Case Report

Herlyn-werner-wunderlich syndrome: A rare case

Sachin Wankhede[1], Priyanka Shelkar[2*], Madhuri Patil[3]

[1]Associate Professor, [2]Sr. Resident, [3]Assistant Professor, Department OBGY, Indira Gandhi Government Medical College, Nagpur, Maharashtra, INDIA.
Email: drsachinwankhede@yahoo.co.in, patilmadhuri303@gmail.com, priyanka.shelkar17@gmail.com

Abstract

The Herlyn–Werner–Wunderlich (HWW) syndrome is a rare variant of mullerian duct anomalies characterised by the combination triad of uterus didelphys with obstructed hemivagina and ipsilateral renal agenesis. The exact etiology of HWWS is still unknown, but it may be caused by the abnormal development of Müllerian and Wolffian ducts1,2. Its estimated occurrence is 0.1%–3.8%1. Reporting a case of 14 year girl who came in emergency with c/o severe pain in abdomen. Her ultrasonography and MRI suggested a diagnosis of Herlyn-Werner-Wunderlich syndrome and for that patient underwent resection of vaginal septum.
Key Words: Herlyn-werner-wunderlich syndrome.

***Address for Correspondence:**
Dr. Priyanka Shelkar, Sr. Resident, Department OBGY, Indira Gandhi Government Medical College, Nagpur, Maharashtra, INDIA.
Email: priyanka.shelkar17@gmail.com
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INTRODUCTION

The Mullerian ducts are the primordium from which the female reproductive tract is derived. Mullerian duct anomalies (MDA) are congenital entities resulting from nondevelopment (agenesis or hypoplasia), defective vertical or lateral fusion, or resorption failure of the Müllerian (paramesonephric) ducts. Herlyn-Werner-Wunderlich syndrome (HWWS) represents a type of MDA associated with mesonephric duct anomalies. HWWS, also known as OHVIRA (Obstructed hemivagina and ipsilateral renal agenesis) syndrome represents a complex female genital malformation characterized by uterus didelphys, obstructed hemivagina, and ipsilateral renal agenesis, all 3 being secondary to mesonephric duct–induced müllerian anomalies[1,2]. Herlyn-Werner syndrome (i.e., renal agenesis and an ipsilateral blind hemivagina) was initially described in 1971 by Herlyn and Werner[3,4]. In 1976, Wunderlich described an association of right renal aplasia with a bicornuate uterus and simple vagina in the presence of an isolated hematocervix. Symptoms usually present after menarche, when haematocolpos develops during menstruation, resulting in dysmenorrhea and a pelvic mass. Ultrasonography, typically demonstrates a pelvic fluid collection, which can stimulate other disease such as ovarian cyst, cystic ovarian malignancy, pyosalpinx, hydrosalpinx. MRI is confirmatory. The potential complications of this syndrome are distinct in acute complications, such as pyohematocolpos, pyosalpinx, pelvicperitonitis and long standing complications such as endometriosis, pelvic adhesions and increased risk of abortion or infertility. Thus diagnosis and treatment at an early stage can relieve acute symptoms and preserve normal fertility.

CASE REPORT

14 year old girl presented with abdominal pain that began with onset of menarche (at the age of 13 year) gradually increase in intensity. The patient also had irregular menses every 2-3 months with heavy flow, soaked 4-6 pads per day and lasted for 6-7 days. She had previously been seen at a private hospital two to three months after

the initial symptoms, but no imaging workup nor gynecological examinations were performed at that time. She was being treated with antispasmodic drug and had little relief with it. On examination, her abdomen was soft with just palpable cystic mass in consistency felt.

Ultasonography Features

Two cornua of uterus are seen. Right cornua – 5.8 x 2.3 x 2.7 cm with Endometrial thickness 1.1 cm. Left cornua – 6 x 2 x 2.6 cm with Endometrial thickness 0.5 cm. Left sided cervix is noted. No free fluid noted in POD. Right ovary – 2.6 x 2 cm. Left ovary - 2.2 x 1.6 cm e/o collection with echoes within of size 10.2 x 6 x 6 cm around 95 cc in volume suggestive haematocolpos. This collection is communicating with right sided cornua of uterus. Right kidney not visualized in abdomen and pelvis ? right kidney agenesis. Left kidney hypertrophied.

