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Case Report

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Herlyn-Werner-Wunderlich Syndrome Complicated by Right Ectopic Atrophic Kidney: A Case Report from a Primary Hospital

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Abstract

Background: Herlyn-Werner-Wunderlich syndrome (HWWS), also termed obstructed hemivagina and ipsilateral renal anomaly (OHVIRA), is a rare congenital Müllerian duct anomaly frequently accompanied by structural abnormalities of the ipsilateral kidney and ureter. Owing to its nonspecific clinical features, which often overlap with common acute abdominal conditions such as pelvic inflammatory disease and appendicitis-HWWS is frequently misdiagnosed or missed in primary healthcare settings, particularly among post-menarchal adolescents with atypical symptoms. This case report presents a 9-year-old girl with HWWS and ipsilateral ectopic atrophic kidney associated with an ectopic ureteral orifice, and discusses the diagnostic reasoning, stepwise imaging strategy, and multidisciplinary management experience gained at a primary hospital.

Case presentation: A 9-year-old girl presented with intermittent lower abdominal distending pain for 15 days. Menarche had occurred four months earlier; menses were irregular, without cyclic dysmenorrhea or vaginal purulent discharge. Initial pelvic ultrasonography demonstrated a mixed echogenic pelvic mass and absent right kidney. Subsequent contrast-enhanced pelvic MRI and CT urography (CTU) confirmed the diagnosis of type I HWWS, characterized by uterus didelphys with double cervix, right vaginal atresia with hematometra and hematocolpos, and a right ectopic atrophic kidney with an ectopic ureteral orifice. After multidisciplinary consultation involving gynecology, urology, and general surgery, the patient underwent combined hysteroscopic resection of the right vaginal oblique septum, laparoscopic resection of the ectopic right kidney and ureter, and cystoscopy. The intraoperative course was uneventful. Postoperative anti-infective therapy and vaginal care were provided; the patient recovered satisfactorily and was discharged on postoperative day 10 without abnormal vaginal discharge.

Conclusion: HWWS exhibits heterogeneous and often atypical manifestations in adolescent girls.

Primary-care physicians should maintain a high index of suspicion and adopt an integrated genitourinary diagnostic mindset. A stepwise imaging strategy combining ultrasonography, MRI, and CTU can significantly improve early recognition. Combined minimally invasive surgery under multidisciplinary collaboration is safe and feasible, and may serve as a useful reference for the management of complex congenital genitourinary malformations in primary hospitals.

Keywords: Herlyn-Werner-Wunderlich syndrome(HWWS); OHVIRA syndrome; ectopic ureteral orifice; renal dysgenesis; case report

1. Introduction

Herlyn-Werner-Wunderlich syndrome (HWWS) is a rare congenital genital tract anomaly resulting from coordinated dysplasia of the Müllerian and Wolffian ducts during embryogenesis. Its reported incidence ranges from 0.1% to 3.8%[1, 2],and approximately 85% of patients have concomitant ipsilateral urinary tract anomalies, among which ipsilateral renal agenesis is the most common[3].The clinical manifestations are insidious and atypical. Coupled with limited awareness among primary-care physicians, misdiagnosis or missed diagnosis at the initial visit exceeds 60%[4],and the condition is frequently misidentified as common acute abdominal entities such as acute pelvic inflammatory disease, appendicitis, or pelvic abscess. Delayed diagnosis and treatment may lead to secondary pelvic infection, endometriosis, and ultimately impaired fertility[5].This report describes a 9-year-old post-menarchal girl admitted with nonspecific lower abdominal pain for 15 days. She had experienced only four menstrual cycles since menarche and had not yet established a regular cycle. She lacked the characteristic features of HWWS, namely, cyclic lower abdominal pain and vaginal purulent discharge-and presented instead with intermittent distending pain accompanied by anal heaviness, which partially resolved after defecation. These features overlapped considerably with those of common acute abdominal conditions, rendering diagnostic confusion highly likely. Drawing on the diagnostic and therapeutic course of this case, we discuss the clinical reasoning and management strategy employed by a primary hospital in dealing with this rare disorder.

2. Case presentation

A 9-year-old girl was admitted to the Department of Gynecology, Dafang County People's Hospital, Guizhou Province, China, with a 15-day history of intermittent lower abdominal pain. Menarche had occurred in October 2025 at the age of 9. She had experienced four menstrual periods, with irregular cycles and scant flow; occasional lower abdominal heaviness during menses had been noted but had not

previously been evaluated.

