CASE REPORT

The Herlyn-Werner-Wunderlich Syndrome: A Rare Case Report

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Abstract

Obstructed Hemivagina and Ipsilateral Renal Agenesis (OHVIRA) is an aberrated and special type of Mesonephric duct malformation and Müllerian duct anomaly. This mostly reports 5% of Müllerian duct anomalies. The majority of cases of OHVIRA are presented alongside uterine didelphys. The presence of uterus didelphys with OHVIRA are the conditions, collectively referred to as the Herlyn- Werner-Wunderlich Syndrome. Its cause is a complete failure of the Müllerian conduits to fuse. Initial symptoms are typically induced by secretions concentrating within the hemivaginal blockage. Patients are usually of reproductive age. Symptoms in patients with this syndrome are mostly complaints of severe dysmenorrhea, pain in the lower abdomen, palpable mass in the pelvis or vagina abrupt urinary retention, fever, or puking, in addition to atypical vaginal secretions, and abnormal vaginal discharge. Cases seldom reveal complaints of infertility in the majority. Here, we focus on the distinctive symptoms of HWW syndrome and the significance of imaging findings in the diagnosis of the condition in a case of a 23-year-old girl who visited our outpatient department primarily with concerns of infertility and periodic pelvic ache. She was initially diagnosed with OHVIRA syndrome through ultrasonography. The outcomes of her MRI are also discussed. The couple was counselled regarding the disease, complications, and treatment options however due to financial constraints they were lost to follow-up.

Keywords: *OHVIRA, uterus didelphys, renal agenesis, hemivagina, infertility.*

INTRODUCTION

Obstructed Hemivagina and Ipsilateral Renal Agenesis (OHVIRA) also comprehended as Herlyn- Werner-Wunderlich Syndrome. It is a strange and one-off kind of Müllerian duct anomaly (MDA). It is an uncommon complex female urogenital contortion [1].

Uterine didelphys is a class III abnormality, according to the American Fertility Society classification, and is interpreted as the partial or complete uterus, cervical, and vaginal duplication [2, 3].

According to research, the prevalence of Congenital Uterine Anomalies is 5.5 percent in the general population, 8 percent in infertile patients, 12.3 percent in patients who had previously experienced a miscarriage, and 24.5 percent in patients who had experienced both miscarriage and infertility [4].

With the OHVIRA Syndrome, each Müllerian duct and hemiuterus are arranged autonomously of one another without evident endometrial attachment of the cavity, and at the level of cervix, a small degree of emulsion is noticed [1, 3, 5, 6].

Mesonephric duct growth is embryonically defective and fails to activate the metanephric blastema, the future kidney. Likewise, the vagina is Wolffian in origin, it also does not form. Also, the loss of mesonephros-derived growth factors interferes with the proper alignment and placement of the paired paramesonephric ducts, leading to nonfusion (uterus didelphys). It includes side fusion deformity in the Müllerian tract's caudal region between 8 and 12 weeks of gestation, as well as malformations accompanied by the absence of immersion from the septum at 20 weeks of gestation [2, 7].

Patients typically first appear during adolescence, not after menarche, and with mostly the symptoms of severe dysmenorrhea, lower pain in the lower abdomen, palpable mass in pelvis or vagina, abrupt urinary retention, fever, or puking, in addition to atypical vaginal secretions. Cases seldom present with infertility in the majority [6, 7].

CASE REPORT

A 23-year-old married female with no known comorbid presented in the outpatient department with complaints of primary infertility and cyclic pelvic pain in the first two days of her cycle, since her menarche at the age of 13 years. She was married for 2 years and has a normal menstrual cycle with bleeding for 4 to 5 days. Although she did not suffer dyspareunia, she expressed minor coitus-related discomfort. She did not use any form of contraception. She took homeopathic drugs for six months for conception. On per abdominal examination, the abdomen was soft and non-tender. No mass was felt. Her secondary sexual characteristics and external genitalia were normal. On pervaginal inspection, the cervix was closed, and a minor protrusion on the lateral wall of the vagina was noticed, her ultrasound and MRI was done findings are discussed below.