Impression: Uterus didelphys with large haematocolpos with mild haematometra in right side with right kidney is not visualized in right renal fossa s/o renal agenesis

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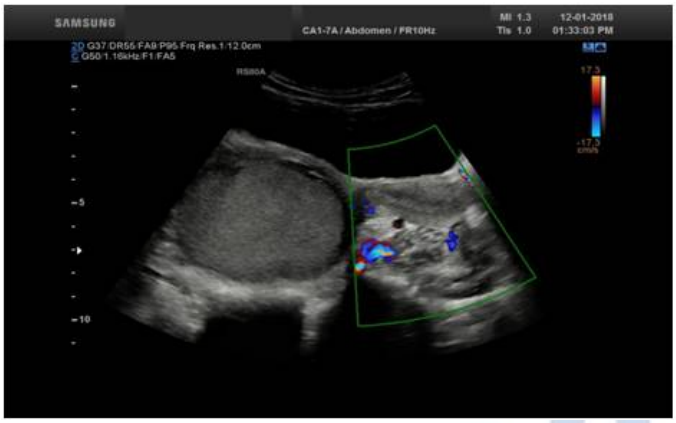

Figure 1:

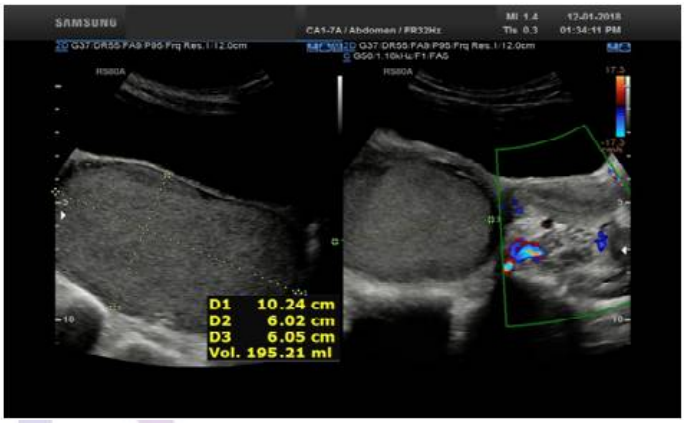

Figure 2:

OBSERVATIONS

There is duplication of uterine horns with their wide separation and without any obvious communication s/o uterine didelphys with T1W and T2W by hyperintense collection within. There is collection of size 7 x 10 x 6.5 cm (approximate volume 230 cc) noted in right hemivagina and right uterine cavity appearing hyperintense on T1W and intermediate signal intensity on T2W sequence s/o hematometra and hematocolpos. Cervix and vagina on left side appears displaced and compressed due to mass effect of right hematocolpos. Bilateral ovaries are visualized appear normal in size and intensity with right ovary measuring 1.5 x 1.8 x 3.2 cm and left ovary measuring 2.1 x 2.2 x 3.8 cm.

Right kidney not visualized in right renal fossa- absent. Left kidney appear normal. Urinary bladder appear adequately filled and reveals signal intensity. No obvious intravesical mass or filling defect seen. Urethra under view appears normal.

Impression

1. Mullerian duct anomaly – uterine didelphys (class III) with right sided completely obstructed hemivagina with resultant hematomera with hematocolpos.
2. Right sided renal agenesis. Features suggestive of Herlyn-Werner-Wunderlich syndrome.

Patient was haemodynamically stable and her laboratory parameters were within normal limit. She was posted for examination under anesthesia. On examination there was a mass of 7 x 7 cm which was bulging into vagina. Cervix could not be visualized. On per rectal examination cystic mass was felt and band like structure felt above the mass on left side. Patient was discharged and called for a follow up visit during her next menses. She was again posted for examination under anesthesia. There was a large bulging swelling seen on right side of vagina. Menstrual blood was seen coming into vagina from left side but cervix was still not visualized. A transverse incision given over the most bulging part of swelling i.e. vaginal septum resection done and around 500 cc hematometra drained out. The incision was extended. After drainage of hematometra cervix was visualized on left side, sounding was done and uterocervical length was three and half inches. On the right side cervix was visible after septum was exiced. Sounding was done on right side and uterocervical length was two inches. Purse string suture was taken around the incision.

DISCUSSION

Mullerian duct anomalies result from non-development, defective fusion or failure of resorption of the Mullerian (paramesonephric) ducts from which originates most of the female genital tract, including the fallopian tubes,

