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Case Report

Ectopic urethral opening in herlyn-werner-wunderlich syndrome: A rare finding ☆

Eesha Akhlaque[a], *Muhammad Faateh Hassan*[a,*], *Irum Akhlaque*[b]

[a] *Department Of Radiology, Lahore Medical and Dental College, Tulspura, North Canal Rd, Lahore, Punjab, Pakistan*
[b] *Department of Radiology, Mayo Hospital, King Edward Medical College, Neela Gumbad Chowk, Lahore, Punjab, Pakistan*

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ABSTRACT

Recorded findings of Herlyn-Werner-Wunderlich Syndrome (HWWS) remain sparse due to low statistical incidence. Ours is also the first known case to present with an ectopic urethral opening, an even rarer complication. A 27-year-old nulliparous woman presented with complaints of cyclical hematuria, and burning micturition with intermittent episodes of urinary retention suprapubic and pelvic pain. After extensive radiological examination, the MRI findings concluded the presence of a uterine didelphys with 2 cervixes and vaginal duplication marked by the compression of the left hemivagina and dilation of the right hemivagina, pointing towards HWWS. The case strongly suggests that the presence of an ectopic urethral opening may be the cause of urinary incontinence and discomfort in an otherwise asymptomatic patient of HWWS.

Introduction

Congenital malformations of the female genital tract, such as those pertaining to the Mullerian ducts, occur in about 2-3% of the global population with the Herlyn-Werner-Wunderlich Syndrome (HWWS) being the least common [1]. The HWWS, also referred to as the obstructive hemivagina and ipsilateral renal agenesis (OHVIRA) syndrome is a rare congenital anomaly defined by a triad of uterus didelphys, obstructed hemivagina and ipsilateral renal agenesis [2]. While most patients show symptoms following the onset of menarche, many remain asymptomatic. Due to the statistically low incidence of this condition, its presenting complaints, such as altered menstrual characteristics due to vaginal obstruction, may be misdiagnosed [3]. The resulting complications may alter the patient's quality of life, and it is therefore vital to recognize, interpret, and manage these clinical manifestations. We present to you the radiological diagnosis of Herlyn-Werner-Wunderlich Syndrome with ectopic urethral opening, an even rarer com-

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* Corresponding author.
E-mail address: syedmuhammadfaateh@gmail.com (M.F. Hassan).

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plication, in a young female after she experienced chronic urinary retention and hematuria. We obtained written informed consent from the patient of this case report for the content and the accompanying images.

Case history

A 27-year-old nulliparous female reported to the outpatient department of Mayo Hospital in October 2022. She complained of cyclical hematuria for the past eleven years, burning micturition with intermittent episodes of urinary retention for the past 3 years, and suprapubic and pelvic pain for the past 6 months.

The age of menarche for the patient was fourteen years with regular menstrual periods. She had cyclical blood-mixed menstruation, as her urine mixed with her regular menstrual blood, which she considered normal. She consulted a physician when there was a sudden onset of moderately intense pelvic pain, with the symptoms as mentioned above.

Apart from a hypoplastic urethra, there were no extraordinary findings upon examination.

For investigation, first, an ultrasound was carried out. The left kidney was not visualized and the sonogram showed uterus didelphys. This was followed by an intravenous urogram (IVU) where the left kidney and ureter were not opacified. The concerned radiologists then concluded that the said kidney was absent.

An MRI of the pelvis without contrast showed multiple findings. The study was multiplanner and multisequential. Two widely spaced uterine bodies, cervices, and vaginas were visualized in the axial imaging as seen in Fig. 1. Additionally, the vaginal septum was visualized in coronal T2 section images as seen in Fig. 2. The right uterus measured 5.7 × 3.1 × 2.9 cm and the left uterus measured 5 × 2.9 × 2.7 cm. Both the uteri showed normal endometrial myometrial ratio, with normal zonal anatomy. There was minimal endometrial and cervical fluid in both uteri with hematometrocolpos.

There was a duplication of the vagina with the right hemivagina mildly dilated with low T1, high T2, and STIR signal intense contents. The left hemivagina appeared compressed, raising the possibility of a partial vaginal septum associated with obstruction.