15 days before admission, the girl developed lower abdominal pain without an identifiable precipitating factor. The pain was intermittent and distending, with paroxysmal exacerbation, accompanied by anal heaviness and occasional dysuria. Symptoms were partially relieved after defecation and micturition, with no other notable discomfort.

She initially presented to the general surgery outpatient clinic. Pelvic ultrasonography demonstrated a mixed echogenic mass in the pelvis measuring approximately 117 mm x 67 mm x 85 mm, with irregular shape, well-defined margins, and peripheral blood flow signals. The right kidney was not visualized.

To further characterize the lesion, contrast-enhanced pelvic MRI was performed. It revealed a uterus didelphys with double cervix and double vagina. The distal segment of the right vagina was atretic; above the atretic segment, the vagina and uterine cavity contained large areas of short T1 and long T2 signal, with locally continuous serpiginous short T1/long T2 signal. The lesion was hyperintense on DWI and slightly hypointense on ADC maps, measuring approximately 11.3 cm x 6.6 cm on sagittal images, with no significant enhancement after contrast administration. The left uterine cavity, cervix, and vagina showed no abnormal enhancement. The right kidney was not visualized, and the right ureter was found to communicate with the right vagina. A small amount of pelvic fluid was noted (**Figure 1**).

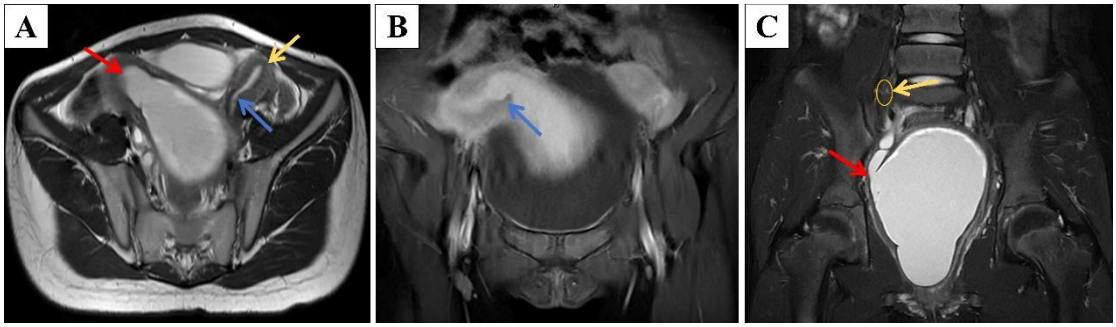


Figure 1: Contrast-enhanced pelvic MRI (plain and enhanced sequences). (A) The red arrow indicates the right uterine horn, the yellow arrow indicates the left uterine horn, and the blue arrow indicates the position of the left cervix. (B) The blue arrow indicates the position of the right cervix. (C) The red arrow indicates the communication between the right ureter and the vagina; the yellow circle and arrow indicate the atrophic right kidney.

After gynecologic admission, physical examination showed normal external genitalia. A pelvic-abdominal mass was palpable, with its upper pole extending between the umbilicus and the pubic symphysis. Rectal examination revealed a soft mass anterior to the rectum in the lower abdomen, with its lower margin approximately 3 cm from the vaginal introitus. The initial diagnosis was: (1) type I

HWWS; (2) suspected right renal agenesis; (3) ectopic ureteral orifice.

To delineate the anatomy of the right kidney and ureter, CT urography (CTU) was performed. It confirmed the didelphic uterus, double cervix, and double vagina, with distal atresia of the right vagina and a large fluid collection above it measuring approximately 11.3 cm x 8.9 cm x 7.6 cm, showing no enhancement. In the excretory phase, the right kidney and the entire right ureter were not opacified. The left kidney was compensatorily enlarged with good excretory function, and the left ureter was well visualized. The bladder was well distended without significant wall thickening (**Figure 2**).

