Herlyn-Werner-Wunderlich syndrome: a case report and literature review


ABSTRACT

Introduction: Uterus didelphys with blind hemivagina and ipsilateral renal agenesis (Herlyn-Werner-Wunderlich syndrome) is a rare congenital anomaly. There is severe dysmenorrhea and palpable mass due to unilateral hematocolpos. A case of Herlyn-Werner-Wunderlich syndrome with left ureteric bud and bladder diverticulum is presented here.

Clinical case: Patient is an eighteen-year-old female who experienced menarche at twelve years of age. Menstrual cycles lasted twenty-eight days with menstrual bleeding for four days and dysmenorrhea. Left salpingo-oophorectomy was carried out three years prior due to hematosalpinx and ovarian cysts. Disease onset presented with dysuria, cloudy and fetid urine, and recurrent urinary infection symptoms that improved with antimicrobial treatment. Physical examination revealed Tanner IV, normal external genitals, and septate vagina. Imaging studies showed left renal agenesis, left ureterocele, and bladder diverticulum. Cystoscopy revealed left ureterocele and was deroofed by endoscopy. Retrograde pyelography revealed left ureteric bud. Open procedures of left ureterectomy and bladder diverticulectomy were later carried out.

RESUMEN

Introducción: El útero bicornio con hemivagina en fondo de saco y agenesia renal ipsilateral (síndrome de Herlyn Werner Wünderlich), es una anomalía congénita poco frecuente. Existe dismenorrea severa y masa palpable, por hematocolpos unilateral. Presentar el caso de síndrome de Herlyn Werner Wunderlich, con yema ureteral izquierda y divertículo vesical.

Caso clínico: Femenino de 18 años de edad. Menarca a los 12 años. Ciclos menstruales 28 por 4, con dismenorrea. Salpingooforectomía izquierda hace tres años, por hematosalpinx y quistes de ovario. Inició su padecimiento con disuria, orina turbia y fétida, cuadros repetitivos de infección urinaria, presentando mejoría con tratamiento antimicrobiano. A la exploración física se encuentra con Tanner IV, genitales externos normales, vagina septada. Por imagen, existía agenesia renal izquierda, ureterocele izquierdo y divertículo vesical. Por cistoscopia se encontró ureterocele izquierdo, destechándose endoscópicamente. La pielografía ascendente mostró yema ureteral izquierda, posteriormente se realizó ureterectomía izquierda y diverticulectomía vesical abierta.
**Discussion:** The Wolffian ducts give rise to the kidneys and induce Müllerian duct fusion; abnormality in Wolffian duct caudal development is the cause of unilateral renal agenesis. The Müllerian duct on the same side as the absent Wolffian duct is laterally displaced without fusing with the contralateral duct. This results in bicornuate uterus and there is no contact with the central urogenital sinus. The contralateral Müllerian duct gives rise to a vagina and the displaced component forms a blind sac.

**Conclusions:** It is important to detect these types of rare abnormalities that cause precocious symptomatology, pelvic endometriosis, and collection infections.

**Keywords:** Renal agenesis, unilateral hematocolpos, bicornuate uterus, Mexico.

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**INTRODUCTION**

Herlyn-Werner-Wunderlich syndrome is a rare congenital malformation of the Müllerian ducts. It is thought to be a developmental abnormality of the Wolffian ducts as well as of the Müllerian ducts. It is also known today as uterine didelphys associated with obstructed hemivagina and ipsilateral renal anomaly (OHVIRA), a broader term, since it includes other types of renal abnormalities. These alterations are significantly more frequent on the right side and there is no clear explanation for this fact. The disease is rare and prevalence is undetermined. In the general population, unilateral renal agenesis is estimated to be from 1 in every 600-1200 individuals. In women, the prevalence of genital abnormalities associated with kidney anomalies is estimated between 25-89%. The alterations of the urinary tract most frequently associated with developmental abnormalities of the Müllerian ducts are renal agenesis, double collecting system, double kidney, and horseshoe kidney.1-6

**CASE PRESENTATION**

Patient is an 18-year-old woman. She began menarche at 12 years of age. Menstrual cycles lasted for 28 days and bleeding lasted for 4 days with dysmenorrhea. Left salpingo-oophorectomy was carried out three years prior due to hematosalpinx and multiple ovarian cysts.

Three years earlier patient presented with dysuria,

*Figure 1.* Excretory urography at 25 minutes with no evidence of left kidney. Adequate contrast medium elimination in right kidney. Filling defect in left lateral wall of the bladder.
cloudy and fetid urine, and recurrent urinary tract infection symptoms that mildly improved with antimicrobial treatment. Due to persistence of symptomatology, patient was sent to the authors’ unit for evaluation. She continued to have the same symptomatology and urine culture was positive for *Escherichia coli*. Physical examination revealed Tanner IV, normal external genitals, and gynecological examination showed unaltered vaginal introitus and septate vagina.

Kidney and pelvic ultrasound revealed one hypertrophied right kidney with regular edges, left renal agenesis, left ureterocele, and bicornuate uterus.

Excretory urography (Figure 1) showed: Right kidney, enlarged kidney silhouette, adequate uptake and elimination of contrast medium, tortuous distal third of right ureter, and no evidence of obstructive pathology. Left kidney was not visible.

Abdominopelvic computed tomography (CT) scan (Figure 2) showed 13 x 6.5 x 6.3 cm right kidney, adequate uptake and elimination of contrast medium, left renal agenesis, left ureterocele, and bicornuate uterus.

Cystourethrography (Figure 3) showed left ureteral reflux and ectasia and bladder diverticulum in left lateral wall.

Cystoscopy reported 350 mL bladder capacity and orthotopic and ejaculating right ureteral meatus. Extrinsic mass was found in left lateral wall compatible with ureterocele, which was deroofed through endoscopy with exit of chocolate-brown matter and

coagulates. Retrograde pyelography showed incomplete left ureteric bud (up to mid-third), and unaltered right ureter.

After retrograde pyelography findings, the open procedures of left ureterectomy and bladder diverticulectomy were carried out.

Patient is currently asymptomatic and being evaluated by the Department of Gynecology, where she is not considered to be a candidate for another surgical treatment at this time. Patient should remain under surveillance.

**DISCUSSION**

The Wolffian ducts, in addition to giving rise to the kidneys, are factors that induce adequate fusion of the Müllerian ducts. Therefore, abnormality in the development of the caudal portion of the Wolffian ducts can be the cause of unilateral renal agenesis associated with imperforated hemivagina. Left renal agenesis presented in the patient described here. The Müllerian duct is laterally displaced on the side in which the Wolffian duct is absent and cannot fuse with the contralateral duct, resulting in a bicornuate uterus with no central urogenital sinus contact. The contralateral Müllerian duct gives rise to the vagina, while the displaced component forms a blind sac, the obstructed or imperforated hemivagina. In the case described here, the patient presented with septate vagina. The vaginal introitus is not involved because it arises from the urogenital sinus. 3

**CONCLUSIONS**

In clinical practice it is important to detect this type of abnormality, despite its rareness, because it is the cause of precocious symptomatology and facilitates early pelvic endometriosis and collection infections (pyocolpos, pyometra, or pyosalpinx). Perhaps the present patient’s hematosalpinx was an early disease manifestation due to endometriosis associated with these types of cases, but this could not be demonstrated because it did not happen when the patient was at the authors’ institution. Simple surgical procedures (septum excision and collection drainage) provide satisfactory results in relation to pain suppression as well as to reproductive capacity. Because these patients have only one kidney, urinary infection prevention is very important.

**REFERENCES**