Ultrasound Pelvis Findings

On ultrasound, two separate uterine cavities and two separate cervical canals were seen as suggestive of Didelphys uterus. The cervical canal of the right uterine horn appeared dilated showing fluid with echoes within it likely blood in it and measures 3.7 cm in AP dimension

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The retroverted left uterine horn measures 8.1 x 3.0 cm (LS x AP). The endometrium of the left uterine horn measures 0.85 cm. The endocervical canal of the left uterine horn appears unremarkable. The right kidney was not visualized in the renal bed **(Fig. 1A-C)**.

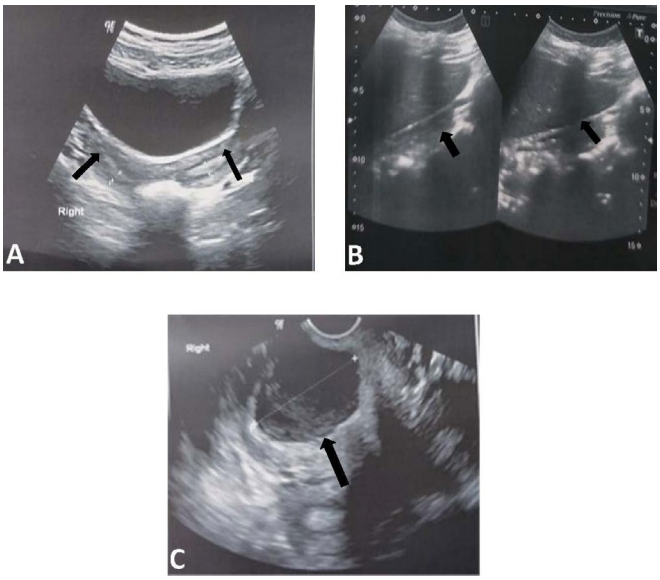

Fig. (1): (A) Shows two separate uterine cavities and two separate cervical canals, **(B)** Shows right renal agenesis, and **(C)** shows dilated right cervical horn.

MRI Pelvis Findings

The above-described ultrasound findings were confirmed on MRI. The uterus was retroverted. It showed two separate uterine cavities that extend to two separate cervical canals with gross dilatation of the cervical canal on the right side secondary to an external Os' stenosis on the right side. The left cervical canal was patent. The diagnosis of Herlyn-Werner-Wunderlich syndrome, also known as OHVIRA SYNDROME, was made taking into consideration the aforementioned findings **(Fig. 2)**.

DISCUSSION

The Herlyn-Werner-Wunderlich syndrome (HWWs) or OHVIRA syndrome is an uncommon inherited female urogenital disorder with the clinical triad of ipsilateral renal agenesis, blocked hemivagina, and uterine didelphys as its defining features [3].

The reported incidence ranges from 0.1-3.8%. 11% of abnormal paramesonephric ducts are caused by uterus didelphys. 5–10% of the paramesonephric duct is caused by a small uterine cavity and an upper vagina that may not exist. Around 43% of patients have associated renal malformation [8].

The pathophysiology of OHVIRA syndrome continues to be unknown [9]. But it is far taken into consideration to result from mixed improper maturation