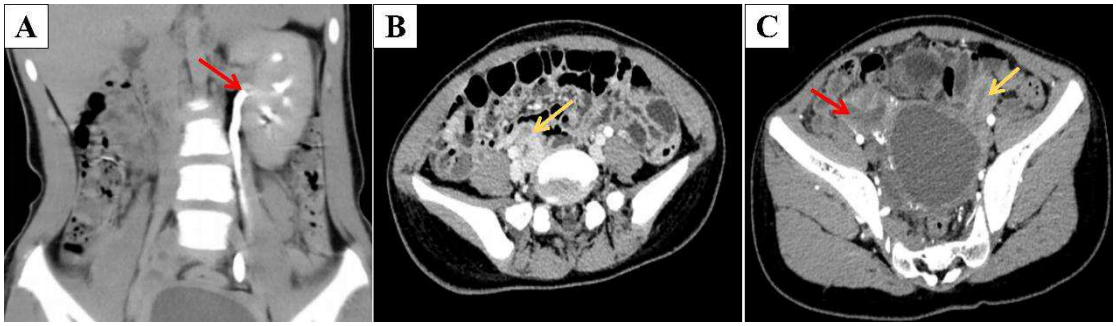


Figure 2: Urinary CT urography (CTU). (A) The red arrow indicates the opacified left kidney and ureter, with no opacification of the contralateral kidney and ureter. (B) The yellow arrow indicates the malformed, atrophic right kidney located in a lower ectopic position. (C) The red arrow indicates the right uterus, and the yellow arrow indicates the left uterus.

After multidisciplinary consultation involving gynecology, general surgery, and urology, the patient underwent hysteroscopic resection of the vaginal oblique septum combined with laparoscopic right nephroureterectomy and cystoscopy.

Hysteroscopy showed a patent left vagina with a normal left cervical os. The right vaginal wall appeared cystically bulging without an orifice. After adequate exposure, the distal aspect of the cyst was incised with a resecting loop, releasing dark, old blood. The vaginal oblique septum was then progressively resected to the level of the left cervical os, revealing a markedly dilated right cervix. Exploration of the right uterine cavity showed no polypoid lesions, and the right ureteral orifice was visualized; the left uterine cavity also showed no polypoid lesions, with the left ureteral orifice visible. Repeated exploration of the right vaginal wall and vaginal vault identified no ureteral orifice, which was attributed to closure of the ectopic right ureteral orifice following decompression of the vagina (**Figure 3**).

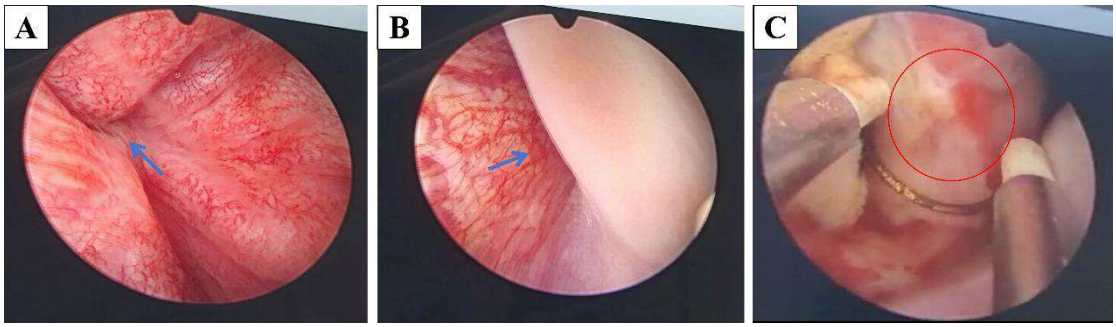


Figure 3: Hysteroscopic views. (A) The blue arrow indicates the left cervical os. (B) The blue arrow indicates the right cervical os. (C) The red circle indicates the vaginal oblique septum.

Cystoscopy demonstrated smooth urethral and bladder mucosa without polyps. The left ureteral orifice was clearly visible with normal urine efflux. No right ureteral orifice was identified, and no ectopic ureteral orifice was found elsewhere in the bladder or urethra.

Laparoscopy showed a bicornuate-appearing uterus. A muscular duct was identified closely adjacent to the right cervix. Dissection along this duct led to an irregular mass measuring approximately 3 cm x 2 cm, located medial to the bifurcation of the right internal and external iliac vessels and below the right infundibulopelvic ligament. The mass was well vascularized; in view of the preoperative imaging findings, it was considered an atrophic right kidney and was completely excised and sent for histopathological examination. Histopathological examination confirmed an atrophic kidney with glomerular sclerosis and extensive hemosiderin deposition, consistent with a long-standing ectopic dysplastic kidney. Dissection was continued distally along the mass and the muscular duct to the vaginal vault. The ureteral orifice at the vaginal vault was sutured closed, and the ectopic ureter was transected. Old blood was identified within the duct lumen, consistent with an ectopic ureter (**Figure 4**).