uterus, upper two-thirds of the vagina and the cervix. The upper two-thirds of the vagina is derived from the Mullerian ducts, while the lower third from the urogenital sinus. Wolffian ducts play an important role in the development of internal genital organs and kidney[6]. The association of uterus didelphys with an obstructed hemivagina is explained by a defect in embryogenesis at the eighth week of gestation affecting the mesonephric and Mullerian ducts, in which there is non-fusion of Mullerian ducts or failed resorption of the uterine/vaginal septum[5]. Renal agenesis predicts an ipsilateral obstructive Mullerian duct abnormality 50% of the time and is ipsilateral to the dilated uterine cavity. The right side is affected nearly twice more frequently than the left one[7,8]. HWWS be classified according to the complete or incomplete obstruction of the hemivagina as follows: Classification 1 -patients with a completely obstructed hemivagina. Classification 2 - patients with an incompletely obstructed hemivagina. The mean age at diagnosis of Classification 1 was also significantly younger than the mean age at diagnosis of Classification 2. In addition, patients with Classification 1 are more prone to hematometra, hematosalpinx and hemoperitoneum. Acute onset of abdominal pain, fever and vomiting are common symptoms several months after menarche. Endometriosis is common complications, if not treated in time; the condition can progress to secondary endometriosis, pelvic adhesion, pyosalpinx and even pyocolpos. Although, patients with Classification 2 mainly complaints of purulent or bloody vaginal discharge and ascending genital system infection years after menarche. Most patients with Classification 2 have normal menstrual cycle, but longer menstrual periods, illness attacks years after menarche. HWWS patients most commonly present at puberty, a few years after menarche, although it can present in adulthood as primary infertility and has also been reported in a neonate, presenting as a mass prolapsing per vaginum[11]. Usually, HWWS patients have a normal onset of menstrual cycles with bleeding from the unobstructed hemiuterus. The primary presenting symptom is cyclical lower abdominal pain evolving into continuous severe pain with increasing distension of the obstructed hemivagina which may be associated with a foul-smelling mucopurulent discharge[12]. On physical examination, a unilateral pelvic mass is usually felt twice as often on the right side than on the left side[12,13,14]. A lower paravaginal bulging mass may be palpable, which could obstruct the contralateral hemivagina. Partially clotted blood collected over a period of time in the hematocolpos with subsequent hematometra and hematosalpinx leads to abdominal and pelvic pain in these patients[14]. In delayed cases, bleeding into the peritoneal space may occur as a consequence of retrograde menstruation. Delay in diagnosis can lead to complications that include endometriosis, adhesions, infertility and infectious complications arising from chronic cryptomenorrhea[9,13,14]. It is not easy to achieve to an early diagnosis, since menstruation is often regular and patients are usually given anti-inflammatory drugs and oral-contraceptives for cyclic dysmenorrhea. This causes a delay in the diagnosis because they reduce or eliminate menses. Although ultrasound and CT scan are commonly used for diagnosis because of their low cost but MRI is the most accurate method for diagnosis up to 100% in some reports[9,13,15]. Ultrasonography is useful in the diagnosis of MDA and the detection of haematocolpos, which appears as a fluid collection with low level echoes and can make the diagnosis of genito-urinary tract anomaly easier, although it may not identify the type of MDA[9,13]. MRI, is a suitable technique for the non-invasive evaluation of female pelvic anatomy because it provides more detailed information regarding uterine contour, the shape of the intrauterine cavity morphology, the continuity with each vagina (obstructed and non-obstructed) lumen, the character of the septum, and fluid content nature, as well as associated aspects such as endometriosis, pelvic inflammation and adhesions and renal anomalies[10,12,13]. Although, laparoscopy is considered the gold standard for the evaluation of female reproductive tract anomaly, in HWWS, it is reserved when the diagnosis is not clear after imaging or when MRI is not available[9,13]. To alleviate symptoms and retain fertility in these patients, the most effective treatment is surgery. Resection of as much of the obstructing vaginal septum as possible is the optimal surgery for these patients. Preservation of the hymen is desirable in younger patients, although not always feasible. Most patients can recover completely after resection of the vaginal septum. The best time for surgery in these patients is approximately at the time of menstruation, as a large distended hematocolpos is easy to visualize and palpate, which aids in resection. Patients who remain with stenosis of the vaginal septum after surgery can safely undergo re-resection of the septum with preserved ability to conceive and maintain pregnancy[16]. Either total or unilateral hysterectomy may be required in cases in which septal resection is not possible[17] and may also be considered in patients with recurrent stenosis and severe endometrial/uterine infection or in patients who do not wish further pregnancies[16]. Prognosis is good, with the major concern being preservation of fertility. Women with uterus didelphys have a high likelihood of becoming pregnant[16,18,19,20], with approximately 80% of patients able to conceive[21,22], but with elevated rates of premature delivery (22%) and abortion (74%). In summary, due to the rare and frequently asymptomatic nature of the HWW

syndrome, gynecologists and other medical providers seeing patients diagnosed with renal agenesis should be aware of the possibility of associated uterine and vaginal abnormalities; conversely, in patients with uterine and vaginal abnormalities, workup for associated abnormalities, especially renal developmental defects, should be performed. Initial workup should include vaginal exam as well as transvaginal ultrasound or pelvic MRI. Referral to a gynecological or pediatric surgeon for vaginal septotomy should be prompt, in order to diminish the risk of endometriosis and infertility. Patients with the condition should be counseled about risks associated with the disease, including a higher likelihood of miscarriages, although around 65% of patients will be able to carry pregnancy to term.

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