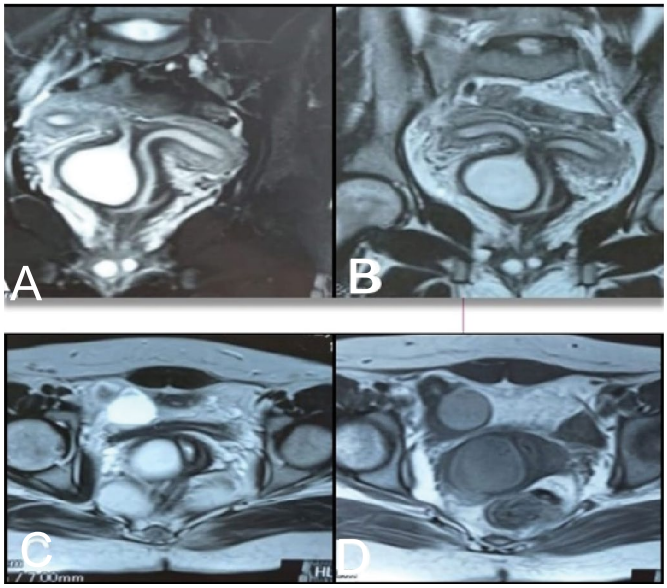

Fig. (2A-D): Coronal and axial cuts show two separate uterine cavities which extend into two separate cervical canals with gross dilatation of the right cervical canal.

of paramesonephric (Müllerian) and mesonephric (Wolffian) ducts [1, 3, 5, 10]. The Wolffian ducts are liable for the formation of kidneys and are also factors for a good enough Müllerian duct combination. Hence, unilateral renal agenesis coupled with imperforate hemivagina may result from a developmental abnormality of one of the Wolffian ducts. A didelphys uterus results from the Müllerian duct being shifted laterally and unable to merge with the contralateral duct on the side where the Wolffian duct is lacking [11-13]. The Müllerian duct is displaced, which prevents it from centrally reaching the urogenital sinus, leading to an imperforate or obstructed hemivagina. The opposite side of the Müllerian duct gives rise to a vagina, whereas the displaced Müllerian duct on the other side creates a blind sac [2, 3].

Renal agenesis is the most common related nongenital anomaly and is usually visible ipsilateral with the obstructed Hemivagina [14-16]. In contrast to the left side, the right side is impacted twice as frequently we also report of right side affected [11, 17]. Different related anomalies encompass renal dysplasia, IVC duplication, ectopic ureter, duplication of the kidneys and ureters, high-riding aortic bifurcation, intestinal malformation, and ovarian malposition [3].

Sufferers of this syndrome are frequently asymptotic up till adulthood as in our case. The majority of the time, a prognosis is determined before menarche because of the lower abdomen pain that is cyclically worsening which was also complained of our patient cyclic pelvic pain in the first two days, and secondary to hematocolpos due

to the long-lasting, partially clotted, retained menstrual blood inside the obstructed Hemivagina [10, 15]. Because of the normal menstrual flow from the patent, unobstructed hemivagina, this syndrome may first go undiagnosed [18].

The most common signs and symptoms are pelvic pain, dysmenorrhoea, and pelvic mass [11, 19] but this case reports the problem of a 23 years old female who visited us with the complaint of primary infertility, It has hardly ever been documented in the literature.

Surgery is used to remove the clogged septum, along with vaginal drainage, as a form of treatment, which lets in the alleviation of acute signs and symptoms. If left untreated, issues such as endometriosis, infertility problems, pelvic adhesion, pyosalpinx, or pyocolpos may result and manifest in the later stages with a significant miscarriage rate [10], there are instances of less common complications such as clear cell carcinoma and adenocarcinoma of the vagina and occluded part of the uterine cervix [11].

CONCLUSION

OHVIRA syndrome is an unprecedented anomaly with possible issues that could arise both soon and later. The prognosis can be overlooked due to the regular menstruation and abdominal pain that is not significant. Highlighting such instances develops consciousness of the disease and enables the acquisition of faster diagnosis and minimizes possible complications because of delayed diagnoses. Vaginal septum-related backflow of genital bleeding can result in endometriosis, pelvic adhesions, or infertility. It is very tough to detect OHVIRA syndrome correctly in the absence of ultrasound and magnetic resonance imaging, and an effective surgical method is to completely separate and remove the obstructing septum.

CLINICAL IMPORTANCE

It is highly rare to have Herlyn-Werner-Wunderlich syndrome. Early detection is crucial for avoiding complications, further research for fertility issues should be done and surgical procedures to ease symptoms are made possible by this.

CONSENT FOR PUBLICATION

Written informed consent was taken from the participants.

CONFLICT OF INTEREST

The authors declare no conflict of interest.

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AUTHOR'S CONTRIBUTION

All authors contributed equally to the work.

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