Both ovaries showed follicular activity and were normal in size with no discrete cystic adnexal masses in the study. There was no evidence of peritoneal free fluid adenopathy in the visualized pelvis. The urinary bladder was partially distended but without any evidence of focal lesions or defective filling. There were no remarkable findings in the visualized small and large bowel loops or any associated gut mass lesions.

The signal voids and pelvic vessels appeared normal, and the pelvic wall soft tissues and bilateral ischio-rectal fossae were clear. The surrounding tissues were also unremarkable.

The MRI concluded that there was a uterine didelphys with 2 cervixes and vaginal duplication marked by the compression of the left hemivagina and dilation of the right hemivagina. The possibility of a partial vaginal septum and associated obstruction with left renal agenesis pointed to the possibility of HWWS. This was a type 2.2 presentation with an incompletely obstructed hemivagina and communicating uteri, as per the classification shown in Table 1.

Keeping the history in mind, a cystoscopy was then advised. The scope was entered into the bladder, and the guide wire and foley passed. The guide wire was then removed. The procedure and subsequent study revealed a urethral opening on the anterior wall of the vagina, almost 1 cm proximal to the external vaginal orifice.

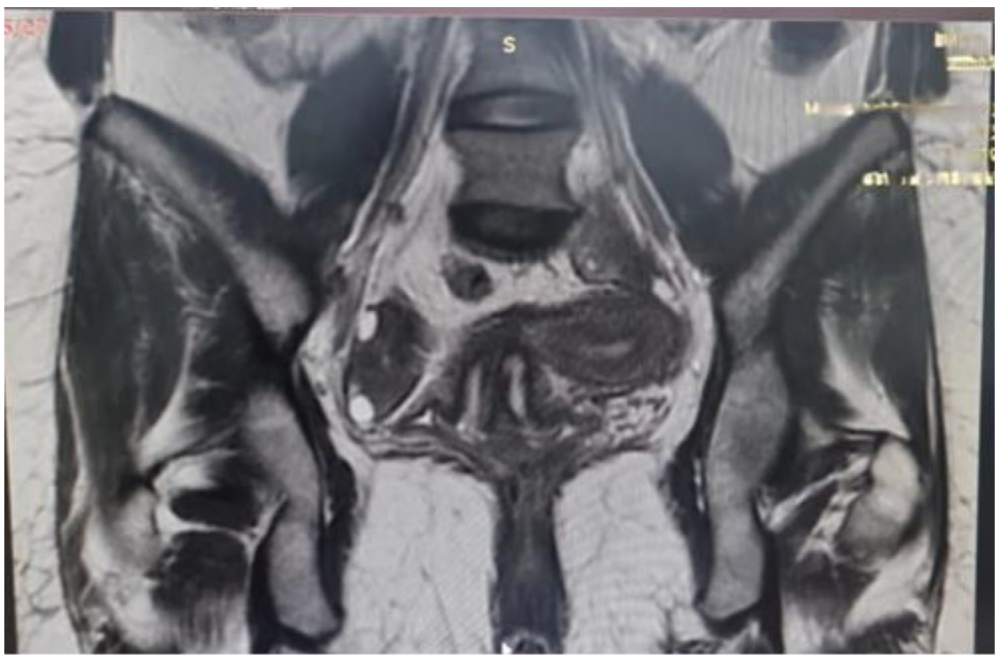

Fig. 1 – MRI: Two widely spaced uterine bodies, cervices, and vaginas on axial T2 images.

