probably due to an interruption in the development of the caudal section of one of the mesonephric ducts, with subsequent effects on the corresponding ipsilateral Müllerian duct, leading to its abnormal location (8). The most common presentation of the Müllerian anomaly was uterus didelphys with blind hemivagina (72.4% of patients), followed by septate uterus with blind hemivagina (11.5%), and bicornuate uterus with blind hemivagina (10.3%). Less common variations include uterus didelphys with unilateral cervical atresia and bicornuate uterus with a septate cervix and obstructed hemivagina, accounting for less than 5% of the cases (4). The mesonephric duct on the affected side is also responsible for the absence of typical ureteral budding and renal differentiation, leading to renal agenesis, along with an abnormal location of the ipsilateral Müllerian duct. Case 5 in this case series showed a completely septate uterus with an incompletely obstructed hemivagina and ipsilateral renal agenesis. Although uterine didelphys is the most common Müllerian anomaly in OHVIRA syndrome, it is important to be aware that a wide spectrum of Müllerian anomalies can be present. While renal agenesis is the most characteristic urological anomaly associated with OHVIRA syn-

Table 1. Patient Characteristics

Patient	Age (Years)	Clinical Presentation	Urological Anomaly	Uterovaginal Anomaly	Accompanied Gynecological Diseases
1	12	Dysmenorrhea	Left renal agenesis	Uterine didelphys with obstructed left hemivagina	Absent
2	36	Intermittent abdominal pain for 10 years	Right renal agenesis	Uterine didelphys with incomplete obstruction of hemivagina (communication at vagina level)	Pelvic endometriosis
3	20	Lower abdominal pain and vaginal bleeding	Left renal agenesis, ectopic ureter	Uterine didelphys with obstructed left hemivagina (suspicious communication at vagina level)	Pelvic inflammatory disease
4	49	Lower abdominal pain and vaginal discharge	Right renal agenesis, ectopic ureter	Uterine didelphys with incomplete obstruction of hemivagina (communication at cervix level)	Clear cell carcinoma of right hemivagina
5	49	Vaginal bleeding	Left renal agenesis	Septate uterus with obstructed left hemivagina (communication at vagina level)	Endometrial cancer in right endometrial cavity

drome, accounting for approximately 95% of cases, there are also several significant variations to consider. Urological anomalies other than renal agenesis have been reported including dysplastic kidney, duplicated ureter, ectopic ureter, and polycystic kidney (6, 8, 9). Two of our patients (Cases 3 and 4) showed renal agenesis with an ectopic ureter communicating with the obstructed hemivagina. Table 1 summarizes the clinical presentations and Müllerian and urological anomalies found in our case series.

A few cases of gynecological malignancies arising from OHVIRA syndrome have been reported (10-19). The reported cases of malignancies in OHVIRA syndrome occurring in the female genital tract, including the uterus, cervix, vagina, and ovaries, are summarized in Table 2. Notably, in ten of the thirteen cases, including ours, malignancies were observed to occur ipsilateral to the obstructed side of the uterine cervix or hemivagina. Two hypotheses explaining carcinogenesis in relation to Müllerian duct anomalies have been postulated. One hypothesis suggests that the tumor may have originated in the paracervix from a columnar epithelium of Müllerian origin that has undergone genetic and hormonal changes over time (14). Another hypothesis proposes that when the blind cervix is exposed to menstrual blood over a prolonged period, basal endometrial fragments are transported into the cervix potentially leading to cancer development from endometriosis (20). In Case 5, the malignancy developed from the opposite site of the obstructed hemivagina, which can be considered a rare occurrence based on the literature reviewed so far. In Case 4, cancer originating from the hemivagina with extension into an ectopic ureter was a unique occurrence and to the best of our knowledge, this was the only case in which the tumor extended into the ectopic ureter. Awareness of the potential coexistence of various urological anomalies with OHVIRA syndrome will enable an accurate diagnosis and effective treatment of the location and extent of these complications in complex cases.

In conclusion, we emphasize the importance of understanding the different anatomical variants and urological anomalies associated with OHVIRA syndrome along with presenting several intriguing clinical cases. Despite its rarity, a comprehensive understanding of the an-