Figure 4: Laparoscopic views. (A) The red arrow indicates the ectopic atrophic right kidney, the yellow arrow indicates the ectopic ureter, and the blue arrow indicates the uterus. (B) The completely resected right atrophic kidney specimen. (C) Histopathological section of the right atrophic kidney (hematoxylin and eosin staining), showing glomerular atrophy and extensive hemosiderin deposition.

The procedure was completed uneventfully. Postoperatively, the patient received anti-infective therapy and vaginal care. On postoperative day 10, she was in good general condition without abnormal vaginal discharge and was discharged. At the 3-month follow-up visit, the patient reported regular menstrual cycles without dysmenorrhea or abnormal vaginal discharge. Pelvic ultrasonography showed no recurrent hematometra or hematocolpos, and the left kidney maintained normal compensatory morphology and function.

3. Discussion

HWWS is a rare Müllerian duct anomaly whose clinical manifestations are nonspecific and substantially overlap with those of common acute abdominal conditions such as pelvic inflammatory disease and appendicitis, contributing to a persistently high misdiagnosis rate in primary healthcare settings[4].Delayed diagnosis and treatment may predispose patients to pelvic infection, endometriosis, and pelvic adhesions, ultimately impairing fertility[5].Based on the anatomical morphology of the vaginal oblique septum, the syndrome is internationally classified into three types. Type I (imperforate oblique septum) represents complete atresia of one hemivagina, with the obstructed uterus completely isolated from the external environment and the contralateral uterus; menstrual blood accumulates in the uterine cavity and vagina behind the septum. Type II (perforate oblique septum) features a small fistulous opening in the septum, resulting in inadequate drainage. Type III (imperforate septum with cervical fistula) involves a fistulous tract between the cervices[6].In a series of 102 patients with HWWS, type I accounted for 36.27%, type II for 49.02%, and type III for 14.71%; patients with type I disease presented predominantly with dysmenorrhea and had the earliest diagnosis and shortest disease course. In the present case, the girl's initial complaint was nonspecific lower abdominal pain. Because her menstrual cycle had not yet become regular after menarche, she lacked the characteristic cyclic dysmenorrhea and purulent vaginal discharge, and this atypical presentation further complicated early recognition, particularly in a primary-care setting.

A notable reflection on the diagnostic course of this case is that the initial ultrasound finding of "absent right kidney" did not prompt sufficient suspicion of a concomitant genital tract malformation[7].In routine primary practice, unilateral renal agenesis is often regarded as an isolated congenital variant, and its intrinsic association with pelvic masses and abdominal pain may be overlooked; this was an important factor contributing to the delayed diagnosis in this patient[8].Embryologically, the Müllerian and Wolffian ducts originate in close proximity and develop under mutual inductive influence, and the two types of

malformations frequently coexist[9].Establishing an integrated “genitourinary” diagnostic mindset during imaging interpretation may therefore be the most cost-effective approach to reducing missed diagnosis of this rare condition at the primary-care level. Guided by this principle, the present case employed a stepwise imaging strategy. Pelvic ultrasonography served as the initial screening modality, adequately demonstrating uterus didelphys, double cervix, vaginal hematocolpos, and renal agenesis[2],although it provided limited detail regarding the precise location of the ectopic atrophic kidney, the abnormal ureteral course, and the classification of the septum. Once suspicious findings were identified on ultrasound, contrast-enhanced pelvic MRI was performed. Its high soft-tissue resolution clearly delineated the location and thickness of the septum, the extent of blood collection, and uterine cavity morphology, and accurately localized the ectopic ureteral orifice, thereby providing reliable anatomical grounds for surgical planning[10].In cases associated with urinary tract anomalies, CT urography should be further performed whenever feasible to comprehensively assess the location, size, vascular supply, and function of the ectopic kidney, and to exclude potential complications such as ureterovaginal fistula[11].In this patient, MRI established the core diagnosis of right vaginal atresia with hematometra and hematocolpos, while CTU further characterized the ectopic atrophic kidney, providing robust imaging support for the subsequent combined surgical procedure.