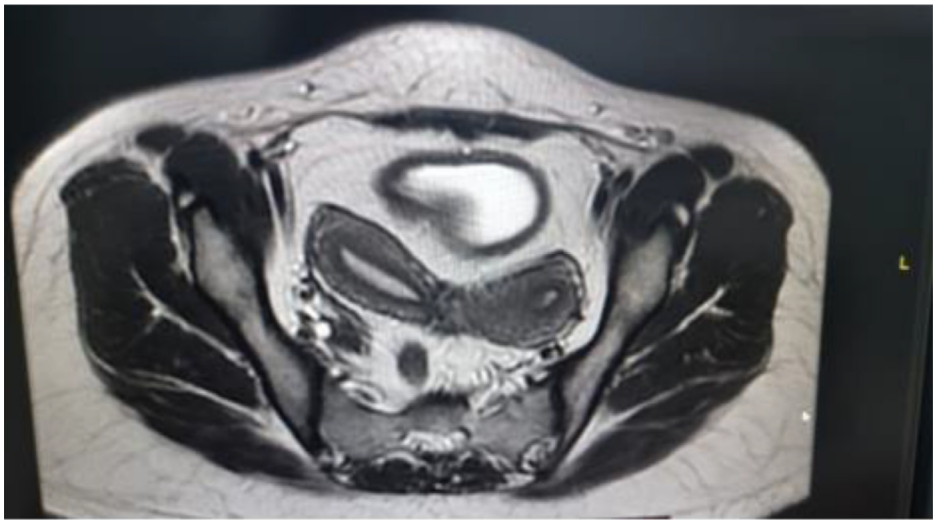

Fig. 2 – MRI: Two vaginas having vaginal septum were seen on coronal T2 section images.

Table 1 – Herlyn-Werner-Wunderlich syndrome.

Class	Class I		Class II	
	Completely obstructed hemi vagina		Incompletely obstructed hemi vagina	
Subclass	1.1	1.2	2.1	2.2
	Blind hemivagina	Cervicovaginal atresia without communicating Uteri	Partial resorption of the vaginal septum	With communicating Uteri

Note: brief classification of HWWS.

Discussion

While primarily a Mullerian duct development abnormality, recent genomic studies have found some association with renal agenesis-related genes such as RET, WNT4, and TRIM32 [4] in affected patients. The incidence of this abnormality is reported to be between 0.1% and 3.8% [5]. Early-stage diagnosis and treatment are usually enough to relieve symptoms and preserve fertility in affected patients. Therefore, radiological investigations play a vital role in this regard: MRI of the pelvis remains the gold standard diagnostic tool for HWWS patients with other less expensive tools including CT scan and ultrasound which can accurately reveal renal agenesis, didelphys and hematometrocolpos [6].

Prompt diagnosis of this condition is usually prevented by its uncommon occurrence. The presence of one unobstructed uterus allowing normal menstrual flow also generally steers diagnosis in the wrong direction. This may cause a significant delay in the accurate diagnosis with many patients simply being put on oral contraceptives or NSAIDs for symptom relief [7]. In our case, the patient was diagnosed at 27 years of age.

Common symptoms include pelvic pain after menarche, dysmenorrhea as well as pelvic mass due to hematometrocolpos from the obstructed vagina. Vaginal obstruction of blood flow is a cause of more serious complications. These might lead to retrograde blood flow which may be a precursor to endometriosis, superimposed infections and pelvic adhesions [5].

Surgical excision of the vaginal septum is usually performed to treat these cases. Our patient underwent urethroplasty, but due to the presence of adequate vaginal space, no other intervention was required.

According to our extensive literature review using PubMed and Google Scholar, ours is an extremely unique case to present with an ectopic urethral opening as well which could be the cause of urinary incontinence.

Conclusion

The case report reinforces the significance of recording and understanding complications associated with rare medical conditions such as HWWS since these can potentially alter the quality of life for the patient and remain symptomatically dormant for a major part of the patient's life. The occurrence of an ectopic urethral opening is also a distinctive manifestation in HWWS, without prior investigative literature specific to this finding present. It is therefore vital that medical professionals consider the incidence of such conditions in patients with any associated presentations to rule out HWWS as a potential causative agent despite the otherwise majorly asymptomatic disposition.

Patient consent

We can confirm that informed consent was obtained from the patient, in a written format, before submitting the manuscript for publication. The patient was made aware of all the shared data, including the age, sex, diagnosis, course, and treatment, along with imaging details. Keeping the privacy of the patient in mind, the name and other personal details, such as residence and contact information, were kept private.

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