Table 2. Reported Cases of Malignancies in OHVIRA Syndrome

References	Age (Years)	Presentation	Urogenital Anomaly	Malignancy	Treatment
Watanabe et al. (2012) (10)	33	Genital bleeding	Uterine didelphys with semiobstructed right hemivagina Right renal agenesis	Endometrioid adenocarcinoma of right cervix (T4aNxM0)	Chemotherapy and RT followed by pelvic exenteration
	53	Vaginal pain and genital bleeding	Uterine didelphys with obstructed right hemivagina Right renal agenesis	Clear cell carcinoma of right vagina (T1NxM0)	RH
Kaba et al. (2013) (16)	49	Menorrhagia	Uterine didelphys with obstructive left hemivagina Dysplastic left kidney	Endometrioid adenocarcinoma of the right uterine cervix (T1b1N0M0)	TAH with BSO, PLND, PALND, omentectomy & peritoneal cytology
Cordoba et al. (2017) (17)	37	Menorrhagia	Uterine didelphys with obstructive left hemivagina Left renal agenesis	Adenocarcinonma of left cervix (T3aN1M0)	PLND + CCRT
Kusunoki et al. (2018) (14)	65	Vaginal spotting	Uterine didelphys with obstructive right hemivagina Right renal agenesis	Clear cell carcinoma of right uterine cervix (T1b2N0M0)	Laparoscopic TH with BSO + adjuvant CCRT
Oka et al. (2020) (12)	38	Dysfunctional uterine bleeding	Uterine didelphys with obstructive left hemivagina (S/P, marsupialization 16 YA) Left renal agenesis	Endocervical adenocarcinoma of left uterine cervix with left vaginal wall invasion (T2a1N1M0)	RH + adjuvant CCRT
Mei et al. (2020) (13)	40	Vaginal bleeding	Uterine didephyis with obstructive left hemivagina Left renal agenesis	Clear cell carcinoma of left vagina (T1N0M0)	Radical surgery + adjuvant CCRT
Tanase et al. (2021) (11)	52	Vaginal spotting	Uterine didelphys with obstructive left hemivagina Left renal agenesis Ectopic left ureter	Clear cell carcinoma of left uterine cervix (T1b1N0M0)	RH with BSO and PLND
Almulhin et al. (2021) (18)	29	Infertility	Uterine didelphys with obstructive right hemivagina Right renal agenesis	Serous borderline tumor of right ovary (T1aNxM0)	Laparoscopic right ovarian cystectomy
Kobayashi et al. (2021) (15)	58	Genital bleeding	Uterine didelphys with obstructive right hemivagina Atrophic right kidney	Endometrioid carcinoma of left uterine corpus (T3aN0M0)	Robot-assisted hysterectomy with BSO and PLND
Mabuchi et al. (2022) (19)	74	Vaginal bleeding	Uterine didelphys with obstructed left hemivagina Left renal agenesis	Clear cell carcinoma of left cervix (T2a1N0M0)	RH with BSO and PLND
Present case (Case 4)	48	Lower abdominal pain and vaginal bleeding	Uterine didelphys with obstructive right hemivagina Right renal agenesis Ectopic right ureter	Clear cell carcinoma of right hemivagina with extension to right ectopic ureter	Primary CCRT
Present case (Case 5)	49	Vaginal bleeding	Septated uterus with incompletely obstructed left hemivagina Left renal agenesis	Endometrioid carcinoma of right uterine corpus (T1aN0M0)	VH with BSO + adjuvant chemotherapy

BSO = bilateral salpingo-oophorectomy, CCRT = concurrent chemoradiation therapy, OHVIRA = obstructed hemivagina and ipsilateral renal anomaly, PALND = paraaortic lymph node dissection, PLND = pelvic lymph node dissection, RH = radical hysterectomy, RT = radiation therapy, TAH = total abdominal hysterectomy, TH = total hysterectomy, VH = vaginal hysterectomy, YA = year ago

atomical intricacies and clinical nuances of OHVIRA syndrome is crucial for prompt intervention.

Author Contributions

Conceptualization, L.E.J., K.H.J.; data curation, C.Y., H.S.S.; formal analysis, K.H.J., H.J.; investigation, L.E.J.; methodology, N.B.; resources, B.S.H.; software, K.H.J.; supervision, L.E.J.; visualization, K.H.J., H.J.; writing—original draft, K.H.J.; and writing—review & editing, L.E.J., H.S.S.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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일측성 질 폐쇄 및 동측 신장기형 증후군에서의 해부학적 변이, 비뇨기계 이상 및 임상적 발현: 증례 시리즈

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일측성 질 폐쇄 및 동측 신장기형 증후군(obstructed hemivagina and ipsilateral renal anomaly; 이하 OHVIRA)은 중복자궁, 반질 폐쇄, 동측 신장 무형성을 특징으로 하는 희귀한 뮐러관 이상 중 하나이다. 이 세 가지는 OHVIRA 증후군에서 가장 흔한 세 가지 구성요소이지만, 다양한 형태의 비뇨생식기계 이상이 존재할 수 있다. 따라서 OHVIRA 증후군의 다양한 형태에 대한 지식은 환자의 적절한 관리 및 치료 계획을 세우는데 중요하다. 본 증례 시리즈에서는 일반적인 신장 무형성이나 중복 자궁 이외의 비뇨생식기 이상을 동반한 다섯 명의 OHVIRA 증후군 환자를 살펴보고 임상의사 및 영상의학과 의사가 주의해야 할 여성 생식기 합병증을 가진 환자의 증례를 제시하고자 한다.

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