The core treatment principle of HWWS is to resect the vaginal oblique septum as early as possible, adequately drain the retained menstrual blood, relieve genital tract obstruction, and preserve fertility to the greatest extent. In patients with a nonfunctioning ipsilateral kidney or an ectopic atrophic kidney, simultaneous nephroureterectomy is recommended to avoid long-term complications such as recurrent pelvic infection and ureteral stump syndrome[12, 13].Guided by this principle, the present case underwent preoperative multidisciplinary consultation and a combined procedure consisting of hysteroscopic resection of the vaginal oblique septum, laparoscopic resection of the ectopic right atrophic kidney and ureter, and cystoscopy. Hysteroscopic resection under direct vision allows maximal preservation of the normal vaginal wall and cervical architecture; particularly in adolescents without sexual activity who require preservation of hymenal integrity, this approach offers the advantages of minimal trauma and preservation of the hymen. Laparoscopic resection of the ectopic atrophic kidney further provides a minimally invasive approach with a clear surgical field and rapid postoperative recovery. Routine intraoperative cystoscopy delineates the position and morphology of both ureteral orifices, effectively preventing iatrogenic injury to the contralateral ureter. In the present case, intraoperative hysteroscopy

initially failed to identify the right ureteral orifice, which was attributed to closure of the ectopic orifice after vaginal decompression and drainage of retained blood. This finding underscores the importance of careful exploration and preoperative imaging localization; relying solely on intraoperative cystoscopy may miss an ectopic ureteral orifice that has already closed. Additionally, the patient experienced menarche at the age of 9, which is earlier than the average menarche age in the general Chinese population. Endocrine evaluation revealed no evidence of central precocious puberty, and breast development was consistent with Tanner stage 3; the early menarche was considered within the normal variation range and did not alter the diagnostic approach.

Adolescent girls presenting after menarche with lower abdominal pain or a pelvic mass should raise a high index of suspicion for HWWS. Once a uterine anomaly is identified in the presence of renal developmental abnormality, a systematic evaluation of the entire urinary tract should be performed to exclude ectopic ureteral orifices and other associated malformations, so as to reduce misdiagnosis. The management of such complex genitourinary malformations requires multidisciplinary collaboration among gynecology, urology, and radiology; the multidisciplinary team model is a cornerstone of successful management of these complex rare conditions at the primary-care level. This experience suggests that primary-care institutions should proactively transcend departmental boundaries and integrate multiple specialties when confronted with cross-disciplinary complex cases; when necessary, linkage with upper-tier hospitals through a medical alliance can complement the limitations of a single department through team-based collaboration.

The combination of HWWS with an ectopic atrophic kidney is clinically rare, and the initial misdiagnosis rate in primary hospitals is generally high. Reviewing the diagnostic and therapeutic course of this case, although primary hospitals face objective constraints related to equipment, technical expertise, and experience in the management of rare diseases, they also possess a unique advantage: as the front line of the hierarchical medical system, primary institutions often encounter patients at an earlier stage of disease. The practical takeaway from this case is that unilateral renal agenesis or ectopic dysplastic kidney in a pubertal girl should prompt systematic evaluation of the genital tract, even when symptoms are nonspecific.

4. Conclusion

This report presents a rare case of type I HWWS complicated by ipsilateral renal dysgenesis and an ectopic ureteral orifice opening into the vagina. The diagnosis was established through a stepwise

imaging workup combining ultrasonography, MRI, and CTU. Following multidisciplinary consultation, the patient underwent hysteroscopic resection of the vaginal oblique septum combined with laparoscopic nephroureterectomy, and she recovered uneventfully. For adolescent girls presenting after menarche with lower abdominal pain or a pelvic mass, systematic evaluation of concomitant genital tract and urinary tract anomalies should be emphasized.

Statements and Declarations

Funding

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Competing Interests

The authors declare that they have no competing interests.

Author Contributions

DHM guided the conception and design of the study. DHM , JW and YD collected clinical data and figures. DHM was responsible for writing the draft. DHM and JW revised the manuscript. DHM, JW and YD confirm the authenticity of all the raw data. All authors have read and approved the final version of the manuscript.

Ethics approval

The requirement of ethical approval was waived by the Institutional Ethics Committee of Dafang County People's Hospital, Guizhou Province, for this case report. This waiver was granted because the study is a retrospective observational report of a single clinical case, and all examinations and treatments involved were part of routine clinical practice without additional invasive interventions or extra biological sample collection.

Consent to participate

The study was conducted in accordance with local legislation and institutional requirements.Written informed consent was obtained from the parents for participation in the study.

Consent to publish

Written informed consent was obtained from the parents for participation in the study and for publication of the case details and accompanying images.

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